JUVENILE NASOPHARYNGEAL ANGIOFIBROMA ROLE OF IMAGING IN DIAGNOSIS, STAGING AND RECURRENCE

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ABSTRACT
Objective: *To describe the C.T features of index and recurrent Juvenile Nasopharyngeal Angiofibroma.*
Study Design: *Case series.*
Setting & Duration: *ENT and Radiology Department of Civil Hospital and Dow University of Health Sciences Karachi from November 2006 to November 2008.*
Methodology: *Forty adolescent male patients.The patients were admitted through ENT OPD with H/O recurrent epistaxis and progressive nasal obstruction were included. Post operative cases with residual diseases also included. Exclusion criteria were adolescent females elderly males and non enhancement on contrast CT scan. All patients had CT scan examination with and without contrast to determine the origin and extent of tumor. It well demonstrates the presence of the residual disease or recurrence on follow up contrast C.T scan. Statistical analysis was done by using MS Excels. Result expressed in Mean, Standard Deviation and Percentage.*
Result: *This study included 40 male patients aged 10–24 years, with an average age of 17.0±5 years. Epistaxis in 40 patients (100%), Nasal obstruction in 35 patients (87.5%). All patients (40 cases) had nasopharyngeal mass (100%). All patients underwent a radiological evaluation by CT. The findings were: Mass limited to nasal cavity / nasopharynx in 2 cases (5%), involvement of maxillary sinus 27 cases (67.5%), ethamoid and sphenoid sinus 15(37.5%) and 5(12.5%) cases respectively. Pterygopalatine fossa and infratemporal fossa involved in 25(62.5%) and 22 (55%) cases respectively. Erosion of skull base out side dura in 3(7.5%) cases, orbit in 2(5%) cases and intracranial extension in 1(2.5%) case noted. Patients had Stage III in 27cases (67.5%), Stage II in 10 cases (25%) and Stage I in 2 cases (5%). Advanced Stage IV in 1 case (2.5%). Follow-up ranged from 1 month to 24 months, with a mean of 12 months Recurrence during follow up period was found in 2 cases (5%).*
Conclusion: *The C.T imaging play vital role in diagnosis and staging of Juvenile Nasopharyngeal Angiofibroma. The success of surgery therefore depends upon accurate assessment of extent of tumor by CT scan examination. CT imaging also helps in post operative surveillance, to show the presence and extent of any recurrence or residual disease.*

KEYWORDS: *C T, Juvenile Nasopharyngeal Angiofibroma, Residual Disease*

INTRODUCTION

Nasopharyngeal Angiofibroma (JNA). It is a relatively uncommon tumor occurring mainly in adolescent males. It accounts for 0.5% of all head and neck tumors.[1] The origin of the tumor is from the posterolateral wall of the nasal cavity near the superior margin of the sphenopalatine foramen.[2] Surgeons avoid biopsy due to vascular nature of tumor and rely upon imaging methods.[3] Once diagnosis established task of imaging is to show the extent and staging of tumor. Final role of imaging is post operative surveillance, to show the presence and extent of any recurrence.[3] Typically, patients present with unilateral nasal obstruction and recurrent epistaxis. As the disease progresses facial deformities and blindness occur.[4] The diagnosis of JNA is essentially based on a careful history, nasal examination and supplemented by imaging studies. Radiological imaging plays a crucial role in the diagnosis, staging and management of Juvenile

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Dr. Sessions and Dr. Alford in 1975 described the radio logic findings seen on plain film with JNA. The sphenoid hole is demonstrated on Water's view or the submental view. This hole represents the eroded space through which the tumor invades. Bowing of the posterior wall of the maxillary sinus or the Holman-Miller sign was also described as one of the most consistently typical features of JNA on plain films X-ray base of skull or waters view for sinuses. These days contrast-enhanced CT scanning and MR imaging are valuable imaging modalities in the work-up of Juvenile Nasopharyngeal Angiofibroma (JNA).[3] C.T is more accurate for bony changes while MRI accurately delineate soft tissues.[5] Angiography has diagnostic and therapeutic value, by embolization of the tumor-feeding vessels at the same sitting. The blood supply of these lesions is primarily from the external carotid artery (ECA) and in some cases from the internal carotid artery (ICA).[6] Surgery is the recommended treatment but with the highly vascular nature of the lesion and there is a risk of extensive blood loss. Preoperative embolization and new surgical approaches facilitate removal without increasing morbidity.[7-12]

Recurrence is growth of residual disease (RD). The choice between contrast-enhanced CT and MR imaging to identify RD is open to debate. Both MR imaging and enhanced CT provide a good definition of the soft tissue interactions and the extent of JNA. Contrast-enhanced CT is better because it depicts the close interactions between the bony anatomy and RD. In addition, helical CT is less prone to motion artifact.[13]

The purpose of the present descriptive study was to evaluate the diagnostic features of index and recurrent JNA on CT. which determine therapeutic approach. Contrast-enhanced follow up CT scan to detect RD / recurrence after surgical excision of JNA.

METHODOLOGY

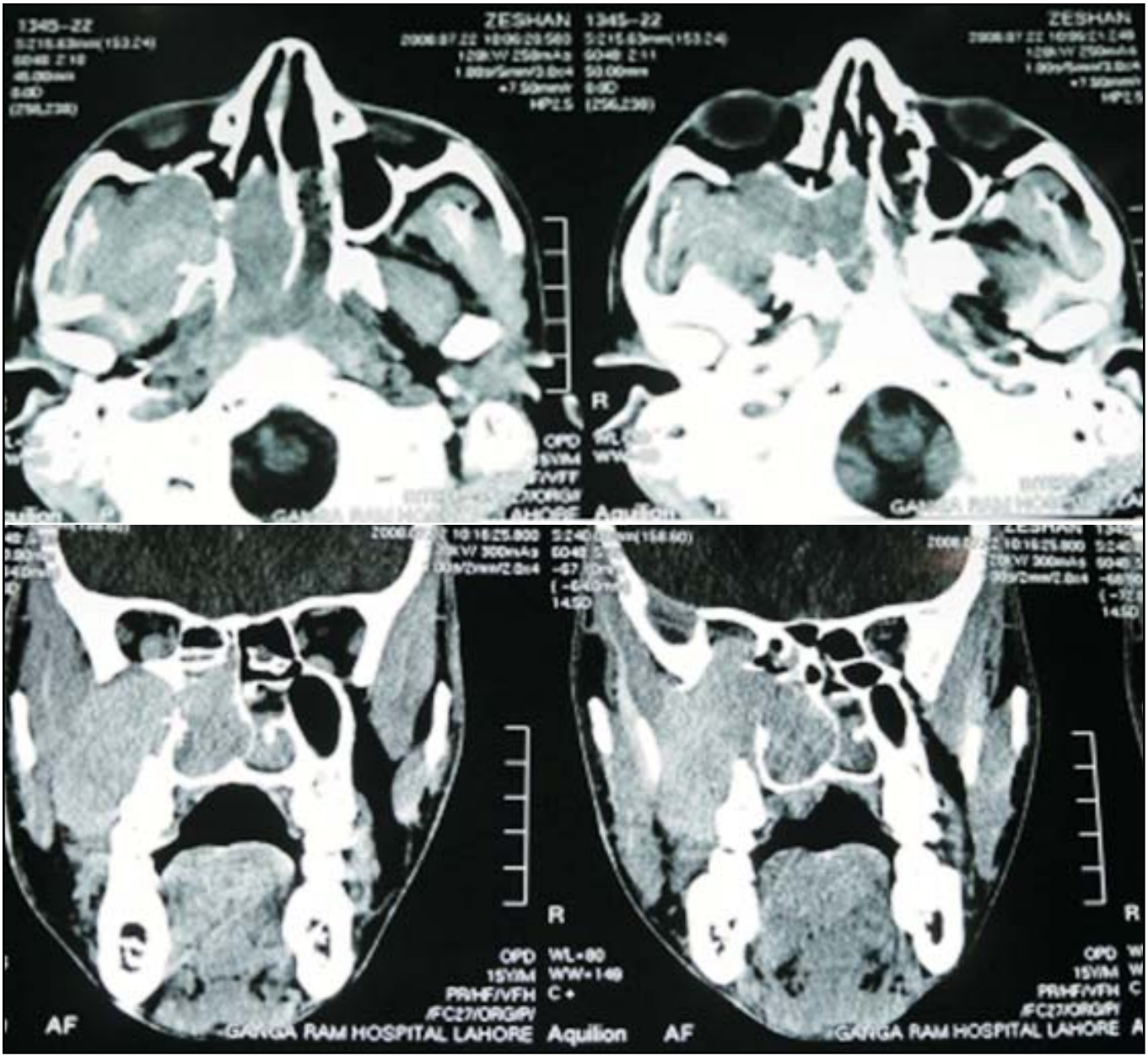
Patients refered through ENT OPD, with H/O repeated epistaxis and nasal obstruction as the main presenting symptom, were admitted in the ENT departmennt. CT reporting was done in Radiology department of Civil Hospital and Dow university of Health Scinces Karachi from November 2006 to November 2008. Inclusion criteria, adolescent male with H/o of recurrent epistaxis, nasopharyngeal mass, nasal obstruction, cheek swelling, proptosis and post operative cases for follow up C.T scan. Exclusion criteria was adolescent female and elderly male. CT exclusion criteria was no enhancement on contrast study. In all patients after clinical diagnosis, origin and extent of the tumor was assessed by means of contrast and non-contrast enhanced CT examination. Patients had a minimum of 1 month 2 years follow-up and had undergone contrast-enhanced CT in the days after apparently complete surgical excision of JNA. Tumor staging is based on CT which demonstrates involvement of sinuses, orbit, intracranial extension, and also illustrate the presence and extension of any recurrence. Statistical analysis done by using MS excels. Result express in Mean, Standard Deviation and percentage.

Chandler 1 staging systems for juvenile nasopharyngeal angiofibroma.

Stage I: Tumor confined to nasopharynx.
Stage II: Tumor extending into nasal cavity or sphenoid sinus.
Stage III: Tumor extending into maxillary antrum, ethmoid, pterygopalatine fossa and Infratemporal fossa.
Stage IV: Intracranial extension of tumor.

Table I. Radiological Findings

Radiological Findings	No. of Patients	Percentage
Maxillary Sinus	27	67.5
Ethamoid Sinus	15	37.5
Sphenoid Sinus	6	15.0
Orbit	2	5
Pterygoplatine fossa	25	62.5
Infra temporal fossa	22	55
Erosion of skull base out side dura	3	7.5
Intra-cranial extension with invasion of optic chiasma	1	2.5
Enhancement on post-operative follow-up scan	2	5



Radiological Findings

RESULTS

This study included 40 adolescent male patients aged 10-24 years, with an average age of 17.0±5 years. Epistaxis in 40 patients (100%), Nasal obstruction in 35 patients (87.5%). All patients (40 cases) had nasopharyngeal mass (100%). All patients underwent a radiological evaluation by CT. The findings of imaging studies are: Mass limited to nasal cavity / nasopharynx in 2 cases (5%), involvement of maxillary sinus 27 cases (67.5%), ethamoid and sphenoid sinus 15(37.5%) and 5(12.5%) cases respectively. Pterygopalatine fossa and infratemporal fossa involved in 25(62.5%) and 22(55%) cases respectively. Erosion of skull base out side dura in 3(7.5%) cases, orbit in 2(5%) cases and intracranial extension in 1(2.5%) case noted, depicted in table-1 and shown in figure. Patients had Stage III in 27cases (67.5%), Stage II in 10 cases (25%) and Stage I in 2 cases (5%). Advanced Stage IV in 1 case (2.5%). Follow-up ranged from 1 month to 24 months, with a mean of 12 months recurrence during follow up period was found in 2 cases (5%). CT findings of recurrence were presence of tumor with positive enhancement on contrast CT on follow up scans. Post contrast axial and coronal CT scan, showing enhancing mass occupies whole nasopharynx, Rt maxillary sinus and Rt. ethamoid sinus with bone erosion extending to pterygopalatine and infratemporal fossa.

DISCUSSION

Radiologic imaging has paramount importance at all stages in the management of juvenile nasopharyngeal angiofibroma. Our experience shows the surgical approach should be selected according to tumor stage, Modern imaging technique allows accurate diagnosis and staging.[14] Once the diagnosis established the task of imaging is to show the extent of tumor, of particular importance is the presence and extent of invasion of the sphenoid, for this is the main determinant of recurrence, a conspicuous feature of the natural history of angiofibroma.[15] Contrast-enhanced CT scanning and MR imaging are valuable imaging modalities in the work-up of juvenile nasopharyngeal angiofibroma (JNA).[3] Basically, the tumor originates from the sphenopalatine foramen and involves both the pterygopalatine fossa and the posterior nasal cavity. [15-16] JNA develops through certain paths of extension expanding by means of bony erosion and displacement of adjacent structures, with possible expansion in the skull base.[15-18]

The CT scan has valuable role in post operative follow up: to show the extent of any recurrence after surgery, or to allow the size assessment after radiotherapy, or to monitor natural tumor involution.[3] Today we rely on CT and MRI. Unfortunately many centers, including our set up do not have MRI facility. So our study was focused on C.T scanning which shows bony changes and offers other additional information about possible intracranial extension. The application of C.T and M.R.I has reduced the need for the routine use of diagnostic angiography. Postoperative contrast-enhanced helical CT was performed with a multidetector-row scanner, scanning extended from the frontal sinus to the inferior limit of the oropharynx (vallecula). RD was suspected when a persistent tumor was seen at a place corresponding to the initial extension of the tumor. Signs of RD are characterized by positive enhancement on radiological follow-up. Arteriography is helpful as a diagnostic and a therapeutic standpoint when we use embolization. Because of vascular nature of JNA, tumor biopsy is usually avoided. Nasopharyngeal mass in an adolescent male patient, Diagnosis is based on history, clinical examination and relay on imaging methods for deciding whether the mass is likely to be an angiofibroma, or a non vascular lesion such as an antrochonal polyp.[3]

Juvenile nasopharyngeal angiofibroma as the name suggests is the disease of young persons.[2,4] In our series the mean age of presentation was 17 years as seen in other studies. Genetic studies have revealed strong androgen receptor expression in JNA suggesting that JNA is an androgen-dependent tumor. This could probably explain the predilection for the male sex.[19]

Most of the patients in our series (67%) belonged to Chandler's Stage III, which is comparable with that, is reported in western literature. The late presentation can be attributed to the poor health services in the rural areas and peripheral centers.

Surgery remains the criterion standard for the treatment of JNA.[17,18,20] Despite new advancements in surgery, high recurrence rates and early recurrence have been reported, especially when the JNA involves the skull base.[15-18]

Recurrence is growth of residual disease (RD). Early detection of such RD may allow for treatment and thus a reduction of recurrences. Radiotherapy is reserved for residual disease after surgery or for palliation of advanced unrespectable stage IV disease. In this study 2 patients were referred to irradiation instead of surgery on the bases of radiological findings A mean recurrence rate of 32% to as high as 40-50% in the case of skull base invasion has been reported.[15,18,22,24] Although in this study recurrence rate are very low only 2 cases 5%.

CONCLUSION

C.T scan has paramount importance in diagnosis and staging of juvenile nasopharyngeal angiofibroma. C.T helps in the proper mod of management and success of surgery therefore depends upon accurate assessment and extent of tumor. CT imaging also facilitate in post operative surveillance, to show the extent of any recurrence or presence of residual disease.

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