CASE REPORT

Juvenile Nasopharyngeal Angiofibroma

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ABSTRACT

Juvenile nasopharyngeal angiofibroma (JNA) is a relatively uncommon neoplasm occurring almost exclusively in the nasopharynx of adolescent males.1 Occasional cases have extended to involve the oral cavity.1 JNA is a histologically benign yet locally aggressive vascular head and neck tumor which affects almost exclusively adolescent boys. JNA is an uncommon tumor, with reported incidence between one in 5000 and one in 60,000 otolaryngology patients. It is estimated to account for only 0.05% of all head and neck neoplasms, but is nevertheless considered the most common benign neoplasm of the nasopharynx. The diagnosis of JNA is based on history, physical examination and radiographic studies. JNA has several characteristic radiographic features.

A case of juvenile nasopharyngeal angiofibroma with intracranial involvement in a 15-year-old boy, its clinical and radiological presentations are reported in this article.

Keywords: Nasal obstruction, Facial swelling, Juvenile nasopharyngeal angiofibroma.

INTRODUCTION

Juvenile nasopharyngeal angiofibroma (JNA) is a highly vascular, aggressive and locally invasive tumor. This tumor has a high potential to cause serious illness from severe epistaxis, involvement of intracranial structures and high-rate of recurrence.

JNA accounts for 0.05% of all neoplasms of the head and neck. Its incidence is relatively higher in India and Egypt than in the United States and Europe. While JNA occurs predominantly in males, there have been reports of its occurrence in females. Some authorities believe that it occurs only in the male and that if a female is involved, chromosomal studies and a reassessment of the histologic structure are necessary. Intraoral JNA is exceedingly rare, two such cases have been reported so far.2

It is characterized morphologically by irregular, proliferating vascular channels within a fibrous stroma, which consists of plump, spindle or stellate cells. It has a peculiar propensity for local extension into the adjacent tissues that often precludes complete surgical resection and likely is responsible for tumor persistence and recurrences in 21 to 34% of the affected patients.3

Topographically, this tumor frequently originates in the posterolateral wall of the nasal cavity, close to the superior margin of the sphenopalatine foramen. Subsequently, JNA may cause bone erosion and displacement of adjacent structures, thus involving the nasopharynx, paranasal, ethmoidal and maxillary sinuses, and orbit and skull base, with possible intracranial extension.3

The following case instills a fear that we need to be aware of this benign rare disease and it should be diagnosed at an early stage to prevent its extension which makes the tumor resection highly difficult.

CASE REPORT

A 15-year-old male patient visited the department of oral medicine and radiology with the chief complaint of swelling on his right side of face since 6 months. On eliciting the complaint, it was found that patient was apparently alright 6 months back when he noticed a swelling which was small initially and gradually increased to attain the present size (Fig. 1). Patient also gave the history of nasal congestion,
frequent episodes of epistaxis, rhinorrhrea and slight difficulty in breathing and swallowing.

The medical, family and personal history were non-contributory.

The general physical examination revealed a moderately built and nourished male patient with gross asymmetry of right side of the face due to presence of a diffuse dumbbell shape swelling, extending superiorly up to the right corner of the eye, inferiorly upto the lower border of mandible, anteriorly up to the ala of nose pushing the nose to left side of face and posteriorly upto the tragus of the right ear. On palpation, swelling was lobulated, nontender, rubbery in consistency, free from overlying skin and fixed to the underlying tissue. A purplish pink soft tissue mass with blood tinged discharge was observed in right nasal vestibule (Fig. 2).

Intraoral examination revealed bulging of right buccal mucosa and presence of a soft tissue mass in the oropharynx just behind the palatoglossal fold measuring around 2 cm in diameter. Hard-tissue examination showed no abnormality except the presence of a grossly destructed 46.

Based on the history and clinical examination a provisional diagnosis of vascular malformation was given and the following lesions were thought of while listing differential diagnosis; hemangioma, AV malformation, nasopharyngeal cyst, juvenile angiofibroma, lymphangioma, rhabdomyosarcoma.

On radiographic examination, orthopantomograph revealed a diffuse hazy radiopacity in the right maxillary sinus area with expansion of pterygomaxillary fissure (Fig. 3), while paranasal sinus view showed diffuse radiopacity in right maxillary sinus area and nasal cavity with deviation of nasal septum to left side.

Doppler ultrasound showed marked vascularity including drainage veins which were dilated and tortuous. There was no effect noticed on compression of lesion (Fig. 4).

Axial and coronal 64 slice CT study revealed a highly vascular nonencapsulated polypoidal mass on right side of cheek/nasopharynx with involvement of right pterygopalatine fossa with destruction of palate and nasopharynx. Anterior and superior extension to right maxillary sinus, almost obliterating it and right nasoethmoidal complex with pushed nasal septum to the left. Superior extension was seen through the inferior orbital fissure to a small portion of right orbit and through the superior orbital fissure to the base of brain and right cavernous sinus (Fig. 5). Nasopharyngeal airway is significantly obliterated.

Feeding vessels were sphenopalatine, internal maxillary, ascending pharyngeal and unnamed arteries (Fig. 6).

Extensive venous channels were seen leading up to neck veins and internal jugular vein.

The mass measures approximately 4.8 × 6.6 × 8.6 cm.

The CT findings were characteristic and consistent with juvenile nasopharyngeal angiofibroma.

Based on the history, clinical examination and various investigations a final diagnosis of juvenile nasopharyngeal angiofibroma of right cheek with intracranial extension was arrived.

The tumor was in stage III A of Sessions et al classification.
DISCUSSION

JNA is an uncommon benign lesion that affects almost exclusively adolescent males. However, cases of JNA have also been reported in elderly and female patients. The actual occurrence of the lesion in women, however, is still debated. Why these lesions occur in a restricted age group and restricted site at the base of the skull is unknown. It is unclear whether they should be considered as neoplasms or hamartomas. Although most authors describe this pathologic entity as neoplasm, the neoplastic nature has been disputed by others who propose a vascular origin, a specific variant of angioma, a reactive overgrowth of connective tissue, and irregular vessels set in a fibrous stroma. Furthermore, Beham et al concluded that JNAs are vascular malformations and not true neoplasms on the basis of their immunohistochemical and electron microscopic findings. Concerning the average age at onset of symptoms, a divergence exists amongst various authors, with a mean average age of 14 to 17 years. The description of JNAs clinical presentation is consistent throughout the literature. Physical examination reveals that JNA appears as a red or red-blue, unencapsulated, infiltrative, ovoid, smooth, lobulated or nodular, sessile or somewhat pedunculated mass that is composed of a rich vascular network within a fibrous stroma. Recurrent and spontaneous episodes of major epistaxis or nasal obstruction are the most common early symptoms encountered in almost all patients. The presence of these two symptoms in an adolescent boy suggests the diagnosis of a JNA until proven otherwise. Additional symptoms, such as eyelid edema, proptosis and visual changes, facial swelling and bulging, headache or otalgia, hearing loss, sinusitis and meningitis, are encountered in fewer patients. Duration of symptoms ranged from 6 months to 5 years before medical evaluation. In this case, there were occasional episodes of epistaxis and unilateral right-sided nasal obstruction. Presentation of the tumor intraorally is rare and is usually related to neglected cases that have expanded medially into the nasal cavity via the sphenopalatine foramen and by erosion of the palatine bone. At this stage, the tumor presents diagnostic features on CT scan. Hora and Weller in 1961 reported a case of angiofibroma that apparently originated from the pterygomaxillary space and was attached to the medial pterygoid plate. The vascular supply of the lesion is primarily derived from branches of the internal maxillary and ascending arteries, both originating from the external carotid artery, although there may also be arterial feeders from the internal carotid artery. The blood supply in the described case originated from the branches of the superior maxillary segment (posterior superior alveolar artery) and branches of the pterygopalatine segment (descending palatine artery). The etiology of this lesion remains uncertain and the pathogenesis unclear. The predilection of JNA for young adolescent males led to a suggested interrelationship between hormones and JNA. Proposed theories include androgens acting on embryonal cartilage, a hamartomatous nidus of hormone-sensitive fibrovascular tissue that was entrapped in the nasopharyngeal periosteum or normal nasopharyngeal fibrovascular stroma located in the nasopharynx. The diagnosis of angiofibroma is made clinically with the history of an adolescent male with nasal obstruction, epistaxis or both and with other aforementioned symptoms. When doubt exists about the clinical diagnosis, transnasal biopsy permits histopathologic diagnosis, however, resultant severe bleeding accompanying JNA biopsy has led surgeons to avoid this procedure in favor of diagnostic methods. Imaging is extremely important in the identification and management of JNA before surgery. CT scan, magnetic resonance imaging and angiography are the primary imaging methods for the evaluation of patients with nasopharyngeal angiofibroma. Of the three, CT scan is most important before surgery. Angiography can be used for
identification of feeder vessels, delineation of lesion size and location before surgical intervention and later, for evaluation of therapy. Staging of the lesion may be helpful in selecting patients for specific modes of treatment. Sessions et al proposed a staging system on the basis of the extent of the lesion to assist in selecting the best surgical approach. In stage IA disease is limited to the posterior nares or nasopharynx and in stage IB disease shows extension into 1 or more paranasal sinuses. Stage IIA lesions have lateral extension through the sphenopalatine foramen, stage IIB lesions occupy the entire pterygomaxillary fossa and displace the posterior wall of the maxillary antrum forward and stage IIC lesions extend through the pterygomaxillary fossa into the cheek and temporal fossa. Stage III lesions show intracranial extension. The case presented was classified as stage IIIA according to Sessions et al classification. Several methods for the management of nasopharyngeal angiofibromas have been suggested over the years. Surgery is considered the treatment of choice for extracranial lesions; however, surgical removal of this tumor carries a potential for extensive morbidity and mortality from hemorrhage. Presence of intracranial extension of the neoplasm is a relative contraindication to surgical removal, with radiation therapy applicable to such cases. The choice of surgical approach and extent of resection is determined with preoperative imaging revealing lesion size and location. As the methods of radiologic imaging have advanced, the accuracy of preoperative assessment has improved. The significance of accurate preoperative assessment of tumor extension and location is emphasized by the observation that incomplete resection leads to high rates of recurrence, exceeding 40%. Batsakis reported a recurrence rate of about 50% but a mortality rate of only 3%. Most recurrences produce symptoms within 1 year after treatment and recurrence is uncommon beyond 2 years after surgery.

REFERENCES