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CASE REPORT Open Access

Atypical angiofibroma: a very rare case report and review of literature

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Abstract

Background: Extranasopharyngeal angiofibroma (ENPA) has been stratified to nodules consisting of blood vessels and fibrous tissue, which occur outside the nasopharynx. ENPA is histologically similar to nasopharyngeal angiofibroma (NA), differing in clinical and epidemiologic characteristics. Since these differences, ENPA considers as a diagnostic challenge and a painstaking appraisal with a high index of doubt is major in determining the true diagnosis and treatment. Fewer than a hundred cases of "atypical angiofibroma" are represented in the international literature, with only 6 cases having been reported previously about ENPA originating from the nasal septum.

Case presentation: We submit a summarized literature review with a case of ENPA emerging from the nasal septum in a 6-year-old child. Under general anesthesia and endoscopically, the mass that emerged from the septum was resected. Histopathology examination confirmed the diagnosis of ENPA. No recurrence over 1 year after surgery. Our patient had an age of onset and site of lesion different from most ENPAs, supporting the rareness of this case.

Conclusion: Our aim is to shed light on a very rare clinical presentation, describe the diagnostic and therapeutic workup, and suggest that ENPA must be regarded as a differential diagnosis with any complaint of nasal occlusion and epistaxis accompanied by a unilateral nasal tumor.

Keywords: Extranasopharyngeal angiofibroma, Atypical angiofibroma, Nasal obstruction, Epistaxis, Case report

Background

Although angiofibroma is a benign vascular neoplasm, it is a locally-aggressive tumor that typically generates from the nasopharynx [1, 2] and may spread into the nasal cavity [3]. Nasopharyngeal Angiofibroma represents 0.05% of all head and neck tumors [1, 4]. The triad of unilateral nasal occlusion, epistaxis, and a nasopharyngeal neoplasm propose the diagnosis [5]. It has been supposed that NA is a tumor dependent on testosterone [6].

Extranasopharyngeal angiofibroma (ENPA) has been stratified to nodules consisting of blood vessels and fibrous tissue, happening outside the nasopharynx [3]. It presents a diagnostic challenge and a painstaking appraisal with a high index of doubt is major in determining the true diagnosis and treatment [1, 3].

Some hundred cases of ENPA have been represented previously and the most affected site is the maxillary sinus (24.6–32%) [6], then the ethmoid, rare in the nasal cavities, inferior and middle concha, sphenoid sinuses, conjunctiva, molar, and retromolar site, tonsil, and larynx [6]. Whereas the nasal septum seems a quite rare site with only 6 cases having been described previously in the international literature [1, 3, 4]. De Vincentiis et al. reported 704 cases of angiofibroma and detected 13 cases outside the nasopharynx, thus suggesting that it is a probable site, though a rare manifestation [3]. Celik et al. proposed that different features other than the conventional angiofibroma indicate "atypical angiofibroma" [3].

We present the case of a 6-year-old child who suffered from progressive epistaxis and mass originating from the nasal septum. Histopathology examination confirmed the diagnosis of ENPA. Our patient had an age of onset and

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site of lesion different from most ENPAs, supporting the rareness of this case. Our aim is to shed light on a very rare clinical presentation, describe the diagnostic and therapeutic workup, and suggest that ENPA must be considered as a differential diagnosis with any complaint of nasal occlusion and epistaxis accompanied by a unilateral nasal tumor.

Case presentation

A 6-year-old child came to the ENT Clinic in June 2021, with 2-month complaint of left side sporadic anterior epistaxis growing in intensity with nasal blockage and snoring. The clinical history was negative for trauma, infection, bleeding disorders, familiar pathologies, medications, and exposure to nasal irritants. ENT checking revealed moderately deviated nasal septum on the right side with reddish well circumscribed non friable mass on the left side, easily bleeding upon manipulation and probing all around except the medial side. Ears, throat, and neck examination was normal. CT scan clarified mass occluding the anterior half of the left nostril, deviating from the septum to the right side. No extension into the paranasal sinuses or the nasopharynx and no bony erosion were noted (Fig. 1).

He was admitted to the ENT department. No anemia or coagulation disorders were detected. Under general anesthesia and endoscopically, the mass emerging from the septum was resected en bloc to its vascular base. Using electrical cauterization, the bleeding was controlled effectively (Fig. 2).

Using a 0o endoscope, we identified adenoid mass and bilateral peritubal tonsils (Fig. 3). A pack was placed in the left nasal fossa only. The period after surgery was uneventful, the pack was ejected on the third postoperative day with no bleeding, and the patient was discharged on the same day. Histological examination was consistent with nasal angiofibroma (Fig. 4). The patient was recalled for follow-up appointments. No recurrence or other nasal abnormalities have been seen (Fig. 5).

Discussion

ENPA has been stratified to nodules consisting of blood vessels and fibrous tissue, happening outside the nasopharynx [3]. Since the spread, gender, age, site of lesion, pathogenesis, clinical and imaging features, and recurrence are totally different from NAs [1, 4], ENPA must be observed as a distinct clinical existence [3]. Older patients are involved, females can also be affected,

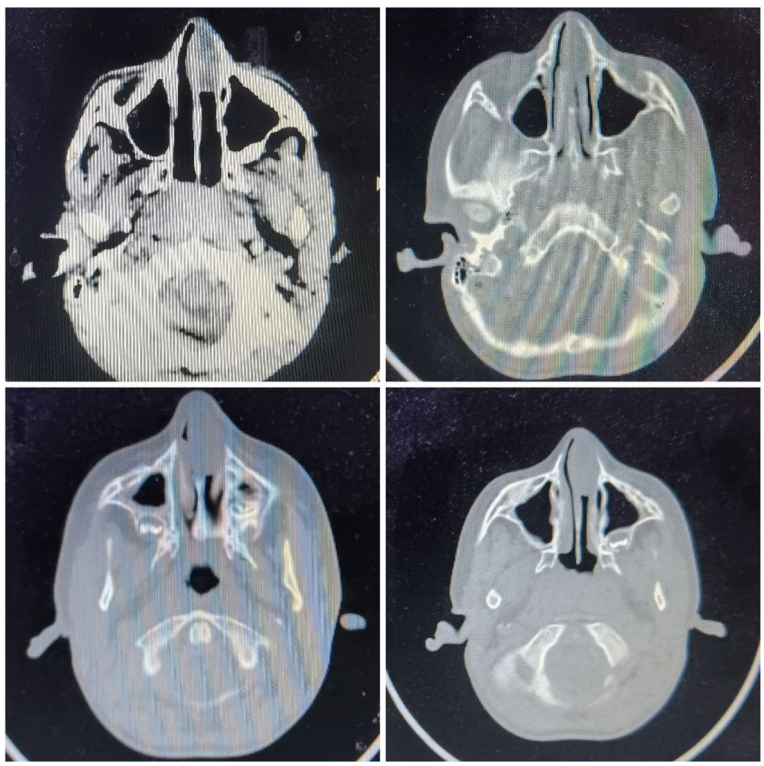

Fig. 1 A CT scan of the nose and paranasal sinuses. A soft tissue mass filling the anterior half of the left nasal fossa and deviating the anterior part of the septum to the right side. The mass was well circumscribed. Difficult to identify its origin. No extension into the nasopharynx or any paranasal sinuses and no bony erosions were noted

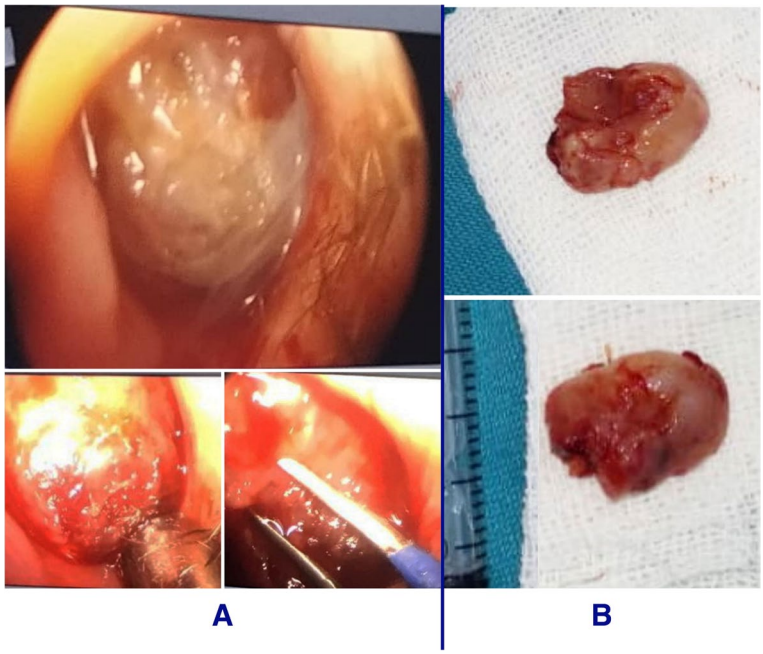

Fig. 2 Surgical excision under general anesthesia and endoscopic guidance. **A** The formation presented of hard consistency, firmly adhering to the septum and bleeding. It was removed up en bloc to its vascular base. Electrical cauterization was sufficient since the poor vascularity of the lesion allowed for removal without any significant intraoperative blood loss and did not require preoperative embolization. **B** The pedicle had been completely transected and the mass sent after resection for a histological examination

symptoms develop more speedily, and hypervascularity is minimal [3]. ENPA presents in an older cohort more than NA, with an average age of 22 years and 17 years, respectively, whereas for nasal vestibule angiofibroma the average age is 43 years [6]. While NA dominantly affects males, ENPA is a major happening in females [6]. The clinical manifestation of ENPA relies on the site and extent [3]. NA rises from the region of the pterygoid plate and the sphenopalatine foramen, and its etiology is debatable. Migration error of the basal fascia is considered as the etiology of ENPA, justifying its existence in diverse sites [4]. The most typical location is the maxillary sinus [4] while the septum is a very rare location [1].

Spinosi et al. reported in 2017 a case of a nasal septum angiofibroma in a 28-year-old man who suffered from right-sided epistaxis, airflow impairment, and nasal swelling [1]. Tasca and Compadretti reported in 2008 a case of a 57-year-old woman with a 1-year history of a right nasal obstruction due to an angiofibroma adhering to the posterior nasal septum [3]. Baptista et al. reported in 2014 a case of an 8-year-old girl who suffered from bilateral nasal obstruction and recurrent epistaxis, worse on the right side, with hyposmia and snoring due to ENPA inserted in the right inferior turbinate [4]. A rare case of angiofibroma of the mandible in a 16-year-old female patient was reported in 2016 by Khaliq et al. [7].

Though the management of ENPA is not totally codified and even the histopathological examination emphasizes the diagnosis, ENPA can be efficiently diagnosed and managed early with imaging, endoscopy, and clinical features, while embolization has been used in selected cases [1]. CT and MRI are essential to define the neoplasm's location and extension, with focusing on skull base involvement, intracranial extent, and relation to significant vessels and neurologic entities [3, 6]. Using a contrast agent with CT and MRI in NA produces a strong and commonly homogeneous enhancement. ENPAs generally have moderate enhancement of contrast or even nothing, due to the usual poor vascularity [3, 6]. Signs of suspected hypervascularity; indicate the need for arteriography before surgery to organize the needful precautions and minimize the risk of severe bleeding throughout biopsy or neoplasm eradication [3]. Using angiography and eclectic embolization must have a high threshold, that is, only if there is radiological proof of widespread, massive and hypervascular lesion [6].

Surgery is the treatment of choice [1, 3] with often preoperative embolization [7]. Although the maxillary artery supplies ENPA such as NA, it might not cause exaggerated bleeding during surgery due to the predominance of fibrous stroma, unlike NA [4]. An endoscopic and endonasal eradication, under general

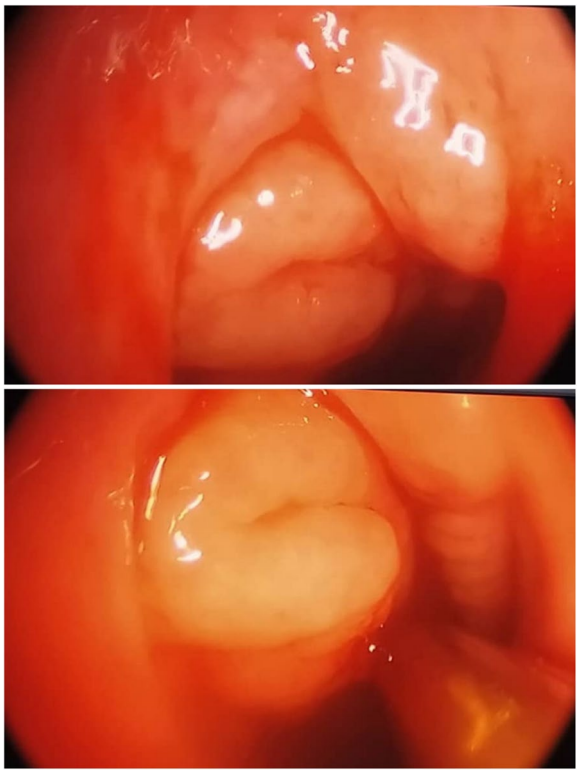

Fig. 3 An inspection of the nasal cavities through a 0o endoscope. Identifying an adenoid mass in the posterior wall of the nasopharynx and bilateral peritubal tonsils

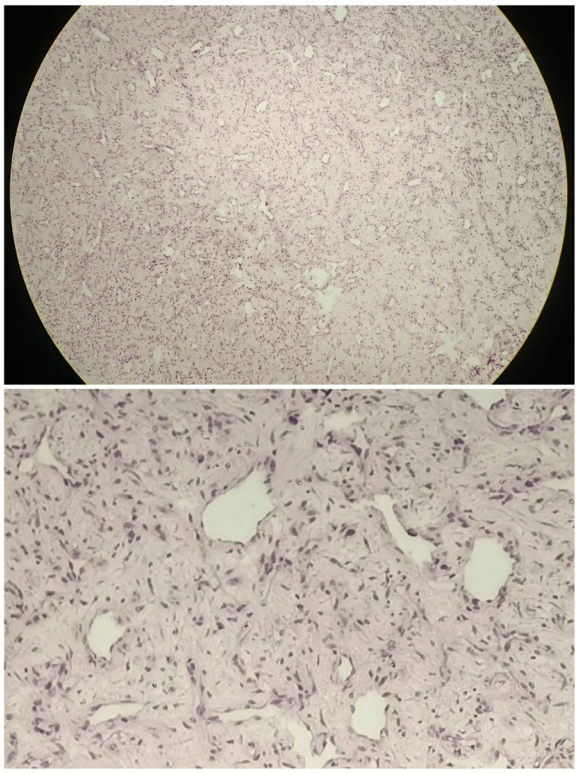

Fig. 4 Histopathological examination. Nasal angiofibroma which revealed (fibrous stroma with plump fibroblastic cells along with scattered capillaries)

anesthesia, must be attempted, though too much bleeding could demand an external incision. Electrical cauterization may not suffice, so an abrasion from the submucosal to the sub-perichondral area must be performed [1]. Other treatment modalities include [7]: sclerosing agents, radiotherapy, preoperative hormone therapy, chemotherapy, and arterial ligation. The outlook therapy should contain intraarterial immunotherapy and specific drugs that inhibit angiogenesis [7].

Differential diagnosis; includes hemangioma, hemangiopericitoma, or pyogenic granuloma [4]. The recurrence ratio of NA ranges from 6 to 27.5%, whereas no recurrence was notified for ENPA [4, 6] because its extrapharyngeal site simplifies total resection [4].

Our patient had an age of onset and the site of lesion different from those of most ENPAs. As usually occurs, epistaxis was the presenting sign. Under general anesthesia and endoscopically, the mass was removed en bloc without any significant intraoperative blood loss and without requiring preoperative embolization. Follow-up appointments confirmed no recurrence over 1 year after surgery.

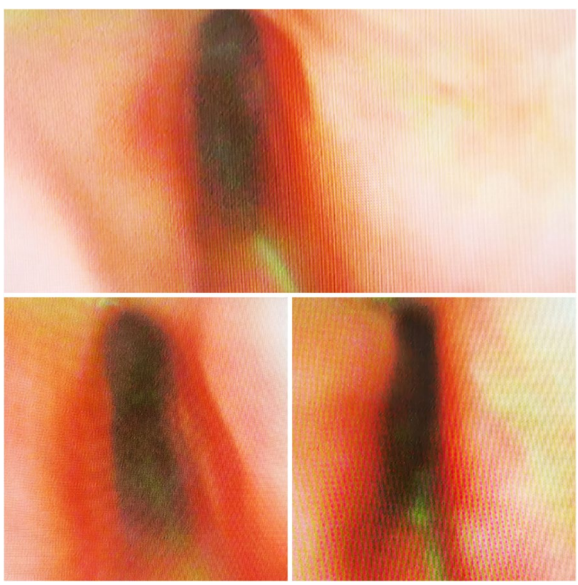

Fig. 5 Follow-up in 1 year after surgery. No recurrence or other nasal anomalies have been visible and nasal endoscopy showed a well-aligned septum

Conclusion

As a conclusion, ENPA, despite being uncommon, should be regarded as a differential diagnosis of nasal vascular neoplasms accompanied by nasal occlusion and epistaxis and the septum must be considered as a possible, although unusual, site of lesion of these tumors [1, 3].

Abbreviations

ENPA: Extranasopharyngeal angiofibroma; NA: Nasopharyngeal angiofibroma.

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Authors' contributions

A.M made the surgery, performed the revision of the manuscript, and edited the final edition of it. T.Y did the conception, design, and writing of the manuscript, performed the literature review, and followed up and collected the patient data. The authors read and approved the final manuscripts.

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Availability of data and materials

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Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

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Competing interests

The authors declare that they have no competing interests.

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