Castleman Disease Presenting as a Parapharyngeal Mass: The First Pediatric Case in the United States

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Introduction

Castleman Disease is a rare nonclonal lymphoproliferative disorder that presents as benign slow-growing masses. Patients with multicentric disease may have systemic symptoms such as fevers, night sweats, fatigue or weight loss, and often have associated HIV or HHV-8. Unicentric disease is often asymptomatic until the masses begin compressing surrounding structures. Though the majority of unicentric cases are in the mediastinum, they can rarely present in the head and neck region, most typically in cervical lymph node chains. Only a very small number of cases have been reported in the parapharyngeal space, typically in adults. We present the first case in the United States to our knowledge of unicentric parapharyngeal Castleman Disease in a pediatric patient.

Case Presentation

A 13-year-old female was referred to our tertiary care center after a large right parapharyngeal mass was identified during outside adenotonsillectomy. Tonsillectomy was aborted and a biopsy showed only lymphocytes and fibrous tissue without malignancy. On our initial evaluation, she had hyponasal voice, biopsy-related throat pain, and moderate snoring.

CT displayed a 7.6cm parapharyngeal mass extending from the skull base to the tongue base, narrowing the oropharyngeal airway. The mass was intimately associated with the carotid artery, but with smooth borders and no infiltration (Fig. 1). A repeat transoral biopsy was performed, which resulted in similar histologic findings as well as brisk intraoperative bleeding (Fig. 2).

Figure 1

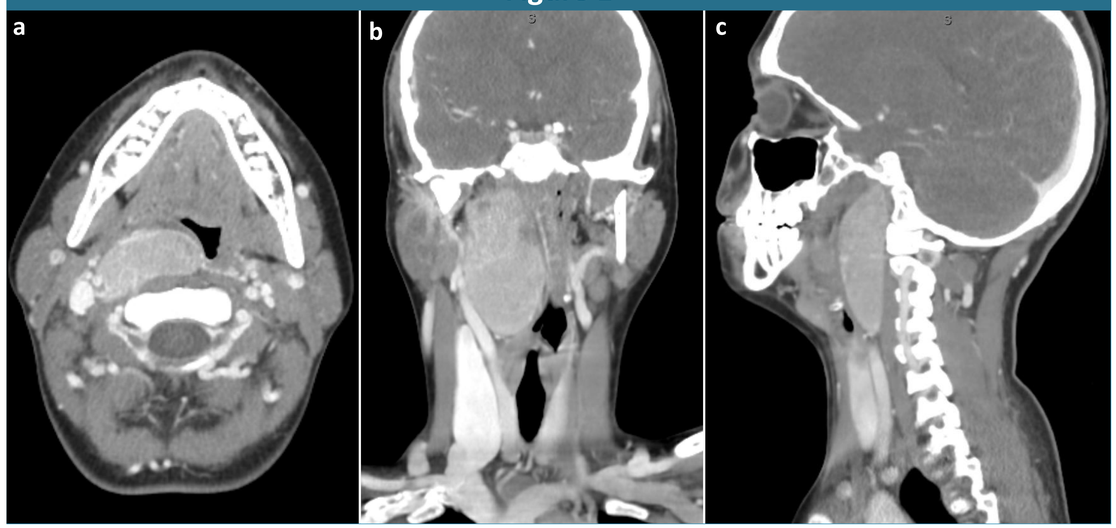

Fig. 1. (a) axial (b) coronal and (c) sagittal CT images with contrast: 7.6cm parapharyngeal mass extending from skull base to base of tongue

Figure 2

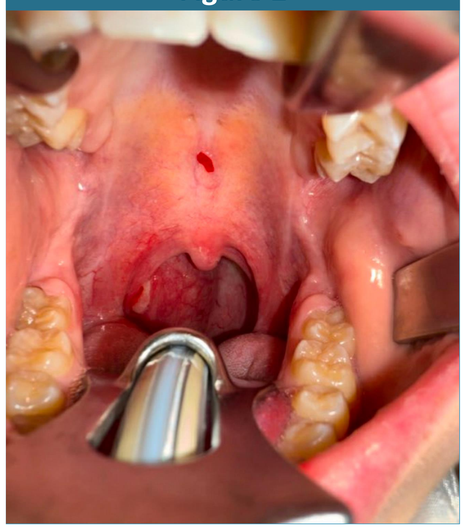

Fig. 2. transoral biopsy of right parapharyngeal mass

Figure 3

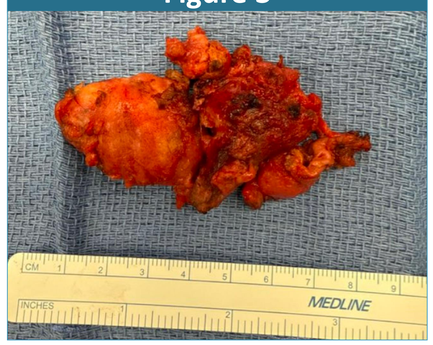

Fig. 3. surgical specimen representing ~90% of lesion

Treatment

Given the lesion's rapid growth and inconclusive biopsies, surgical resection was pursued with pre-operative embolization of ascending pharyngeal artery branches with Interventional Neuroradiology.

~90% of the lesion was resected transorally using a Dingman retractor with endoscopic assistance (Fig. 3). Intraoperative frozen section showed only lymphocytes. With no evidence of sarcoma or other malignant etiology, a small amount of lesion was left for safety along the carotid artery at the skull base.

Final pathology was consistent with Castleman Disease, hyaline vascular variant. CT Chest, Abdomen, Pelvis confirmed there was no multicentric involvement. HIV and HHV-8 labs were predictably negative.

Once tolerating a liquid diet, the patient was discharged on post-operative day 3. She remains under surveillance without further intervention or progression of the residual lesion to date.

Discussion

This case represents and highly uncommon presentation of Castleman Disease, particularly within the pediatric population. Ten prior cases of parapharyngeal unicentric disease have been identified in the literature, including three pediatric cases from China, Spain, and Italy, all of which had curative outcomes. One pediatric patient received pre-operative embolization similar to our patient. All patients underwent surgical resection. One patient experienced a recurrence requiring radiation for definitive management. Almost all cases involved a transcervical approach, with the only reported surgical complications being transient nerve weaknesses (Table 1).

In this patient, sufficient resection was achieved with a minimally invasive transoral approach due to use of a wide retractor, piecemeal resection starting with the middle third in order to visualize the full inferior and superior extent of tumor, preservation of involved critical structures, and pre-operative embolization for improved hemostasis throughout the case.

Patient	Lesion	Presentation	Treatment	Pathologic subtype	Outcome	Source
20 y.o. M, Turkey	9cm, jugular foramen to vocal folds	Dysphagia x4 years	Total resection, transcervical approach, pre-operative diagnostic angiography	Hyaline vascular	No recurrence, transient post-operative left vocal fold immobility	Karakoc et al. 2011
40 y.o. F, New York, NY	Post-styloid parapharyngeal space	Incidental, mild globus sensation	Total resection, transcervical infratemporal fossa approach	Hyaline vascular	No recurrence, unremarkable post-op course	Clain et al. 2013
8 y.o. F, China	4cm, parapharyngeal, extension into parotid region	Incidental, mild facial asymmetry from parotid extension	Total resection, cervical-parotid approach, included parotidectomy and level II neck dissection	Hyaline vascular	No recurrence, unremarkable post-op course	Wang et al. 2023
53 y.o. F, Tunisia	Parapharyngeal, 5cm retropharyngeal	Parotid swelling, otherwise asymptomatic	Total resection, cervical-parotid approach, EBRT for recurrence	unspecified	Retropharyngeal recurrence 1 year post-op requiring radiation	Jbali et al. 2022
38 y.o. M, Cincinnati, OH	4c,m post-styloid parapharyngeal	Transient facial numbness, self resolved, presumed unrelated	Total resection, transcervical approach, included submandibular gland excision	Hyaline vascular	No recurrence, unremarkable post-op course	Choi et al. 2019
10 y.o. F, Spain	8cm, nasopharynx to hypopharynx, contacting skull base, partially obliterating pharynx	Neck swelling, progressive dysphagia	Total resection, transcervical approach pre-op embolization	Hyaline vascular	No recurrence, unremarkable post-op course	Raboso et al. 2019
34 y.o. F, South Korea	8cm, retropharyngeal and parapharyngeal, skull base to hyoid	Painless oropharyngeal and neck swelling	Total resection, transcervical approach	Hyaline vascular	No reported complications	Shin et al. 2000
32 y.o. M, China	3cm, tonsil	Tonsillar hypertrophy	Total resection via tonsillectomy	Hyaline vascular	No recurrence, unremarkable post-op course	Li et al. 2019
42 y.o. F, Colombia	6cm, parapharyngeal	Globus sensation, neck swelling	Total resection, transcervical approach	Hyaline vascular	No recurrence, unremarkable post-op course	Ozgursoy et al. 2009
16 y.o. M, Italy	7cm, para-pharyngeal, nasopharynx to vallecula	Submandibular swelling	Total resection, transcervical approach	Hyaline vascular	Mild transient CN IX, X, X, XII weakness	Ottaviani et al. 1999

Table 1. summary of prior reported cases of unicentric parapharyngeal Castleman Disease

Teaching Points

Castleman Disease can present in rare cases in an isolated fashion in the parapharyngeal space, both in adult and pediatric patients, and can be successfully treated with surgical resection alone

Once unicentric diagnosis is confirmed, debulking is a reasonable surgical option when masses lie in close proximity to critical structures

Pre-operative embolization can be a useful adjunct to transoral resection

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