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Systematic Review

Pediatric Tracheobronchial Granular Cell Tumors: Case Series and Systematic Review

Theresa Tharakan [1,*], Carla V. Valenzuela [1], David S. Leonard [1], Selby Chu [2] and Katherine Dunsky [1]

[1] Department of Otolaryngology-Head and Neck Surgery, Washington University School of Medicine, Saint Louis, MO 63110, USA
[2] Department of Pediatrics, University of Texas Southwestern Medical Center, Dallas, TX 75390, USA
* Correspondence: tharakant@wustl.edu

Abstract

Background/Objectives: To report two cases of pediatric tracheobronchial granular cell tumor (GCT) and present a systematic review on the management and outcomes of pediatric tracheobronchial GCTs treated with endoscopic or open resection. **Case Presentations**: Case 1: A 10-year-old female presenting with severe obstructive sleep apnea was found to have a near-completely obstructing tracheal mass. Excisional biopsy revealed a GCT. She underwent definitive tracheal resection and re-anastomosis. There was no evidence of recurrence at 5 months postoperatively. Case 2: A 9-year-old female presenting with respiratory failure was found to have an endobronchial GCT. She underwent thoracotomy with right upper lobe resection and bronchoplasty, with no evidence of recurrence at 21 months postoperatively. **Systematic Review Methods and Results**: A medical librarian searched the published literature for records discussing "child OR pediatric" AND "granular cell tumor OR granular cell myoblastoma" AND "bronchial OR tracheal OR airway OR subglottis." The primary outcome was recurrence following endoscopic or open resection. Of 152 records screened and an additional 3 records found from citation searching, 27 total articles met inclusion criteria. The majority of GCTs were managed with open resection (n = 19, 68%), while 9 were managed endoscopically. Tumor recurrence was reported in 4 patients who underwent endoscopic resection. **Conclusions**: GCT is rarely reported arising from the tracheobronchial tree. Both endoscopic and open approaches have been utilized for management, but recurrences have only been reported with endoscopic resection. We propose a clinical pathway for management and surveillance of pediatric tracheobronchial GCTs.

Keywords: granular cell tumor; pediatric tracheal resection; tracheobronchial tumor; case report

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1. Introduction

Tracheobronchial tumors typically present with non-specific airway symptoms such as wheezing, cough, chest pain, and respiratory failure and may initially be misdiagnosed as asthma, pneumonia, or sleep apnea. The incidence of pediatric tracheobronchial tumors is approximately 0.0049 per 100,000 children, or about 0.2% of all tumors in children [1,2]. In adults, tracheobronchial tumors are most commonly malignant (approximately 90%), with squamous cell carcinoma (SCC) and adenoid cystic carcinoma (ACC) accounting for about two-thirds of tumors [3]. In pediatric tracheobronchial tumors, the reported rate of malignancy varies from 10% to 62% [1,3]. Papilloma and infantile hemangioma are the two most common pediatric tracheobronchial tumors, but the differential can include

rarer, benign or malignant tumors, such as carcinoid tumor, mucoepidermoid carcinoma, granular cell tumor, inflammatory myofibroblastic tumor, and rhabdomyosarcoma [4].

Tracheobronchial granular cell tumors (GCT) are especially rare in pediatric patients, with fewer than 30 cases reported in the literature. Diagnosis is confirmed by pathology. There have been no cases of malignant transformation of GCT in the pediatric literature, and surgical resection (endoscopic or open) is the treatment modality of choice [4].

In the adult literature, it has been recommended that large tracheobronchial GCTs undergo open resection, while small tumors can be resected endoscopically [5]. Ipakchi et al. estimate a local recurrence rate of approximately 50% based on eight cases of endoscopically resected tracheobronchial GCTs with follow-up ranging from six weeks to nine years [6]. A systematic review by Daniel et al. reports that GCTs larger than 8 mm should raise suspicion for transmural extension and be considered for tracheal resection [7]. In the pediatric population, Finck et al. propose an algorithm for management that includes open resection for large tumors, endoscopic resection for small tumors, and a minimum of five years of follow-up regardless of tumor size. They recommend treating tumor recurrences with resection and tracheobronchial reconstruction [8]. However, this remains expert opinion, and evidence-based recommendations are needed. In this systematic review, we provide a comprehensive list of pediatric tracheal and bronchial GCT cases with available management and recurrence data, including two patients at our institution. The aim of this review is to identify and synthesize published cases of tracheobronchial GCTs in children, with particular emphasis on their clinical presentation, diagnostic characteristics, management, surveillance, and outcomes.

2. Case Presentations

We reviewed the medical records of two patients diagnosed with tracheobronchial GCT at our institution. Records between 2018 and 2021 were available for review. Case histories, management, and outcomes were reported. The CARE checklist is in the Supplementary Materials (Supplementary File S1).

2.1. Case 1

2.1.1. Patient Information

A 10-year-old female with a diagnosis of mild persistent asthma since the age of nine years and no pertinent family history was seen in the otolaryngology clinic for noisy breathing and snoring for one year. Polysomnography revealed severe obstructive sleep apnea (OSA) with an apnea-hypopnea index (AHI) of 111.6 events/hour, obstructive AHI of 106.8 events/hour, and lowest oxygen desaturation to SpO_2 52%. In the clinic, she was noted to have biphasic stridor at rest with unlabored breathing and small, non-obstructing tonsils. Flexible nasal laryngoscopy revealed a tracheal mass.

2.1.2. Diagnostic Assessment

A computed tomography (CT) of the neck and chest demonstrated a 2.0 × 1.3 × 1.8 cm intraluminal tracheal mass with near-complete airway obstruction. She was admitted to the pediatric intensive care unit and taken to the operating room (OR) for direct laryngoscopy with rigid bronchoscopy (DLB) and biopsy. Upon induction of anesthesia, the patient developed airway obstruction, desaturation, and bradycardia requiring <30 s of cardiopulmonary resuscitation. The airway was stabilized first with bag-mask ventilation, then with ventilation via an airway exchange catheter passed beyond the mass. The tumor was biopsied and then debulked using a CO_2 laser and microdebrider to re-establish patency of the trachea. She was extubated and discharged on postoperative day two. Surgical pathol-

ogy demonstrated a GCT staining positive for S100, cluster of differentiation 68 (CD68), transcription factor E3 (TFE3), inhibin, and SRY-related HMG-box 10 (SOX10).

Three weeks later, chest magnetic resonance imaging (MRI) with contrast demonstrated a 1.1 × 1.2 × 1.5 cm T2-hyperintense polypoid mass arising from the right posterolateral trachea at the level of vertebral bodies C7 to T2 with significant airway compromise and apparent invasion of the tracheal wall.

2.1.3. Therapeutic Intervention

The patient was taken to the OR for tracheal resection with primary anastomosis. Intraoperative frozen section demonstrated negative margins. Final pathology confirmed a GCT measuring 1.8 × 1.2 × 0.7 cm with negative margins.

2.1.4. Follow-Up and Outcomes

Repeat DLB one week later showed granulation tissue with grade I tracheal stenosis. Repeat DLBs at one, two, and five months postoperatively demonstrated no evidence of recurrence. At the two-month and five-month postoperative DLBs, she was found to have grade I tracheal stenosis, which was treated with balloon dilations and steroid injections.

2.2. Case 2

2.2.1. Patient Information

As previously published by our institution, a 9-year-old female presented with respiratory failure and right lung opacification on chest radiograph [9].

2.2.2. Diagnostic Assessment

After a trial of antibiotics failed to relieve her symptoms, a chest CT was obtained and revealed a 3.7 cm endobronchial tumor obstructing the right mainstem bronchus with rightward tracheal deviation. She underwent DLB with biopsy and debulking. Pathology confirmed a diagnosis of GCT, which was negative for S100.

2.2.3. Therapeutic Intervention

She underwent a second debulking one month later with KTP laser. Repeat chest CT two months afterward demonstrated regrowth of the mass. She subsequently underwent right thoracotomy with right upper lobectomy and bronchoplasty for complete resection of the tumor.

2.2.4. Follow-Up and Outcomes

The patient is asymptomatic and disease-free with 21 months of follow-up and surveillance chest CT studies.

3. Systematic Review

3.1. Search Methodology

The methodological requirements of a systematic review were fulfilled using Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines where applicable [10]. No review protocol was published. Articles were selected using the PICOS (population, interventions, comparators, outcomes, and study design) format. We defined our population to include patients less than 18 years of age with GCT located in the subglottic larynx, trachea, or bronchi with the intervention of open resection in comparison to those who underwent endoscopic resection. The outcome of interest was tumor recurrence. Included study designs were limited to case reports. Tumor recurrence

(yes/no) was chosen as the outcome measure to determine whether open versus endoscopic approach leads to similar recurrence rates. We included pathology-proven GCTs in the subglottic larynx only if there was extension to the trachea. GCTs limited to the subglottic larynx only are described separately. Our search strategy excluded patients aged 18 years or older and any manuscripts not published in the English language.

The published literature was searched using strategies created by a medical librarian for tracheal or bronchial GCTs in children. The search strategies were implemented in Ovid MEDLINE 1946-, Embase 1947-, Scopus 1823-, Cochrane Central Register of Controlled Trials, and ClinicalTrials.gov, and were established using a combination of standardized terms and keywords including, but not limited to: (child OR pediatric) AND (granular cell tumor OR granular cell myoblastoma) AND (bronchial OR tracheal OR airway OR subglottis). We completed initial searches in March 2021. There were 278 results exported to a reference manager software (EndNote X8; Clarivate Analytics, Philadelphia, PA, USA). EndNote's automatic duplicate finder identified 155 duplicates, resulting in 125 unique citations included in the project library. The search was updated on 17 September 2026, which yielded 48 new results (326 total records reviewed). There were 19 duplicates removed (174 duplicates total) and 29 new citations were added to the pool for screening, for a total of 152 records screened. Three additional records were added from citation searching. Fully reproducible search strategies for each database are provided in the Supplementary Materials in the online version of this article. The PRISMA 2020 checklist is in the Supplementary Materials (Supplementary File S2).

3.2. Study Selection

Two authors (C.V.V. and T.T.) independently screened the records for eligibility (Figure 1). In cases of discordance, a third author (S.C.) independently screened the articles as a tiebreaker. Records were screened using Rayyan Web (Rayyan Systems, Inc.: Cambridge, MA, USA). The titles and abstracts were first reviewed to determine whether they were eligible for study inclusion. Records were excluded if: (1) the article was not an original case report/series, (2) tumor location was outside of the larynx, trachea, or bronchi, (3) it included patients aged 18 years or older, (4) the record was not published in English. After this initial screening, full-text articles were then reviewed. Articles were excluded if any of the previously defined record exclusion criteria were met and additionally: (1) the tumor was not anatomically located in either the trachea or bronchi, (2) the study did not include pediatric patients, (3) the study did not include individual patient clinical information, (4) the study did not include a follow-up assessment to determine if there was tumor recurrence, or (5) the article was not published in English. The two cases presented here, one of which was previously published by Chen et al. [9], were also included in the analysis.

3.3. Data Extraction and Summary Measures

Our primary outcome variable was any tumor recurrence at follow-up. Our secondary outcome variables were length of follow-up time and method used to identify recurrence. Descriptive statistics were used to describe the number of cases with tumor recurrence based on open versus endoscopic resection, length of follow-up time, and tumor size. Demographics were also reported. All analyses were performed in Microsoft Excel Version 16 and R (R version 2025.05.01+513, R Foundation for Statistical Computing, Vienna, Austria).

3.4. Quality Assessment

A risk of bias assessment could not be performed due to the inherent bias in case reports and the lack of a standardized assessment of bias for case reports.

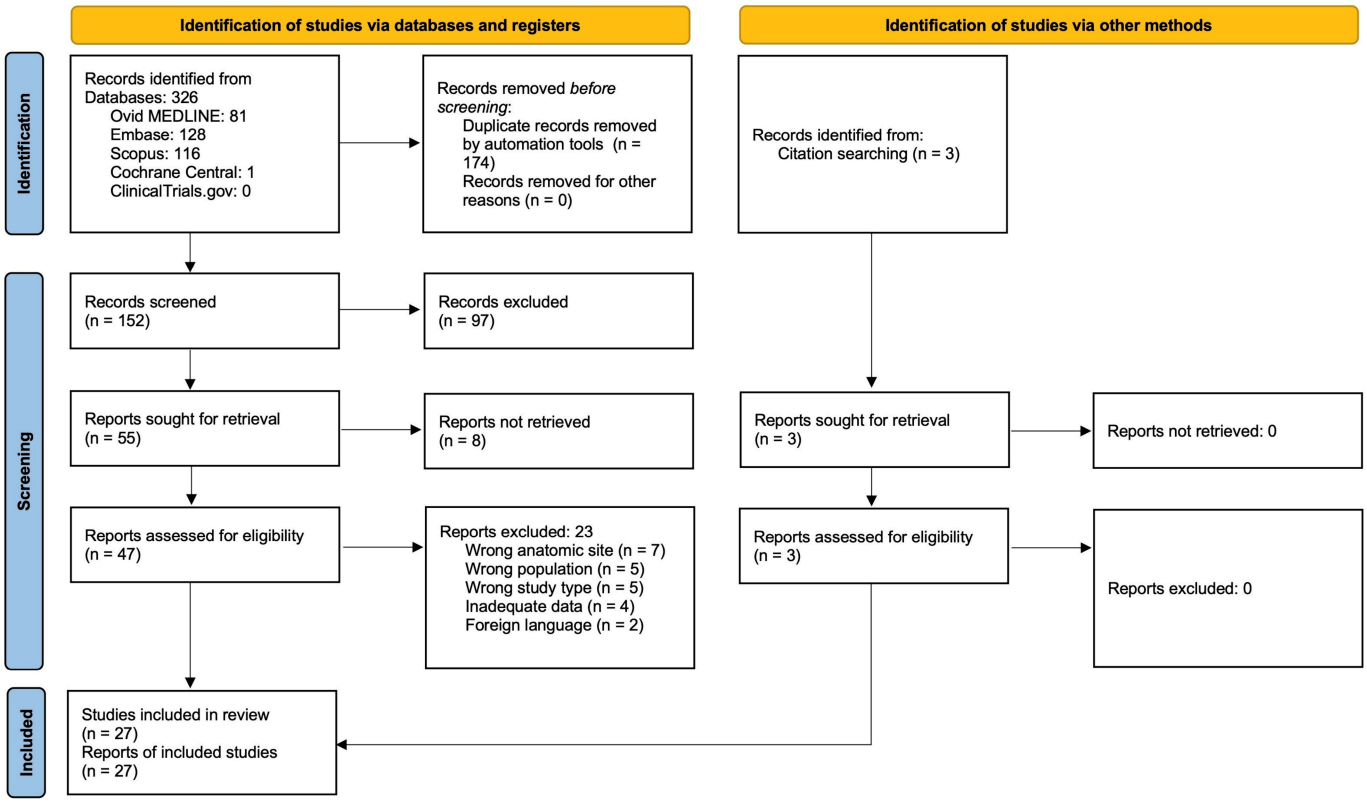


Figure 1. PRISMA Flow Diagram for Systematic Review Article Selection.

3.5. Ethical Considerations

This case series and systematic review did not meet the definition of human subjects research as described in 45 CFR 46.102(1) and were deemed not to be under the purview of an institutional review board (IRB). Therefore, informed consent was not requested.

3.6. Results

We reviewed 27 articles reporting 27 cases of GCT spanning from 1939 to 2026 that met our inclusion criteria (Figure 1). The present, previously unpublished Case 1 was added to the analysis for a total of 28 cases. Characteristics of each case are summarized in Tables 1 and 2. The median patient age was 11 years, and the majority of the cases were female (n = 21, 75%).

Presentation. Of the 28 cases, 25 reported one or more presenting symptoms. Clinical presentations included recurrent pneumonia (n = 11/25, 44%), asthma (n = 6/25, 24%), acute respiratory distress (n = 4/25, 16%), obstructive sleep apnea (n = 2/25, 8%), dysphonia (n = 4/25, 16%), difficult intubation (n = 1/25, 4%), and incidental radiologic finding (n = 1/25, 4%); sum of percentages is greater than 100% to account for multiple symptoms listed.

Tumor characteristics. Tumor locations were listed in all 28 cases. The median maximum diameter measured on CT or diagnostic endoscopy was 2 cm [0.5–7.7 cm]. Tumor locations included trachea alone (n = 12), bronchus alone (n = 10), trachea with subglottic larynx (n = 5), and trachea with bronchi (n = 1). One patient had a separate, concurrent glottic GCT in addition to a tracheal GCT. Diagnosis of GCT was made by pathology in all cases.

Table 1. Characteristics of endoscopic cases (n = 9).

Authors	Year	Age (y)	Sex	Race/Ethnicity	Country	Presentation	Site	Max Diameter (cm)	Initial Management	Definitive Management	Recurrence/Persistence	Follow-Up (mo)	Surveillance Method
Sambel et al. [11]	2025	16	F	NR	Turkey	Progressive asthma	Left mainstem bronchus	1.5	Rigid bronchoscopy with excision	Same	No	3	Endoscopy
Ahn et al. [12]	2020	6	M	NR	Korea	Barking cough	Posterior subglottis and trachea	2.2	Suspension laryngoscopy and cup forceps	None	Yes	7	NR
Jichlinski et al. [13]	2018	15	F	NR	USA	Recurrent pneumonia	Distal trachea	NR	Flexible bronchoscopy and biopsy	Endoscopic sleeve resection	NR	NR	NR
Sjogren & Sidman [14]	2013	11	M	NR	USA	NR	Anterior trachea	NR	NR	CO_2 laser debulking	Yes	8	No symptoms reported
Xu et al. [15]	2012	17	F	Indian	Singa-pore	Recurrent pneumonia	Multiple: Bronchus intermedius, left main bronchus, left lower lobe orifice	NR	Flexible bronchoscopy and biopsy	Nd YAG laser debulking	No	1.5	Endoscopy
Leboulanger et al. [16]	2008	12	F	NR	France	Dysphonia	Multiple: Right cervical trachea, left vocal fold	0.5	Rigid endoscopy and biopsy	CO_2 laser debulking	No	2	Endoscopy
Pernas [17]	2006	11	F	Black	USA	Dysphonia	Subglottis and cervical trachea	4	Endoscopic debulking and biopsy	Debulking multiple times, ultimately laryngofissure	Yes	4	Endoscopy

Table 1. *Cont.*

Authors	Year	Age (y)	Sex	Race/Ethnicity	Country	Presentation	Site	Max Diameter (cm)	Initial Management	Definitive Management	Recurrence/Persistence	Follow-Up (mo)	Surveillance Method
Torsiglieri [18]	1991	15	F	NR	USA	Dysphonia	Subglottis and cervical trachea	1.5	Direct laryngoscopy, frozen biopsy, CO_2 laser debulking	None	Yes	1.5	Endoscopy
Kramer [19]	1939	15	F	NR	USA	Pneumonia	Right lower lobe bronchus	NR	NR	Endoscopic cold debulking	No	5	Endoscopy

mo = months, NR = Not Reported, y = years.

Table 2. Characteristics of open surgery cases (n = 19).

Authors	Year	Age (y)	Sex	Race/Ethnicity	Country	Presentation	Site(s)	Max Diameter (cm)	Initial Management	Definitive Management	Recurrence/Persistence	Follow-Up (mo)	Surveillance Method
Tharakan et al. (Case 1)	*	10	F	Black	USA	Obstructive sleep apnea	Cervical trachea	2	Rigid endoscopy and debulking	Tracheal resection	No	5	Endoscopy
Chen et al. (Case 2) [9]	2020	9	F	Non-Hispanic	USA	Respiratory distress, pneumonia	Right upper lobe bronchus	3.7	Intubation, rigid endoscopy, KTP laser debulking	Lobectomy and bronchoplasty	No	21	CT
Chen et al. [20]	2019	15	F	Asian	China	Recurrent pneumonia	Trachea	NR	Flexible bronchoscopy and biopsy	Tracheal resection	No	12	CT
Esterson et al. [21]	2017	15	F	NR	USA	Shoulder pain, pneumonia	Right segmental bronchus	7.7	Flexible bronchoscopy, biopsy	Lobectomy	NR	NR	NR

Table 2. *Cont.*

Authors	Year	Age (y)	Sex	Race/Ethnicity	Country	Presentation	Site(s)	Max Diameter (cm)	Initial Management	Definitive Management	Recurrence/Persistence	Follow-Up (mo)	Surveillance Method
Gonik et al. [22]	2014	13	F	NR	USA	Respiratory distress, asthma	Subglottis and cervical trachea	3	Rigid endoscopy, debulking	Laryngofissure, partial cricotracheal resection, hemithyroidectomy	No	12	No symptoms reported
Torre et al. [23]	2010	4	F	White	Chile	Stridor, laryngitis	Subglottis and cervical trachea	NR	Rigid endoscopy	Cricotracheal resection	No	42	No symptoms reported
Kutuya et al. [24]	2010	16	F	Asian	Japan	Incidental	Right segmental bronchus	1.3	Rigid endoscopy, biopsy	Lobectomy	NR	NR	NR
Vaos et al. [25]	2006	10	F	NR	Greece	Recurrent pneumonia	Left upper lobe bronchi	2	Rigid and flexible bronchoscopy	Lobectomy	No	12	Endoscopy
Stieglitz et al. [26]	2005	14	F	NR	Germany	Pneumonia	Carina	1.5	Rigid endoscopy, biopsy	Carina resection with reconstruction, bronchoplasty	No	36	Endoscopy
Amar et al. [27]	2002	9	M	NR	Canada	Difficult intubation, dysphonia	Posterior trachea	4.5	Rigid bronchoscopy, emergent tracheostomy	Laryngotracheal resection and reconstruction	No	6	Ultrasound, PFTs
Prasad & Keller [28]	2002	10	F	NR	USA	Stridor, asthma	Right trachea	1.5	Rigid endoscopy, biopsy	Tracheostomy, tracheal fissure and excision	No	12	Endoscopy

Table 2. *Cont.*

Authors	Year	Age (y)	Sex	Race/Ethnicity	Country	Presentation	Site(s)	Max Diameter (cm)	Initial Management	Definitive Management	Recurrence/Persistence	Follow-Up (mo)	Surveillance Method
Abdulhamid & Rabah [29]	2000	8	F	Black	USA	Recurrent pneumonia	Right mainstem bronchus	2.5	Rigid endoscopy, biopsy	Lobectomy and bronchoplasty	No	72	NR
Desai et al. [30]	1999	10	F	NR	USA	Stridor, respiratory distress, asthma, obstructive sleep apnea	Posterior trachea	NR	N/A (single stage)	Rigid endoscopy, biopsy, tracheostomy, tracheal fissure and excision	No	72	NR
Spandow & Lindholm [31]	1994	12	M	White	Sweden	Dyspnea, asthma	Cervical trachea	2	Tracheostomy, tracheoscopy, biopsy	L hemithyroidectomy, tracheal resection and reanastomosis	No	18	CT and endoscopy
Burton (Personal Communication—Grillo) [5]	1992	14	F	Black	NR	NR	Multiple: Cervical trachea	NR	NR	Tracheal resection	No	4	NR
Solomons [32]	1988	10	M	NR	South Africa	Respiratory distress, stridor, asthma	Cervical trachea	NR	Rigid endoscopy, tracheostomy, biopsy	Laryngofissure, partial tracheal resection	No	2	Deceased/autopsy
Lack et al. [33]	1983	10	M	White	USA	NR	Left upper lobe	NR	Rigid endoscopy, biopsy	Lobectomy	No	36	NR

Table 2. *Cont.*

Authors	Year	Age (y)	Sex	Race/Ethnicity	Country	Presentation	Site(s)	Max Diameter (cm)	Initial Management	Definitive Management	Recurrence/Persistence	Follow-Up (mo)	Surveillance Method
Sawada et al. [34]	1981	5	F	NR	Japan	Tuberculosis pneumonia	Trachea, right mainstem bronchus	1.8	Rigid endoscopy, biopsy	Lobectomy, adjuvant radiation and interstitial brachytherapy	No	144	NR
Oparah & Subramanian [35]	1976	10	M	Black	USA	Recurrent pneumonia	Left segmental bronchus	NR	Rigid endoscopy	Lobectomy	No	36	No symptoms reported

CT = computed tomography scan, mo = months, NR = Not Reported, PFT = pulmonary function test, y = years * Present case report, previously unpublished.

Management. The modality of treatment was reported in all patients. Six patients required airway interventions before definitive management and included endoscopic debulking (n = 3/6, 50%), tracheostomy (n = 3/6, 50%), and/or intubation (n = 1/6, 17%). Table 1 describes initial and definitive management. There were 9/28 (32%) patients who underwent an endoscopic approach and 19/28 (68%) patients who underwent an open approach for tumor resection. In two cases of endoscopic debulking, treatment was described as intentionally leaving residual tumor and therefore not considered "definitive".

Surveillance. Follow-up information was reported in 25 (89%) of the 28 cases (Table 3). Median follow-up time among these 25 cases was 12 months [range 1.5–144 months]. Surveillance methods included the following: serial endoscopy (n = 10/25, 40%), CT scan (n = 2/25, 8%), both serial endoscopy and CT (n = 1/25, 4%), reported symptoms (n = 4/25, 16%), pulmonary function tests and ultrasound (n = 1/25, 4%), and pathology upon death (n = 1/25, 4%). A surveillance method was not reported in 9/25 (36%) cases. In the cohort with follow-up data, tumor recurrence or persistence was identified in 4/8 patients (50%) who underwent an endoscopic approach and 0/17 patients (0%) who underwent an open approach. Mortality was reported in one patient who underwent an open approach.

Table 3. Follow-up data for patients treated with endoscopic or open resection.

Approach	Median Age (Years)	Median [Range] Follow-Up Time (Months)	Median [Range] Tumor Maximum Diameter (cm)	Number of Cases with Recurrence/Persistence	Number of Cases with Follow-Up Data
Endoscopic (n = 9)	15	3.5 [1.5–8]	1.5	4	8
Open (n = 19)	10	18 [2–144]	2 [1.5–4.5]	0	17

4. Discussion

GCTs typically present in the third to sixth decade of life, with over half found in the head and neck, and the majority of these arising in the tongue [8]. GCTs in children are most frequently seen as congenital epulis arising from the maxillae. Pediatric tracheobronchial GCTs are very rare, with limited evidence-based recommendations for management. We provide a comprehensive summary of tracheobronchial GCT cases reported in the literature.

4.1. Presentation of Tracheobronchial Granular Cell Tumors

Among the cases reported, typical presentations involved acute respiratory distress, persistent asthma, and/or recurrent pneumonia. Many patients have an insidious onset of symptoms and are initially misdiagnosed. Our first case presented with a history of asthma and sleep apnea, both of which appeared to resolve with tumor management. Diagnosis often occurs only after significant airway symptoms manifest. Initial management requires airway stabilization. The goal of initial endoscopic biopsy and debulking is to establish a diagnosis and alleviate acute airway symptoms, followed by immediate or staged open resection.

4.2. Clinical Management of Tracheobronchial Granular Cell Tumors

We propose a clinical pathway for management and surveillance of pediatric tracheobronchial GCTs (Figure 2). This is a preliminary proposal based on limited case-based evidence and is not a validated clinical guideline. Although uncommon, tracheal tumors should be on the differential for pediatric patients presenting with recurrent pneumonia,

persistent asthma, respiratory distress, and severe OSA. Initial management of any patient requires airway evaluation and stabilization. Awake intubation past a soft, compressible mass may be possible in a cooperative adolescent. Temporizing measures, such as endoscopic debulking or tracheostomy, may be required if airway occlusion is imminent. Frozen section can be obtained via endoscopic debulking to permit pathologic diagnosis before definitive management. Open resection is the preferred definitive management for many tumors because it minimizes the risk of tumor regrowth and can establish clear tumor margins. However, an endoscopic approach offers the advantages of a less invasive, shorter procedure, and avoids any risk of subsequent graft failure compared to open tracheobronchial resection and reconstruction [4]. Open resection may follow endoscopic management immediately after frozen biopsy or in a staged manner.

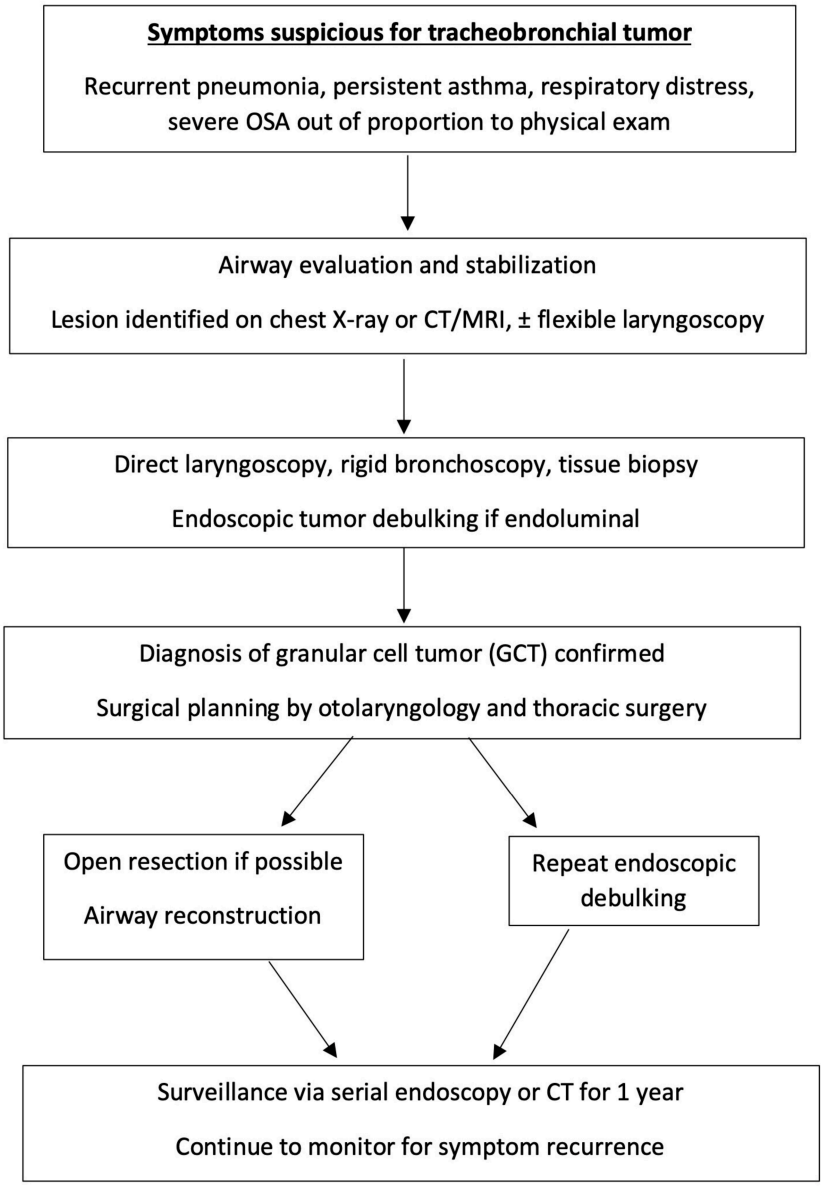


Figure 2. Proposed clinical pathway for management of pediatric tracheobronchial granular cell tumor derived from case-based evidence.

Regarding surveillance, we suggest that endoluminal tumors managed with tracheal or bronchial resection be monitored with serial endoscopy if the anastomotic site is endoscopically accessible. Endoscopy can also be used to assess airway anastomosis. If the

tumor site is not accessible endoscopically, such as in our Case 2, CT surveillance may be more appropriate.

4.3. Considerations for Subglottic Tumors

A recent review of laryngeal GCTs by Mur et al. identified 18 (49%) of 38 reported cases involved the subglottis [36]. Our search criteria excluded subglottic GCTs that did not have tracheal involvement. Exclusively subglottic tumors may be treated similarly to those of the proximal cervical trachea via endoscopic laser debulking or open resection [37–39]. Some reports of subglottic GCTs treated endoscopically found recurrence within 6 weeks to 3 years, requiring repeat endoscopic treatment and, in some cases, eventual resection by laryngofissure [17,18,39].

4.4. Study Limitations

Limitations of the study include a small sample size because pediatric tracheobronchial GCT is rare and tumor recurrence is an even rarer event due to a limited follow-up period. The small sample sizes of both the endoscopic and open resection groups limited the ability to perform inferential statistics. Furthermore, most articles did not describe the rationale for proceeding with endoscopic versus open management, limiting comparisons between groups. Case reports and case series present inherent bias and were the only article types included, prohibiting any meta-analysis. These case reports spanned over 80 years of pooled cases (1939–2026), as diagnostic and surgical management of pediatric airway tumors have evolved. This heterogeneity decreases the validity of comparison across cases.

Future reports of tracheobronchial tumors should ideally include demographic information, such as age and race/ethnicity, as well as presenting symptoms, tumor size, initial airway management, definitive management, and duration and modality of surveillance (i.e., radiologic, endoscopic, or symptom reporting).

5. Conclusions

Tracheobronchial tumor should be considered in the differential diagnosis of pediatric patients presenting with recurrent pneumonia, persistent asthma, and severe OSA out of proportion to upper airway examination. This highlights the need for a high index of suspicion in all cases when disease presentation is atypical or unresponsive to standard treatment. Tracheobronchial GCTs should be treated with airway stabilization and definitive surgical resection. Since tumor recurrences have only been reported following an endoscopic approach, our proposed management pathway suggests open resection when possible. Smaller tumors may be more amenable to an endoscopic resection. The risk of recurrence necessitates postoperative surveillance by endoscopy or imaging.

Supplementary Materials: The following supporting information can be downloaded at: https://www.mdpi.com/article/10.3390/ohbm7020039/s1, File S1: CARE checklist; File S2: PRISMA 2020 checklist.

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Institutional Review Board Statement: This case series and review did not meet the definition of human subjects research as described in 45 CFR 46.102(L) and was deemed not to be under the purview of an IRB.

Informed Consent Statement: Patient consent was waived due to use of retrospective de-identified data and determination to not meet the definition of human subjects research by the IRB.

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Abbreviations

The following abbreviations are used in this manuscript:

GCT	Granular cell tumor
OSA	Obstructive sleep apnea
CT	Computed tomography
AHI	Apnea-hypopnea index
PFT	Pulmonary function test

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