Extranasopharyngeal Angiofibroma in an Adult: A Rare Presentation

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Abstract Juvenile nasopharyngeal angiofibromas are benign but locally aggressive tumors of nasopharynx seen exclusively in adolescent males. Angiofibromas arising in sites other than nasopharynx is very rare. Maxilla is the most frequent site of origin. Extra nasopharyngeal angiofibromas of adult is extremely rare. We present a case of extra nasopharyngeal angiofibroma arising from maxillary sinus in an adult patient which was excised successfully.

Keywords Angiofibroma · Adult · Maxilla

Introduction

Angiofibromas are well documented benign tumors of the adolescent males. They account for .5% of all head and neck neoplasm, usually arising in the nasopharynx. However, angiofibromas rarely may be encountered in sites other than nasopharynx [1]. On review of world literature we found 65 cases of extranasopharyngeal angiofibromas. Maxilla was the most common site, followed by ethmoids, nasal septum and nasal cavity [2]. A high level of suspicion is needed for prompt diagnosis and treatment of such lesions.

We report here an unusual case of adult extranasal angiofibroma, originating from the maxillary sinus which was successfully removed in toto.

Case Report

A 28 year old male was referred to the department of ENT, PGIMER, Chandigarh, India, with history of profuse bleeding during Caldwell luc procedure. The patient first presented to the Government hospital, Patiala with complaints of gradually progressive swelling of the right side of cheek and mild protrusion of right eye for 1 month. He also complained of mild dull aching pain over the right side of cheek. There was no history of nasal bleed, nasal obstruction, nasal discharge or loss of sense of smell abandoned. The patient underwent Caldwell luc procedure during which he has profuse bleeding. The maxilla was packed and procedure abandoned. He was subsequently referred to our centre. After admission, a complete ENT examination was done which included anterior and posterior rhinoscopy which was normal (Figs. 1, 2).

CECT nose and paranasal sinus was done which revealed contrast enhancing mass involving right maxillary sinus with erosion of the anterolateral and posterior wall of maxilla and extending superiorly into the orbit with erosion of inferior orbital wall. The posterior wall of maxilla was pushed posteriorly with erosion at some places. Nasal endoscopy and biopsy was done local anaesthesia, there was pinkish mass in the middle meatus, biopsy as taken which was reported as inflammation, nonspecific.

The excision of the mass was done by lateral rhinotomy approach using Weber Fergusson incision. A 5 × 4 cm pinkish mass of variegated consistency was removed from the right maxilla and nasal cavity in toto. The origin of tumor was traced and nasopharynx was found to be free from tumor. The tumor was seen to be attached to lateral wall of maxilla and had expanded the whole of maxillary sinus including its posterior wall. Blood loss was approx 100 ml.

This material has never been published and is not currently under evaluation in any other peer-reviewed publication.

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Fig. 1 CECT scan of nose and PNS—showing contrast enhancing mass in the right maxillary sinus with destruction of anterolateral wall and medial wall of maxilla

Fig. 2 Postoperative scan—no mass lesion seen

Histopathological examination of excised mass showed sub-epithelium containing numerous vascular spaces of varying sizes lined by endothelium intricately admixed with fibrous stroma. The intervening stroma contained haphazardly arranged spindle cells scattered in loose myxoid stroma. Histopathological diagnosis of angiofibroma was made.

Patient came for follow up 2 months after surgery. Patient is asymptomatic. A repeat CT scan didn't show no residual lesion.

Discussion

Juvenile nasopharyngeal angiofibromas occur exclusively in adolescent males (10–20 years). The origin is believed

to be a vascular nidus in the posterolateral wall of nasal cavity near superior margin of sphenopalatine foramen [3, 4]. The origin of extranasopharyngeal angiofibromas is outside this area. On review of world literature we found 65 cases of extra nasopharyngeal juvenile angiofibromas. In 1980 De Vincentalis and Pinelli reviewed 704 cases of angiofibromas; 13 cases were extranasopharyngeal in origin. Maxilla was found to be the most common site (26.4%), followed by ethmoid, nasal cavity and nasal septum [5]. Habert (1964) recorded eight cases of extranasopharyngeal juvenile angiofibromas, seven cases had origin in the maxillary sinus [6]. Chakrabarty et al. suggested that maxillary extra nasopharyngeal angiofibromas may be caused by unequal development of skull bones coupled with sex and age of puberty [7]. Windfurh et al. reported swelling of cheek, epistaxis and nasal obstruction to be the commonest presentation in maxillary sinus angiofibromas [2]. However, our patient had no episode of nasal bleed. The absence of bleeding in our patient can probably be due to excess fibrous element in the tumour. A very strong index of suspicion is required to diagnose an extranasopharyngeal angiofibroma. Computed tomography scan and MRI are sufficient to diagnose such lesions. Nasal biopsy is to be avoided as may cause profuse bleeding. Surgical excision is the treatment of choice for such tumors.

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