

Article

SMARCB1 (INI-1)-Deficient Sinonasal Carcinoma: A Systematic Review and Pooled Analysis of Treatment Outcomes

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Supplementary Methods

Literature search strategy and criteria

(((((((((((SMARCB1[title/abstract]) OR SMARCB-1[title/abstract]) OR INI1[title/abstract]) OR INI-1[title/abstract]) OR INI?[title/abstract]) OR INI*[title/abstract]) AND sinonasal[title/abstract]) OR paranasal[title/abstract]) OR paranasal sinuses[title/abstract]) OR nasal[title/abstract]))) AND ("0001/01/01"[Date - Publication] : "2022/03/31"[Date - Publication]))

Publications excluded from the systematic review

1. Agaimy, A. The expanding family of SMARCB1(INI1)-deficient neoplasia: implications of phenotypic, biological, and molecular heterogeneity. *Adv. Anat. Pathol.* **2014**, *21*, 394–410.

Reason: It is a general overview article which describes SMARCB1-deficient sinonasal carcinoma as a new disease entity of sinonasal malignancy with a different genetic and molecular characteristics, but without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma.

2. Agaimy, A. Poorly differentiated sinonasal tract malignancies: A review focusing on recently described entities. *Cesk. Patol.* **2016**; *52*, 146–153.

Reason: It is a general overview article which describes SMARCB1-deficient sinonasal carcinoma as a new disease entity of sinonasal malignancy with a different genetic and molecular characteristics, but without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma.

3. Bishop, J.A. Newly described tumor entities in sinonasal tract pathology. *Head Neck Pathol.* **2016**, *10*, 23–31.

Reason: It is a general overview article which describes SMARCB1-deficient sinonasal carcinoma as a new disease entity of sinonasal malignancy with a different genetic and molecular characteristics, but without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma.

4. Bishop, J.A. Recently described neoplasms of the sinonasal tract. *Semin. Diagn. Pathol.* **2016**, *33*, 62–70.

Reason: It is a general overview article which describes SMARCB1-deficient sinonasal carcinoma as a new disease entity of sinonasal malignancy with a different genetic and molecular characteristics, but without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma.

5. Barresi, V.; Branca, G.; Raso, A.; Mascelli, S.; Caffo, M.; Tuccari, G. Atypical teratoid rhabdoid tumour involving the nasal cavities and anterior skull base. *Neuropathol.* **2016**, *36*, 283–289.

Reason: It is a general overview article of atypical teratoid rhabdoid tumour in the nasal cavities and anterior skull base, without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma.

6. Agaimy, A.; Weichert, W. SMARCA4-deficient sinonasal carcinoma. *Head and Neck Pathol.* **2017**, *11*, 541–545.
Reason: It is a general overview article of SMARCB4-deficient sinonasal carcinoma, without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma.
7. Stelow, E.B.; Bishop, J.A. Update from the 4th Edition of the World Health Organization Classification of head and neck tumours: tumors of the nasal cavity, paranasal sinuses and skull base. *Head and Neck Pathol.* **2017**, *11*, 3–15.
Reason: It is a general overview article which describes SMARCB1-deficient sinonasal carcinoma as a new disease entity of sinonasal malignancy, without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma.
8. Schaefer, I.M.; Agaimy, A.; Fletcher, C.D.; Hornick, J.L. Claudin-4 expression distinguishes SWI/SNF complex-deficient undifferentiated carcinomas from sarcomas. *Mod. Pathol.* **2017**, *30*, 539–548.
Reason: It is a review article which described claudin-4 expression in SWI/SNF complex-deficient (SMARCB1-deficient) undifferentiated carcinoma, without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma.
9. Pawel, B.R. SMARCB1-deficient tumors of childhood: a practical guide. *Pediatr. Dev. Pathol.* **2018**, *21*, 6–28.
Reason: It is a general overview article of SMARCB1-deficient tumour of childhood, without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma.
10. Thompson, L.D.R.; Franchi, A. New tumor entities in the 4th edition of the World Health Organization classification of head and neck tumors: Nasal cavity, paranasal sinuses and skull base. *Virchows Arch.* **2018**, *472*, 315–330.
Reason: It is a general overview article which describes SMARCB1-deficient sinonasal carcinoma as a new disease entity of sinonasal malignancy, without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma.
11. Laco, J.; Kovarikova, H.; Chmelarova, M.; Vosmikova, H.; Sieglova, K.; Bubancova, I.; Dunder, P.; Nemejcova, K.; Michalek, J.; Celakovsky, P.; et al. Analysis of DNA methylation and microRNA expression in NUT (nuclear protein in testis) midline carcinoma of the sinonasal tract: a clinicopathological, immunohistochemical and molecular genetic study. *Neoplasma* **2018**, *65*, 113–123.
Reason: It is a journal article which analysed DNA methylation and microRNA expression in NUT midline carcinoma of the sinonasal tract, without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma.
12. Agaimy, A.; Weichert, W.; Haller, F.; Hartmann, A. Diagnostic and predictive molecular pathology of head and neck neoplasms. *Pathologe* **2018**, *39*, 42–47.
Reason: It is a review article without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma. Besides it is not written in English but in German.
13. Agaimy, A.; Haller, F.; Hartmann, A. Sinonasal tumors: News from the WHO with special reference to mesenchymal entities. *Pathologe* **2018**, *39*, 18–26.
Reason: It is a review article without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma. Besides it is not written in English but in German.
14. McCuiston, A.; Bishop, J.A. Usefulness of NKX2.2 Immunohistochemistry for Distinguishing Ewing Sarcoma from Other Sinonasal Small Round Blue Cell Tumors. *Head Neck Pathol.* **2018**, *12*, 89–94.

Reason: It is a journal article which described the use of NKX2.2 immunohistochemistry to distinguish Ewing sarcoma from other sinonasal malignant without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma.

15. Rooper, L.M.; Bishop, J.A.; Westra, W.H. INSM1 is a Sensitive and Specific Marker of Neuroendocrine Differentiation in Head and Neck Tumors. *Am. J. Surg. Pathol.* **2018**, *42*, 665–671.

Reason: It is a journal article without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma.

16. Schaefer, I.M.; Hornick, J.L. Diagnostic Immunohistochemistry for Soft Tissue and Bone Tumors: An Update. *Adv. Anat. Pathol.* **2018**, *25*, 400–412.

Reason: It is a review article without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma.

17. Silveira, H.A.; Almeida, L.Y.; Nonaka, C.F.W.; Alves, P.M.; Ribeiro-Silva, A.; León, J.E. Myoepithelial carcinoma with rhabdoid features in the maxillary sinus: Immunohistochemical and in situ hybridization analysis of a rare case. *Oral Oncol.* **2019**, *93*, 116–119.

Reason: It is a journal article which described myoepithelial carcinoma in the maxillary sinus, without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma.

18. Guilmette, J.; Sadow, P.M. High-grade sinonasal carcinoma: classification through molecular profiling. *Arch. Pathol. Lab. Med.* **2019**, *143*, 1416–1419.

Reason: It is a review article only without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma.

19. Tang, S.J.; Zhai, C.W.; Yuan, C.C.; Zhang, J.H.; Want, S.Y. SMARCB1 (INI1)-deficient sinonasal carcinoma: a clinicopathological analysis of six cases. *Zhonghua Bing Li Xue Za Zhi.* **2020**, *49*, 47–51.

Reason: It is not written in English but in Chinese.

20. Neves-Silva, R.; Almeida, L.Y.; Silveira, H.A.; Colturato, C.B.N.; Duarte, A.; Ferrisse, T.M.; Silva, E.V.; Vanzolin, B.F.; Bufalino, A.; Ribeiro-Silva, A.; et al. SMARCB1 (INI-1) and NUT immunoexpression in a large series of head and neck carcinomas in a Brazilian reference center. *Head Neck.* **2020**, *42*, 374–384.

Reason: It is a journal article which described SMARCB1 (INI-1) tumours outside the sinonasal region or paranasal sinuses.

21. Agaimy, A.; Franchi, A.; Lund, V.J.; Skálová, A.; Bishop, J.A.; Triantafyllou, A.; Andreasen, S.; Gnepp, D.R.; Hellquist, H.; Thompson, L.D.R.; et al. Sinonasal undifferentiated carcinoma (SNUC): from an entity to morphologic pattern and back again – a historical perspective. *Adv. Anat. Pathol.* **2020**, *27*, 51–60.

Reason: It is a review article without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma.

22. Agaimy, A.; Jain, D.; Uddin, N.; Rooper, L.M.; Bishop, J.A. SMARCA4-deficient sinonasal carcinoma: a series of 10 cases expanding the genetic spectrum of SWI/SNF-driven sinonasal malignancies. *Am. J. Surg. Pathol.* **2020**, *44*, 703–710.

Reason: It is an article on SMARCB4-deficient sinonasal carcinoma, without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma.

23. Chen, Z.W.; Fang, S.G.; Ren, J.Q.; Qian, L.J.; Shi, H.Q. Clinical and pathological analysis of 2 cases of SMARCB1(INI-1)-deficient sinonasal carcinoma. *Zhonghua Er Bi Yan Hou Tou Jing Wai Ke Za Zhi.* **2020**, *55*, 400–402.

Reason: It is not written in English but in Chinese.

24. Kezlarian, B.E.; Lin, O.; Dogan, S. SMARCB1-deficient carcinomas of the head and neck region: a cytopathologic characterization. *J. Am. Soc. Cytopathol.* **2020**, *9*, 494–501.
Reason: It is a review article without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma.
25. Heft Neal, M.E.; Birkeland, A.C.; Bhangale, A.D.; Zhai, J.; Kulkarni, A.; Foltin, S.K.; Jewell, B.M.; Ludwig, M.L.; Pinatti, L.; Jiang, H.; et al. Genetic analysis of sinonasal undifferentiated carcinoma discovers recurrent SWI/SNF alterations and a novel PGAP3-SRPK1 fusion gene. *BMC Cancer.* **2021**, *21*, 636.
Reason: It is a review article without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma.
26. Glöss, S.; Jurmeister, P.; Thieme, A.; Schmid, S.; Cai, W.Y.; Serrette, R.N.; Perner, S.; Ribbat-Idel, J.; Pagenstecher, A.; Bläker, H.; et al. IDH2 R172 Mutations Across Poorly Differentiated Sinonasal Tract Malignancies: Forty Molecularly Homogenous and Histologically Variable Cases With Favorable Outcome. *Am. J. Surg. Pathol.* **2021**, *45*, 1190–1204.
Reason: It is a review article without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma.
27. Wang, J.Y.; Bai, Y.P.; Xing, L.; Piao, Y.S.; He, X.J.; Yue, C.L.; Zhao, X.L.; Liu, H.G. Clinicopathological characteristics of SMARCB1(INI1)-deficient sinonasal carcinoma. *Zhonghua Bing Li Xue Za Zhi.* **2021**, *50*, 1240–1245.
Reason: It is not written in English but in Chinese.
28. Liu, Z.; Seshadri, M.; Gupta, V.; Papanicolau-Sengos, A.; Merzianu, M. INI1-deficient thyroid carcinoma is an aggressive disease with epithelioid and rhabdoid phenotype. A case report, survey of INI1 expression in thyroid lesions and literature review. *Head Neck Pathol.* **2021**, *15*, 1246–1252.
Reason: It is a journal article on INI1-deficient thyroid cancer but not on SMARCB1-deficient sinonasal carcinoma.
29. Agaimy, A. Moving from “single gene” concept to “functionally homologous multigene complex”: The SWI/SNF paradigm. *Semin. Diagn. Pathol.* **2021**, *38*, 165–166.
Reason: It is a review article without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma.
30. Agaimy, A.; Bishop, J.A. SWI/SNF-deficient head and neck neoplasms: An overview. *Semin. Diagn. Pathol.* **2021**; *38*, 175–182.
Reason: It is a review article without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma.
31. Lopez, D.C.; Wadley, A.E.; London, N.R. Jr. Emerging concepts in sinonasal tumor research. *Curr. Opin. Otolaryngol. Head Neck Surg.* **2022**, *30*, 33–39.
Reason: It is a review article without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma.
32. Sakthivel, P.; Raveendran, S.; Rajeshwari, M. Sinonasal small round blue cell tumors: What the clinician must know! *Oral Oncol.* **2022**, *126*, 105735.
Reason: It is a review article without any individual patient data and survival outcomes of SMARCB1-deficient sinonasal carcinoma.
33. Gonçalves, J.M.; Scarini, J.F.; Gondak, R.; Altemani, A.; Mariano, F.V. Oral involvement of sinonasal undifferentiated carcinoma: A case report and immunohistochemical study of a challenging case. *Oral Oncol.* **2022**, *126*, 105779.
Reason: It is a case report describing a SMARCB1-positive sinonasal carcinoma.

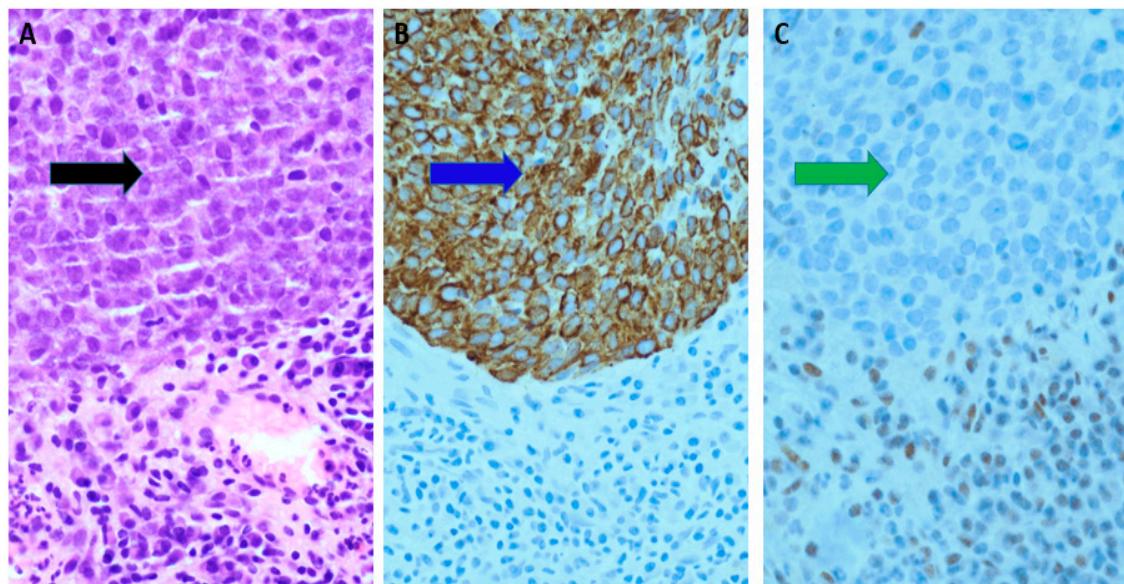


Figure S1. The tumor of our patient (Case number 124 in the Table S2) shows (A) a predominant basaloid cell pattern (haematoxylin and eosin $\times 400$, black arrow). (B) These tumor cells are strongly immunoreactive to pan-cytokeratins (immunohistochemical staining for cytokeratins MNF116, blx400, blue arrow). (C) Loss of expression of SMACRB1 is demonstrated, contrasting with retained signals in the stromal inflammatory cells (immunohistochemical staining for INI-1, $\times 400$, green arrow).



Patient Name	Tumor Type
	Nasopharynx and paranasal sinuses undifferentiated carcinoma
Date of Birth	Medical Facility
Sex	Ordering Physician
FMI Case #	Additional Recipient
Medical Record #	Medical Facility ID #
Specimen ID	Pathologist

ABOUT THE TEST:

FoundationOne™ is a next-generation sequencing (NGS) based assay that identifies genomic alterations within hundreds of cancer-related genes.

PATIENT RESULTS**TUMOR TYPE: NASOPHARYNX AND PARANASAL SINUSES UNDIFFERENTIATED CARCINOMA**

3 genomic findings

0 therapies associated with potential clinical benefit

0 therapies associated with lack of response

10 clinical trials

Genomic Alteration Identified[†]*SMARCB1* loss**Additional Findings[†]***Microsatellite status* MS-Stable*Tumor Mutational Burden* TMB-Low; 1 Muts/Mb

[†] For a complete list of the genes assayed and performance specifications, please refer to the Appendix

THERAPEUTIC IMPLICATIONS

Genomic Findings Detected	FDA-Approved Therapies (in patient's tumor type)	FDA-Approved Therapies (in another tumor type)	Potential Clinical Trials
<i>SMARCB1</i> loss	None	None	Yes, see clinical trials section
<i>Microsatellite status</i> MS-Stable	None	None	None
<i>Tumor Mutational Burden</i> TMB-Low; 1 Muts/Mb	None	None	None

Note: Genomic alterations detected may be associated with activity of certain FDA-approved drugs; however, the agents listed in this report may have little or no evidence in the patient's tumor type. Neither the therapeutic agents nor the trials identified are ranked in order of potential or predicted efficacy for this patient, nor are they ranked in order of level of evidence for this patient's tumor type.

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Patient Name
Report Date
Tumor Type

FOUNDATION ONE®

20 July 2018
Nasopharynx and paranasal sinuses undifferentiated carcinoma

GENOMIC ALTERATIONS

GENE ALTERATION	INTERPRETATION
● SMARCB1 loss	<p>Gene and Alteration: The SMARCB1 gene encodes the SNF5 protein, also known as INI1, one of three core subunits of the SWI/SNF family of chromatin remodelling complexes¹. SNF5, as a member of the remodelling complex, plays a key role in cell cycle control and can act as a tumor suppressor². Alterations in SMARCB1 that remove or disrupt the DNA binding domain (amino acids 106-183) or protein-protein interaction domains (amino acids 186-318)^{14,5}, as observed here, are predicted to be inactivating.</p> <p>Frequency and Prognosis: In a genomic analysis of 56 nasopharyngeal carcinoma cases, alterations of SMARCB1 was not detected in any of the samples⁶. Loss of SMARCB1 has been reported in all three sinonasal basaloid carcinomas analyzed in one study⁷. Additionally, the loss of SMARCB1 protein expression was reported in 6% of 142 sinonasal carcinomas in another study, with genomic loss of SMARCB1 identified in 75% (6/8) of patients in that subset⁸. These studies suggest that SMARCB1 loss may indicate a sinonasal carcinoma characterized by poor differentiation and aggressive clinicopathologic features.</p> <p>Potential Treatment Strategies: There are no therapies that directly target SMARCB1 loss or inactivating mutations. Preclinical evidence suggests that tumors with loss of SMARCB1 may be sensitive to a variety of targeted therapies, including EZH2 inhibitors^{9,10}, inhibitors of the hedgehog pathway¹¹, CDK4/6 inhibitors^{12,13}, proteasome inhibitors¹⁴, inhibitors of fibroblast growth receptors (FGFRs)¹⁵, and Aurora kinase inhibitors¹⁶. Several of these agents are being tested in clinical trials. A clinical trial of the Aurora kinase A inhibitor alisertib for pediatric patients with atypical teratoid rhabdoid tumors, which are characterized by SMARCB1 loss or inactivation, reported disease stabilization or regression in 100% (4/4) of patients, and 2 patients achieved disease regression lasting more than 1 year¹⁷. EZH2 inhibitor tazemetostat (EPZ-6438) has shown efficacy, including several complete responses, in adult and pediatric patients with INI1-negative tumors in early phase trials (Gounder et al., 2017; ASCO Abstract 11058 , Chi et al., 2017; AACR-NCI-EORTC Abstract 175, Taliano et al., 2015; ECC Abstract 302), with one study of INI1-negative epithelioid sarcoma reporting 4 partial responses and 18 stable disease versus 7 progressive disease, with median progression-free survival of 5.7 months (Gounder et al., 2017; ASCO Abstract 11058). Clinical data suggest that CDK4/6 inhibitors, such as ribociclib, abemaciclib, and palbociclib, have efficacy for the treatment of solid tumors^{18,19,20,21,22}.</p>
● Microsatellite status MS-Stable	<p>Gene and Alteration: Microsatellite instability (MSI) is a condition of genetic hypermutability that generates excessive amounts of short insertion/deletion mutations in the genome; it generally occurs at microsatellite DNA sequences and is caused by a deficiency in DNA mismatch repair (MMR) in the tumor²³. Defective MMR and consequent MSI occur as a result of genetic or epigenetic inactivation of one of the MMR pathway proteins, primarily MLH1, MSH2, MSH6, or PMS2^{23,24,25}. The tumor seen here is microsatellite-stable (MSS), equivalent to the clinical definition of an MSS tumor: one with mutations in none of the tested microsatellite markers^{26,27,28}. MSS status indicates MMR proficiency and typically correlates with intact expression of all MMR family proteins^{23,25,27,28}.</p>

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Electronically Signed by Jo-Anne Vergilio, M.D. | Jeffrey S. Ross, M.D., Medical Director
| 20 July 2018
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Sample Preparation: 7010 Kit Creek Road, Morrisville, NC 27560 / CLIA: 34D2044309
Sample Analysis: 150 Second St., 1st Floor, Cambridge, MA 02141 / CLIA: 22D2027531

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Figure S2. Comprehensive genomic profiling with FoundationOne® of the tumour in our patient (Case number 124 in the Table S2) revealing a complete genomic loss of SMARCB1.

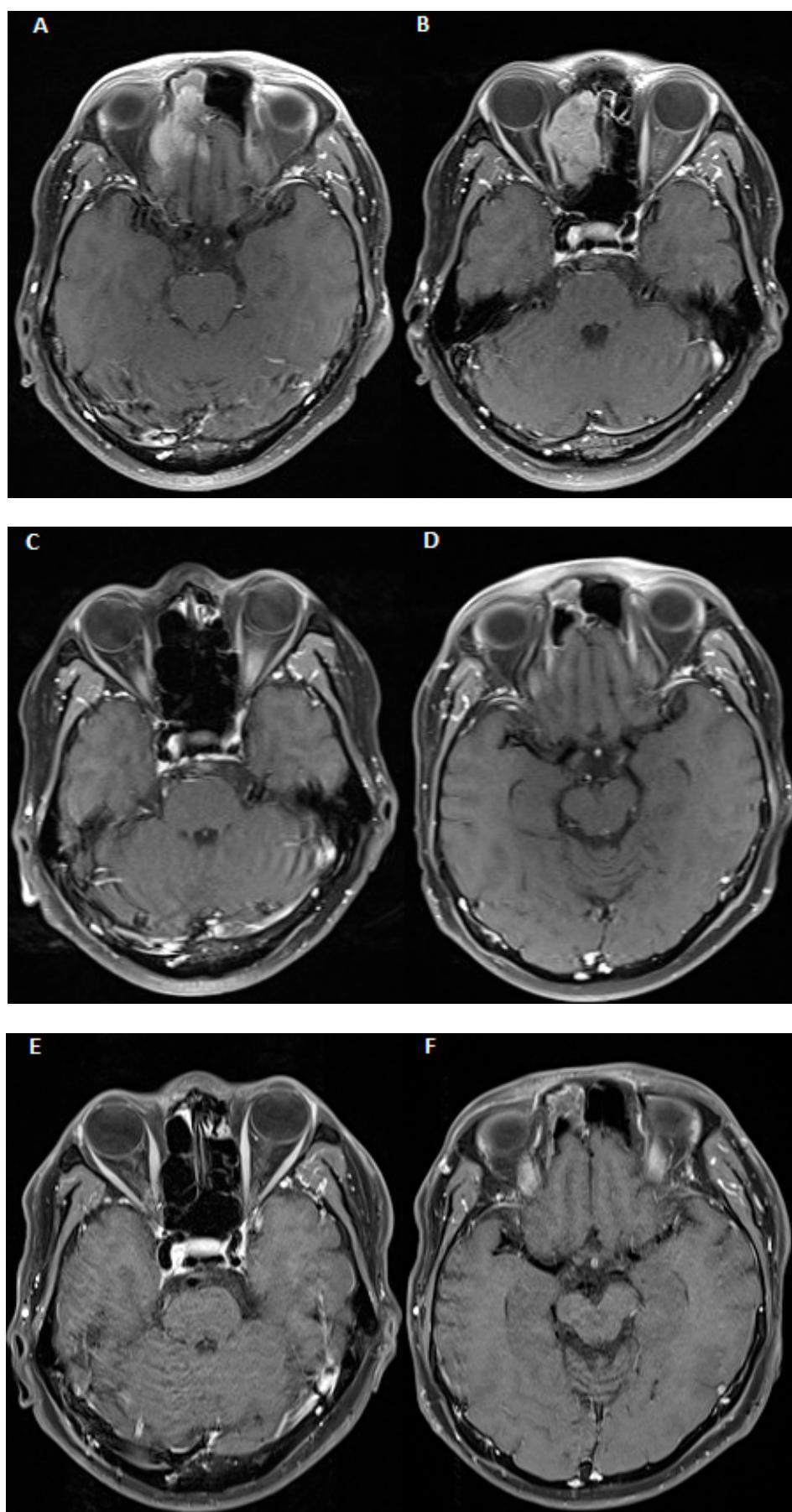




Figure S3. Radiological images of our patient (Case number 124 in the Table S2) treated at our institution. (A) and (B) Magnetic resonance imaging of the paranasal sinuses showing a T1-weighted fat-saturated contrast-enhancing soft tissue mass in the right ethmoid and frontal sinuses invading the lamina papyracea and compressing the right medial rectus muscle at the time of histological diagnosis of SMARCB1-deficient sinonasal carcinoma. (C) and (D) Magnetic resonance imaging of the paranasal sinus at 12 weeks after completion of induction chemotherapy with docetaxel, cisplatin and 5-fluorouracil (TPF) and radical concurrent chemoradiation with photons of 60Gy in 30 fractions and 2 cycles of 3-weekly cisplatin demonstrating a complete radiological response. (E) and (F) Magnetic resonance imaging at 6 months after induction chemotherapy and concurrent chemoradiation demonstrating persistent complete response. (G) and (H) Contrast-enhanced computed tomography of the paranasal sinus at 2.5 years after induction chemotherapy and concurrent chemoradiation demonstrating persistent complete response.

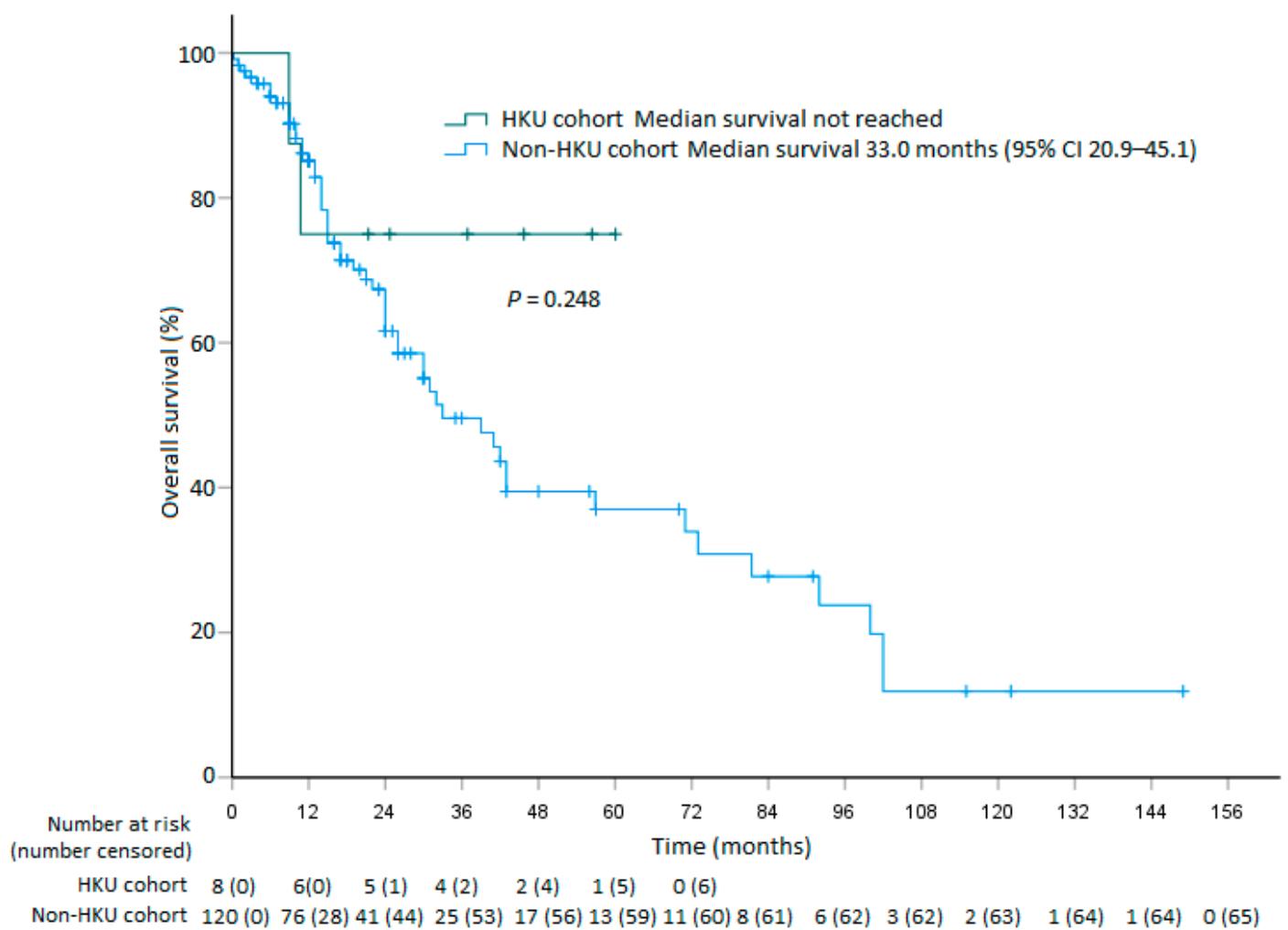


Figure S4. Kaplan-Meier estimates of overall survival of patients with SMARCB1-deficient sinonasal carcinoma identified from systematic review and those treated at our institution.

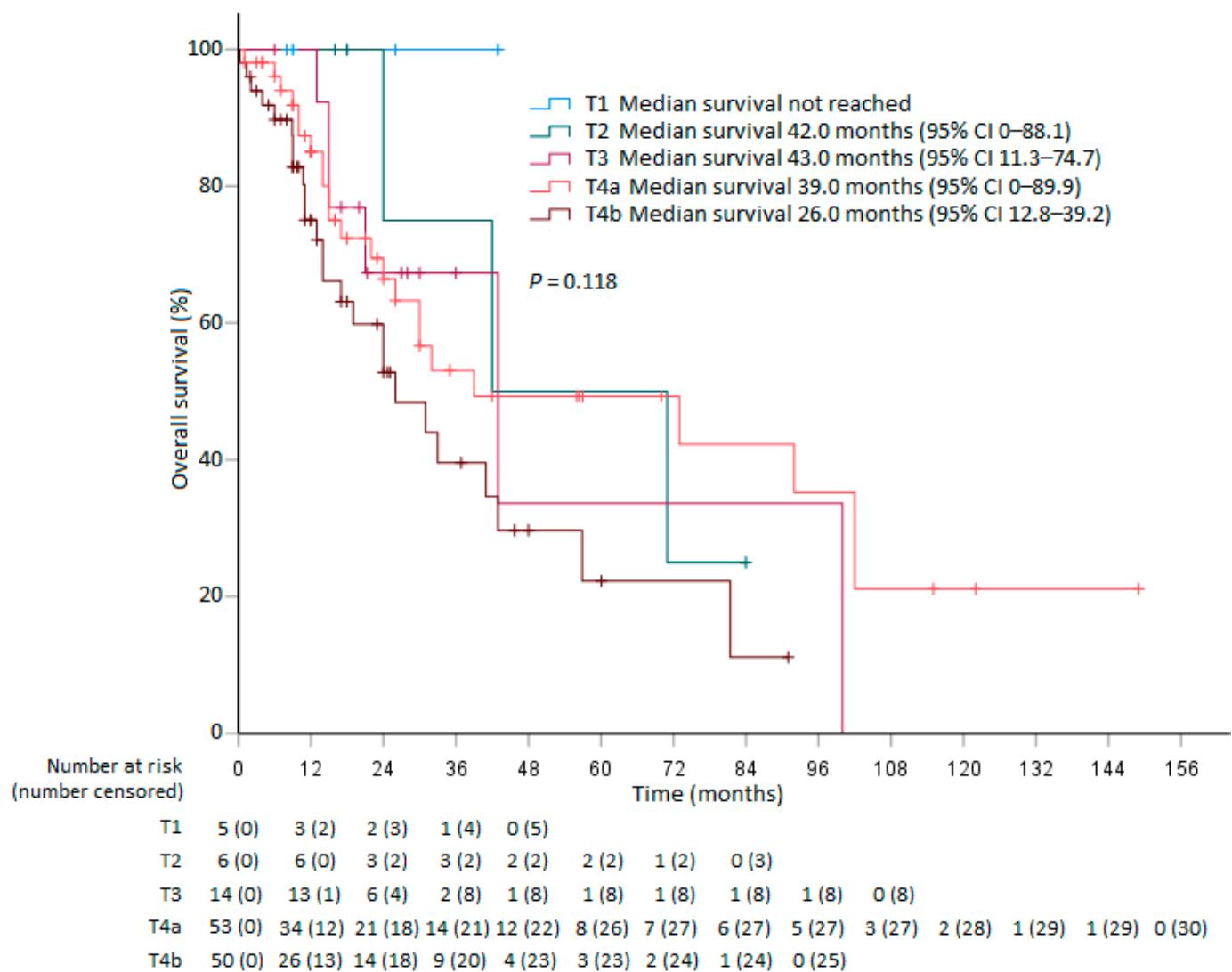


Figure S5. Kaplan-Meier estimates of overall survival of patients with SMARCB1-deficient sinonasal carcinoma stratified by T-categories.

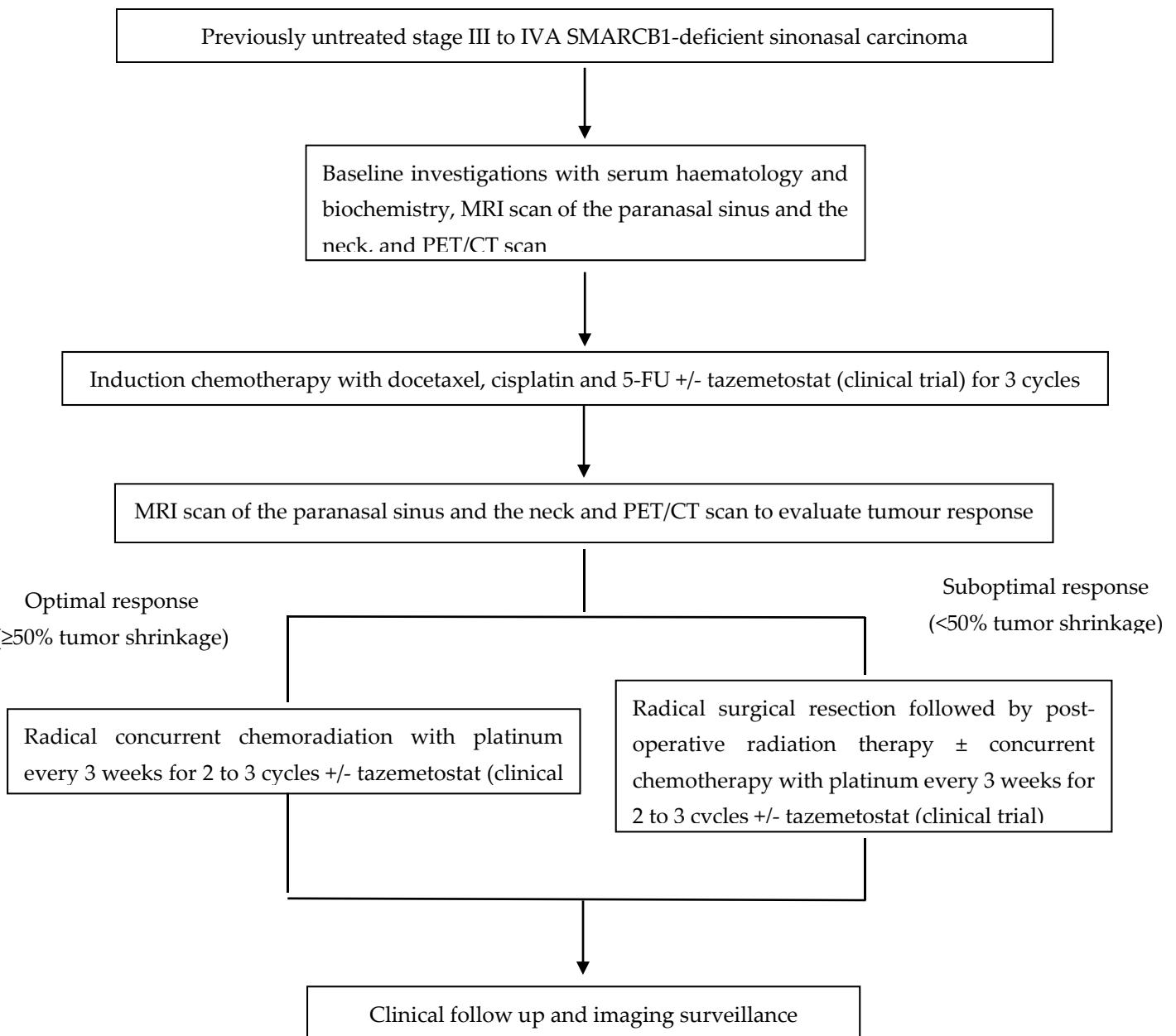


Figure S6. Our proposed treatment strategy for previously untreated stage III to IVA SMARCB1-deficient sinonasal carcinoma.

Table S1. T and N categories for tumors of the nasal cavity and paranasal sinuses according to the 8th edition of American Joint Committee on Cancer/Union for International Cancer Control TNM classification system.

T category	Maxillary sinus	Nasal cavity and ethmoid sinus	N category	Lymph node metastasis
Tis	Carcinoma in-situ	Carcinoma in-situ	N0	No regional lymph node metastasis
T1	Tumor limited to the mucosal lining	Tumor limited to the mucosal lining	N1	Metastasis in a single ipsilateral lymph node, 3cm or less in greatest dimension
T2	Bone erosion or destruction limited to the hard palate and middle meatus	Tumor at 2 subsites or adjacent nasoethmoid sites	N2a	Metastasis in a single ipsilateral lymph node 3–6cm
T3	Bone erosion or destruction of the posterior bone of maxillary sinus, floor and medial bone of orbit. Tumor growth into the pterygoid fossa or ethmoid sinus	Bone erosion of lamina papyracea or floor of orbit, maxillary sinus, palate and cribriform plate	N2b	Metastases in multiple ipsilateral lymph nodes <6cm
T4a	Tumor growth into the anterior orbit, pterygoid plates, infratemporal fossa, cribriform plate, frontal sinus, sphenoid sinus, or skin of cheek	Tumor growth into the anterior orbit, anterior cranial fossa, pterygoid fossa, pterygoid plates, frontal sinus, sphenoid sinus, or skin of nose or cheek	N2c	Metastases in bilateral or contralateral lymph nodes <6cm
T4b	Tumor growth into the orbital apex, dura mater, brain middle cranial fossa, cranial nerves other than V2, nasopharynx, or clivus		N3	Metastasis in a lymph node >6cm

Table S2. Details of all 128 patients with SMARCB1-deficient sinonasal carcinoma.

Case number	Author*	Year	Age at diagnosis	Sex (0=male; 1=female)	T-stage	N-category	M-category	Tumour extent	Treatment	Preoperative treatment			Multimodality treatment and postoperative treatment	Relapse (0=no; 1=yes)	Local relapse (0=no; 1=yes)	Regional relapse (0=no; 1=yes)	Distant relapse (0=no; 1=yes)	Survival status (0=alive; 1=dead)	Duration of survival (months)	Evidence of disease (0=alive; 1=survival; 2=persistent disease; 3=died of disease; 4=died of other causes)	Predominant histological features (if available)
										Induction treatment (0=no; 1=yes)	Adjuvant treatment (0=no; 1=yes)	operation (0=no; 1=yes)									
1	Agaimy et al, ¹	2014	52	0	4a	1	0	Nasal cavity, left frontal and ethmoidal sinuses	Radical resection of the sinonasal lesion and neck dissection followed by postoperative concurrent chemoradiation	0	1	0	1	1	0	1	0	1	102.0	2	-
2	Agaimy et al, ¹	2014	28	1	2	0	0	Nasal cavity, right frontal sinus and anterior ethmoid sinus	Multiple local resections	0	0	0	0	0	0	0	0	0	84.0	0	-
3	Agaimy et al, ¹	2014	35	1	4a	0	0	Right anterior ethmoidal sinus with invasion into the frontal sinuses and	Radical resection of the tumor with enucleation of the ocular bulb and resection of	0	1	0	1	1	0	0	1	0	70.0	1	-

Ref	Author	Year	n	Site		Treatment		Follow-up		Locoregional failure		Distant metastasis		Overall survival		Cause-specific survival		Histology		
				Primary tumor	Nodal status	Treatment	Time	n	Failure	Failure	Death	n	Failure	Death	n	Failure	Death			
4	Bishop et al, ²	2014	54	1	4a	0	0	Nasal cavity, ethmoidal and maxillary sinuses	Radical resection followed by postoperative chemoradiation	0	1	0	1	0	1	0	0	15.0	2	Undifferentiated
5	Bishop et al, ²	2014	71	1	3	0	0	Maxillary sinus	Radical resection followed by postoperative radiation therapy	0	1	0	1	1	1	0	0	15.0	2	High-grade non-intestinal adenocarcinoma
6	Bishop et al, ²	2014	33	0	4a	0	0	Nasal, ethmoidal and frontal sinuses	Radical resection followed by postoperative radiation therapy	0	1	0	1	1	1	0	1	30.0	2	Undifferentiated carcinoma
7	Bishop et al, ²	2014	60	0	4a	0	0	Ethmoidal sinus	Radical resection followed by postoperative radiation therapy	0	1	0	1	0	0	0	0	10.0	0	Squamous
8	Bishop et al, ²	2014	67	1	4a	0	0	Ethmoidal sinus	Radical resection followed by chemoradiation	0	1	0	1	1	1	0	0	16.0	0	Undifferentiated
9	Bishop et al, ²	2014	46	1	4a	0	0	Frontal and ethmoid sinuses	Resection followed by radiation therapy	0	1	0	1	0	0	0	0	57.0	0	Squamous

10	Jamshidi et al, ¹¹	2014	32	0	4b	0	0	Sphenoid sinus with compression of bilateral optic nerves	Debulking surgery followed by chemotherapy TPF x 2 cycles and concurrent weekly cisplatin	0	1	0	1	1	1	0	1	1	6.0	2	Squamous
11	Allison et al, ¹³	2016	77	0	4b	1	0	Posterior ethmoidal sinus invading into skull base and orbit	Radical chemoradiation	0	0	0	1	0	1	1	1	1	24.0	2	Plasmacytoid/rhabdoid
12	Zeng et al, ¹⁴	2016	86	0	3	0	0	Left nasal cavity, left maxillary sinus with invasion of maxillary bone	Radical resection	0	0	0	0	0	0	0	1	21.0	0	Basaloid	
13	Shatzkes et al, ¹⁵	2017	77	0	4b	0	0	Nasal cavity, ethmoidal, maxillary and sphenoidal sinuses	Radical resection followed by postoperative chemoradiation	0	1	0	1	1	0	0	1	17.0	2	Plasmacytoid/Rhabdoid	
14	Shatzkes et al, ¹⁵	2017	51	1	4a	0	0	Sphenoidal and ethmoidal sinuses	Radical resection followed by postoperative chemoradiation	0	1	0	1	1	0	1	1	24.0	2	Undifferentiated carcinoma	
15	Shatzkes et al, ¹⁵	2017	33	1	4b	0	0	Nasal cavity and	Radical resection followed by	0	1	0	1	1	0	0	1	33.0	2	Germ cell tumour	

		chemoradiatio n																		
23	Shatzkes et al. ¹⁵	2017	72	0	4a	0	0	Nasal cavity	Radical resection followed by postoperative chemoradiatio n	0	1	0	1	0	0	0	12.0	0	Undifferentiated carcinoma	
24	Shatzkes et al. ¹⁵	2017	45	0	4b	0	0	Nasal cavity and ethmoidal sinus	Radical resection followed by postoperative chemoradiatio n	0	1	0	1	0	0	0	0	48.0	0	Adenocarcinoma
25	Shatzkes et al. ¹⁵	2017	75	0	4b	0	0	Nasal cavity and ethmoidal sinus	Preoperative chemoradiatio n followed by surgery and postoperative radiation therapy	1	1	1	1	1	1	0	0	12.0	UD	Basaloid
26	Shatzkes et al. ¹⁵	2017	78	1	1	0	0	Nasal cavity	Radical resection	0	0	0	0	0	0	0	0	26.0	0	Plasmacytoid/rhabdoid
27	Shatzkes et al. ¹⁵	2017	44	0	4b	0	0	Nasal cavity and ethmoidal sinus	Radical resection followed by postoperative radiation therapy	0	1	0	1	0	0	0	0	18.0	0	Basaloid
28	Shatzkes et al. ¹⁵	2017	32	0	4b	0	0	Nasal cavity and ethmoidal sinus	Radical resection followed by postoperative radiation therapy	0	1	0	1	1	0	0	0	24.0	1	Undifferentiated carcinoma
29	Shatzkes et al. ¹⁵	2017	64	1	4b	0	0	Frontal and ethmoidal sinuses with invasion into the orbit	Radical resection followed by postoperative chemoradiatio n	0	1	0	1	1	0	0	0	13.0	1	Undifferentiated carcinoma

30	Agaimy et 201 al, ¹⁶	7	60	0	4a	0	0	Ethmoidal sinus	Radical resection followed by postoperative radiation therapy	0	1	0	1	0	0	0	0	1	10.0	UD	Basaloid
31	Agaimy et 201 al, ¹⁶	7	24	1	4a	0	0	Nasal cavity, ethmoidal and sphenoid sinuses	Chemotherapy	0	0	0	0	0	0	0	0	1	1.0	2	Plasmacytoid/rhabdoid
32	Agaimy et 201 al, ¹⁶	7	61	0	3	0	0	Nasal cavity and ethmoidal sinus	Preoperative chemoradiation followed by radical resection	1	0	0	1	1	1	1	1	1	43.0	2	Basaloid
33	Agaimy et 201 al, ¹⁶	7	35	1	4a	0	0	Right anterior ethmoidal and bilateral frontal sinuses	Radical resection followed by postoperative chemoradiation	0	1	0	1	1	0	1	1	1	93.0	2	Basaloid SCC
34	Agaimy et 201 al, ¹⁶	7	52	0	4a	2	0	Ethmoidal and left frontal sinuses	Radical resection followed by postoperative chemoradiation	0	1	0	1	1	0	1	0	1	102.0	2	Plasmacytoid/rhabdoid
35	Agaimy et 201 al, ¹⁶	7	21	0	4a	UK	UK	Nasal cavity	Radical resection	0	0	0	0	0	0	0	0	1	6.0	2	Basaloid
36	Agaimy et 201 al, ¹⁶	7	67	1	3	0	0	Right ethmoidal, sphenoidal, and frontal sinuses	Radical resection followed by postoperative chemoradiation	0	1	0	1	1	0	0	1	1	100.0	2	Basaloid
37	Agaimy et 201 al, ¹⁶	7	24	0	4a	UK	UK	Frontal sinus	Radical resection followed by postoperative	0	1	0	1	1	1	0	1	1	14.0	2	Basaloid

		chemoradiatio n																																					
		Radical resection																																					
38	Agaimy et al, ¹⁶ 2017	77		0		4a		0		0		Ethmoidal sinus		followed by postoperative chemoradiatio n		0		1		0		1		1		0		0		1		17.0		2		Plasmacytoid/rhabdo id			
		Nasal cavity,		Radical resection																																			
39	Agaimy et al, ¹⁶ 2017	54		1		4a		0		0		ethmoidal and maxillary sinuses		followed by postoperative chemoradiatio n		0		1		0		1		1		1		0		0		1		15.0		2		Basaloid	
		Left maxillary, sphenoidal and frontal sinuses		Radical resection																																			
40	Agaimy et al, ¹⁶ 2017	67		0		4a		1		0		Maxillary sinus		followed by postoperative radiation therapy and palliative chemotherapy		0		1		0		1		1		1		0		1		1		22.0		2		Basaloid	
		Nasal cavity,		Radical resection																																			
41	Agaimy et al, ¹⁶ 2017	71		1		3		0		0		Maxillary sinus		followed by postoperative radiation therapy		0		1		0		1		1		1		0		0		1		15.0		2		Basaloid, spindled, adenoid	
		Nasal cavity,		Radical resection																																			
42	Agaimy et al, ¹⁶ 2017	33		0		4a		0		0		Nasal cavity, ethmoidal and frontal sinuses		followed by postoperative radiation therapy		0		1		0		1		1		1		0		1		1		30.0		2		Basaloid	
		Nasal cavity, maxillary and ethmoid sinuses with		Preoperative radiation therapy																																			
43	Agaimy et al, ¹⁶ 2017	86		0		4b		0		0		maxillary and ethmoid sinuses with		followed by radical resection		1		0		0		1		1		1		1		1		1		9.0		2		Plasmacytoid/rhabdo id	

intracranial extension																			
44	Agaimy et al, ¹⁶ 2017	89	0	3	0	0	Nasal cavity, sphenoidal and ethmoid sinuses	Radical resection followed by postoperative radiation therapy	0	1	0	1	1	0	0	6.0	2	Plasmacytoid/rhabdoid	
45	Agaimy et al, ¹⁶ 2017	40	1	4a	0	0	Left nasal cavity and middle turbinate, ethmoidal and frontal sinuses	Radical chemoradiation	0	0	0	1	1	0	0	24.0	1	Plasmacytoid/rhabdoid	
46	Agaimy et al, ¹⁶ 2017	38	1	4a	0	0	All paranasal sinuses with skull base invasion	Biopsy followed by chemotherapy	0	0	0	0	0	0	0	4.0	1	Plasmacytoid/rhabdoid	
47	Agaimy et al, ¹⁶ 2017	70	0	4a	0	0	Ethmoidal and sphenoidal sinuses	Chemotherapy	0	0	0	0	1	1	0	0	9.0	1	Basaloid
48	Agaimy et al, ¹⁶ 2017	64	0	1	UK	UK	Nasal cavity	Radical chemoradiation	0	0	0	1	0	0	0	18.0	0	Basaloid	
49	Agaimy et al, ¹⁶ 2017	46	0	1	0	0	Frontal sinus	Preoperative chemoradiation, followed by radical resection and postoperative chemotherapy	1	1	1	1	0	0	0	0	43.0	0	Basaloid
50	Agaimy et al, ¹⁶ 2017	28	1	4a	0	0	Anterior ethmoidal and right frontal sinuses	Radical resection followed by postoperative chemoradiation	0	1	0	1	1	0	0	0	115.0	0	Basaloid

51	Agaimy et al, ¹⁶ 2017	52	0	4a	UK	UK	Nasal cavity and ethmoid sinus	Radical resection	0	0	0	0	1	1	0	0	12.0	2	Basaloid
52	Agaimy et al, ¹⁶ 2017	78	1	4a	0	0	Nasal cavity and ethmoidal sinus	Radical resection	0	0	0	0	0	0	0	0	26.0	0	Plasmacytoid/rhabdoid
53	Agaimy et al, ¹⁶ 2017	76	1	4a	0	1	All paranasal sinuses	Radical resection followed by postoperative chemoradiation	0	1	0	1	1	1	0	0	4.0	2	Plasmacytoid/rhabdoid
54	Agaimy et al, ¹⁶ 2017	26	0	2	UK	UK	Ethmoidal sinus	Radical resection followed by postoperative chemoradiation	0	1	0	1	0	0	0	0	16.0	0	Basaloid with focal clear cell features
55	Agaimy et al, ¹⁶ 2017	87	1	4a	UK	UK	Nasal cavity and all paranasal sinuses	Radical resection followed by postoperative chemoradiation	0	1	0	1	0	0	0	0	11.0	0	Basaloid
56	Agaimy et al, ¹⁶ 2017	44	0	4a	0	0	Ethmoidal sinus	Radical resection followed by postoperative chemoradiation	0	1	0	1	0	0	0	0	23.0	0	Basaloid
57	Agaimy et al, ¹⁶ 2017	45	0	4a	0	0	Nasal cavity and left frontal sinus	Radical resection followed by postoperative chemoradiation	0	1	0	1	0	0	0	0	42.0	0	Plasmacytoid/rhabdoid with adenoid features
58	Agaimy et al, ¹⁶ 2017	46	1	4a	0	0	Frontal and ethmoidal sinuses	Radical resection followed by	0	1	0	1	0	0	0	0	57.0	0	Basaloid

postoperative radiation therapy																				
59	Wasserman et al., ¹⁷ 2017	34	0	4a	0	0	Maxillary sinus invading into the orbital floor	Preoperative chemoradiation followed by radical resection	1	0	0	1	1	0	0	1	1	26.0	2	
60	Wasserman et al., ¹⁷ 2017	56	1	4b	0	0	Sinonasal mass with intracranial extension and cavernous sinus invasion	Radical concurrent chemoradiation	0	0	0	1	0	0	0	0	12.0	0	Rhabdoid	
61	Dogan et al., ¹⁹ 2017	54	0	4a	0	0	Nasal cavity and ethmoidal sinus	Radical resection	0	0	0	0	1	0	0	1	1	39.0	2	-
62	Dogan et al., ¹⁹ 2017	47	0	4b	0	0	Ethmoid sinus	Preoperative chemoradiation followed by surgery	1	0	0	1	1	1	0	0	0	7.0	1	-
63	Dogan et al., ¹⁹ 2017	54	0	4b	0	0	Nasal cavity and ethmoidal sinus	Induction chemotherapy followed by surgery	1	0	0	1	1	1	0	1	0	23.0	1	-
64	Laco et al., ²¹ 2018	76	0	2	0	0	Nasal cavity	Radical resection followed by post-operative radiation therapy	0	1	0	1	0	0	0	1	1	71.0	3	Basaloid and rhabdoid
65	Laco et al., ²¹ 2018	27	0	4a	0	0	Nasal cavity	Radical resection followed by post-operative chemoradiation	0	1	0	1	1	1	0	0	1	73.0	2	Basaloid and rhabdoid

66	Laco et al, ²¹	2018	51	0	4b	0	0	Nasal cavity followed by post-operative radiation therapy	0	1	0	1	1	1	0	0	1	14.0	2	Basaloid and rhabdoid
67	Laco et al, ²¹	2018	54	1	4a	0	0	Nasal cavity followed by post-operative radiation therapy	0	1	0	1	1	1	0	0	0	122.0	0	Basaloid and rhabdoid
68	Allard et al, ²²	2018	87	0	4b	UK	UK	Nasal cavity, ethmoidal and maxillary sinuses with intracranial extension	UK	UK	UK	UK	UK	1	1	1	1	9.0	2	Basaloid
69	Allard et al, ²²	2018	51	1	4b	0	1	Sphenoid sinus with invasion into the temporal fossa	UK	UK	UK	UK	UK	0	0	0	1	24.0	2	Basaloid
70	Allard et al, ²²	2018	62	0	4b	UK	UK	Nasal cavity and ethmoidal sinus	UK	UK	UK	UK	UK	1	1	1	1	43.0	2	Rhabdoid
71	Allard et al, ²²	2018	32	0	4b	UK	UK	Nasal cavity, ethmoidal and frontal sinuses with orbital extension	UK	UK	UK	UK	UK	0	0	0	0	9.0	0	Basaloid
72	Allard et al, ²²	2018	75	0	3	1	0	Nasal cavity and ethmoidal sinus	UK	UK	UK	UK	UK	1	0	1	0	30.0	1	Basaloid

73	Allard et al. ²²	2018	64	1	3	0	1	Frontal sinus	UK	UK	UK	UK	UK	1	0	0	1	0	36.0	1	Rhabdoid
74	Kakkar et al. ²³	2019	58	0	1	0	0	Left nasal cavity	Left lateral rhinotomy excision followed by chemoradiation	0	1	0	1	0	0	0	0	0	8.0	0	Basaloid
75	Kakkar et al. ²³	2019	23	0	3	0	0	Left nasal cavity, maxillary and ethmoidal sinuses with destruction of bony walls and floor of maxillary sinus	Preoperative chemotherapy followed by radical maxillectomy and postoperative radiation therapy	1	0	0	1	0	0	0	0	0	17.0	0	Basaloid
76	Kakkar et al. ²³	2019	20	0	4b	0	0	Nasal cavity, right maxillary sinus extending to the infratemporal fossa, skull base, and orbit	No treatment	0	0	0	0	1	1	0	0	0	2.0	2	Basaloid
77	McHugh, et al. ²⁴	2019	53	0	4b	0	0	Ethmoidal sinus with invasion into the left medial orbit and frontal cranial fossa	Radical resection with orbital exenteration and infratemporal fossa dissection	0	0	0	0	1	0	0	1	0	9.0	1	Basaloid

78	Gomez-Acevedo et al. ²⁵	2019	42	1	3	0	0	Paranasal sinuses with invasion into the trigeminal nerve	Radical resection followed by gammaknife stereotactic radiosurgery	0	1	0	1	1	1	0	0	1	13.0	2	Undifferentiated carcinoma
79	Trieu et al. ²⁶	2019	53	0	4b	0	0	Rt posterior ethmoidal sinus with orbital apex n	Induction chemotherapy followed by radical concurrent chemoradiation with proton therapy	1	0	0	1	1	0	0	1	1	13.0	2	Basaloid
80	Vaziri Fard et al. ²⁷	2019	75	0	4a	0	0	Large sinonasal mass with bilateral orbital and skull base involvement	Radical chemoradiation	0	0	0	1	0	0	0	0	7.0	1	Plasmacytoid/rhabdoid	
81	Vaziri Fard et al. ²⁷	2019	61	0	4b	2	1	Large sinonasal mass with orbital and brain extension	Radical chemoradiation	0	0	0	1	0	0	0	0	17.0	1	Plasmacytoid/rhabdoid	
82	Vaziri Fard et al. ²⁷	2019	61	0	4b	0	1	Large sinonasal mass with brain invasion	Surgery followed by chemoradiation	0	1	0	1	0	0	0	0	11.0	1	Plasmacytoid/rhabdoid	
83	Levitian et al. ³¹	2020	36	0	4b	0	0	Ethmoidal and frontal sinuses with invasion into the right orbit and dura	No treatment	0	0	0	0	0	0	0	1	0.3	2	-	
84	Shanti et al. ³²	2020	63	1	4b	0	0	Left maxillary	Induction chemotherapy	1	0	0	1	1	1	0	0	8.0	0	Rhabdoid	

85	Chitguppi et al. ³³ 2020	UD	0	4b	UD	UD	Paranasal sinuses with invasion into the orbital cavity	Radical resection followed by postoperative chemoradiation	0	1	0	1	UD	UD	UD	UD	1	1.4	2
86	Chitguppi et al. ³³ 2020	UD	0	4b	UD	UD	Paranasal sinuses with invasion into the orbital cavity	Radical resection followed by postoperative chemoradiation	0	1	0	1	UD	UD	UD	UD	1	2.1	2
87	Chitguppi et al. ³³ 2020	UD	1	4b	UD	UD	Paranasal sinuses with invasion into the orbital cavity	Induction chemotherapy followed by radical concurrent chemoradiation	1	0	0	1	UD	UD	UD	UD	1	56.9	0
88	Chitguppi et al. ³³ 2020	UD	1	4b	UD	UD	Paranasal sinuses with invasion into the orbital cavity	Induction chemotherapy followed by radical concurrent chemoradiation	1	0	0	1	UD	UD	UD	UD	1	81.4	0
89	Chitguppi et al. ³³ 2020	UD	0	4b	UD	UD	Paranasal sinuses with invasion into the orbital cavity	Induction chemotherapy followed by radical concurrent chemoradiation	1	0	0	1	UD	UD	UD	UD	0	9.7	2

90	Chitguppi et al., ³³ 2020	UD	0	4b	UD	UD	Paranasal sinuses with invasion into the orbital cavity	Induction chemotherapy followed by radical concurrent chemoradiation	1	0	0	1	UD	UD	UD	UD	0	25.1	0	-
91	Libera et al., ³⁶ 2021	70	0	4b	0	0	Maxillary sinus	UK	UK	UK	UK	UK	UK	UK	UK	UK	1	4.0	UD	-
92	Libera et al., ³⁶ 2021	66	0	4b	0	0	Ethmoidal sinus	UK	UK	UK	UK	UK	UK	UK	UK	UK	1	11.0	UD	-
93	Libera et al., ³⁶ 2021	40	0	4b	0	0	Maxillary sinus	UK	UK	UK	UK	UK	UK	UK	UK	UK	1	14.0	UD	-
94	Libera et al., ³⁶ 2021	72	0	4b	0	0	Frontal sinus	UK	UK	UK	UK	UK	UK	UK	UK	UK	1	19.0	UD	-
95	Libera et al., ³⁶ 2021	48	0	4b	0	0	Maxillary sinus	UK	UK	UK	UK	UK	UK	UK	UK	UK	1	26.0	UD	-
96	Libera et al., ³⁶ 2021	42	1	4b	0	0	Frontal sinus	UK	UK	UK	UK	UK	UK	UK	UK	UK	1	41.0	UD	-
97	Libera et al., ³⁶ 2021	43	0	2	0	0	Ethmoidal sinus	UK	UK	UK	UK	UK	UK	UK	UK	UK	1	42.0	UD	-
98	Libera et al., ³⁶ 2021	76	0	4b	0	0	Ethmoidal sinus	UK	UK	UK	UK	UK	UK	UK	UK	UK	0	91.0	UD	-
99	Libera et al., ³⁶ 2021	46	1	4a	0	0	Ethmoidal sinus	UK	UK	UK	UK	UK	UK	UK	UK	UK	0	149.0	UD	-
100	Li et al., ³⁷ 2021	55	0	4a	0	0	Right nasal cavity invading into maxillary and sphenoidal sinuses and right orbital wall	Radical resection followed by postoperative radiation therapy	0	1	0	1	0	0	0	0	0	6.0	0	Yolk sac differentiation
101	Ayyanar et al., ³⁹ 2021	61	0	2	0	0	Frontal and ethmoidal sinuses	Radical resection followed by postoperative	0	1	0	1	1	1	0	0	1	24.0	2	Small cells

102	Ayyanar, et al. ³⁹	202 1	35	1	4b	0	0	Nasal cavity, right ethmoidal sinus with intracranial extension	Anterior crano-facial resection of right sided nasal mass and right orbital exenteration followed by chemoradiatio	n	0	1	0	1	1	1	0	0	0	5.0	1	Pseudoalveolar pattern, pseudoglandular pattern, empty vacuoles			
103	Ayyanar, et al. ³⁹	202 1	35	1	3	0	0	Left nasal cavity with extension into sphenoidal and posterior ethmoid sinuses	Radical resection followed by postoperative chemoradiatio	n	0	1	0	1	0	0	0	0	0	20.0	0	Basaloid			
104	Hazir et al. ⁴⁰	202 1	55	0	4a	0	0	Left maxillary sinus invading into the left orbit	Preoperative chemotherapy followed by surgery with orbital exenteration and postoperative chemoradiatio	n	1	1	1	1	1	1	0	1	1	12.0	2	Basaloid and squamoid			
105	Su et al. ⁴¹	202 1	54	1	4b	0	0	Left nasal cavity with invasion into the ethmoidal, maxillary and sinuses and orbital cavity and	Left lateral rhinotomy, left maxillectomy, and pansinusectomy	n	0	0	0	0	0	0	0	0	1	11.0	0	Oncocytoid and rhabdoid			

		right medial orbital wall																			
ID	Author et al, ⁴²	Year	Age	Sex	Tumor stage	Number of involved sinuses	Location	Treatment		Follow-up (months)		Local control (%)		Regional control (%)		Distant metastasis (%)		Overall survival (%)			
								Left ethmoidal and sphenoid sinuses	Endoscopic resection followed by postoperative radiotherapy	0	1	0	1	1	0	0	1	1	32.0	2	-
106	Wang et al, ⁴²	2022	47	1	4a	0	0	Right ethmoidal sinus	Endoscopic surgery followed by postoperative radiotherapy	0	1	0	1	1	0	0	1	1	32.0	2	-
107	Wang et al, ⁴²	2022	54	1	4a	0	0	Bilateral ethmoidal sinuses	Induction chemotherapy followed by radical chemoradiation	1	0	0	1	1	0	0	1	1	7.0	2	-
108	Wang et al, ⁴²	2022	59	1	4a	0	0	Bilateral ethmoidal sinuses	Preoperative chemotherapy followed by resection and postoperative chemoradiation	1	1	1	1	1	1	1	1	1	9.0	2	-
109	Wang et al, ⁴²	2022	38	0	4b	2	0	Bilateral ethmoidal sinuses	Radical resection followed by postoperative chemoradiation	0	0	11	1	1	1	1	1	31.0	2	-	
110	Wang et al, ⁴²	2022	64	0	4a	0	0	Left maxillary sinus	Radical endoscopic resection followed by postoperative chemoradiation and chemotherapy	0	1	0	1	1	1	0	11	1	14.0	2	-
111	Wang et al, ⁴²	2022	59	0	4a	0	0	Right ethmoidal sinus	Radical endoscopic resection	0	0	0	0	0	0	0	0	18.0	0	-	
112	Wang et al, ⁴²	2022	38	0	3	0	0	Left ethmoidal and maxillary sinuses	Radical endoscopic resection	0	0	0	0	0	0	0	0	28.0	0	-	

113	Wang et al, ⁴²	2022	78	0	4a	0	0	Right maxillary, ethmoidal and sphenoid sinuses	Radical endoscopic surgery and open surgery	0	0	0	1	1	1	0	0	0	56.0	1	-
114	Wang et al, ⁴²	2022	43	1	4a	0	0	Right frontal and ethmoid sinuses	Radical endoscopic resection followed by postoperative chemoradiation	0	1	0	1	0	0	0	0	0	3.0	0	-
115	Wang et al, ⁴²	2022	65	1	2	0	0	Right ethmoidal sinus	Radical endoscopic resection followed by postoperative chemotherapy	0	1	0	1	0	0	0	0	0	18.0	0	-
116	Wang et al, ⁴²	2022	53	1	3	UD	UD	Bilateral ethmoidal sinuses	Radical endoscopic resection followed by postoperative radiotherapy, open surgery and chemotherapy	0	1	0	1	1	1	0	0	0	27.0	1	-
117	Wang et al, ⁴²	2022	25	0	4a	0	0	Right frontal and ethmoidal sinus	Preoperative chemotherapy followed by open surgery and postoperative chemoradiation	1	1	1	1	1	1	0	1	0	35.0	1	-
118	Wang et al, ⁴²	2022	53	0	4a	0	0	Bilateral ethmoidal and frontal sinuses	Preoperative chemotherapy followed by resection	1	0	0	1	0	0	0	0	0	30.0	0	-

119	Wang et al, ⁴²	2022	42	0	4a	2	1	Bilateral ethmoidal and sphenoid sinuses, right frontal and maxillary sinuses	Palliative chemotherapy	0	0	0	0	1	0	1	0	0	21.0	1	-
120	Wang et al, ⁴²	2022	41	1	4a	0	0	Left ethmoidal sinus	Radiation therapy followed by surgery	1	0	0	1	0	0	0	0	0	16.0	0	-
121	HKU cohort (current study)	2022	45	0	4b	0	0	Nasal cavity with invasion into the maxillary sinus, ethmoid sinus, orbital floor and postoperative intracranial cavity	Preoperative chemotherapy TPF × 3 cycles followed by craniofacial resection and orbital exenteration and postoperative intracranial chemoradiation	1	1	1	1	0	0	0	0	1	8.9	0	Undifferentiated carcinoma
122	HKU cohort (current study)	2022	51	0	4b	0	0	Nasal cavity with invasion into the maxillary sinus, ethmoid sinus and orbital floor	Preoperative chemotherapy PF × 3 cycles followed by craniofacial resection and postoperative chemoradiation with carboplatin	1	1	1	1	1	0	0	1	1	10.7	2	Undifferentiated carcinoma
123	HKU cohort (current study)	2022	58	1	4a	0	0	Nasal cavity with invasion into the maxillary sinus and	Craniofacial resection followed by chemoradiation with proton therapy	0	1	0	1	1	1	0	0	0	56.4	0	High-grade non-intestinal adenocarcinoma

											orbital floor and intracranial cavity											
128	HKU	cohort	202	53	0	4b	0	0	0	n	Preoperative Nasal cavity chemotherapy with TPF × 3 cycles invasion followed by into the radical maxillary craniofacial sinus and resection and ethmoidal postoperative sinus chemoradiatio	1	1	1	1	0	0	0	0	24.7	0	Undifferentiated carcinoma
	(current study)																					

*References are in the same order as those cited in the manuscript.

PF cisplatin and 5-fluorouracil, TPF docetaxel, cisplatin, and 5-fluorouracil, TPX docetaxel, cisplatin, and capecitabine, UD undetermined, UK UnknownReferences

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