Recurrent Pediatric Extranasopharyngeal Angiofibroma of the Epiglottis: Case Report

Case Report

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Abstract

Angiofibroma is a non-encapsulated, highly vascular tumor that usually originates in the nasopharynx. Laryngeal cases of extranasopharyngeal angiofibroma (ENA) are a very rare pathology, especially in children. Only eight ENA laryngeal cases have been described in the literature, and only one of them is a pediatric case. In this report we present an 11-year-old child with epiglottic ENA resected with transoral endoscopic ultrasonic surgery (TOUSS) with review of the literature. Because of reccurrence after five months he underwent re-excision with CO_2 laser. Recurrences in ENA are infrequent, but as demonstrated in our case, close endoscopic follow-up is mandatory in this location. Endoscopic hemostatic procedures like TOUSS and CO_2 laser ensure bloodless surgery for the management of this type of vascular laryngeal tumors.

Keywords: Angiofibroma, epiglottis, ultrasonic surgical procedures, laser therapy, pediatric, case report

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Introduction

Angiofibroma is a non-encapsulated, highly vascular tumor that usually originates in the nasopharynx. Head and neck angiofibroma with an origin other than the nasopharynx [extranasopharyngeal angiofibroma (ENA)] is highly uncommon. Laryngeal cases are particularly unusual, especially in children (1).

Case Presentation

An 11-year-old male was referred to our institution with a six-week history of increased snoring, mild dysphagia and dysphonia without dyspnea. Flexible nasolaryngoscopy (FNL) revealed a 3-cm smooth, round, reddish lesion in the supraglottis (the laryngeal side of the epiglottis). An urgent computed tomography (CT) scan confirmed the presence of a polypoid mass likely originating from the laryngeal side of the epiglottis (Figure 1a).

Due to the patient's age, the possibility of endoscopic surgical access, CT findings, endoscopic evidence that it was a pediculated mass, and the availability of an ultrasonic scalpel for resection, we concluded that invasive diagnostic procedures such as angiography were not necessary.

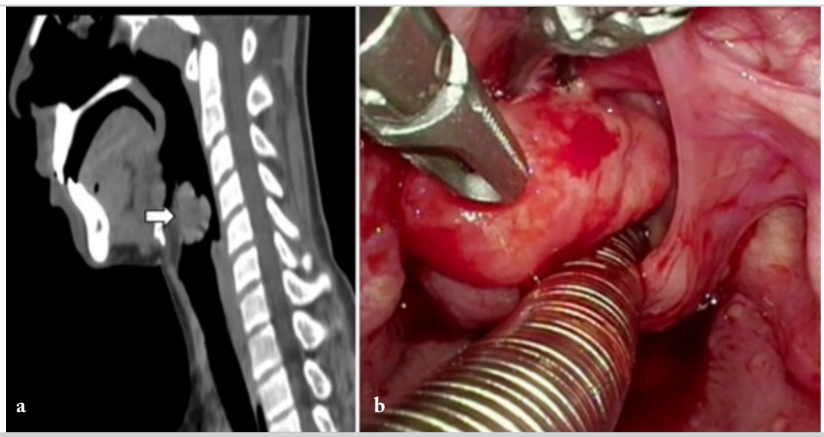

Figure 1. Original epiglottic tumor. a) Sagittal computed tomography with contrast that shows a polypoid solid lesion of 20.6x21.1x23.7 mm (white arrow) with an hyperdense periphery and a less dense center without epiglottic cartilage or pre-epiglottic fat involvement and without pathological adenopathy; b) Intraoperative endoscopic view of the epiglottis attached mass obstructing the supraglottic airway being resected with the grasping tip of the ultrasonic scalpel

We performed a complete resection using transoral endoscopic ultrasonic surgery (TOUSS) without a previous biopsy because of the child's airway compromise and the risk of profuse bleeding, bearing in mind that resection was going to be performed with a hemostatic method (Figures 1b, 2a). The postoperative course was uneventful and the patient was discharged after 24 hours once FNL confirmed the absence of laryngeal edema or airway compromise.

The histological description was a 3x2x2 cm diameter polypoid mass lined by smooth mucosa with focal ulceration that covered a soft tissue proliferation with numerous blood vessels with an attenuated smooth muscle layer and a thin endothelial layer with no atypia. The vessels often presented a staghorn shape, resembling that of a solitary fibrous tumor or hemangiopericytoma. Between the vessels, there was a dense, collagenous, fibrous stroma containing scattered plump, epithelioid or stellate fibroblasts with occasional mild nuclear atypia and no mitotic activity. Stromal cellularity varied and hyalinized nearly acellular areas were present in the center area. No malignant transformation, necrosis, mitotic activity or high cellularity were seen. Immunohistochemically, the stroma cells were positive for Actina HHF-35 and negative for desmin, CD34, STAT-6, S100, SOX10, myogenin, SM actin, beta catenin and estrogen or progesterone receptors (Figures 2b, c, d).

A possible recurrence of eight mm was found five months after the resection when FNL once more showed a supraglottic lesion. Revision surgery was performed by direct laryngoscopy with CO_2 laser. Again, the patient was discharged after 24 hours of uneventful postoperative follow-up (Figures 3a, b). Histopathological diagnosis was ENA again. After two years of follow-up, the patient remains disease free.

Informed consent form was signed by the parents of the patient to publish this case.

Discussion

ENA is an uncommon tumor whose clinical presentation differs from classic juvenile nasopharyngeal angiofibroma (JNA); therefore, they should be regarded as separate clinical entities. ENA can originate anywhere in the upper aerodigestive tract and there is a much more balanced gender ratio (2.13:1). It predominantly affects the nasal septum and often presents with faster-progressing symptoms (1).

An extensive Medline and Google systematic research by Windfuhr and Vent (1) in 2017, which included papers published up to 31 December 2015, reported 174 patients with ENA, and a male preponderancy of 66.1% and a median age of 23 years at diagnosis. Their findings included only 22 patients aged under 12 years, only one of them was epiglottic (2, 3). To our knowledge, only seven cases in children younger than 12 years have been published since Windfuhr and Vent's (1) review: two nasal septum ENA reports by Singh et al. (4) and Ganguly et al. (5), an oropharyngeal case by Gupta et al. (6), two inferior turbinate cases by Kim and Choi (7), an ethmoid case by Uwents et al. (8), and an inferior turbinate and lateral wall of nasopharynx case by Yan et al. (9).

The most frequent locations in pediatric cases were the nasal septum (30%), the maxilla (15%), the inferior turbinate (11%), and the ethmoid (11%).

Only eight ENA laryngeal cases have been described in the literature so far and only one of them is a pediatric case (in an 8-year-old child) described by Gołąbek et al. (2). In their case, the location was also epiglottic, but the tumor was smaller in size compared to our case (1.5 cm). Both patients developed the typical clinical manifestations of laryngeal involvement

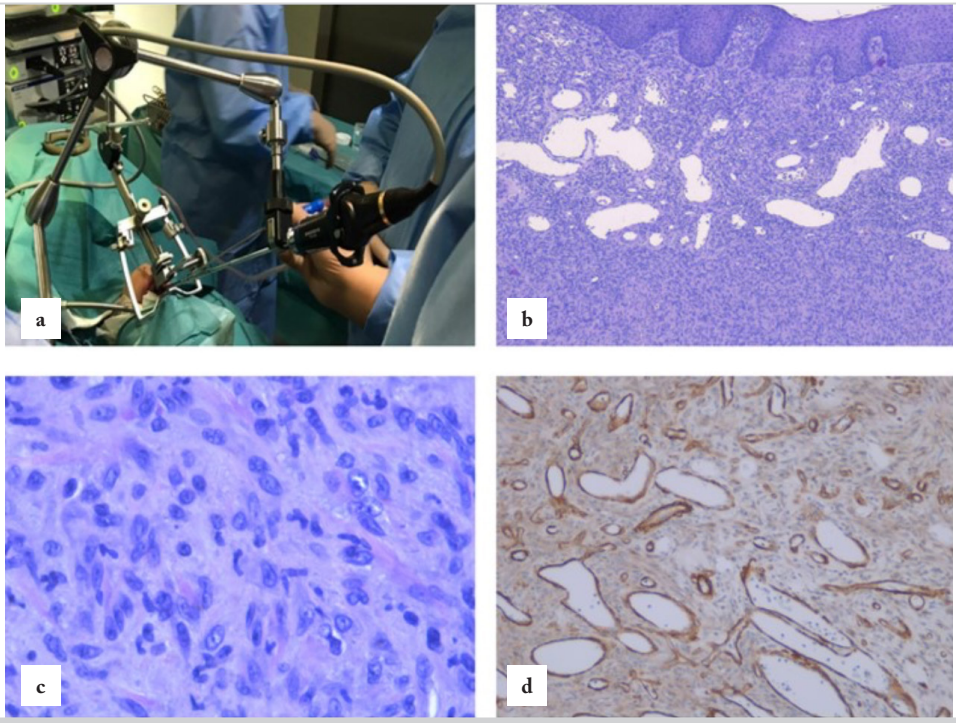

Figure 2. Transoral endoscopic ultrasonic surgery (TOUSS) and histological findings. a) TOUSS setup with retractor and scope holder; b) Hematoxylin & Eosin (H&E) (10X): Extranasopharyngeal angiofibroma presenting as a polypoid lesion. Sharply demarcated mass containing multiple vascular profiles. A non-specific superficial zone with squamous mucosa covers the luminal aspect of the tumor. Hemangiopericytoma-like abundant vascular pattern. c) H&E (40X): A higher magnification shows spindled cells containing uniform nuclei with delicate nucleoli. A coarse collagenous matrix is a consistent finding; d) IHQ for CD34 shows positivity in vascular channels and is negative in fusocellular components

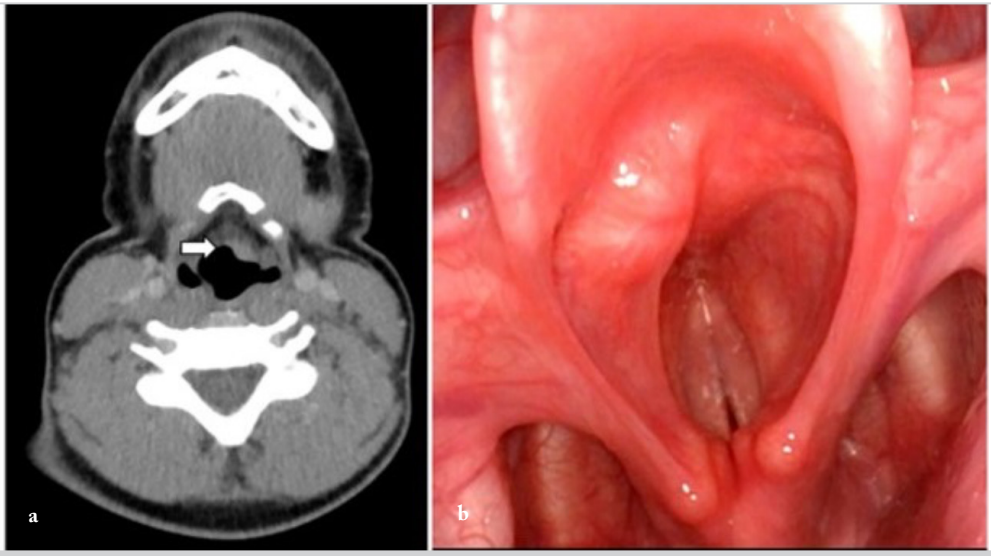

Figure 3. Fifth month recurrent tumor. a) Axial computed tomography with contrast showing a lesion of 5x5x8 mm with a moderate contrast enhancement in laryngeal epiglottic side (small white arrow); b) Endoscopic view of the recurrent lesion in outpatient control

such as hoarseness, muffled voice or dysphagia, as well as quick progressing symptoms [four weeks in Gołąbek et al.'s (2) case and six weeks in our case] unlike the two and a half years of progressive symptomatology reported in the previous supraglottic ENA cases in adults (10).

Hemostatic endoscopic procedures were used in our case. The original tumor was resected with the transoral endoscopic ultrasonic surgery technology, a non-robotic endoscopic, three-handed method using an ultrasonic scalpel as a resection tool (a 35-cm ThunderbeatTM) with high-definition 2D-3D endoscopic imaging with Olympus ENDOEYE Flex 5-mm 2D/10 mm 3D deflecting tip video laparoscopes (Figure 2a).

The recurrence was successfully excised with a CO_2 laser. Given that this recurrence was smaller in size (8 mm) than the original tumor (3 cm) and had a very sessile epiglottic

attachment, we thought that this limited the benefits of using an ultrasonic scalpel grasping tip and probably we would not be able to excise it without the risk of breaking the surgical piece and felt that a monobloc resection with laser was easier. There were previous successful excisions with this method in this location described in literature, too (2, 3).

The patient is now disease-free for two years after this second surgery with a close follow-up: first three months post-recurrence resection with monthly controls, then quarterly up to one year of control, and later biannual, all of them with FNL.

In contrast with nasopharyngeal angiofibroma, ENA recurrences were reported only in four patients included in Windfuhr and Vent's (1) systematic research; all of them within 12 months after diagnosis and only one in a pediatric case as early as two weeks after surgery (11).

We consider that our patient had the recurrence because there was a tumor growth following a complete monobloc excision. However, we know that the prevalent opinion in the recent JNA series is that most postsurgical recurrences are in fact cases of persistent disease due to incomplete excision and we cannot rule out that possibility in our case, even when the time to diagnosis was considerably long (five months) or having performed several endoscopies without evidence of the tumoral presence in this period.

Conclusion

Recurrences in ENA are infrequent but, as demonstrated with our case, a close follow up with FNL is mandatory in epiglottic locations until further experience is acquired with these rare tumors to help develop standardized protocols for their management.

Informed Consent: Informed consent form was signed by the parents of the patient to publish this case.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: M.H., A.L., R.S., J.A.P., M.F-F., Concept: M.H., A.L., C.A., R.S., J.A.P., M.F-F., Design: M.H., C.A., R.S., M.F-F., Data Collection and/or Processing: M.H., R.S., M.F-F., Analysis and/or Interpretation: M.H., A.L., C.A., J.A.P., M.F-F., Literature Search: M.H., A.L., J.A.P., Writing: M.H., A.L., M.F-F.

Conflict of Interest: There is no conflict of interest to disclose.

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Main Points

- Laryngeal cases of extranasopharyngeal angiofibroma (ENA) are a very rare pathology, especially in children.
- Recurrences in ENA are infrequent but, as demonstrated with our case, a close follow-up with flexible nasolaryngoscopy is recommended in epiglottic locations.
- In published epiglottic ENA cases, children have faster progressing symptoms than adults.
- Hemostatic surgical methods such as transoral endoscopic ultrasonic surgery or CO_2 laser may allow us to avoid invasive previous tests like angiography or partial biopsy in this type of vascular tumor.
- Given that recurrence is uncommon in this benign tumor and developed standardized protocols are not published, we decided to follow up the patient for two whole years.

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