

Angiofibroma masquerading as a cheek swelling - A diagnostic dilemma

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ABSTRACT

Case report presenting an unusual site of angiofibroma is described. Extranasopharyngeal angiofibromas are rare. The most common site of presentation of extranasopharyngeal angiofibromas is the maxillary sinus. We report a case of angiofibroma of the buccal mucosa. The purpose of this article is to put forth an unusual presentation of an Angio fibroma in the oral cavity as reports of such cases involving the oral cavity are very few in literature.

INTRODUCTION

Vascular and fibrous lesions are common in the head and neck area including both benign and malignant subtypes. Angiofibromas are highly vascular benign tumors that characteristically arise within the nasopharynx and are predominantly seen to occur in young adolescent males. Although it is the most common benign neoplasm of the nasopharynx, it accounts for 0.05- 0.5% of all head and neck neoplasms.[6] The term extranasopharyngeal angiofibroma has been applied to vascular, fibrous nodules occurring outside the nasopharynx. As on 2009, 56 extra nasopharyngeal fibromas have been reported, with the most common site of presentation



Fig 1: Gross Specimen

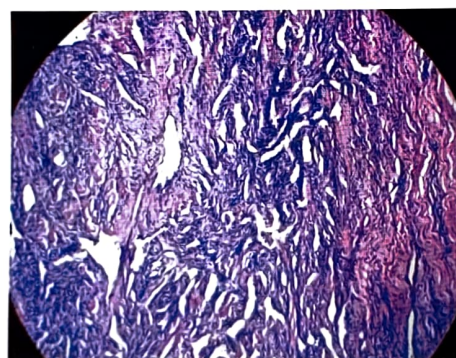


Fig 2: H & E (20X) picture showing numerous vascular channels with fibrocellular background

being the maxillary sinus.[2] The buccal space is an uncommon location for these tumors, with only three previously reported cases of buccal space extra nasopharyngeal angiofibromas.[2,3,5]

The clinical characteristics of extra nasopharyngeal angiofibromas do not conform to that of nasopharyngeal angiofibromas. Therefore, they can present diagnostic challenges. Here, we present a rare case of angiofibroma in the buccal mucosa, thus emphasizing that angiofibroma, although rare and unusual, should be included

in the differential diagnoses of such lesions. Furthermore, it is suggested that angiofibroma should be included as one of the differential diagnosis of soft tissue swellings in the oral cavity.

CASE HISTORY

The patient was a 51 year old male who reported to the institution with the complaint of swelling in the left cheek making it difficult for him to eat. There were no symptoms related to the mass such as pain, rapid increase in size, inability to open the mouth or bleeding. His past medical history revealed that he was a known case of epilepsy for the last 20 years and was on and off the drugs on his own will. The last episode of seizure was about one and half months back. Review of symptoms was negative for relevant constitutional symptoms such as weight loss, fever or loss of appetite. Physical examination revealed a swelling in the left buccal mucosa, about 5x4 cm with slight bluish tinge extending from posterior region of the maxillary tuberosity till about 2-3 cm from corner of mouth. It had a lobulated surface. An intraoral and external examination with bimanual palpation revealed a well defined border, soft to firm in consistency, mildly tender and failed to reveal any obvious breach of the oral mucosa or adherence to the overlying skin, which clinically correlated with a lesion of the buccal space. Examination of the head and neck region, as well as the cranial nerve

examination, was normal.

Further diagnostic work up included a fine needle aspiration cytology, which was non-diagnostic. Roentgenographic examination was not helpful. A ultrasound was done which revealed a mixed heterogenous mass, 2.4x2.8 cm in left cheek deep to subcutaneous plane. A diagnostic and therapeutic excision was planned. The mass was excised via an intra oral approach (**Fig 1**). The final pathology revealed a well encapsulated lesion, periphery of the lesion showed dense vascularity. Numerous vascular channels lined by a single layer of endothelial cells with fibrocellular background were noticed (**Figs 2 and 4**).

The stromal component was made up of spindle type and angulated cells with occasional round cells in the background of collagenous fibrous tissue which showed hyalinization and many bundles of collagen fibres. No inflammatory component was noticed throughout the lesion (Figure 3), the histopathology was suggestive of Angiofibroma.

DISCUSSION

Nasopharyngeal angiofibroma is a well-defined entity sharply localized in time, space, and sex. In 1980, De Vincentiis and Pinelli reviewed a series of 704 cases of angiofibroma and found that 13 cases manifested outside the nasopharynx, thus suggesting that extra nasopharyngeal localization of this tumour is a possible, although rare occurrence.[5] Angiofibromas rarely originate outside the nasopharynx.[6] In 2004, Windfuhr and Remmert reviewed the literature and compiled 65 cases of extra nasopharyngeal angiofibromas in which four cases had oropharyngeal origin and the maxilla was the most commonly affected site.[6] Besides the different location, typical clinical characteristics of extranasopharyngeal angiofibromas, such as,

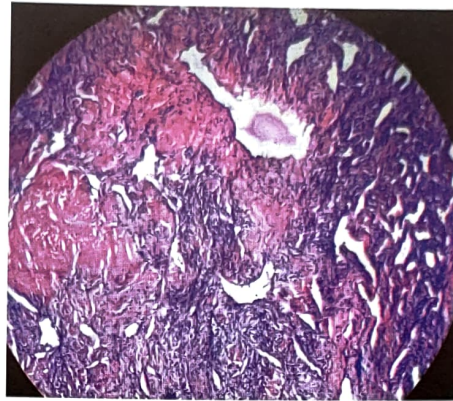


Fig 3: H & E (20X) picture showing background of collagenous fibrous tissue showing Hyalinization

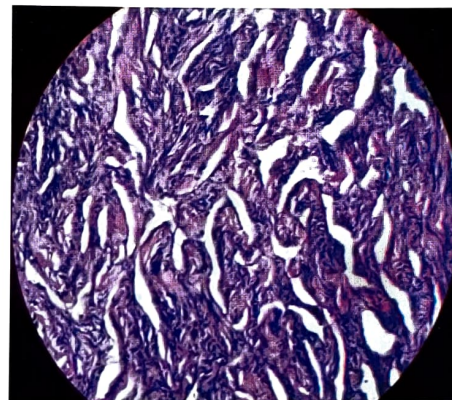


Fig 4: H & E (40X) picture showing numerous vascular channels with fibrocellular background

symptoms, age, sex, do not conform to a great extent with that of nasopharyngeal angiofibroma. This fact has led to doubt as to whether extranasopharyngeal angiofibromas, though structurally similar, should be considered as being different from nasopharyngeal angiofibroma.[4] Recently, Celik et al also proposed that patients, who have different characteristics other than the classical angiofibromas, should be referred to as "atypical angiofibroma". Due to these different features, extranasopharyngeal angiofibromas can present a diagnostic challenge and a meticulous evaluation with a high index of suspicion is essential in establishing the correct diagnosis and treatment. Angiofibromas are histologically composed of a proliferating vascular component set in a fibrous stroma. The

former is characterized by blood vessels of different size and smooth muscle content. The stroma consists of plump spindle, angular or stellate shaped cells and a varying amount of collagen fibers. Immuno histochemical analysis has shown that stromal cells have strong cytoplasmic reactivity for vimentin and are generally immunonegative for smooth muscle actin. For an experienced pathologist although accurate diagnosis of an angiofibroma is not too difficult, but when its location is an extremely rare one, a methodic evaluation and a high index of suspicion are essential in establishing the proper diagnosis and treatment.