International Journal Dental and Medical Sciences Research
Volume 3, Issue 5, Sep-Oct 2021 pp 885-888 www.ijdmsrjournal.com ISSN: 2582-6018

Extra Nasopharyngeal Angiofibroma: A Review

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Submitted: 01-10-2021 Revised: 12-10-2021 Accepted: 15-10-2021

ABSTRACT
The term extranasopharyngeal angiofibroma has been used to define vascular, fibrous nodules occurring outside the nasopharynx. It is quite common in females, as against its other variant, nasopharyngeal angiofibroma. The maxillary sinus is the most common site involved. Computerized tomography scan and magnetic resonance imaging are used to determine the tumour site and its extension. Surgical excision of the mass is the treatment of choice, and recurrence is rare.
KEYWORDS: Extranasophayrngeal angiofibroma, maxillary sinus, vascular tumour, sphenoid bone, epistaxis

I. INTRODUCTION

Nasopharyngeal angiofibroma (NA) is a rare, vascular tumor affecting adolescent males which represents 0.05 % of all head and neck tumors.[1] Angiofibroma originates from the sphenoid foramen at the junction of the root of the sphenoid process of palatine bone, horizontal ala of vomer and pterygoid process of sphenoid bone.[2] When it occurs out of this site, it is termed as extranasopharyngeal Angiofibroma (ENA). This rare tumor usually displays variable clinical presentation and was recently termed as "atypical angiofibroma" due to its distinct characteristic features. Thus, ENA can constitute a challenge in terms of diagnosis and treatment. To our knowledge, there have been only a few ENA case reports in the literature. It is extremely rare in females. Although it sometimes involves the sphenoid sinus, it rarely originates from this site.

II. DISCUSSION

The most common primary extra-nasopharyngeal site for these tumours is the maxillary sinus.[3] Other primary extra-nasopharyngeal sites reported are ethmoid and sphenoid sinuses, nasal septum, frontal recess, middle and inferior turbinates, tonsil, parapharyngeal space, ear, trachea, larynx, middle cranial fossa, infratemporal fossa, tonsil, retro molar region and conjunctiva.[3] The mean age of presentation of the tumour in extra nasopharyngeal sites is 22 years and it occurs more commonly in females.[4] In contrast nasopharyngeal angiofibroma presents almost exclusively in adolescent males with a mean age range between 14 and 17 years; female presentation is very rare.

A case of an extra nasopharyngeal angiofibroma was histopathologically diagnosed in a 15-month-old boy at an atypical site, anterior and medial to the lacrimal sac. The tumour was resected via an endonasal, micro-endoscopic approach avoiding an external incision.

Angiofibromas, in general, have various theories of origin. Ringertz (1938) suggested that it originated from periosteum of nasopharyngeal vault. Bensch and Ewing (1941) thought that it arose from embryonic fibro cartilage between basi-occiput and basi-sphenoid, whereas Brunner (1942) suggested origin from conjoined pharyngobasilar and buccopharyngeal fascia.[5]

Anna et al., presented a case series of 10 patients, wherein 7 patients had nasal origin, nonetheless to say that it the most common site of origin. One patient in this study had a tumor originating from the tonsil, which is quite an uncommon presentation.[6]

Cases of tumors arising in the following locations have been also been reported: external nose, hard palate, external ear, lacrimal sac, carotid bifurcation, esophagus, trachea, facial nerve, middle cranial fossa and infratemporal fossa.[7,8]

Symptoms of extranasopharyngealangiofibromas are numerous and varied, and depends on the site of the tumour. Tumors originating from the nasal cavity will have symptoms characteristic for NA. Nasal obstruction and epistaxis occur with nasal angiofibroma. Less common symptoms include rhinolalia, headache and mucopurulent nasal discharge. Clinical presentation of laryngeal lesions includes hoarsness, dysphagia, dyspnea and stridor[9] ENAs

located in other areas will have more confusing clinical presentation and the delay between the onset of symptoms and the diagnosis may be longer. Tumors originating from one of the paranasal sinuses may manifest with pain, fever, rhinorrhea, swelling of the cheek, proptosis, headache, progressive nasal obstruction, occasional epistaxis.

Angiofibromas are rarely reported in women, most large series from prominent institutions do not contain any cases. The first reported case was by Osbourne and Sokoloski in 1965[10].Massimo ralli et al.,[11] reported NA in an elderly female and concluded that even when the sex and age of the patient are rare for this condition one should warrant suspicion of NA in any patient presenting with a nasopharyngeal mass and recommend of an angiographic examination to prevent dangerous intra- and postoperative bleeding. There are few other reported cases of NA in literature with nearly the same conclusion as above.[12,13]

No attempt should be made to biopsy these lesions in outpatients nor indeed in inpatients as serious bleeding may occur. Diagnosis can be made with almost complete certainity radiologically, which additionally allows accurate assessment of tumour extent and vascularity.

Imaging studies play a major role in diagnosis, staging and management of an angiofibroma. A combination of CT and MRI adds detail to the previously known classical Hollman Miller sign, also known as antral sign. This radiological sign consists of anterior bowing of posterior wall of maxilla sinus seen on lateral plain radiograph and more recently on CT Scan, but this is seen in a classical case of NA rather than ENA. Angiography might be additionally undertaken if it's a large tumour to delineate tumour blood supply. It also allows tumor embolization, which reduces intraoperative bleeding.

According to histopathology ENAs constitute a more heterogeneous group. The predominance of the vascular component in the fibrous stroma characteristic for NA has been stated only in a few patients by few authors.[14]Therefore, classic radiological findings characterizing NAs are not shared by ENAs. Most ENAs enhance after contrast medium injection, however, enhancement is not a constant sign.[15,16] Unlike NAs, radiological presentation of extranasopharyngealangiofibromas is much more variable also due to their various locations. From their point of origin, tumors may spread to adjacent areas by widening of natural foramina and fissures or by erosion of bony structures.Contrast enhanced CT scan (CECT) and magnetic resonance imaging (MRI) are the key investigations to determine tumor site and extension, with special attention on skull base involvement and intracranial spread and relationship to important surrounding structures. Bone erosion can be more easily revealed by CT scan and MRI is generally adequate in demonstrating cortical erosion and cancellous replacement by tumour. [17].T1 weighted MRI show a typical "salt and pepper" appearance caused by the increased vascularity of the tumour.[18]. Alvi et al. considered CT scan to be sufficient for the diagnosis of extranasopharyngeal angiofibroma, as it clearly delineates and identifies the tumour.[19]

Macroscopically, angiofibromas vary from small, smooth multi-lobulated masses varying from grey white to red-purple and from as little as 2-3 cm to 10 cm or more. Microscopically they show varying degrees of blood vessels and fibrous components with the vascular structures ranging widely from capillaries to compressed slit like spaces and ectatic sinusoidal areas.[20]

Histopathology shows richly vascular lesion which has variable-sized thin-walled vessels surrounded by a fibroblastic stroma.[21]. The vessels have a single endothelial cell lining without a muscularis layer. Immunohistochemical staining should be performed for AE1/3, vimentin, SMA, desmin, S-100, CD34, CD31, CD117, CD99 BCL-2, and ki-67. The histomorphology and immunohistochemical staining profile supports the diagnosis of angiofibroma.[22]

Pre operative embolization is generally not required as the size of the tumour is small when detected early and owing to less vascularity compared to NAs. Surgical excision of the mass is the treatment of choice and recurrence rate is generally low if excised in toto.

III. CONCLUSION

In closing our reviewwe would like to cinclude thatextranasopharyngeal angiofibroma is a rare entity. It should always be kept in mind as a differential diagnosis of a nasal mass irrespective of the status of the vascularity, age or sex of the patient until histopathologically proven. Extranasopharyngeal sites should be regarded as potential, though exceptional, places of origin for these neoplasms. Atypical presentations of extranasopharyngealangiofibromas can pose a considerable diagnostic and surgical challenge for clinicians.

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