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Nasal Angiofibroma - A Rare Presentation

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Abstract

Angiofibroma is a fibrous and vascular tumour that commonly occurs in adolescent males and involves the nasopharynx. Hence the name juvenile nasopharyngeal angiofibroma. The incidence of juvenile angiofibroma is less than 0.5% of head and neck tumours. Radiological evidence, drawn from over 20yrs, reveals the probable origin of this tumour to be the recess behind the pterygopalatine ganglion at the exit aperture of the pterygoid canal. It develops almost exclusively in adolescent males, though there are reports of this tumour being found in children, the elderly and pregnant women. We report a rare presentation of a middle aged female patient who came with right sided nasal mass which was found arising from nasal septum and was diagnosed as angiofibroma on postoperative histopathological evaluation.

Introduction

Nasopharyngeal angiofibroma is a slow growing vascular tumour arising in the sphenopalatine foramen. The tumour expands laterally via the pterygopalatine fossa to the infratemporal fossa, nasopharynx and choanal space and thereby may extend into the anterior nasal cavity. It can have intracranial extensions into middle and anterior cranial fossa. Angiofibroma arising from sites other than nasopharynx have rarely been reported.[1] Here is a case of 50 years old female with a septal mass which turned out to be angiofibroma on histopathology.

Case Report

A 50yr old female patient came with complaints of right sided nasal obstruction and recurrent spontaneous epistaxis since 6 months. Blood loss of about 10-15 cc was reported every time. Three months later she noticed nasal mass on the right side. On anterior rhinoscopy, a pinkish brown mass was seen occupying the whole of right nasal cavity. It was soft to firm in consistency. On probing, there was minimal bleeding and the probe could be passed all around except medially. Left nasal cavity was normal. Posterior rhinoscopy and paranasal sinus examination was normal. CT scan was suggestive of a soft tissue mass anteriorly in right nasal cavity. [Fig 1 and 2] There was no evidence of extension of mass in nasopharynx or sinuses. There was no evidence of bony erosion. Patient's haemoglobin was 7.5gm% for which 3 pints of blood was transfused to bring haemoglobin levels to 10gm%.

Endoscopic excision of nasal mass was done under GA. The mass was confined to the right nasal cavity and was arising from the nasal septum. The mass appeared firm in consistency and was removed in toto with cauterisation of base. Intraoperative bleeding was minimal and anterior nasal packing was done after excision of the mass. Nasal pack was removed after 48 hours. Postoperatively, patient experienced great improvement in symptoms. There was no epistaxis or nasal blockage thereafter. Surprisingly, the histopathology was suggestive of angiofibroma which showed loose fibrous stroma with scattered blood vessels. One year after surgery the patient showed no signs of residual disease or recurrence.

Discussion

Angiofibroma accounts for less than 0.5% of all head and neck tumours. It develops almost exclusively in adolescent males, though there are reports of this tumour being found in children, the elderly and pregnant women.[3] 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.[4] Recent immunocytochemical techniques have been used to show that androgen receptors are present in atleast 75% of tumours, these receptors being present in both the vascular and stromal elements. A much smaller proportion of tumours also have some progesterone receptors. In contrast, oestrogen receptors have not been demonstrated. [5]

There are various theories to explain the etiology of nasopharyngeal angiofibroma. They postulate that nasopharyngeal angiofibroma (a) arises from the periosteum of bones developing from the embryonic occipital plate, (b) is a distinct type of hemangioma, (c)

is a desmoplastic or connective tissue response of the nasopharyngeal periosteum to a hamartomatous ectopic nidus of vascular tissue, probably of the IT type, and (d) is a specific type of fibromatosis.[4] Recurrent severe epistaxis accompanied by progressive nasal obstruction are the classical symptoms of juvenile angiofibromas at the time of presentation. The pathology behind this is that the vessel walls lack elastic fibres and have incomplete or absent smooth muscle that will account for their tendency to bleed.[6]

As it grows, the tumour extends into the nasopharynx, nasal cavity , pterygopalatine and infratemporal fossae. Larger tumours can involve the orbit and cavernous sinus. Angiofibroma is locally invasive, though few have been reported to behave in malignant fashion.[3] The investigations required in a clinically suspected angiofibroma are CT and angiography. A trans-nasal biopsy is avoided as it can provoke brisk haemorrhage. Diagnostic angiography is undertaken to evaluate the blood supply to the tumour and as a prelude to selective embolization [3]. Preoperative embolisation is used to reduce intraoperative bleeding.[1] Several staging systems have been proposed but that of Fisch is the most robust and practical. It defines clearly which tumours can be resected by endonasal techniques and those that would be better tackled by more open or infratemporal fossa/neurosurgical approaches.[7]

Endoscopic sinus surgery techniques have advanced significantly over the past two decades and have allowed the intranasal access and nasal anatomy to be approached in an extremely competent fashion. Several authors have published their experience of successful endoscopic resection of such tumours. Several reports of endoscopic resection have reasonable follow-up durations (13 patients in 23 months) and low recurrence rates (8%) [1].

The patient who presented to our hospital was middle aged female who presented with right sided nasal mass and epistaxis. Endoscopic excision of the nasal mass arising from the nasal septum was done. The mass was limited to the right nasal cavity without eroding the nasal septum or the lateral wall of the nose. The nasopharynx was normal. The histopathology of the mass was suggestive of angiofibroma.

Conclusion

A differential diagnosis of angiofibroma should be considered for any patient coming with complaints of nasal mass and spontaneous, recurrent epistaxis irrespective of age, sex, site of origin and extension of the mass.

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Illustrations

Illustration 1

Fig 1: CT scan showing soft tissue mass arising from the nasal septum

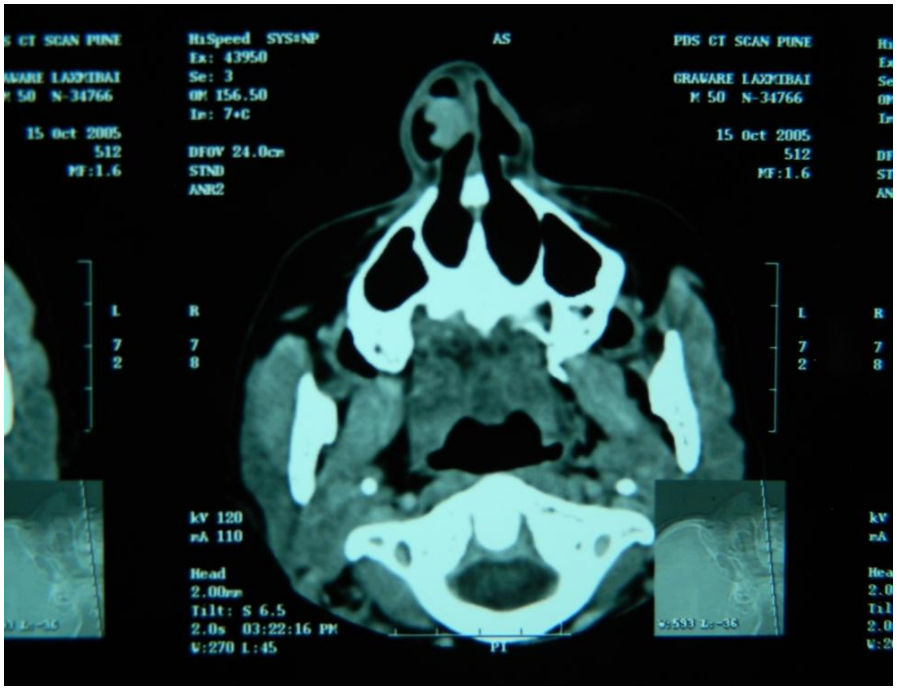
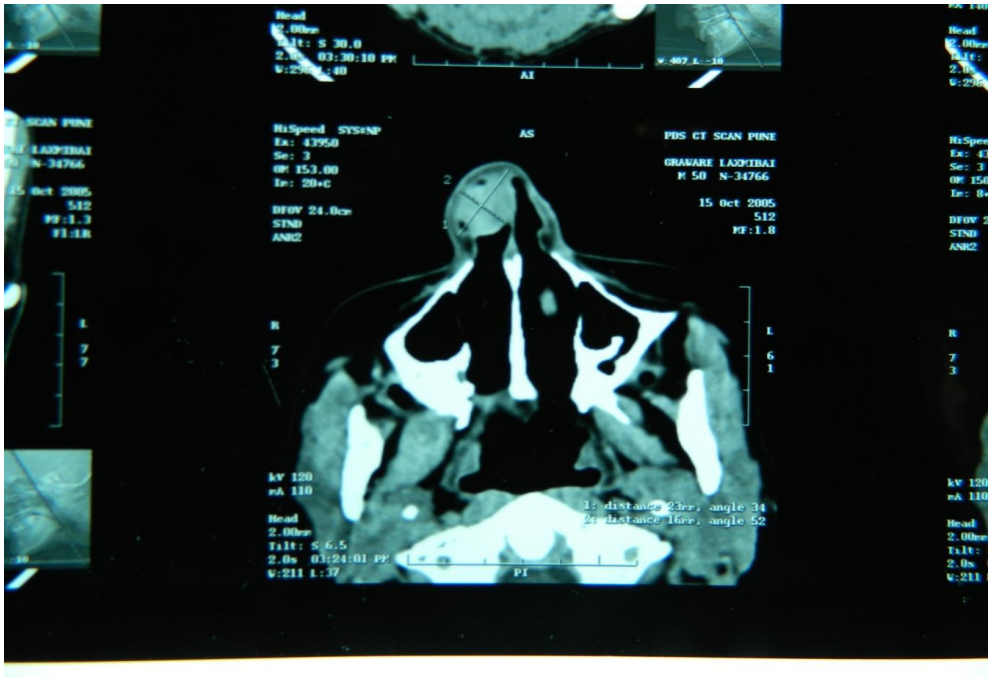


Illustration 2

Fig 2: soft tissue mass occupying anterior aspect of nasal cavity



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