

Juvenile Nasopharyngeal Angiofibroma: A Case Report

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Received: May 06, 2022; **Published:** June 06, 2022

Abstract

Juvenile Nasopharyngeal Angiofibroma (JNA) is a rare, benign, locally invasive, vascular neoplasm that accounts for less than 0.5% of all head and neck tumors. Other terms used for this tumor are nasopharyngeal angiofibroma, bleeding fibroma of adolescence, fibroangioma. In this paper, a rare case of Juvenile Nasopharyngeal Angiofibroma in a 19 years old male patient has been discussed along with mini review of pathogenesis and histological features and treatment modalities.

Keywords: Juvenile nasopharyngeal angiofibroma (JNA); Masson's trichrome stain; vascular tumor; male tumor; fibrous lesion

Introduction

Juvenile nasopharyngeal angiofibroma (JNA) is relatively uncommon benign neoplasm found in head and neck region. Hippocrates described this tumor in 5th century and friedberg first used the term 'angiofibroma' in 1940. JNA occur almost exclusively in the nasopharynx of adolescent males [1]. The site of origin of JNA remains controversial. According to some authors, it originates from the superior tip of the sphenopalatine foramen at the junction of the pterygoid process of the sphenoid bone and the sphenoid process of the palatine bone while, others believe that it arises from the bone of the vidian canal [2].

Few theories has been proposed by authors which firstly includes a hormonal theory that has been suggested due to the lesion's occurrence in adolescent males, other theories include a desmoplastic response of the nasopharyngeal periosteum or the embryonic fibrocartilage [1].

The blood supply to these benign tumors is most commonly from the internal maxillary artery, but may also be supplied by the external carotid artery, the internal carotid artery, the common carotid artery, or the ascending pharyngeal artery.

Case Report

A 19 years old male patient complained of difficulty in breathing since 6 months and bleeding from the nose since 1 month. Patient was relatively alright 6 months back when he felt difficulty in breathing from nose and recurrent nasal infections. He also complained of epistaxis since 1 month which was recurrent, painless, and severe which stops of its own. On clinical examination, there was a nasal mass protruding from left side of nasal cavity which was bleeding. CT scan revealed, asoft tissue shadow seen in left maxillary sinus causing deviation of nasal septum. The shadow was extending upto posterior ethmoid, pharynx, sphenoidal sinus and to maxillary tuberosity area.

Based on clinical examination and CT Scan the provisional diagnosis was made as nasopharyngeal angiofibroma. After routine hematological investigation, the patient underwent for embolization followed by surgical excision.

The excised single soft tissue specimen received was whitish in color. Gross examination revealed a soft tissue specimen which was oval in shape, of approx. 5.5 x 5 cm in size and rubbery in consistency and smooth surface texture.



Figure 1: Gross Specimen.

On microscopic examination, the H&E stained sections showed, dense connective tissue stroma interspersed with numerous dilated endothelial lined blood capillaries. These vessels were of varied caliber, irregularly shaped and empty. Angulated dilated capillaries were also present. The large vessels were lined by single layer of endothelial cells and at some places these vessels were lined by thicker cell layer.

The connective tissue stroma was fibrocellular. Collagenous fibrils of connective tissue stroma were arranged in an unoriented pattern or parallel fashion. The dense paucicellular fibrous tissue showed cells which were cytologically bland (stellate or spindle shaped fibroblasts). Some of the fibroblast also showed hyper chromatic nuclei.

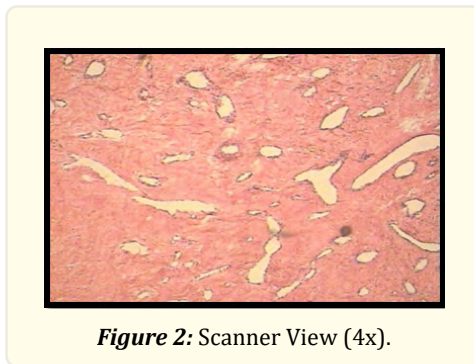


Figure 2: Scanner View (4x).

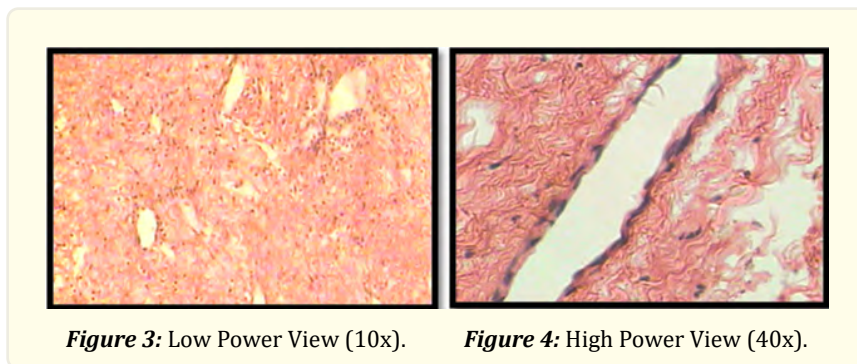


Figure 3: Low Power View (10x).

Figure 4: High Power View (40x).

To confirm the collagenous nature of the connective tissue Masson's trichrome staining was done which revealed greenish colour of collagen fibers Based on clinical, radiological and histopathological findings, diagnosis of "nasopharyngeal angiofibroma" was made.

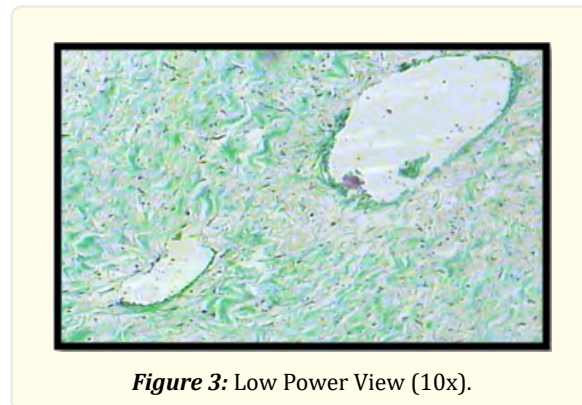


Figure 3: Low Power View (10x).

Discussion

Historical review of the Juvenile Nasopharyngeal Angiofibroma

Chelins (1847) was the first investigator to observe that the fibrous nasal polyp often occurred in patients at puberty. Legouest in 1865 called attention to the specificity of juvenile nasopharyngeal angiofibroma for males. Gosselin in 1876 noted that these fibrous polyps tend to occur in the nasal passage and undergo spontaneous regression after puberty. Chaurean (1906) probably was the first investigator to use the term angiofibroma [5].

Pathogenesis of Juvenile Nasopharyngeal Angiofibroma

Juvenile Nasopharyngeal Angiofibroma is unique entity and the tissue of origin remains obscure. There are various theories proposed, some of which are as follows.

1. Bensch (1878)-fibroblast theory: This theory proposes abnormal response or growth of the connective tissue such as the embryonic occipital plate.
2. Ringertz hypothesis (1938): It states that Juvenile Nasopharyngeal Angiofibroma arises from the ventral periosteum of the posterior pharyngeal wall.
3. Brunner hypothesis (1942): suggests the origin from the fascia basalis.
4. Hugh's theory: It states that the tumor arises from the remnants present in the craniopharyngeal duct.
5. Coenen's hypothesis: is implying that chondrocranium forms the matrix of the tumour.
6. Willi's hypothesis: stresses that Juvenile Nasopharyngeal Angiofibroma is a type of immune response.
7. Sternberg's hypothesis: Juvenile Nasopharyngeal Angiofibroma is a variant of hemangioma.
8. Harma hypothesis: It is a hyperplastic tissue reaction.
9. Osborne's hamartoma hypothesis: It is of hamartomatous origin.
10. Martin and Able hypothesis: According to it, Juvenile Nasopharyngeal Angiofibroma is formed as a result of oestrogen-androgen imbalance showing deficiency of androgen and overactivity of oestrogen.
11. Dane's hypothesis: He hypothesized that it is a result of increase in androgen and not oestrogen activity.
12. New concept-Mild and Mauris hypothesis: It arises from the midline erectile tissue. It is a hamartoma and simulator to that of the erectile tissue of the penis. Period of development of tumour coincides with that of the erectile tissue of penis. Both develop during sexual development days. Both occur in the midline of the body, representing presenting sequestration of erectile tissue growing during puberty under the influence of male hormone.

13. Schiff's theory: This theory is also based on the hormonal imbalance. The alteration in the pituitary-androgen-estrogen axis most probably causes overactivity of the pituitary. Therefore in maturing males, absolute serum oestrogen level increases but seldom to produce feminizing effects. The plasma androgens increases which stimulates the growth of the tumour cells. The normal nasopharyngeal tissues containing androgen receptor cells, may have escaped their normal control, thus giving rise to Juvenile Nasopharyngeal Angiofibroma
14. Juvenile Nasopharyngeal Angiofibroma may be caused by an ectopic nidus of cells which are androgen dependent.
15. Girgis and Fahmy have suggestion of having relation with paragangliomata as the JNA occurs in relation to large vessels. The nutrient artery of JNA is terminal part of the internal maxillary artery. Paraganglionic tissue with large vacuolated cells, is the most probable origin of the tumour. This theory explains the increased incidence of extrapharyngeal extension, as these tumours starts immediately outside the nasopharynx and push their way into the nasopharynx or rather the reverse is not known. Histologically, Zellenballen cells is suggestive of paraganglionoma relation [5].

Clinical Features

Nasopharyngeal angiofibroma are benign tumors that are life threatening because of the vascularity and location. They account for 0.05–0.5% of all head and neck neoplasms [1, 4]. The tumor develops almost exclusively in adolescent boys [3]. Average age of onset is 15 years which ranges from 7-19 yrs [1]. Clinically, the tumor is a red-blue polypoid tissue which may protrude into the anterior nasal cavity or into the pharynx. On examination, tumor may be noticed as nasal mass (80%), orbital mass (15%), proptosis (10-15%). The tumor does not cause pain but it is manifested by nasal obstruction (80-90%), epistaxis (45-60%), sinusitis & headache (25%), facial swelling (10-18%). Intra-Oral Manifestations of JNA can be palatal or tonsillar mass with nasal obstruction. Occasionally, swelling of posterior portion of maxilla may also be seen. Secondary Complications includes otitis media, mastoiditis, dacryocystitis, radiation osteomyelitis, brain abscess or aspiration pneumonitis [5].

On Gross Examination

Tumors are generally rounded or nodular masses in appearances having either a sessile or slightly pedunculated base, red-pink to tan-gray in appearance. Tissue usually has a rubbery consistency and smooth surface texture.

Histopathology

The tumor is unencapsulated and consists of two important components of a

Vascular Network

The vessels vary in number and configuration, appearing slit like or dilated. The peripherally located vessels are often larger and arterial type with visible elastic laminae, However, the smaller vessels in the central portion typically lack the elastic laminae, which may explain the propensity for spontaneous or surgically induced haemorrhage.

A Connective Tissue Stroma

Made up of dense paucicellular fibrous tissue. The cells in the fibrous tissue are cytologically bland and may be stellate or spindle shaped, with nuclei that lack hyperchromatia and have small nucleoli. These cells are interspersed in both fine and coarse collagenous fibrils interweaving in an irregular, unoriented pattern or parallel fashion arranged in connective tissue stroma. Multinucleated giant cells are sometimes present. Under Electron Microscope, stromal cells are mostly fibroblast and show intensive immunostaining for vimentin. Slit like capillaries apparently being obliterated by stromal compression are not uncommon [5].

Treatment Options

Main treatment is surgery. Various approaches like Transpalatine-Wilson's approach, Transpalatal and sublabial approach, Trans-

palatal transantral-Denker's procedure, Transmandibular pterygoid approach, Transzygomatic, Lateral rhinotomy and medial maxil-lotomy can be considered depending upon the feasibility or surgeon's choice.

Radiation therapy may be used adjuvant to the surgical procedure with doses approximately 3000 to 3500 cGy over 3 weeks. Indications of radiotherapy usually include orbital involvement, intracranial extension, few cases of recurrences, residual disease or when indicated preoperatively to shrink tumours.

Chemotherapy advised for Juvenile Nasopharyngeal Angiofibroma are

- Doxorubicin-60 mg/m² IV 1st day
- Vincristine-2 mg/m² IV once a week for 12 weeks
- Dactinomycin-0.015 mg/kg daily IV infusion for 5 days every 3 months.
- Sacarbazube
- Cyclophosphamide-2.5 mg/kg daily orally for 7 days every 6 weeks

Hormone therapy (Oestrogen therapy) helps in making the field of surgery less vascular. It also decreases tumour size and helps in maturity of the tumour, i.e., smaller embryonic vessels become bigger (less bleeding) and induces fibrosis.

Embolization is reserved for symptomatic intracranial extension and multiple cranial nerve palsy cases commonest being internal maxillary and ascending pharyngeal artery. Embolisation should be followed by surgery within 48-72 hours as collaterals develops as time passes [6].

Conclusion

Angiofibroma is a rare, benign, vascular tumor found almost exclusively in young males. Surgery is the gold standard with a trend towards endoscopic approaches. Frequent follow-up after treatment is necessary.

References

1. Rajendran R and Shivapathasundharam B. Shafer's text book of oral pathology, 5th ed. India: Elsevier (2006)
2. Garrett Hauptman, MD SeckinUlualp, Grand Rounds Presentation, UTMB, Dept. of Otolaryngology (2007).
3. Robert E Marx and Diane Stern. "Oral & Maxillofacial Pathology". A Rationale for Diagnosis & Treatment, 2nd ed. (2012).
4. Walvekar, et al. "Extra-nasopharyngeal angiofibroma of the buccal space: surgical management and review of literature". Oral Surgery 2 (2009): 83-86.
5. Harrison DFN. "The natural history, pathogenesis and treatment of juvenile angiofibroma". Archotolaryngol Head Neck Surg (1987): 936-42.
6. Radkowski D, McGillT, Healy GB, Ohlms L and Jones DJ. "Angiofibroma: changes in staging and treatment". Archotolaryngol Head Neck Surg 122.2 (1996): 122-9.

Volume 1 Issue 5 June 2022

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