Otolaryngology Case Reports 22 (2022) 100392

Contents lists available at ScienceDirect

Otolaryngology Case Reports

journal homepage: www.elsevier.com/locate/xocr

Retropharyngeal lipoblastoma causing severe pediatric obstructive sleep apnea

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ARTICLE INFO

Keywords:
Obstructive sleep apnea
Head and neck neoplasm
Lipoblastoma
Retropharyngeal mass

ABSTRACT

Lipoblastomas are benign neoplasms of white adipose which usually occupy the trunk and limbs in pediatric patients. They can seldomly involve the head and neck, usually as a lateral cervical mass. Magnetic resonance imaging is the most useful modality for identifying these lesions. Treatment involves complete excision, and patients are followed with serial exams and/or imaging as recurrence is more common in the head and neck. Here we present an exceedingly rare case of retropharyngeal lipoblastoma in a two-year-old male causing severe obstructive sleep apnea which was identified during adenotonsillectomy. As this mass can be mistaken for other more common masses of the retropharyngeal space, we review the differential diagnoses, imaging, and histopathologic features of this neoplasm, which was responsible for upper airway obstruction in this case.

1. Introduction

Lipoblastomas are benign, lobulated neoplasms of white adipose tissue which predominantly occupy the trunk and limbs of pediatric patients under 3 years of age [1]. Rarely are these tumors identified within the head and neck. In these circumstances, the majority present as quickly enlarging, painless masses occupying the lateral neck and are managed with complete excision [1–3]. Herein we report one of the first cases of retropharyngeal lipoblastoma [3], the diagnosis of which remained elusive until histopathologic analysis. This report emphasizes the importance of differential diagnoses for upper airway obstruction and keen surgical awareness when unexpected anomalies are identified in routine procedures such as adenotonsillectomy.

2. Case presentation

A 2-year-old male was referred for evaluation of severe obstructive sleep apnea (OSA). Polysomnography demonstrated an apnea-hypopnea index of 18.5/hr (central 0.1/hr) and oxygen nadir of 69%. He had no other medical or surgical history. Exam demonstrated a healthy weight, normal-appearing child with 1+ tonsils and Friedman I palate. We decided to pursue sleep endoscopy followed by adenotonsillectomy with possible supraglottoplasty and/or lingual tonsillectomy. Endoscopy demonstrated protuberant adenoid tissues obstructing the nasopharynx without laryngomalacia or lingual tonsillar hypertrophy. Extracapsular tonsillectomy was performed. Upon transoral evaluation of the nasopharynx in preparation for adenoidectomy, a midline mass was identified as the primary source of nasopharyngeal obstruction rather than adenoid hypertrophy. The mass was submucosal and soft, with discrete edges and no mucosal abnormality. The adenoidectomy was deferred. Magnetic resonance imaging (MRI) demonstrated a T2 hyperintense, submucosal, cystic lesion with thin septations in the midline prevertebral space of the nasopharynx measuring 1.5 × 2.6 × 2.9 cm. Heterogeneous internal T1 signaling and few internal septations were seen (Fig. 1A and B). The mass displaced the adenoid pad and soft palate and significantly narrowed the nasopharyngeal airway. Imaging showed no diffusion restriction, intradural or intracranial communication, vertebral periosteal reaction, or lymphadenopathy. The leading differential diagnosis was a Thornwaldt cyst. The patient then underwent complete transoral excision of the mass, which demonstrated a cystic, well-circumscribed, tan-white, semisolid mass underlying the buccopharyngeal fascia (Fig. 2A–D). The postoperative course was unremarkable. Histopathologic examination revealed irregular lobules of adipocytes of varying size and maturation lying within a blue myxoid background separated by connective tissue septa of varying thickness. (Fig. 3A-C). The final diagnosis was a retropharyngeal encapsulated lipoblastoma. A six-month follow-up MRI neck/nasopharynx with and without contrast demonstrated strands of fat density within the nasopharynx but no clear

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https://doi.org/10.1016/j.xocr.2022.100392
Received 14 December 2021; Accepted 3 January 2022
Available online 5 January 2022

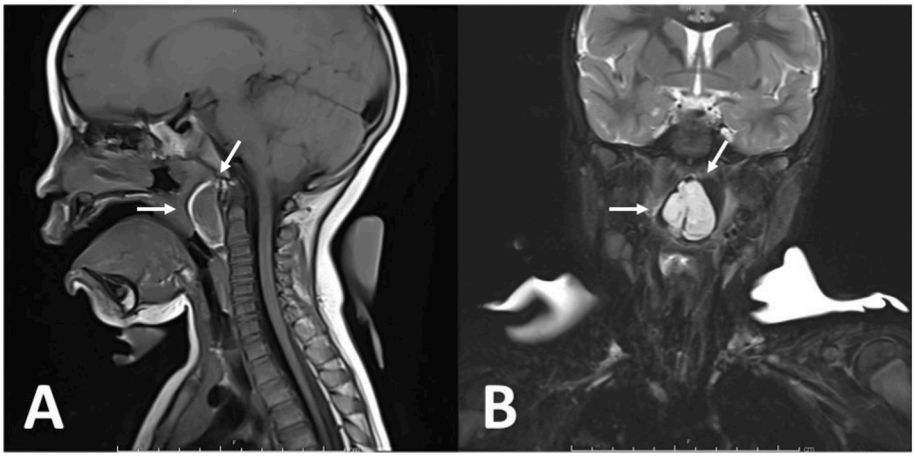

Fig. 1. (A) Mid-sagittal T1-weighted MRI revealing intermediate intensity, submucosal, cystic lesion in the midline prevertebral space of the nasopharynx measuring 1.5 × 2.9 cm. The mass displaces the adenoid tonsil and soft palate and narrows the nasopharynx. There is no intradural or intracranial communication nor periosteal reaction. **(B)** Coronal T2-weighted MRI with fat suppression revealing a 2.6 × 2.9 cm, non-enhancing, midline cystic mass with internal heterogeneous intensity and few thin internal septations.

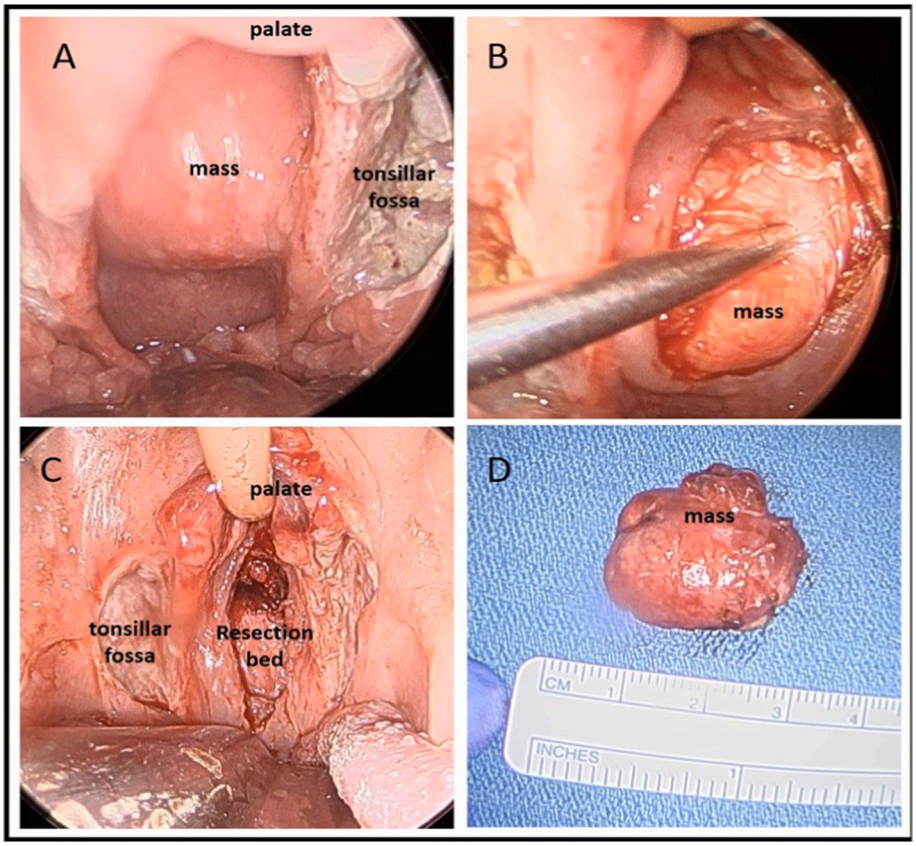

Fig. 2. All images are obtained transorally with a 30-degree endoscope and McIvor mouth gag. **(A)** Transoral view of oropharynx demonstrating the submucosal mass protruding inferiorly from the nasopharynx. **(B)** Submucosal mass with overlying mucosa incised. **(C)** Resection bed after transoral excision of the lesion. **(D)** Well-circumscribed, smooth, and lobulated mass after excision.

evidence of residual or recurrent disease.

3. Discussion

The differential for a benign-appearing, cystic mass in the retropharyngeal space is broad. As demonstrated in this case, visualization of the posterior nasopharynx in pediatric patients is difficult unless sedated under general anesthesia. In this case, transnasal flexible endoscopy without retraction of the palate yielded an incomplete assessment of the nature of the obstruction. Furthermore, diagnosis without imaging and pathology is difficult, especially as signs and symptoms of lesions in this area are relatively nonspecific [4].

Thornwaldt cysts are well-defined, rounded nasopharyngeal cysts resulting from pharyngeal bursa occlusion secondary to inflammation or surgical trauma [4]. They are rare in the 1st decade of life and appear within the submucosal, midline, posterior nasopharynx with T1 hypointensity and T2 hyperintensity on MRI without contrast enhancement [5]. These lesions, however, are lined with respiratory epithelium with minimal lymphoid tissue within the cyst [6]. This contrasts with adenoid retention cysts, which are usually small, isolated, ovoid cysts located eccentrically in the adenoid bed with similar imaging findings of Thornwaldt cysts [4,5]. Histopathology instead demonstrates abundant

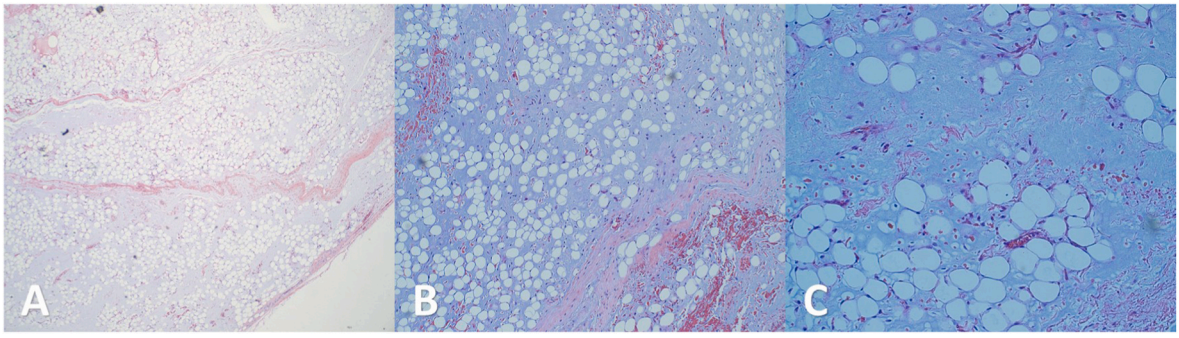

Fig. 3. **(A)** Low power view demonstrating irregular lobules of mature and immature adipocytes in a myxoid background separated by connective tissue septa of varying thickness (H&E 4x). **(B)** A closer view of adipocytes of varying size and maturation surrounded by a myxoid matrix. A small amount of pink connective tissue that is creating lobulations is present in the lower right corner (H&E 10x). **(C)** High power view of adipocytes of varying size and maturation surrounded by a blue myxoid matrix. A plexiform vascular pattern is also apparent (H&E 20x). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

lymphoid tissue and mucus center surrounded by granulation tissue [4]. Nasopharyngeal branchial cleft cysts appear laterally near the Eustachian tube and are enhancing with T1-hypointensity and T2-hyperintensity [7]. They are lined with stratified squamous or respiratory epithelium and filled with seromucinous fluid [8].

Lipoblastomas are benign tumors of embryonal white adipocytes which did not fully mature and lack malignant potential. They are further categorized as encapsulated or diffuse. The more common encapsulated form, as seen in this case, are well-circumscribed, non-invasive, and unifocal. The diffuse form, also known as lipoblastomatosis, are multicentric and infiltrate surrounding structures [1–3]. The vast majority of these lesions present within the trunk and extremities of pediatric patients, making head and neck lipoblastomas incredibly rare [1]. Almost no reports exist of retropharyngeal or nasopharyngeal lipoblastomas, as most appear within the lateral neck [1–3]. Males predominate (55%–68%) with average presentation from age 2–3 years [1–3]. Although benign, they can rapidly increase in size to cause compression and mass effect. Symptoms vary depending upon location involved, although over half present asymptomatically followed by respiratory distress [1].

The workup for nasopharyngeal masses should begin with visualization through nasopharyngoscopy unless incidentally identified on imaging first. Imaging involves computed tomography (CT) or MRI and is important to evaluate for malignancy such as nasopharyngeal carcinoma [4]. CT is quite non-specific for lipoblastomas, but will demonstrate a non-enhancing, lobulated, heterogenous mass with density similar to adipose [2]. MRI provides the greatest detail and will identify a non-enhancing, encapsulated, lobulated mass with heterogeneous T1-weighted signal (less intense than mature adipose) and hyperintense T2-weighted signal. T2-weighted fat suppression sequences are particularly useful to identify lipoblastoma [2].

Although biopsy is feasible for benign-appearing lesions, complete surgical excision is recommended for symptomatic cases with preservation of critical structures to prevent morbidity. Histologically, they appear as a well-circumscribed mass of lobulated immature adipocytes divided by fibrous septations without evidence of nuclear atypia (as would be seen in liposarcoma) [1]. Recurrence within the first year of excision is 27%, slightly higher than trunk and extremity lipoblastomas. Therefore, follow up for these patients is important with repeat MRI imaging and/or endoscopy over at least five years. However, recurrences have been identified ten years after initial excision [1].

4. Conclusion

This case highlights the importance of endoscopic evaluation and surgical awareness in the setting of pediatric obstructive sleep apnea. Although adenoidectomy and/or tonsillectomy are the mainstay of treatment, remaining cognizant of alternative causes of upper airway obstruction is important to perform targeted surgical intervention and achieve the best outcomes.

Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Ethical statement

This investigation was deemed exempt from Institutional Board Review. Written consent for production of this case report was obtained from the primary caregiver after review of its content and images.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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