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Original Research Paper

Pathology

A CLINICOPATHOLOGICAL STUDY OF NASOPHARYNGEAL ANGIOFIBROMA

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ABSTRACT

Juvenile nasopharyngeal angiofibroma is a rare, histologically benign and a highly vascular tumour. It is aggressive in nature and usually affects young, adolescent males. The tumour has a tendency for massive bleeding even with minimal manipulation. Nasopharyngeal angiofibroma needs to be differentiated histopathologically from other vascular lesions like lobular capillary hemangioma and hemangiopericytoma.

A retrospective and prospective study of 70 cases of nasopharyngeal angiofibroma was carried in one of the teaching institutes in Mumbai, over a period of 8 and half years. Fisch classification was used to classify the cases clinically. Criteria outlined in the WHO classification of Head and Neck tumours 2017 was used for the diagnosis of nasopharyngeal angiofibroma.

All patients were male except for one female patient and 60 of the cases presented in the second decade. Radiology findings were available in 35 cases and 20 of these were categorised in stage I according to Fisch classification. Histologically it showed a highly vascular tumour composed of cellular fibrotic stroma and collagen along with variably sized vessels. Surgical excision is the primary treatment with incomplete resection being the major cause of recurrence.

Nasopharyngeal angiofibroma is a rare, benign vascular tumour occurring in young males with recurrent biological behaviour and needs to be differentiated histopathologically from other vascular and benign nasal polypoidal lesions.

KEYWORDS : angiofibroma, vascular tumour, young male

INTRODUCTION

Juvenile nasopharyngeal angiofibroma is a rare benign tumour and accounts for less than 1% of all head and neck neoplasms[1]. Histologically it is a benign fibrovascular lesion arising in the nasopharynx of prepubertal and adolescent males with a median age of diagnosis being 15 years[2].The tumour mostly arises from the posterolateral wall of nasopharynx adjacent to the sphenopalatine foramen and may extend to the paranasal sinuses, the orbit and the pterygopalatine fossa[3].It derives its blood supply mostly from the internal maxillary artery. It exhibits a strong tendency to bleed and majority of patients present in late stage with symptoms of nasal obstruction, epistaxis and rhinorrhoea. With the tumour expanding to surrounding areas, frog face deformity can be seen along with proptosis[4].

Unilateral nasal obstruction, epistaxis and mass in nasopharynx form a classic triad for diagnosis of nasopharyngeal angiofibroma[3]. CT and MRI help in diagnosis and establish the extent of tumour and degree of surrounding tissue destruction. Radiologically, an anterior bowing of the posterior wall of the maxillary sinus, can be seen in most patients[1].

Grossly the tumour ranges from 3 to 5 cm in dimension and appears encapsulated, lobulated, rubbery- firm mass with a sessile or pedunculated base. Histology shows abundant fibrous stroma along with plump stellate shaped fibroblasts surrounding numerous blood vessels lined by plump endothelial cells but having very little or no smooth muscle or elastic fibers. This lack of muscle contributes for massive haemorrhage in the tumour caused by even minimal manipulation[5].

Among the various classification systems used for nasopharyngeal angiofibroma, the Fisch classification is the most widely used which helps in deciding the appropriate surgical approach[6].

The presence of androgen, testosterone and dihydrotestosterone receptors with the absence of estrogen and progesterone receptors correlates with the epidemiological data of tumour having a predilection for adolescent males[7][8].

Surgical excision remains the preferred treatment for JNA with concomitant use of diagnostic imaging techniques[2]. Extensive haemorrhage and intracranial extension signifies the prognosis which is overall excellent with less than 1% mortality[1].

MATERIAL AND METHODS

A retrospective and prospective study of 70 cases of nasopharyngeal angiofibroma was carried in one of the teaching institutes in Mumbai, over a period of 8 and half years. For clinical records, age, sex, clinical presentation of the patient and investigations including radiological examination were noted. The clinical staging was noted as per Fisch classification based on radiological findings as follows:

TABLE 1: FISCH CLASSIFICATION	
Stage I	tumour limited to nasal cavity with no bony destruction;
Stage II	tumour invading pterygomaxillary fossa and paranasal sinuses with bony destruction;
Stage III	tumour invading infratemporal fossa, orbit and/or parasellar region remaining lateral to the cavernous sinus;
Stage IV	tumour invading the cavernous sinus, optic chiasmal region and/or pituitary fossa.

The tissue specimens were processed routinely after fixing in 10% formalin and stained with haematoxylin and eosin stains.

RESULTS

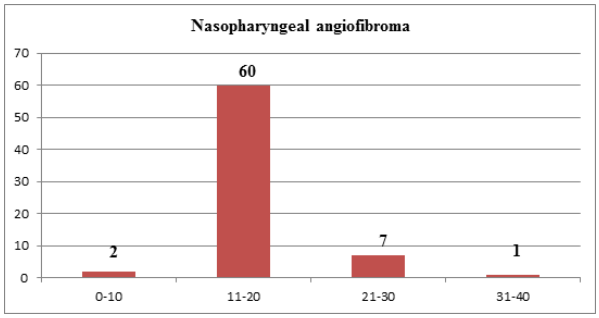

TABLE 2: Age distribution in Nasopharyngeal angiofibroma

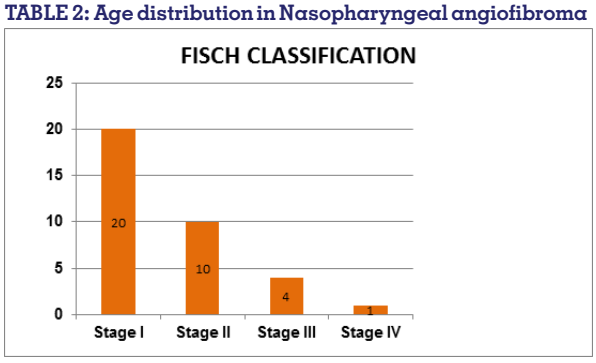

TABLE 3: Fisch classification for staging of Nasopharyngeal angiofibroma

All 70 cases in the study were male except for one 22 years old female patient. The age distribution ranged from 1st decade to 4th decade with 85.71% cases (60 cases) being in the second decade. In fact, the oldest patient who presented at the age of 39 years had a recurrent lesion with long standing history. The site involved in all the cases was nasopharynx with nasal obstruction and epistaxis being the most common symptom. Radiological findings were available in 35 cases and these details were used to stage the tumour according to the Fisch classification of staging. Majority of these cases (20 cases) were limited to the nasal cavity without any bony destruction and were thus categorised in stage I.

Histological examination showed a richly vascular neoplasm with a variable amount of cellular fibroblastic stroma and collagen. There were variably sized vessels lined by plump endothelial cells with little smooth muscles. The vascular spaces seen are usually defects in the stroma lined by single layer of endothelial cells but thick walled blood vessels devoid of elastic tissue may also be present. 11.42% (8) of our cases showed presence of thick walled blood vessels. Multinucleated stromal cells may be seen in nasopharyngeal angiofibroma and cells resembling the ganglion like cells of proliferative myositis may also be found. Such multinucleated stromal cells were seen in 4.28% (3) of cases whereas ganglion like cells were evident in 14.28% (10) of cases.

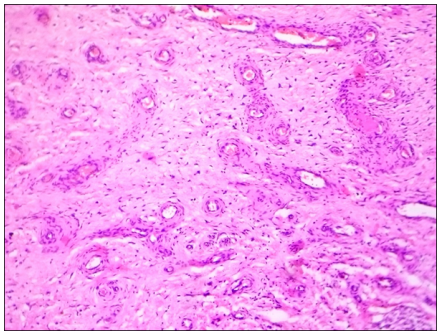

FIGURE 1: Nasopharyngeal angiofibroma histology showing cellular fibrotic stroma and collagen along with variably sized blood vessels.

DISCUSSION

Nasopharyngeal angiofibroma is a rare tumour accounting for <1% of head and neck tumours and has a strong tendency to bleed. It occurs exclusively in pre pubertal and adolescent males with rare case reports of females being affected are published in literature[9][10]. The occurrence of any nasopharyngeal angiofibroma in females is so rare that some authors believe that sex chromosome studies are indicated if this diagnosis is confirmed in a female, considering the hypothesis of a female phenotype with a 46, XY genotype, as in male pseudohermaphroditism[11].

The increase in circulating hormones during puberty results in the pathogenesis of nasopharyngeal angiofibroma owing to the presence of androgen, testosterone and dihydrotestosterone receptors in the tumour cells[7]. Consistent with these findings many attempts at use of hormonal therapy have been made. Recently, Thakar et al.[12] observed that prepubertal and postpubertal patients have different responses to flutamide and that only postpubertal patients demonstrated flutamide-induced partial regression of nasopharyngeal angiofibroma. Gates[13] in a pilot study demonstrated hormonal reduction of nasopharyngeal angiofibroma with the use of flutamide which acts by interfering with binding of testosterone.

Epistaxis and nasal obstruction are the most common presenting complaints encountered by the patients. Clinical aggressiveness and rapid growth of the tumour results in facial deformity and may result in secondary bone resorption due to compression of adjacent bony structures leading to bone destruction[11].

The tumour comprises of blood vessels of varying calibre admixed in a myxoid to densely arranged fibrous stroma with some vessels comprising of thick muscular walls. Compressed small vessels are usually seen in the relatively cellular areas. Stellate and angulated fibroblasts and some giant, binucleated cells are commonly seen in histology[11]. Nasopharyngeal angiofibroma needs to be differentiated from other vascular lesions like lobular capillary haemangioma, hemangiopericytoma and solitary fibrous tumour. Benign lesions like inflammatory polyps or antrochloanal polyp with stromal atypia also needs to considered as differential diagnosis. However distinct radiological features combined with appropriate age and sex of the patient points towards a diagnosis of nasopharyngeal angiofibroma[1]. Nasopharyngeal angiofibroma shows high vascularisation which is also confirmed by immunohistochemistry. This vascularisation is directly related to the vascular endothelial growth factor (VEGF) associated with the proliferation and high vascular density of the tumour regions. It has been found localized on both endothelial and stromal cells. However, neither the proliferative index nor VEGF expression in cases of nasopharyngeal angiofibroma seem to correspond in any relation to its degree of aggressiveness[14].

The mainstay of treatment consists of complete surgical excision. Preoperative angiographic embolisation is recommended as an important adjunct to surgical removal[2]. Incomplete surgical resection is the major cause of recurrence in cases of nasopharyngeal angiofibroma which are further treated with surgery or radiation therapy[1]. The recurrence rate of nasopharyngeal angiofibroma is about 20%[15]. Regardless of its aggressive nature, the mortality rate continues to be less than 1%[16].

CONCLUSION:

Nasopharyngeal angiofibroma is a rare, benign vascular tumour occurring in young males which needs to be differentiated histopathologically from other benign nasal

polypoidal lesions considering its recurrent biological behaviour.

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