Extra Nasopharyngeal Angiofibroma Of Anterior Ethmoid In A Middle Aged Woman

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Citation

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Abstract

Angiofibroma is a benign but locally destructive vascular tumor of nasopharynx occurring commonly in adolescent males. Its occurrence in other than this site is known as extra nasopharyngeal angiofibroma (ENA). ENA of head and neck in elderly woman is extremely rare. We present an exceptional case of nasal angiofibroma originating from ethmoid bulla in a 36 years old female presented with nasal block and blood stained nasal discharge. The tumor was completely removed with endoscopic sinus surgery. With best of our knowledge only 8 cases of ENA arising from ethmoid has been reported in English literature, of which only two patients were females.

INTRODUCTION

Angiofibroma is a vascular tumor seen among young boys[1]. The nasopharyngeal angiofibroma (NA) is well known, well described and commonest benign tumor of nasopharynx. It originates from the sphenopalatine foramen at the junction of the sphenoid process of the palatine bone and the horizontal ala of the vomer and the root of the pterygoid process of the sphenoid[2,3]. Its occurrence in other than this site is known as extra nasopharyngeal angiofibroma. The ENA is extremely rare clinical entity in both sex and all ages with variable clinical presentation. Its clinical characteristic is not identical to the NA in all cases[4,5].

There are very few reported cases of ENA in English literature. Its occurrence in female is extremely rare[2]. We report a rare case of ENA of left nasal cavity originating from the ethmoid bulla in a middle aged female. The tumor was treated successfully with endoscopic sinus surgery. With best of our knowledge only 8 cases of ENA arising from ethmoid has been reported in English literature, of which only two patients were female.

CASE REPORT

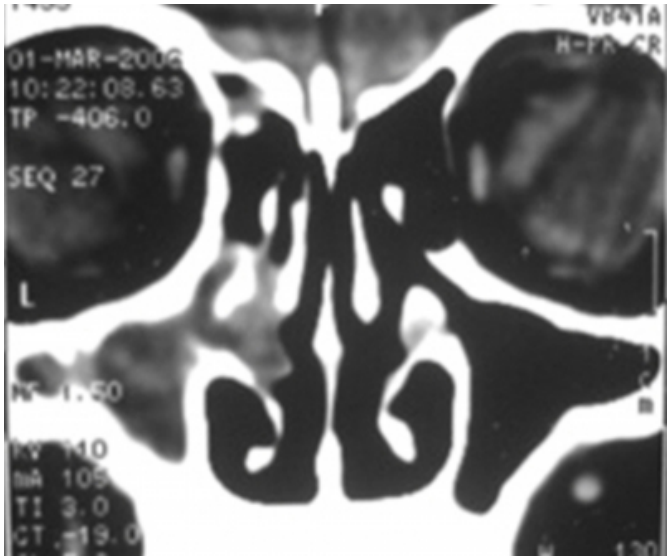
A 36-year-old female presented to the out patient Department of B P Koirala Institute of Health Sciences, Dharan, Nepal, with 5-months history of left-sided nasal obstruction and a 3-months history of blood stained mucoid nasal discharge. There was no past history of any other medical and surgical illness except the history of allergic nasal rhinitis.

Examination of the nose revealed no external nasal deformity. The spatula test showed left sided decrease nasal patency. Anterior rhinoscopy discovered a grayish white colored, smooth, round, polypoidal mass occupying left side of nasal cavity in middle meatus area. Posterior rhinoscopy was normal in appearance. The results of blood, urine examination and X-ray result of the chest were within normal limits. The rest of the ear, nose, throat, head, neck and systemic examinations were unremarkable. On the basis of the above findings a pre-operative provisional diagnosis of ethmoid polyp was made.

The patient was advised for computed tomography (CT) scan of nose and paranasal sinuses, which revealed soft tissue density lesion along the lateral nasal wall on the left side at the region of middle meatus. The lesion appeared to be arising from the anterior ethmoid and extended along the widened infundibulum in to maxillary sinus laterally, and the nasal cavity anteriorly (Figure1). There was no orbital, posterior ethmoid, intracranial and nasopharyngeal extension.

Figure 1

Figure 1: Computed tomographic scan of nose and paranasal sinuses showed soft tissue density lesion along the lateral nasal wall on the left side at the region of middle meatus.

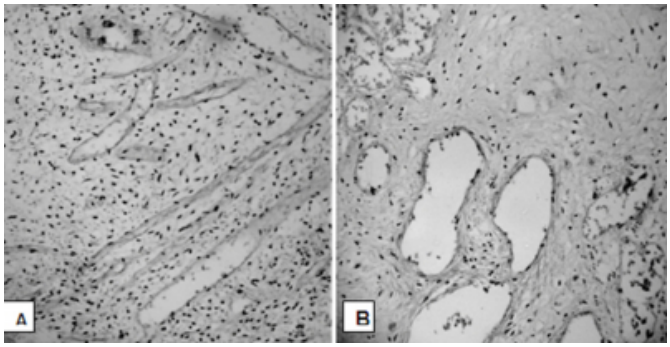


Under general anesthesia, the endoscopic uncinectomy, ethmoidectomy, middle meatus antrostomy was performed in left side. The specimen was sent for histopathological examination. The tumor was attached to the ethmoid bulla extending to infundibulum. The maxillary antrum also contained mucoid collection with healthy looking mucosa. The bleeding during procedure was found to be more than the usual sinus surgery for polyp. Total blood loss was about 150 ml. There were no other complications of surgery and anterior nasal packing was done with ribbon gauze soaked with antibiotic ointment. The pack was removed after 48 h. Post operative recovery was uneventful. Patient was discharged after three days with advice to follow-up after two weeks with histopathological report.

Histopathological report surprisingly showed dilated, cavernous vascular spaces lined by endothelial cells, separated by fibrostroma with stromal cell nuclei. These findings were compatible with a diagnosis of angiofibroma (Figure 2 A, B). The subsequent follow-ups for past six months revealed well healed, epithlialized cavity with no sign of recurrence.

Figure 2

Figure 2: Nasal angiofibroma showing loose fibro-collagenous tissue with interspersed slit-like and gaping vascular channels. [H&E, 20x, 40x]



DISCUSSION

Angiofibroma is the commonest tumor of nasopharynx account for 0.5% of all head and neck tumors[1,4]. It is an age, sex and site linked disease of human being classically appear in the nasopharynx of a young boy in first or second decades of life[2,4]. Isolate ENA is a rare clinical entity. Windfuhr J P and Remmert S reviewed 65 cases of extra nasopharyngeal angiofibroma from published international literature[3]. The mean age of presentation was 22.9 years with oldest of 79 years. They observed only 17 (26%) female patients in their study. Huang et al has done similar review of published literature over 55 cases and got similar results[2]. Thus, the ENA is distinct from NA as it occurs in slightly older age and also has been reported in females. The present case is of a report of ENA in 36 years old female.

The commonest extra nasopharyngeal site of angiofibroma is maxillary sinus followed by ethmoid sinus and sphenoid sinuses[2]. The other reported sites are nasal cavity, nasal septum, larynx, external ear, cheek, conjunctiva, oropharynx, hypopharynx, retro molar area, middle and inferior turbinate. An isolate case has been published in the trachea, lacrimal sac, carotid bifurcation, esophagus, hard palate, facial nerve, external ear, external nose, and infratemporal fossa[2,3,4]. In our patient, the tumor originated from the ethmoid bulla.

A number of theories have been propounded from time to time to explain the etiopathogenesis of nasopharyngeal angiofibroma, such as developmental, hormonal and genetic[1, 3]. The concept of the origin from the fascia basalis is only implicable for those arising nasopharynx [5]. Its abnormal extension to the septum through perpendicular plate of ethmoid was speculated as the tissue of origin for septal angiofibroma. The presence of this tumor in the place where no fascia basalis is found indicated that the origin may

be from ectopic tissues located further away to the usual place[1,3,5].

The clinical manifestation is variable and depends on the site of involvement[6]. They mimic various other types of lesion and lack the classical clinical features of angiofibroma. Clinically, the lesion appears as a reddish-purple nodular mass[1]. The nasal obstruction, recurrent epistaxis and deformities are the sequential presentation of sino-nasal and nasopharyngeal angiofibroma[2,4,6]. The tumor of an intranasal site may present earlier due to the limited space in nasal vault. On the other hand, the presentation of the maxillary sinus tumor is insidious[2]. In our patient, the main complaints were that of left sided nasal obstruction and blood stained nasal discharge. The mass was grayish white in color.

Radiological evaluation of the region involved is crucial for establishing diagnosis, to identifying the feeding vessel, and to determine extent of the lesion and to make the treatment plan including selective arterial emobilization[1,3,4]. The Computerize tomography, Magnetic resonance imaging, and angiography are the prime radiological modalities available for such evaluation of any angiofibroma[1,2]. The selective angiography has been considered as the useful investigation procedure. It clearly demonstrates the blood flow dynamics and vascular pattern of the tumor. However, the absence of hypervascularity in angiography does not exclude the ENA[3]. Due to the advantage of clear delineation and identification of the tumor, the CT scan is considered being the sufficient for the diagnosis of ENA[3,6]. It is readily available and most economical all other radiological investigations in establishing the diagnosis. Our patient was poor and could not afford to pay for the MRI and angiography. Although the incision or punch biopsy is consider as frequent pre treatment diagnostic procedure in most of the head and neck tumors. It is not frequently recommended in case of angiofidroma with the fear of life threatening bleeding[3]. Our clinical diagnosis was ethmoidal polyp. The mass was small in size and confined to the limited areas of nose and paranasal sinuses without any orbital and intra cranial invasion. Thus, we prefer to do endoscopic sinus surgery.

Histologically, it is benign and unencapsulated neoplasm composed with rich vascular network and fibrous stroma. The vascular space is classically lined by endothelial cells and separated by dense fibrous stromal cells with lack of muscular layer. The proportion and cellularity of each component is highly variable[1,5].

The recommended treatment of choice for angiofibroma is the complete surgical excision of the lesion. However, successful endoscopic excision of nasal ENA has been reported in English literature[1,5,7]. The other treatment modalities include radiotherapy, cryosurgery, embolization, hormone therapy, chemotherapy, arterial ligation, sclrotherapy and watchful observation with the hope of spontaneous regression. Radiotherapy for ENA is however not as effective as compaired to NA[3].

Our case was unique in view that the patient was a middle aged female and site of origin of the mass was ethmoid bulla. The scanty blood mixed nasal discharge is an unusual presentation of angiofibroma, which usually present with moderate to massive bleeding. The mass was completely and successfully removed by endoscopic sinus surgery. The clinical presentation, radiological features and unusually little amount of bleeding during surgery never made a suspicion of angiofibroma till the appearance of histopathological features. The unusual presentation of this case might make the first move to discuss that the ENA and NA are the same or different clinical entity.

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