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Case Report

Rare case of extra nasopharyngeal angiofibroma arising from tonsil-case report

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ABSTRACT

Angiofibroma is a highly vascular tumour. It occurs in adolescent males mainly in nasopharynx. Due to aggressive local growth, skull base location and risk of profound haemorrhage, management is challenging. Angiofibromas have been sporadically described in extra nasopharyngeal locations. Presentation varies depending upon the location of origin. Here we present a case of 7 year old boy presenting with difficulty in breathing with lump in the throat for 3 years, which has progressively increased in 4 months period with recurrent chocking while sleeping. Initially he was diagnosed with polyp arising from the tonsil. Polyp along with tonsil excised completely. Histopathology confirmed extra nasopharyngeal angiofibroma (ENA). To the best of our knowledge, only 3 cases arising from tonsil had been reported in English literature.

Keywords: Angiofibroma, Extra nasopharyngeal, Tonsil

INTRODUCTION

Nasopharyngeal angiofibroma (NA) is a highly vascular benign tumour arising from nasopharynx. It represents less than 0.05% of tumours of the head and neck.[1] It is exclusively reported only in males between 12 and 14 years of age.[1] Tumour usually arises from the posterior part of nasal cavity close to the sphenopalatine foramen. Nasal obstruction and epistaxis are common presentations. Even though it is benign, bony erosions of sphenoid, pterygoid and clivus are common. Tumour can spread to orbit via inferior orbital fissure. Intracranial spread can occur through superior orbital fissure and through roof of infratemporal fossae. Tumour is mainly supplied through sphenopalatine and maxillary arteries. Diagnosis is mainly clinical and supported by radiology and histopathology.

Angiofibromas arising from other locations are called as extra NAs (ENA) or atypical angiofibromas.[2] Aetiology of ENA remains unclear. It was suggested that embryonic ectopic remnants during the development of nasal septum was the cause for the origin of septal ENA.[3] ENAs are reported to occur in females also. Management is mainly surgical. Radiotherapy and chemotherapy may be considered for recurrent and residual tumours.[4] Here, we present a case of ENA arising from tonsil which is removed completely with tonsil and confirmed by histopathological examination.

CASE REPORT

Seven years old otherwise healthy boy presented with history of lump in throat for 3 years. He was diagnosed with a tiny lump over the left tonsil by ENT physicians and paediatricians during the visit for treatment of respiratory infections over the course of 3 years prior to the current presentation. The child remained asymptomatic for three years. However, for the past 4 months, child started feeling foreign body sensation in

throat with progressively increasing dysphagia. Child became progressively dyspnoeic with intermittent stridor for the past one month with frequent apnoeic episodes at night. The child's past medical history and family history were unremarkable.

Clinical examination revealed pedunculated, pinkish, oblong polyp arising from the superior pole of left tonsil. It was found to be blocking a major part of oropharynx (Figure 1). Inferior pole of the polyp was visualised by flexible pharyngoscopic examination. It was found to be reaching until the laryngeal inlet. Otherwise, ENT, head and neck examination (especially right tonsil) was unremarkable. Diagnosis of polyp arising from left tonsil was made and radiological investigations were considered not indicated.

Tonsil along with pedunculated polyp was surgically removed in toto (Figure 2 and 3). Surgery was uneventful with minimal bleeding. Child made a normal postoperative recovery.

Gross examination of the specimen showed the pedunculated polyp measuring 3.2x1.5x1.5 cm. The attached tonsillar tissue measured 3x2x1 cm. Histopathological examination revealed tissue lined by stratified squamous epithelium. Sub-epithelium showed prominent lymphoid follicles of varying sizes. Tonsillar crypts are seen with minimal fibrotic bands separating the lymphoid collections. Deep to the lymphoid collections and partly involving the tonsil are vascular channels of varying sizes lined by flat endothelial cells separated by spindly fibrous stroma. The vascular endothelial cells are positive for CD 31 (Clone: JC70A), CD 34 (Clone: QBEND 10) and most are also positive for D2-40 (clone: Polyclonal). Stromal cells show patchy nuclear positivity for estrogen receptor (ER) (Clone: 1D5) and androgenic receptor (AR). Cytoplasmic beta-catenin is seen in epithelial cells, but no nuclear localization noted. Stromal cells are negative for beta-catenin (Clone: 14) (All controls show appropriate reactivity) (Figures 4-9) Above histological and immunohistochemical features confirmed the diagnosis of extra NA.

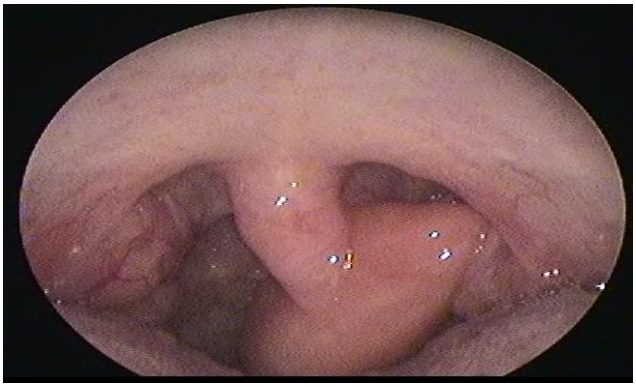

Figure 1: Preoperative picture showing pinkish polyp arising from left tonsil occupying oropharynx and extending down.

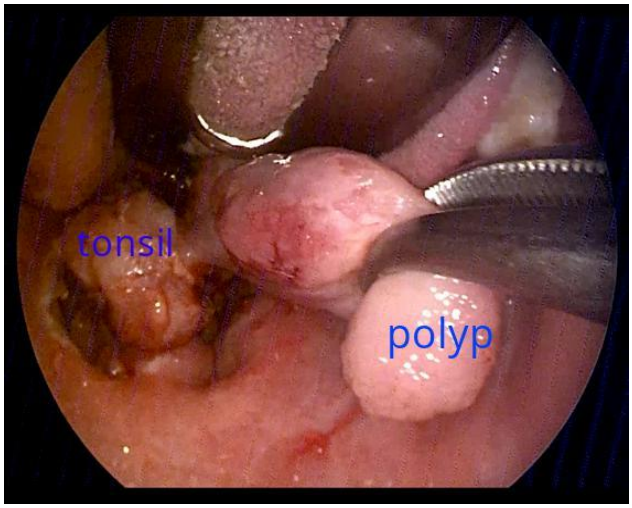

Figure 2: Intraoperative picture showing removal of tonsil along with polyp.

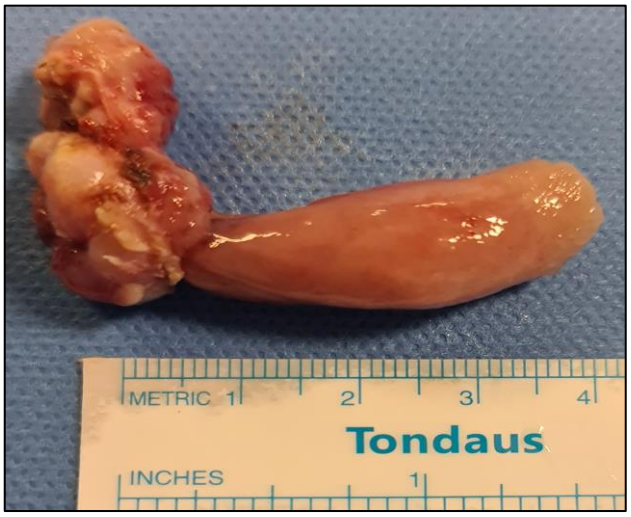

Figure 3: Specimen-excised tonsil with polyp.

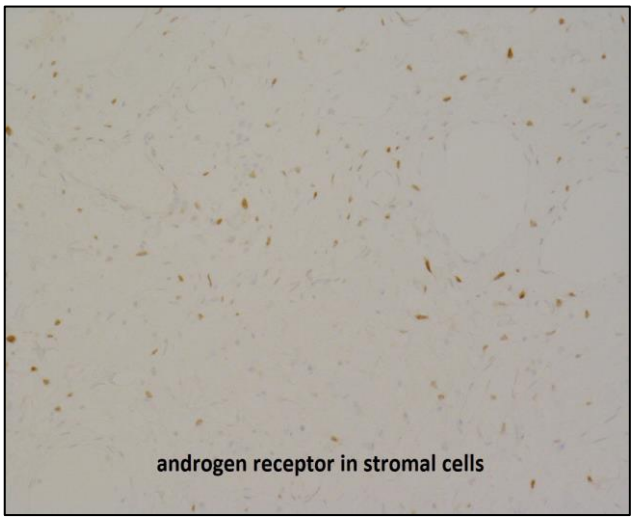

Figure 4: Immunohistochemistry showing androgen receptors in stromal cells (x200 magnification).

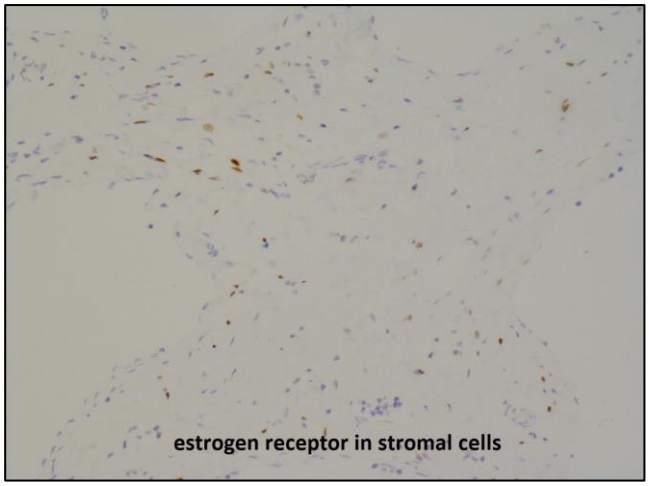

Figure 5: Immunohistochemistry showing estrogen receptors in stromal cells (x200 magnification).

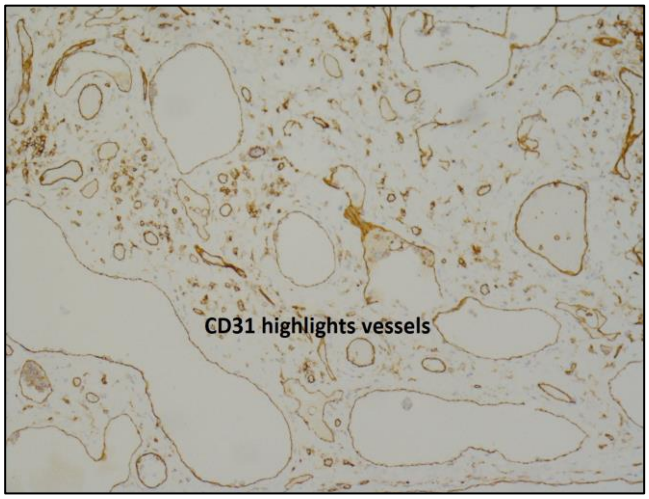

Figure 6: Immunohistochemistry showing CD31 highlighting vessels (x100 magnification).

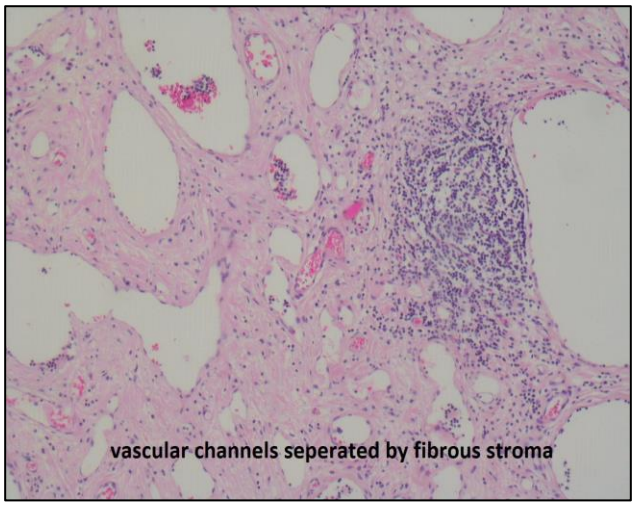

Figure 7: Eosin/ hematoxylin stain showing vascular channels separated by fibrous stroma (x100 magnification).

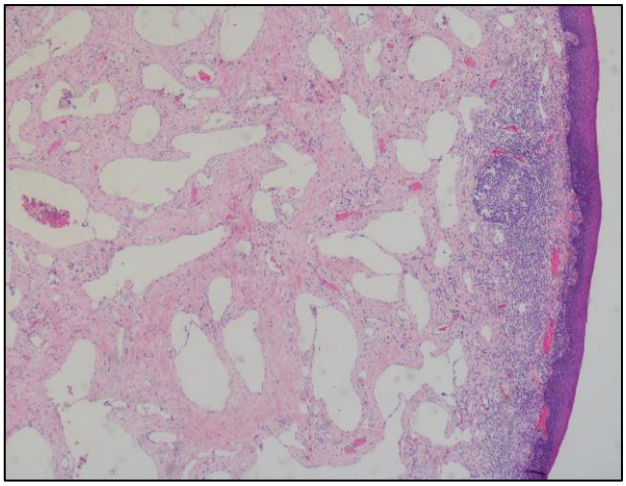

Figure 8: Eosin/ hematoxylin stain showing squamous epithelium lining lymphoid collection and vascular channels (x40 magnification).

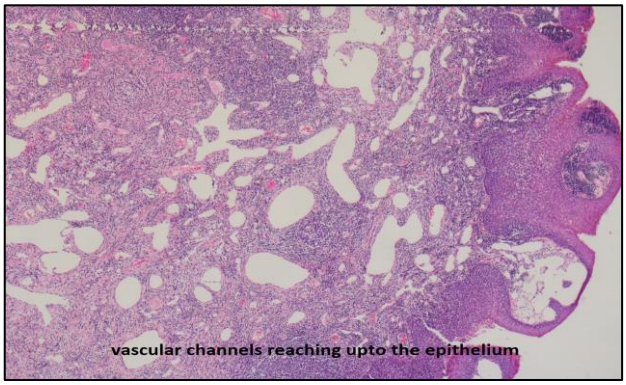

Figure 9: Eosin/ hematoxylin stain showing vascular channels reaching up to epithelium (x40 magnification).

Since, pedunculated polyp was excised along with entire tonsil, no further treatment was indicated. Follow-up of the child did not show any recurrence.

DISCUSSION

Majority of angiofibromas of head and neck originates in nasopharynx. Many theories of origin of NA are postulated like developmental, genetical and hormonal, but none is proved. Since, the tumour exists almost exclusively in males, strong hormonal disorder of pituitary, androgen-estrogen system is suggested. NA has typical clinical and radiological features. Common presentation is a young boy with progressively worsening nasal obstruction and epistaxis.[5] Radiological features are due to the growth pattern of the tumour with extensive vascular component. NA arises from the superior margin of sphenopalatine foramen, extending to nasopharynx and laterally to pterygopalatine fossa. Typical radiological features are strongly enhancing mass in nasopharynx with widened pterygopalatine fossa with erosion of pterygoid process.[6,7] With advancing disease, involvement of nasal

fossa, infratemporal fossa extension and invasion of paranasal sinuses with erosion of walls happen.

ENA are extremely rare and most commonly arises from maxillary sinuses (24.6%).[8] Tumour can originate from ethmoid sinus, nasal cavity, nasal septum, sphenoid sinus, cheek, conjunctiva, oropharynx, retromolar area, middle turbinate and inferior turbinate.[1,3,9-13] Tumours arising in other locations like external ear, external nose, hard palate, lacrimal sac, parapharyngeal region, oesophagus, trachea, middle cranial fossa and infratemporal fossa had been reported.[5,14-16] 4 cases of ENA involving larynx in aryepiglottic fold, vocal cord, interarytenoid region, laryngopharyngeal junction had been reported.[5] Only three cases of ENA involving tonsil had been reported.[5]

In contrast to NA, presentation of ENA is varied and depending upon the site of origin. Tumours arising from nose present earlier due to nasal obstruction, foul smell and epistaxis. ENA arising from larynx present with hoarseness, stridor and dysphagia.[16] Tumours arising from paranasal sinuses may present with pain, cheek swelling, nasal discharge, fever, visual disturbances with eye swelling, headache, nasal obstruction and epistaxis.[17,13]

Characteristic histopathology of NA contains numerous wide, irregular vessels with a single layer of endothelial cells, embedded in fibrous stroma. Extensive vascular component is the reason for profuse bleeding, strong contrast enhancement in CT scan/MRI scan, signal void in MRI and intensive vascular blush of angiography.[6,18] CT/MRI scan with angiography are important for preoperative evaluation. Selective angiography is much more valuable especially for tumour embolization to reduce intraoperative bleeding. Preoperative biopsy is not recommended due to extensive bleeding.

Histopathology of ENA is more varied compared to NA. Predominance of vascular component in fibrous stroma is not seen in all ENA.[16] In our case vascular channels of varying sizes were seen separated by spindly fibrous stromal cells. These stromal cells are hallmark for the diagnosis of ENA and are not expected to be seen in hemangioma or lymphangioma. The stromal cells characteristically express Androgen receptor in 40-75% cases and Estrogen receptor in 0-90% cases.[19] The 70-90% of cases show nuclear positivity for beta catenin.[20] Our case showed positivity for estrogen receptor and androgen receptor, but beta catenin was negative. A vascular hamartoma is also close morphological differential, but no adipose tissue was seen in the stroma. Classical radiological features of NA are not found in ENA. Contrast enhancement is not a constant feature of ENA. Radiological features may differ depending upon the site of origin and extension. Blood supply of ENA depends upon its point of origin, but for NA it is mainly maxillary artery.

Treatment for NA and ENA is surgical resection. Radiotherapy or chemotherapy may be used for unresectable advanced lesions. Majority of ENA in literature are small lesions and easily resectable with minimal bleeding. There is no recurrence of ENA mentioned in literature.[8]

CONCLUSION

Clinical and radiological presentation of ENAs are highly varied compared to NAs, due to the varied site of origin/vascularity. They are less vascular, present in all age groups and in females. This case is being published due to the rarity of the occurrence.

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