Case Report

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Juvenile Nasopharyngeal Angiofibroma with Intracranial Extension : A Case Report

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ABSTRACT

Background: *Juvenile Nasopharyngeal Angiofibroma (JNA) is a benign but aggressive neoplasm, shows propensity for destructive local spread and often extends to the base of the skull and into the cranium. It generally presents as painless, unilateral or bilateral nasal obstruction with or without epistaxis. Diagnosis is aided by clinical examination and specialized imaging techniques such as CT scan & MRI, and confirmed by excision and histopathology. In this article, we aimed to present the diagnostic dilemma of an invasive nasopharyngeal mass with massive extension.*

Case Report : *A 16-year-old male presented with a bilateral nasal obstruction, recurrent epistaxis, hearing and breathing difficulties. After total clinical, radiological and histopathological evaluation it was diagnosed as juvenile nasopharyngeal angiofibroma.*

Conclusion: *The approach to its diagnosis and management was a combined multi-disciplinary effort of otorhinolaryngology, radiology and pathology.*

Key words : Andrew-Fisch staging; Intracranial extension; Juvenile nasopharyngeal angiofibroma.

Introduction

Juvenile Nasopharyngeal Angiofibroma (JNA) is a benign, locally invasive highly vascular tumor, accounts for 0.05-0.5% of all head-neck neoplasms. It is exclusively seen in adolescent males and the average age of presentation is 14 years[1,2]. As the name implies, it develops in the nasopharynx, more specifically within pterygopalatine area. The choanae and nasopharyngeal vault are also common sites of origin[3,4].10%-20% of JNAs have the potential to extend intracranially and into orbit through the infraorbital fissure but dural invasion is very rare[5]. Histologically angiofibromais always composed of an intricate mixture of irregular blood vessels and fibrous stroma. Its propensity to bleed torrentially and ability to spread along cranial fossa and base of skull makes the tumor surgically challenging[6]. Here, we report a case of 16 year old male patient diagnosed as juvenile nasopharyngeal angiofibroma with intracranial and orbital extension.

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Case Report

A 16 years old male patient was admitted in the Department of Otolaryngology, Chittagong Medical College Hospital in December 2019 with the complaints of nasal obstruction for last nine months. Initially it was on the right nasal side and with time progressed into bilateral nasal obstruction. He also complained of occasional per-nasal bleeding. Gradually he developed hearing impairment and difficulty in breathing which worsen at bed time for last 4 months. He did not have any visual or neurologic complaints. He had no noteworthy past medical history, family history or drug history. Rhinoscopic examination revealed a red fleshy mass innasal cavity that occluded the air passage. The oro-dental examination revealed nothing abnormal. He also had mild swelling on right cheek with normal appearing overlying skin. There was mild proptosis of the right eye. The visual acuity was detected 10/10 in ophthalmology consultation and the eye movements were all normal. The patient has also underwent a neurosurgical consultation. The neurological evaluation was unremarkable with normal cranial nerves' function. General examination revealed no significant abnormality. Routine haematological investigations were found to be within the normal limits.

The Computed Tomography scan (CT scan) of paranasal sinus and brain revealed a large mass in the right sino-nasal cavity with well-defined margins [Figure 1]. Bony destruction was noticed at superior, medial and infero-lateral aspect of right maxillary sinus. The mass extended into the ethmoidal sinus, right orbit and cheek. Extension into the cranium at left side of sphenoid bone

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with erosion at the lesser wing through sphenoidal sinus was also evident. Left nasal fossa was almost obliterated [Figure 2].

This massive extension of sino-nasal tumor put the surgeons in diagnostic dilemma and the patient was advised for surgical evaluation.A soft, reddish fleshy mass with lobulated surface was removed measuring 5x3x2.5 cm. After removal of the tumor, surgeons' provisional diagnosis was juvenile nasopharyngeal angiofibroma, Andrew-Fisch stage- IIIB[7,8].

Histopathological examination revealed a proliferative connective tissue stroma consisting plump fibroblasts-ovoid to spindle shaped, inter spersed with a dense vascular network [Figure 3]. Vascular component consist numerous blood vessels of different sizes and shapes lined bysingle layer of plump endothelial cells with little or no smooth muscle or elastic fibers [Figure 4]. This microscopic finding confirms the diagnosis- juvenile nasopharyngeal angiofibroma.

The neurologic functions and visual acuity was intact post operatively. The post-operative period was unremarkable and he was discharged at 6th post-operative day. He also came for follow up 6 months later and was well enough with no residual disease.

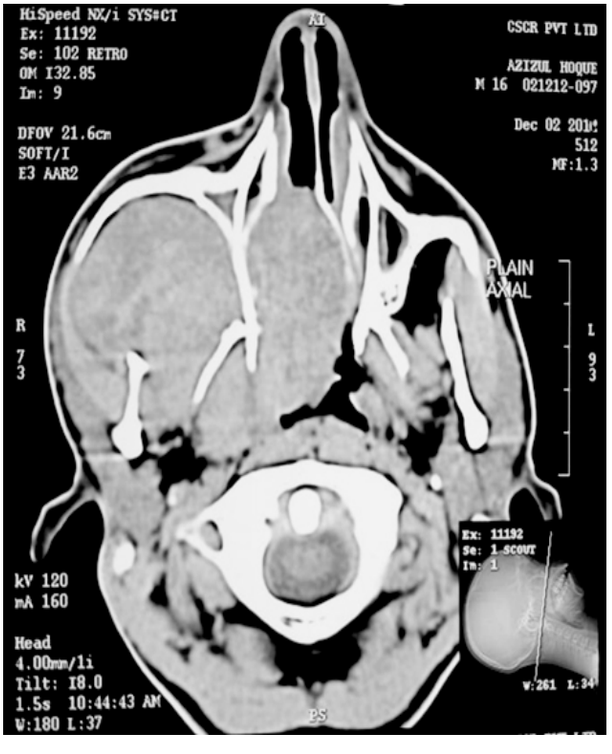

Figure 1 Axial view of CT scan of paranasal sinus and brain showing huge mass in the right sino-nasal cavity

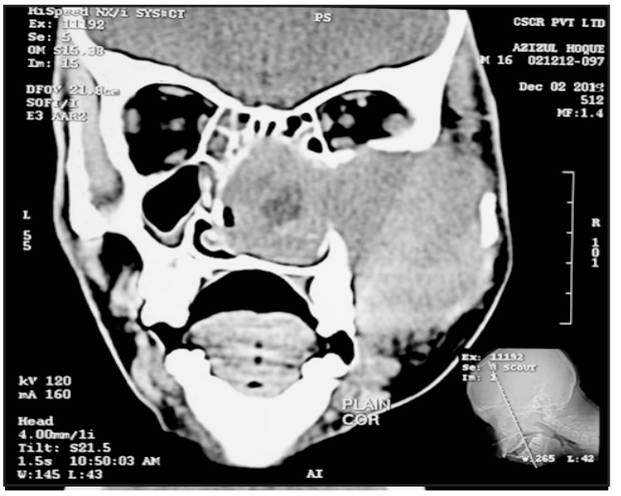

Figure 2 Coronal view of CT scan. Bony destruction was noticed at superior, medial and infero-lateral aspect of right maxillary sinus. The mass extended into the ethmoidal sinus, right orbit and cheek

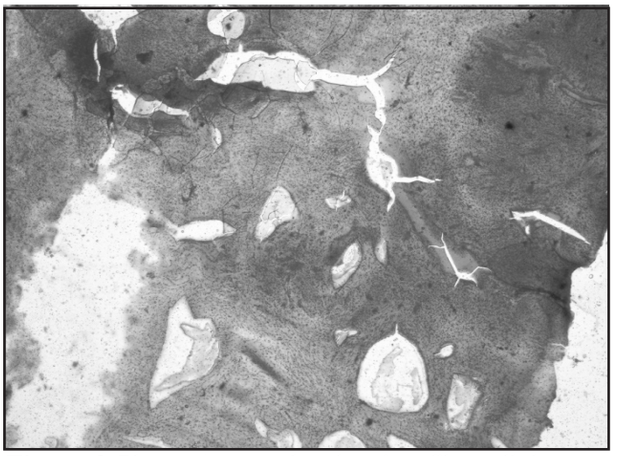

Figure 3 Photomicrographs showing fibrocellularstroma and numerousstaghorn appearances of the blood vessels (H&E stain, ×20 view)

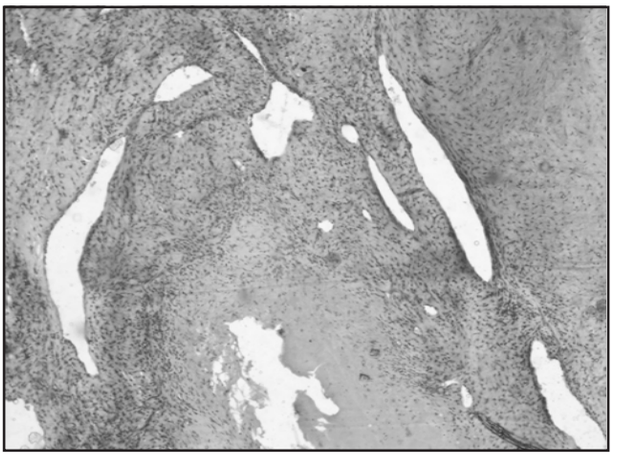

Figure 4 Blood vessels lined by endothelial cells but no smooth muscle and compressed by fibrous tissue (H&E stain, ×40 view)

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Table I Andrews- Fisch classification

Type I	Tumor limited to the nasopharynx, limited or negligible bone erosion of the sphenopalatine foramen.
Type II	Tumor invading the pterygopalatine fossa or the maxillary, ethmoid or sphenoid sinus with bone destruction.
Type IIIa	Tumor invading the infratemporal fossa or orbit without intracranial involvement.
Type IIIb	Tumor invading the infratemporal fossa or orbit with intracranial parasellar extradural involvement.
Type IVa	Intracranial intraduraltumour without involvement of the cavernous sinus, pituitary fossa or optic chiasm.
Type IVb	Intracranial intraduraltumour with involvement of the cavernous sinus, pituitary fossa or optic chiasm.

Discussion

Juvenile Nasopharyngeal Angiofibroma (JNA) is a locally expansive highly vascular benign neoplasm. In the fifth century (AD) Hippocrates described this tumor, but did not name it. In 1897 Chelius mentioned a nasopharyngeal benign vascular tumor as polyp that took place during the adolescence & early adulthood. The term angiofibroma was first used by Friedberg in 1940[2,4].

The pathogenesis and development of JNA is not fully understood. Haemartoma and vascular malformation theories are some of popular hypothesis. The histologic origin of JNA contains vascular endothelial cells and fibroblasts. Simultaneous growth of these two main components in JNAs suggests a vascular haemartoma/ pathologic angiogenesis[8,9]. Selectivity of age and sex indicate that its development is hormone dependent. Several authors studied androgen receptor, progesterone receptors, and Estrogen Receptors (ERs) in JNA and found discrepancies in the results[10]. However, no apparent alteration in serum hormone levels is observed. So, the hormonal influence in JNA remains controversial.

Clinical history and physical examination usually give important clue in patient evaluation.Classically JNAs present with triad of unilateral nasal obstruction, epistaxis and nasopharyngeal mass in adolescent males. Other common presentations are swelling of the cheek, hearing loss, dacrocystits, rhinorrhea, anosmia etc. Advanced lesions may cause proptosis, diplopia, headache, cranial neuropathy and other less uncommon symptoms[11].

Plain radiographs are no longer use for the diagnosis of JNA. Advanced imaging techniques such as Computerized Tomography (CT scan) and Magnetic Resonance Imaging (MRI) help in determining the precise invasion of JNA lesion and the feeding blood vessels, making it possible for angiographic embolization[12]. Staging/ classification of JNA is based on its location and extensionseen on imaging (CT and/or MRI). Different authors proposed a variety of staging criteria for evaluating JNAs, although none is universally accepted. Some of the more commonly used classification systems are-Radkowski, Andrews- Fisch, Chandler, Johns, Onerci and Sessions[11,13]. Andrews- Fisch classification is described in Table I.

Staging helps in choosing the surgical approach. A modern staging, the UPMC (University of Pittsburgh Medical Center) staging system, is supposed to be superior to other staging systems for prediction of residual vascularity of the tumor following embolization as a prognostic factor and tumor recurrence, but it has some potential limitations in clinical use[13].

The definitive treatment of angiofibroma is surgical excision. Different surgical approaches arepracticed nowadays- endoscopic, endoscopy-assisted or open surgical approaches. Open approaches include lateral rhinotomy, transpalatal, transmaxillary, mid faciald egloving, Le Fort I-III, Denker, infratemporal and various combinations of approaches. Tumors that involve multiple compartments are challenging to access surgically. Radiotherapy is usually reserved for inoperable tumors[14,15].

To comfirm the initial diagnosis and to exclude malignancy,histopathological examination is always justified. JNA shows a fibrocellular stroma with spindle cells and haphazardly arranged collagen interspersed with an irregular vascular pattern. The vascular component is made up of slit like or dilated vessels of different calibers with little or no smooth muscle or elastic fibers. This lack of muscle undoubtedly contributes to massive hemorrhage following minimal manipulation on tumor.The fibrous component is made up of a collagen stroma, where the essential cellular element is the fibroblast. Myofibroblasts and myxoid areas may present interspersed within the tumor. Mitosis is rare. Inflammatory cells such as mast cells and T lymphocytes are occasionally seen[16,17].

Limitations

Long-term follow up and surveillance for recurrence could not be done.

Conclusion

The classification and clinical staging of JNA helps to plan treatment and determine prognosis. Preoperative embolization and surgical resection is the treatment of choice. Early diagnosis, accurate staging and adequate excision are essential in the management of this lesion.With a well-executed surgical management and a correct clinical follow-up, the prognosis for this tumor is generally favorable.

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Recommendation

All the JNA cases diagnosed in Chittagong Medical College Hospital should be recorded briefly and long term follow up should be ensured to know about tumor recurrence.

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Disclosure

All the authors declared no competing interest.

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