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Original Research Article

Magnetic resonance imaging in evaluation of juvenile nasopharyngeal angiofibroma in term of its diagnosis and pattern of spread

Puran, Narendra Kumar Kardam*

Department of Radiodiagnosis, R.N.T Medical College, Udaipur, Rajasthan, India

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***Correspondence:**
Dr. Narendra Kumar Kardam,
E-mail: nkadamdr@gmail.com

ABSTRACT

Background: Juvenile nasopharyngeal angiofibroma (JNA) is a tumor that shows a predictable spreading pattern. This study showed the characteristic MRI findings and its pattern of spread of JNA for diagnosis.
Methods: This study consisted of 6 cases of JNA for diagnosis and pattern of spreading by using MRI as modality of choice.
Results: The total patients included in study were 6 with age range from 9-20 years and all were male. The classification system used in the study was Radkowski and Onerci system. The tumor showed isointensity to muscle and hyperintensity on T1 and T2 sequences respectively on MR imaging. All lesions had internal signal void regions and showed intense enhancement after IV contrast administration. Diffusion restriction was not an associated features however high values of ADC were noted. The MR angiography of three patients showed blood supply of tumor was mainly from the internal maxillary branch of external carotid artery. The treatment was surgical excision of tumor which was decided on MRI Diagnosis and pattern of Extension described in Radiological report.
Conclusions: MRI is a modality of choice for diagnosis and extension of JNA based on clinical and radiological findings, without performing a biopsy.

Keywords: Angiography, Juvenile nasopharyngeal anfiofibroma, Nasopharynegeal mass, Spread pattern

INTRODUCTION

Juvenile nasopharyngeal angiofibroma (JNA) is a rare tumor that predominantly occurs in adolescent and young males,the average age being 15years. It is the most common benign tumor of nasopharynx and consist of approx 0.05 percent of all head and neck masses. Histopathologically its a being tumor of vascular origin that is locally invasive and aggressive in course causes erosion of adjacent anatomical structures particularly of the bones and may result in intracranial extension of the tumor.

There are different theories regarding pathogenesis of JNA in literature's. For high prevalence of the disease in adolescent boys Schick et al suggested role of hormones; however recent advances challenge this theory.[1-5]

While Patrocinio et al and Madhavan Nirmal et al in their study showed that it is not only the disease of adolescent and young boys but it was being diagnosed in older and females.[6-9]

Malignant transformation is a rare finding and associated with recurrent radiotherapy used for treatment in advance tumor with intracranial extention.[10]

JNA is thought to be originate from sphenoplatine foramen (posterio-superior part of it) located in postero-lateral wall of nasal cavity.

The adjacent anatomical structures includes ptergoplaltine fossa, infratemporal fossa, maxillary sinus, nasal cavity, orbit, sphenoid sinus and skull base all mentioned structures can be involved by the extension of tumour and which can further extend to involve intracranial structures.[3,11]

The clinical symptoms include progressive nasal blockage and recurrent nasal bleeding in 80%-90% and 45-60% of patients respectively.[12] For accurate diagnosis, clinical history physical examination along with radio-logical investigation plays a pivotal role. Computed tomography (CT) and magnetic resonance imaging (MRI) are the preferred modalities to detect and to determine the extent of the disease, without the need of the biopsy.

Treatment consist of surgical excision with preoperative embolization which is gold standard treatment and in case of advanced tumour due to complex regional anatomy of the involved structures, chemotherapy, radiotherapy and hormonal therapy is added to the treatment protocol in these cases.

The risk of recurrence is largely depending on extent of the tumor at the time of diagnosis and by success of treatment protocol used which depends on accurate delination of location and extension by radiological investigation. In this study we retrospectively evaluated the role of MRI in the diagnosis and extension of JNA.

METHODS

The study was retrospective in design so histopathologically proved cases of JNA were included in the study who had their preoperative MR studies at our department at R.N.T Medical College Udaipur from January 2017 to December2018.

MRI Protocol

MR imaging were performed on 1.5 Tesla philips achieva maching using adequate standard protocol. For all six patients T1 weighted images in Axial and saggital planes, axial T2 weighted and fat saturated coronal T1 weighted images; coronal shot tau inversion recovery (STIR) images; and fat saturated contrast enhanced T1 weighted images in axial coronal and saggital planes were included in protocol. All patients had their additional ADC and Diffusion weighted images. Three patients also undergone for MR angiography.

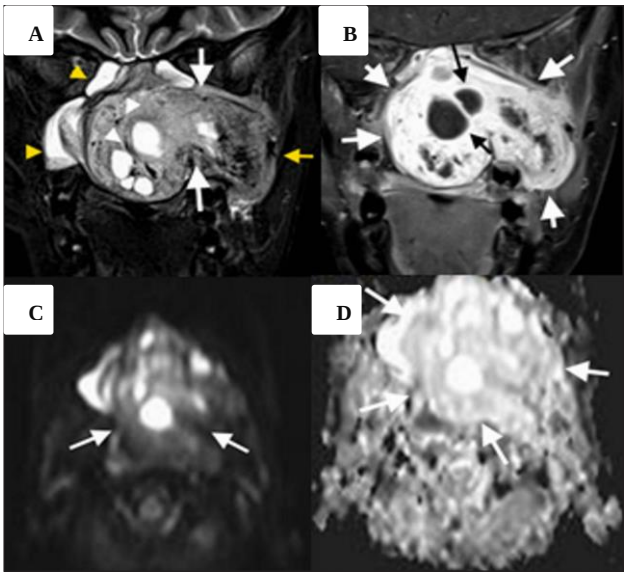
Images analysis

The lesion's primary location its extension, contour, signal intensity characteristic on different sequences contrast enhancement pattern and feeding vessels were analyzed and additional lesion associated findings if present were analysed. In all patients; DWI and mean ADC of lesion were also analysed. All lesions were staged according to the onerci and Radkowski stagging system.

RESULTS

All patients were young adolescent and all were male with age varies from 9-20 years. The primary presenting symptoms was nasal obstruction in three patients; and epistaxis in two patients while one patient had both epistaxis and nasal obstruction on presentation. The tumor was primarily located on the right side in five patients and on the left side in one patient. All tumour had lobulated contours. Tumor extension (into nasopharynx, nasal cavity and into the pterygoplatine fossa) was a common feature in all patents. The infratemporal fossa was involved in five cases, the maxillary sinus was involved in two, the sphenoid and ethmoid sinuses were involved in four cases. The cheek region was involved in two patients, and two patients had orbital involvement. The tumour extended into cavernous sinus in three patients. All patients demonstrated medullary bone marrow edema.

Holman Miller sign (Antral sign) i.e. anterior bowing of the posterior wall of maxillary antrum was noted in three cases.



(a, white arrows) and extends into the temporal fossa (a, yellow arrow). The mass is hyperin- tense on T2-weighted image (a) and exhibits significant contrast enhancement (b). Diffusion-weighted images (c) showed no diffusion restriction, and the lesion has high signal intensity on the ADC map (d). The tumor demonstrates internal cystic components (b, black arrows) and signal-void regions (a, white arrowheads). Image a demonstrates inflammatory signal changes in maxillary and sphenoid sinuses (yellow arrowheads).

Figure 1: 13-year-old male patient (A) coronal t2-weighted precontrast, (B) coronal t1-weighted postcontrast, (C) DWI, and (D) ADC map demonstrate a left-sided nasopharyngeal mass which enlarges the ipsilateral pterygopalatine fossa.

Heterogeneous MRI signal intensity of the tumour were noted in all cases. Tumors were largely isointense to muscle on T1 weighted images and hyper-intense on T2 weighted images. All lesions had internal signal void and all shows intense enhancement after intravenous contrast injection.

Diffusion restriction was not an associated feature (Figure 1). ADC values for these tumour were high which is consistent with benign lesions.

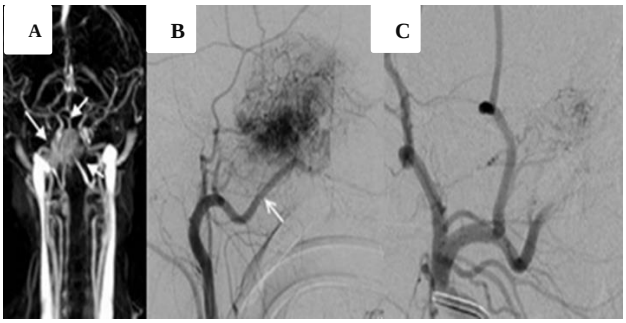
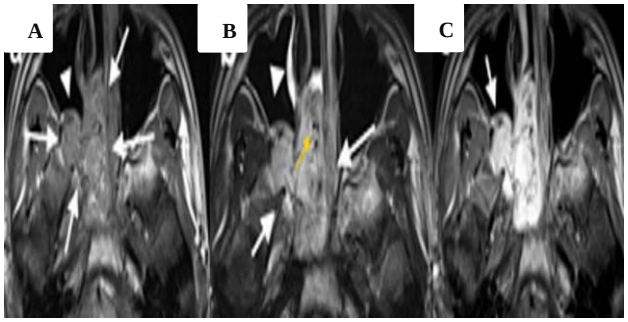


Figure 2: 10-year-old male patient. (A) TWIST-MR angiography of the patient showed bilobule hypervasculer mass on the right side (white arrows). (B) Selective right carotid artery angiography shows that JNA is supplied with the right internal maxillary artery (white arrow). (C) It is observed that the opacification of JNA mainly disappeared in the angiography display obtained after the internal maxillary artery was embolized with microcoil.

The MR angiography of three patients showed hyper vascular mass (Figure 2) having blood supply from the internal maxillary branch of the external carotid artery.



Enlarges the ipsilateral pterygopalatine fossa (a and b white arrows). The mass is isointense with the muscle on T1-weighted image and hyperintense on T2-weighted image (b) and exhibits significant contrast enhancement (c). The posterior wall of the maxillary sinus demonstrates anterior bowing with resultant Holmann Miller sign (a, white arrowhead; c, arrow). The tumor demonstrates signal-void regions (b, yellow arrow) and inflammatory signal changes in maxillary sinuses (b, white arrowhead).

Figure 3: 15-year-old male patient. (A) axial T1-weighted precontrast, (B) axial T2-weighted precontrast, and (C) axial T1-weighted postcontrast MR images demonstrate a right-sided nasopharyngeal mass.

In all patients, the diagnosis was based on MR images, and a surgical excision was planned. To reduce the intraoperative blood loss and morbidity, five patients underwent a preoperative selective embolization procedure; however, in one patient the embolization was not performed.

Magnetic resonance imaging findings of the patients are listed in Table 1, and extension areas of the tumors are listed in Table 2.

Table 1: Magnetic resonance imaging findings of the patients.

Patient No	1	2	3	4	5	6
Tumor size (mm)	43 ×40 × 47	72 × 70 × 64	52 × 62 × 45	38 × 43 × 33	101 × 72 × 81	54 × 58 × 48
T1WI	Hyperintense	Isointense	Iso-/Hyperin-tense	Isointense	Isointense	Iso-/Hypointense
T2WI	Hyperintense	Iso-/Hyperin-tense	Hyperintense	Iso-/Hyperin-tense	Hyperintense	Iso-/Hyperintense
Contrast enhancment	Intense	Intense	Intense	Intense	Intense	Intense
Flow-voids	+	+	+	+	+	+
DWI	–	–	–	–	–	–
ADC value	high	high	high	high	high	high
Antral sign	+	–	–	+	–	+
Cystic component	–	+	+	–	+	+
Vascular compression	–	–*	–	–	–	–
*Surrounds the internal carotid artery without compression.						

The tumor were supplied with the internal maxillary artery in three patients which was also showed by MR angiography, by sphenopalatine artery in one patient and by internal as well as external carotid systems in one patient.

Table 2: Extension areas of the tumors.

Patient no.	1	2	3	4	5	6
Localization	Right	Right	Right	Right	Left	Right
Nasopharynx	+	+	+	+	+	+
Nasal cavity	+	+	+	+	+	+
Pterygopalatine fossa	+	+	+	+	+	+
Infratemporal fossa	–	+	+	+	+	+
Maxillary sinus	–	+	–	–	+	–
Ethmoid sinus	–	+	+	–	+	+
Sphenoid sinus	–	+	+	–	+	+
Cheek	–	+	–	–	+	+
Orbit	–	+	–	–	+	–
Cavernous sinus	–	+	–	–	+	+
Optic chiasm	–	+	–	–	–	–
Skull base erosion	–	+	+	–	+	+
Middle cranial fossa	–	+	+ dural	–	+ dural	–
Bone marrow edema	+	+	+	+	+	+

Table 3: Preoperative stages of the patients, according to Radkowski and Onerci staging systems.

Patient no.	Radkowski	Onerci
1	IIB	II
2	IIIB	IV
3	IIIA	III
4	IIC	II
5	IIIA	IV
6	IIIA	IV

Table 4: Summary of the staging systems.

Radkowski et al (1996)
IA tumor limited to nasal cavity/nasopharynx IB extension into one or more paranasal sinuses IIA minimal extension into pterygopalatine fossa IIB invasion of the pterygomaxillary fossa (with or without erosion of the orbital bones IIC tumor extension into infratemporal fossa, with or without the involvement of the cheek or the pterygoid plates IIIA erosion of the skull base, with minimal extension into cranial fossa IIIB prominent intracranial extension, with or without invasion of the cavernous sinus
Onerci et al (2006)
Minimal extension into nasal cavity, nasopharynx, ethmoid-sphenoid sinuses, or pterygomaxillary fossa Extensive invasion of maxillary sinuses and the pterygomaxillary fossa, extension into anterior cranial fossa and limited extension into infratemporal fossa Marrow involvement of the body and the greater wing of sphenoid bone and base of the pterygoid; extensive involvement of the infratemporal fossa, pterygoid plate, or orbital region; obliteration of cavernous sinus Intracranial extension between the pituitary and ICA, tumor located lateral to ICA, extension into middle cranial fossa

The Table 3 shows preoperative stages of the patients, according to Radkowski and Onerci staging systems. The Table 4 shows summary of the staging systems of Radkowski and Onerci.

The patients were followed for maximum two years, only three patients who were diagnosed with advance stage of tumor showed residual lesion on subsequent imaging. All three cases of recurrence received radiotherapy.

DISCUSSION

Juvenile nasopharyngeal angiofibroma is a locally aggressive, polypoidal neoplastic lesion that originate from sphenopalatine foramen. It is a highly vascular tumour and predominantly occurs in adolescent males. Owing to its intense vascular supply, diagnostic biopsies are mostly avoided. Thus, the patient's clinical history, physical examination and characteristic imaging findings play an important role in diagnosis. In this study the age group and clinical presentation was consistent as mentioned in various literatures. All cases of this study was diagnosis and tumour evaluation in terms of its extension and recurrence was done on the basis of MR findings. The gold standard treatment of JNA is surgical excision with preoperative embolization of feeding vessels. However, in case of locally advanced tumour the surgical approach is difficult so other methods like radiotherapy alone or combined with surgery is used.

The reported recurrence rate are between 20-50 percent. The rate of recurrence is high in advanced stage or large sized lesions. For all the above mentioned reasons, recognition of the tumour's imaging characteristic and its extension pathways is essential for accurate pre-operative staging and determining the appropriate treatment protocol which may include radiotherapy.[1,3]

Histopathologically the Juvenile nasopharyngeal angiofibroma (JNA) is a benign tumour, but its locally aggressive nature causes its extension into adjacent foraminas, paranasal sinuses and even intracranial extension.[1,14] Although it may extend into unexpected locations, it usually follows a predictable spread pattern. Originating from sphenopalatine foramen, JNA mostly extends medially and laterally, largely because it encounters less resistant barriers in these directions.[6]

The tumour usually invades the nasopharynx, the nasal cavity, and the maxillary and ethmoid sinuses medially. Laterally, it invades the pterygopalatine fossa and causes an anterior bowing of the posterior wall of maxillary sinus (Holmann-Miller sign) (Figure 3). Through the pterygomaxillary fissure, the tumour extends into the infratemporal fossa and into the cheek.[1,6]

Through the skull base, JNA may extend posteriorly into pterygoid canal, pterygoid processes, and parapharyngeal space which may erode the pterygoid processes and reach the greater wing of sphenoid bone through the involvement of medullary bone marrow. Also, from the infratemporal fossa, the tumour may extend into orbit throught the inferior orbital fissure.

Further from the orbit, it may extend into the middle cranial fossa and parasellar region through the superior fissure, where it may encase the internal carotid artery and invade the cavernous sinus. Invasion of the roof of sphenoid sinus and medial extension from the cavernous sinus may result in intracranial involvement as well as the invasion of the.

Roof or the infratemporal fossa, foramen rotundum or foramen ovale. Rarely, the tumour may reach the intracranial space through the invasion of the anterior ethmoid cells and the base of the anterior cranial fossa. The intracranial extension of the tumour is frequently limited to the extradural compartment.[1,6,15,16]

The pterygopalatine fossa, nasopharynx and nasal cavity were invaded in all patients in this study. Two case predominantly exhibited a lateral extension pattern with the involvement of the infratemporal fossa. Three cases in the study had advanced stage tumour and exhibited both lateral and medial extensions

CT and MRI are the two primary imaging modalities used in the diagnosis and staging of JNA. CT or MR angiography studies may also be used to detect the feeding vessels.[13,16,17]

CT is superior in demonstrating the bone erosion and the invasion of the sphenoid bone. However, MRI is modality of choice because of its superior soft tissue resolution ability to detect marrow edema and intracranial extention.[13] MRI is the preferred modality for post-treatment followup and recurrences.[9]

MRI is helpful in differentiating the postoperative granulation tissue from a recurrent lesion. On MR images, JNA demonstrates low signal intensity on precontrast T1 weighted sequences and medium to high signal intensity on T2 weighted sequence.1 However, most of the cases in our study showed isointensity to muscle on T1 weighted images and one case exhibited high signal intensity. On T2 weighted images, the tumour were hyperintense. Intramural signal voids and intense enhancement of the tumour after i.v contrast(gadolinium) injection are characteristic MRI features of JNA, and we observed these findings in all of our patients. None of the lesions in this study demonstrated diffusion restriction on DW images, and their mean ADC values were high, as expected in hypocellular benign tumours. The lesion's lateral extension were better appreciated from axial and coronal images, while saggital and coronal reformation for superior extension of the tumour.[1,13] Although the presence of intratumoral degernative cystic components are a rare radiological findings of JNA, observed this feature in four out of six patients. Medullary marrow edema of the sphenoid bone is a feature that is known to negatively affect the success rate of surgery however high contrast resolution of MRI and the use of fat suppressed sequences have facilitated the better detection of this entity. Another advantage of MRI is that fluid collections and the mucosal diseases of the paranasal sinuses are easily distinguished from the intrasinusoidal component of the disease.[6,14,15]

Inflammatory mucosal thickening of the paranasal sinuses were readily distinguishable from the adjacent intrasinosoidal component of the JNA in this study.

MRI plays an essential role in detecting the intracranial and intracavernosal extension of the tumour and demonstrates the relation of tumour with internal carotid arteries and the pituitary gland. However, detecting the dural involvement may be challenging. Dural contrast enhancement on postcontrast T1 weighted images is an important sign of dural invasion. Also, contrast enhanced FLAIR sequence are reported to be more sensitive in detecting leptomeningeal spread.[18]

The expansion of the pterygopalatine fossa due to presence of a nasopharyngeal mass causes anterior bowing of the posterior wall of maxillary sinus, and this sign(antral sign or Holmann-Miller sign) is one of the characteristic findings of JNA.[6,17] Three out of six patients in this study exhibited Holmann-Miller sign, and the pterygopalatine fossa was expanded in all of our cases. JNA is frequently supplied by the sphenopalatine and the maxillary arteries, especially in its early stage. Vascular supply from other external carotid artery's branches of from the vertebral and internal carotid artery's branches is also possible.[11] In this study, three patients were evaluated with MR angiography and the vascular supply of the tumour from internal maxillary branch of external carotid artery.

The differential diagnosis includes nasopharyngeal carcinoma, lymhoma, rabhdomyosarcoma, sinonasal polyp and adenoid hypertrophy. Staging systems for JNA is mostly based on the tumour location and its extension. Various staging system are described by various aurthors, including Radkowski, Fisch, Andrew and onerci.[19-22] In this study we used Radkowski and Onerci staging systems.

The recurrence risk is largely determined by the stage of the tumour and by the surgical approach that is used. Preoperative staging of the tumour is of utmost important in surgical planning and directly affects the surgical approach and success of the treatment protocol. Thus, the imaging studies are expected to provide accurate information regarding the extent of the lesion and to guide the surgeon.

JNA exhibits characteristic imaging findings and its spread pathways are largely predictable. MRI is the most commonly used modality for these tumour due to lack of ionizing radiation, its multiplanar image acquisition capability, high soft tissue contrast resolution, high sensitivity in demonstrating medullary marrow edema, tumour vascularity and its ability to differentiating the mucosal inflammation-fluid retentions from tumoral invasion and in differentiating granulation tissue and residual lesion after surgery. All these feature makes MRI as a modality of choice compare to computed tomography. MRI is also preferred modality for the postoperative follow up.

CONCLUSION

Despite the fact that our study consist of low number of subject, we concluded that MRI accurately delineates the extent of these tumors and aid in staging. The diagnosis can be established based on the characteristic imaging findings without performing a biopsy. The differential diagnosis includes nasopharyngeal carcinoma, lymhoma, rabhdomyosarcoma, sinonasal polyp and adenoid hypertrophy. DWI and their mean values ADC values can be used for distinguishing between benign and malignant tumour. DWI and their mean ADC values can be used for distinguishing between benign and malignant tumors. MRI is the modality of choice for diagnosis, to know the extent, and in postoperative followup.

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