Original Research Article

Juvenile nasopharyngeal angiofibroma in our experience

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Abstract

Introduction: Juvenile nasopharyngeal angiofibroma (JNA) is a highly vascular histologically benign, locally aggressive neoplasm of the nasopharynx. It accounts for 0.5% of all head and neck neoplasms with a high incidence of persistence and recurrence.
Materials and methods: This was a retrospective observational study whereby the medical records of 32 patients treated for Juvenile angiofibroma between May 2010 and August 2016 were reviewed.
Results: This study included patients aged between 13-25 years average was 19 years. More than 10 cases were in 14-18years group. This age group exhibits rapidity of growth high incidence of recurrence and early involvement of neibouring structures such as pterygopalatine fossa and infratemporal region and cheek. In 16 cases out of 32 there were attachments of tumor on the roof of nasopharynx. And 8 cases had attachments on the choana. Nasal obstruction and epistaxis is the most common symptom observed in study. CECT was the most common imaging modality utilized for diagnosis and staging of JNA and it was required in all cases. MRI was utilized as an additional investigation in cases with intracranial extention and was required in 37.5% of cases. Postoperative complications were not serious and only included malar defect, ectropion, epiphora and palatal defect. Total complication rate in all was about 28.2%, seen in 11 out of 32 patients. 7 patients were diagnosed with recurrence, with a recurrence rate of about 30%. These recurrences were diagnosed at varying intervals of follow-up 4 out of 13 cases with Stage IIIa and 3 out of 4 cases with Stage IVb diagnosed with recurrence.
Conclusion: Radiologic follow-up is essential in the early identification of residual or recurrent disease.

Key words

Juvenile, Nasopharynx, Angiofibroma.

Introduction

The angiofibroma is benign yet biologically aggressive tumor. It originates almost exclusively from the posterior nasal and nasopharyngeal region in adolescent males. Thus it has been known as juvenile Angiofibroma, although cases have been reported in older adults and in females as well. The term Angiofibroma denotes a vascular swelling presenting in the Nasopharynx of pre-pubertal and adolescent males and exhibiting tendency to bleed. Much of the previous literature concerning etiology and treatment was speculative and controversial, but a clearer picture of the nature of these swellings, their site of origin, behavior, and safe management has emerged in recent years.

At one time, the impression existed that the prevalence of angiofibromata was higher in certain parts of the world, for example the middle East and America, than in North Western Europe, where it was considered to be quite low. It is probable that such an assumption was arrived at by equating larger reported series from specific centers with high Geographical prevalence. The usual clinical behavior of the tumor is one of expansive growth with a potential for intracranial extension. Histologically Angiofibroma is composed of fibrous connective tissue interspersed with variable proportions of endothelium lined spaces. A preponderance of fibrous stroma may indeed allow surgical removals with advances in surgical and arterial embolization techniques; the intraoperative blood loss is still a major concern. The capacity for spontaneous regression of Angiofibroma at sexual maturity is doubtful. The suggestion that total regression occurs in the late teens or early twenties has never been convincingly demonstrated. Although most authorities concede that some shrinkage, hardening and loss of vascularity of the swelling occur with age.

Treatment of this tumor is matter of interest. Rare occurrence of this tumor is probably the reason for such varying experiences and opinion regarding the best method of therapy before 1920, surgical treatment used to be more hazardous, with a high recurrence rate. Radiation and Hormonal therapy were tried in an attempt to reduce the size and vascularity of the tumor. External transfusion is always used replace the blood inevitably lost during surgical removal. The disillusionment, and dangers of surgery led to search for alternative and safer methods of treatment has gathered momentum in recent years, largely because of improvements in pre-operative assessment and a better understanding of the condition.

Thus study of Juvenile Angiofibroma contains a review of its historical aspect, anatomy and physiology, of the nasophaynx, pathology symptoms and signs with method of diagnosis and treatment.

Materials and methods

This was a retrospective observational study whereby the medical records of 32 patients treated for Juvenile Angiofibroma between May 2010 and August 2016 were reviewed. Since the time period was prolonged and diagnostic and therapeutic protocols had undergone many changes. Fourteen patients belonged to the 10-14 years age group, 15 to the 15-19 years age group, and three patients belonged to the 20 + years age group. The mean age was 16 years (range 11-35 years). All patients were males.

The tumors were staged according to the criteria laid down by Chandler, et al. [1]. The mean duration of the following investigations were carried out to all patients Urine examination to exclude diabetes and UTI, CBP to exclude any hemotological disorder, ESR, Blood grouping to replace the blood loss that occurring during surgery, HPE whenever the diagnosis was doubt.

Radiological investigation were done as

- X-Ray PNS; In cases there as haziness of the antrum reported as sinusitis.
- X-Ray Nasopharynx: This X-Ray films shows soft tissue shadow in the nasopharynx near the roof.
- X-Ray base of Skull: No erosion of the base of skull seen in any of the cases studied.
- Angiography: Reveals the vascularity of the tumor and feeding vessels.

In 2 cases CT scan PNS and brain was done. In case No 7 suggestive of nasopharyngeal fibroma with extension to the anterior cranial fossa, infratemporal fossa and pterygopalatine fosssa on right side, in case No 11 suggestive of massive nasopharyngeal fibroma with intracranial extension and protrusion into nasoropharynx with proptosis of right eye ball.

In this study surgical removal of is tumor forms the main mode of treatment is Wilson's transpalatal approach and Lateral Rhinotomy approach. Follow-up was 30.3 months (range 6-140 months). Six patients were lost to follow-up (18%). The age distribution, disease patterns, management approaches and treatment outcomes of patients in the two groups were recorded. Statistical analyses were done using students 't' test and test for proportion.

Results

All the 32 cases were males no female case was noticed in this study. Age extremes of presentation: lowest age of presentation was 9 years and highest age of presentation was 26 years.

This study included patients aged between 13-25 years average was 19 years. More than 10 cases were in 14-18 years group. This age group exhibited rapidity of growth high incidence of recurrence and early involvement of neighboring structures such as pterygopalatine fossa and infratemporal region and cheek.
In 16 cases out of 32, there were attachments of tumor on the roof of nasopharynx and 8 cases had attachments on the choana out of which one case had attachment on superior and lateral margin of choana and anterior extension into the nose. One case had pedicle found attached to apheopalatine foramen with widening of the foramen. 3 cases had orbital and cheek extension.

Nasal obstruction and epistaxis was the most common symptom observed in study (**Table – 1**).

Table - 1: Symptoms presented in patients.

Symptomatology	Present in patients (%)	Absent in patients (%)
Nasal obstruction	32(100%)	0
Epistaxis	32(100%)	0
Facial swelling	16(50%)	16(50%)
Proptosis	12(37.5%)	20(62.5%)
Diminution of vision	8(25%)	26(75%)
Protruding nasal mass	4(12.5%)	28(87.5%)
Headache	1(3.1%)	31(96.9%)
Voice	1(3.1%)	31(96.9%)

CECT was the most common imaging modality utilized for diagnosis and staging of JNA and it was required in all cases. MRI was utilized as an additional investigation in cases with intracranial extension and was required in 37.5% of cases (**Table – 2**).

Post-operative complications were not serious and only included malar defect, ectropion, epiphora and palatal defect. Total complication rate in all was about 28.2%, seen in 11 out of 32 patients. All the cases were followed for a minimum of 6 months, and longest follow-up

was of more than 3 years. All the cases underwent nasal endoscopy and check-CECT on follow-up (**Table – 3**).

Out of the 32 cases of nasopharyngeal angiofibroma diagnosed and followed for a minimum of 6 months, seven patients were diagnosed with recurrence, with a recurrence rate of about 30.7%. These recurrences were diagnosed at varying intervals of follow-up 4 out of 13 cases with Stage IIIa and 3 out of 4 cases with stage IVb diagnosed with recurrence (**Table – 4**).

Table - 2: Imaging modalities required for Diagnosis.

Imaging modalities	Number of patients	Percentages
Contrast enhanced computed tomography (CECT)	**20**	62.5%
CECT + magnetic resonance imaging (MRI)	**12**	37.5%

Table - 3: Post-operative complications in study.

Post-operative complications	Number of patients	Percentage
Palatal fistula	2	6.2%
Malar defect	3	9.3%
Ectropion	4	12.5%
Epiphora	2	6.2%

Table - 4: Correlation between stage of tumor and recurrence.

Stage	Total no of patients	Total no of patients with recurrence	Recurrence %	P- Value
I	0	-	-	0.001
II	10	0	0	
IIIa	13	4	30.7	
IIIb	2	0	0	
IVa	2	0	0	
IVb	4	3	75%	

Discussion

Juvenile nasopharyngeal angiofibroma (JNA) is a highly vascular histologically benign, locally aggressive neoplasm of the nasopharynx that exclusively affects male adolescents, with an average age of onset being 14 years. It accounts for 0.5% of all head and neck neoplasms with a high incidence of persistence and recurrence.

In this study, a total of 32 cases of JNA were studied angiofibroma is essentially a disease of adolescent males and peak age of presentation is 15 years. Earlier Stage (I and II) presentations are relatively rarer signifying that there is a significant delay in diagnosis. About one-quarter of angiofibroma patients have intracranial extension at presentation. Nasal obstruction and epistaxis are the most common presentations of angiofibroma and a high index of suspicion is required for diagnosis. CECT is the most common imaging modality utilized for diagnosis and staging of JNA and MRI is utilized as an additional investigation in cases with intracranial extension. A combination of various surgical approaches is used to remove angiofibromas. Radiologic follow-up is essential in the early identification of residual or recurrent disease. The incidence of recurrent or residual disease is high and is about 30%.

Many of the older reports implied that patients suffering from the condition displayed signs of delayed maturity, as judged by secondary sexual characteristics, and that tumor pathogenesis was somewhat linked to this [2-7]. The regression which was observed with age or supposedly

under the influence of hormones was cited as evidence of hormonal etiology but never supported by objective bio-chemical signs, of hormonal insufficiency. The lack of complete regression could well explain the inclusion of older patients in some of earlier reports.

This tumor has been exclusively observed in males by a large number of authors. Female cases were reported by a few. The earliest report was that of Pryor SG [8] reported 12 cases in females, but many of them were outside the accepted age incidence, recent reports also suggest the same [9].

Geographical prevalence: At one time the impression existed that the prevalence of angiofibromata was higher in certain part of the world, for example the middle east and the American, then in Northwestern Europe where it was considered to be quite low.

A few cases have been reported from Japan and Ceylon [10, 11]. It is also common in Asian countries and many cases have been reported from India. It is commonly observed in patients of low social economic living. Juvenile angiofibroma is a relatively rare tumor, histologically benign, but clinically troublesome neoplasm occurring in male adolescents. Now general agreement that, this is exclusively a disease of male and that the mean age at presentation is around 14 years. The range is however wide and varies between 7 and 19 years with Isolated cases presenting.

Conclusion

Nasopharyngeal fibroma (Juvinile fibroma) Occurs exclusively in male adolescents. Nasal obstruction and epistaxis are almost constant features. Majority of the tumors arise from the nasopharynx. Out of 32 cases one case had on sphenopalatine foramen, and others were attached to choanal margins and roof of the nasopharynx and posterior end of the septum. Radiologic follow-up is essential in the early identification of residual or recurrent disease. The incidence of recurrent or residual disease is high and is about 30%.

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