

# Juvenile primary extranasopharyngeal angiofibroma, presenting as cheek swelling

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## Abstract

Angiofibroma is a locally advancing immensely vascular tumor that essentially arises from the nasopharynx. The clinical characteristics of extranasopharyngeal angiofibroma (ENA) do not accord to that of nasopharyngeal angiofibroma and can present a diagnostic confront. We describe a case of primary juvenile ENA in a 19-year-old patient who presented with a rapidly enlarging mass of the cheek region. The case is unusual because of its anatomic location. The diagnostic and management particulars are sketched.

**Keywords:** Cheek swelling, extranasopharyngeal angiofibroma, masticator space, vascular tumor

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## INTRODUCTION

Angiofibroma is histologically benign, but locally aggressive highly vascular tumors that virtually always arise from the nasopharynx in the region of the sphenopalatine foramen and pterygopalatine fossa, accounting for 0.05%–0.5% of all head-and-neck neoplasms.<sup>[1]</sup> Angiofibromas have been sporadically described in extranasopharyngeal locations that occur in other areas apart from the nasopharynx. As on 2004, a total of 65 patients with atypical localizations of nasopharyngeal angiofibroma have been reported, with the maxillary sinus being the most commonly involved site.<sup>[2]</sup> The masticatory space is an uncommon location for these tumors, and our research indicates that no case has been reported in that location.

The clinical characteristics of extranasopharyngeal angiofibroma (ENA) do not conform to that of nasopharyngeal angiofibroma. Therefore, they can present

diagnostic challenges. We, therefore, present a rare case of juvenile ENA that was confined predominantly to the masseter muscle region, with neither sphenopalatine foramen nor nasopharynx involvement, presenting as a cheek swelling.

## CASE REPORT

A 19-year-old male patient reported to an outpatient Department of Oral Medicine and Radiology in a dental college and hospital, with a 2-month history of rapidly enlarging painful swelling in the left cheek region. There was a history of trauma on the left side of the face before a couple of months due to an external force and had immediate pain. A small swelling was noticed around 15 days later by his friends and neighbors, and this gradually grew to reach the present dimension. The swelling seems to increase in size after waking up from the bed and while bending downward and mildly reduced in size while under

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heavy work. The patient consulted various local physicians, and there was no reduction in the size of the swelling in spite of taking medications prescribed by them. His past medical, surgical and dental history was not contributory.

On extraoral examination, there was a diffused swelling on the left side of the face [Figure 1], which measured about 8 cm × 6 cm that extended superoinferiorly from temple region to 1 cm short of the base of the mandible and anteroposteriorly 1 cm short of ala of the nose to tragus of ear, which was mildly warm, tender and had variable consistency, i.e. firm to hard in center and soft to firm at the periphery. The right submandibular lymph node was palpable and tender. On intraoral soft-tissue examination, erosive areas were noted on the posterior part of buccal mucosa and vestibule from 25 to 28 regions and obliteration of buccal vestibule with tenderness evident in the 25–28 regions. On hard-tissue examination, grade I mobility was present from 24 to 27.

Routine hematological investigations were found to be within the normal limits.

Ultrasonogram [Figure 2a] of the left cheek showed a hypoechoic focus of size 4 cm in intramuscular plane in masseter muscle extending up to the buccal space. Minimal color uptake was noted on color Doppler.

Contrast-enhanced computed tomography (CT) [Figure 2b] showed a lobulated, 6 cm soft-tissue dense mass in the left masticator space, abutting lateral pterygoid and masseter muscles, with extension craniocaudally from the infratemporal fossa to the buccal space, anteriorly to the lateral wall of orbit and zygomaticus major, medially to the pterygomaxillary fissure and para maxillary space; posteriorly, it is confined within the masseter muscle.



**Figure 1:** Clinical photograph of the patient showing swelling of the left cheek

The mass showed heterogeneous enhancement with minimal central necrosis in the inferior aspect. Bowing of lateral wall of the left maxillary sinus was noted (Holman–Miller sign).

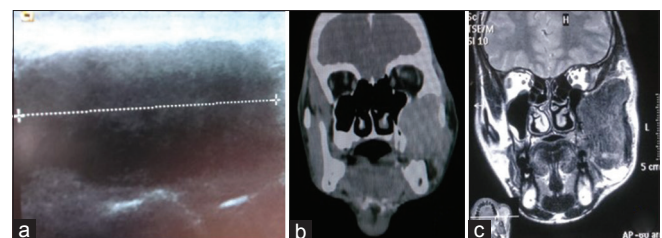
Magnetic resonance imaging (MRI) [Figure 2c] performed a month later showed a relatively well-defined 7 cm isointense mass with T1-weighted images, isotense to hyperintense in T2-weighted images and short tau inversion recovery (STIR). The epicenter and extent of the mass were almost the same as noted in CT. There were multiplanar linear flow voids within the lesion. Postcontrast, lesion shows intense heterogeneous enhancement. On digital subtraction angiography, the left superficial temporal artery was prominent and few branches from it supplied the lesion.

Fine-needle aspiration was nondiagnostic as it showed only formed elements of blood.

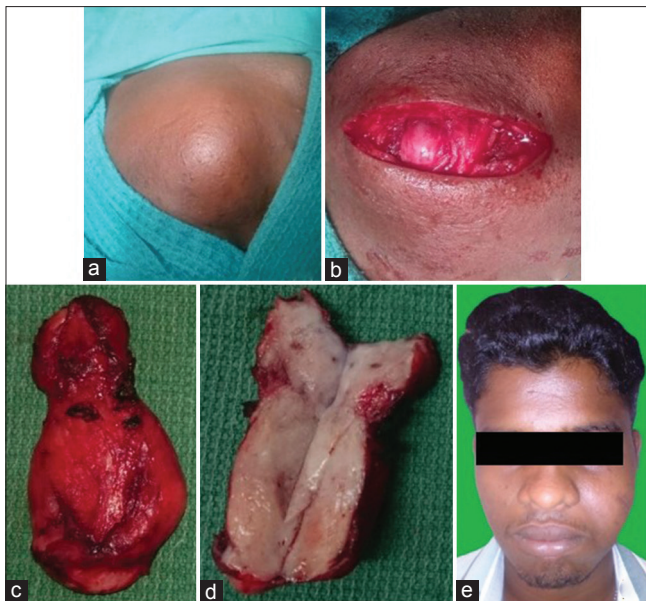
After obtaining the consent, photographs were taken and diagnostic and therapeutic excision was executed on July 2015 under general anesthesia through extraoral approach, and the mass was removed in toto by applying countertraction [Figure 3a-e]. Owing to massive bleeding which was met intraoperatively of about 300 ml, a unit of fresh blood transfusion was made. Postoperative recovery was uneventful.

Histopathological examination showed parts of tumor composed of thin- and a few thick-walled ectatic vascular channels lined by attenuated endothelial cells set in a fibroblastic stroma composed of uniform spindle-shaped cells. There is no increase in mitotic figures or evidence of atypia [Figure 4a and b], and the features were compatible with the pathohistological diagnosis of angiofibroma.

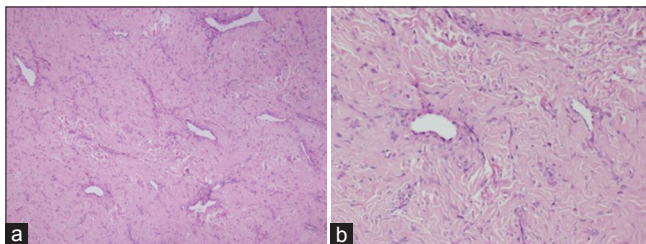
Immunohistochemistry showed positive for vimentin [Figure 5a], and CD 34 highlights the vascular channels [Figure 5b].



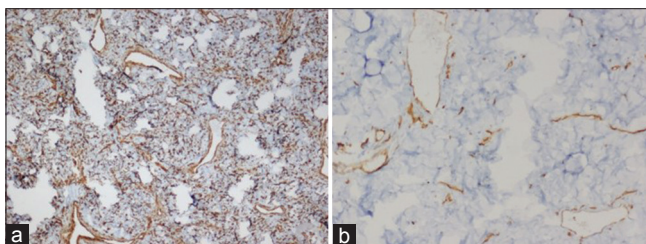
**Figure 2:** (a) Ultrasonogram shows 4 cm hypoechoic foci. (b) Computed tomography shows soft-tissue mass with positive Holman–Miller sign in the lateral wall of the left maxillary sinus. (c) Magnetic resonance imaging shows isointense soft-tissue mass in T1-weighted images in the left masticator space



**Figure 3:** (a) Preoperative patient preparation. (b) Extraoral incision and exposure of the mass. (c) Soft-tissue mass after excision. (d) Cut section of the excised mass. (e) Postoperative profile after 2 months



**Figure 4:** (a and b) Histopathological picture showing fibrous tissues along with scattered vascular channels  $\times 50$  and  $\times 100$ , respectively



**Figure 5:** (a) Immunohistochemical stain of angiofibroma showing vimentin-positive fibroblastic stroma  $\times 100$ . (b) Immunohistochemical stain of angiofibroma showing CD 34 shows positive for vascular channels  $\times 100$

The patient is under regular follow-up and no recurrence was noted.

## DISCUSSION

Majority of the studies have reported that the head-and-neck region is the common site for vascular and fibrous tumors whether benign or malignant. Hippocrates described a vascular tumor that predominantly had fibrous stroma in the 5<sup>th</sup> century BC, but Chauveau first used

the term juvenile nasopharyngeal angiofibroma (JNA) in 1906.<sup>[3]</sup> JNA is a relatively rare tumor with an incidence ranging between 1:6000 and 1:55,000 of the population.<sup>[4]</sup> Although they occur almost exclusively in male adolescent, there have been occasional reports of juvenile angiofibroma occurring *de novo* in adult men and women.<sup>[2]</sup>

Although many theories have been put forward for its origin, current evidence indicates that angiofibroma does not originate in the nasopharynx, as assumed, but rather from a fibrovascular nidus in the posterolateral wall of the nasal cavity in the vicinity of the sphenopalatine foramen.<sup>[5]</sup>

Despite their characteristically benign histopathologic appearance, juvenile angiofibroma tends to be locally aggressive tumors that have a propensity to extend some distance from their primary site of origin from the nasopharynx, nearby the area of sphenopalatine foramen, often encountered extending well beyond the nasopharynx along natural tissue planes, foramina, and fissures that interconnect the defined compartments of the head and neck typically by submucosal extension. Lateral extension from the pterygopalatine fossa into the pterygomaxillary fissure and infratemporal fossa is also frequent whereas intraorbital and superior intracranial invasion is less frequently observed but not uncommon.<sup>[6]</sup>

In a review of 704 cases of angiofibroma by Tasca and Compadretti, 13 cases presented outside the nasopharynx, suggesting extranasopharyngeal possibility.<sup>[7]</sup> In 2004, Windfuhr and Remmert reviewed the literature and compiled 65 cases of ENAs where four cases had an oropharyngeal origin and the maxilla was the most commonly affected site.<sup>[2]</sup> Reports of primary ENAs, especially those originating in the buccal space<sup>[8,9]</sup> and infratemporal space,<sup>[10-14]</sup> have appeared exceptionally in the literature. Besides the different location, Celik *et al.* also opined that typical clinical characteristics of ENAs, such as symptoms, age and sex, do not conform to a great extent with that of nasopharyngeal angiofibroma and henceforth should be referred to as “atypical angiofibroma.”<sup>[15]</sup>

Angiofibroma is composed of a rich vascular network within a fibrous stroma. The smaller vessels in the central part of the lesion typically lack muscular elastic laminae, and the absence of muscular coat contributes to the capacity for massive bleeding that occurs with angiofibroma.<sup>[16]</sup> This feature of massive bleeding in angiofibroma was encountered intraoperatively in our case by blood loss of about 300 ml which necessitated transfusion of a unit of whole blood. There is immunohistochemical evidence

to suggest that juvenile angiofibroma has abundant noradrenergic innervation<sup>[17]</sup> and appears to contain an angiogenic growth factor. Vascularization arises most frequently from the maxillary artery branch of external carotid, with background vascularization arising from blood vessels in the ascending pharyngeal artery and internal carotid artery.<sup>[18]</sup> In our case, the tumor was supplied by the superficial temporal artery.

Although atypical in location (masticator space), our case showed typical and characteristic features and radiologic finding, including strong contrast enhancement on CT and MRI, signal-void areas representing tumor vessels visible on MRI images, as well as intensive vascular blush. CT and MRI scans have been advocated as optimal diagnostic techniques for evaluation of ENAs.<sup>[19]</sup> However, signs of suspected hypervascularity indicate the need for arteriography and carotid angiography before surgical procedures to arrange the necessary precautions, such as embolization, and reduce the risk of brisk bleeding during tumor removal.

Although surgical approach with preoperative transarterial embolization through super-selective catheterization of supplying arterial pedicles with polyvinyl alcohol particles remains the standard conventional endovascular therapeutic approach to these tumors, direct puncture techniques with liquid adhesives injected directly into the tumor neovasculature have been successfully used.<sup>[20]</sup> Other methods of treatment that have been employed are irradiation, hormone therapy, cryotherapy, arterial ligation and use of sclerosing agents. Future therapy for JNA may include intra-arterial immunotherapy and the use of certain agents that inhibit angiogenesis.<sup>[18]</sup>

## CONCLUSION

ENA is a very rare tumor to be found within the masticator space. By virtue of their location, they can have variable presenting signs and symptoms, which can present a diagnostic challenge. However, it must be taken into consideration in the differential diagnosis of cheek swelling suggestive of vascular nature and should be regarded as a potential, though exceptional, site of origin for these neoplasms. A very strong index of suspicion is required to diagnose an ENA.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not

be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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## Conflicts of interest

There are no conflicts of interest.

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