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CLINICORADIOLOGICAL STUDY IN JUVENILE NASOPHARYNGEAL ANGIOFIBROMA IN 10 PATIENTS: A RETROSPECTIVE CASE STUDY

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ABSTRACT

Juvenile nasopharyngeal angiofibroma (JNA) is a rare, benign, highly vascular, and locally aggressive tumor seen in adolescent males. Juvenile nasopharyngeal angiofibromas occur almost exclusively in males and usually in adolescence (~15 years). They account for only 0.5% of all head and neck tumors. Usually, the presenting symptom is a painless nasal obstruction or epistaxis; however, other symptoms may develop depending on the size and extent of the tumor mass. Due to the vascularity of the tumor, biopsy is not usually indicated. The diagnosis is dependent on multiplanar imaging modalities like Computed Tomography (CT), Magnetic Resonance Imaging (MRI), and Angiography. These imaging modalities help in assessing the tumor mass, pre-operative embolization of the feeder vessel, and treatment planning. We have done a retrospective case study of 10 patients in JNA in Department of ENT and Radiology.

KEYWORDS

Embiolization, epistaxis, Angiography

INTRODUCTION

Juvenile nasopharyngeal angiofibroma (JNA) is a highly vascular, benign, yet locally aggressive tumor with a propensity toward skull base erosion and intracranial extension. JNA is noted to be the most common benign tumor originating in the nasopharynx. The incidence of JNA is approximately 1/150,000 and almost exclusively affects adolescent males between the ages of 10 and 25 years.

Many theories regarding the pathogenesis of JNA have been proposed. Given the high prevalance of JNA among adolescent boys, one theory suggests that the tumor may be hormone dependent. Although the genetic studies have shown the expression of estrogen-androgen receptors by JNAs, an association with increased serum hormone levels hasn't been proved, and the influence of the hormones on these tumors remains debatable.

Malignant transformation of the tumor is also a rare finding and reported to be associated with recurrent radiotherapy .

JNA is thought to originate from the posterior-superior part of the sphenoplatine foramen, which is located on the posterolateral wall of the nasal cavity. The tumor is located in close relationship with the pterygoplatine fossa, and although growing slowly, it may extend into the infratemporal fossa, the maxillary sinus, nasal cavity, orbita, sphenoid sinus, skull base, and into the cranial cavity.

The typical symptoms of JNA are unilateral progressive nasal obstruction (80%–90%) and recurrent epistaxis (45%–60%).

Recognition of the characteristic imaging findings of this tumor, along with the clinical history and physical examination findings of the patients, is important for the diagnosis. Computed tomography (CT) and magnetic resonance imaging (MRI) are the two preferred modalities that are used to detect and to determine the extent of these tumors. In most cases, a diagnosis of JNA can be established based on the characteristic imaging findings, without performing a biopsy .

In this study, we aimed to report the characteristic clinical and radiological findings and the spread patterns of this rare tumor. We retrospectively evaluated the clinical and radiological findings of these tumors and their extension pathways.

MATERIAL AND METHODS

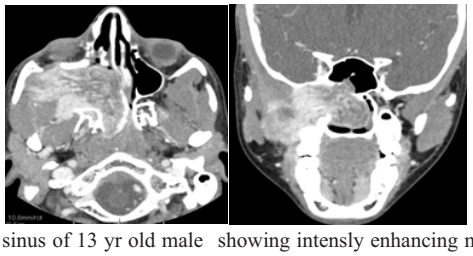
All identified cases of pathologically confirmed JNA in children were assessed from a gender, imaging and embolization findings, tumor stage, surgical approach, and clinical outcomes. We retrospectively analysed imaging data of 10 primary cases of JNA for sites involved and pathway of spread.

RESULTS

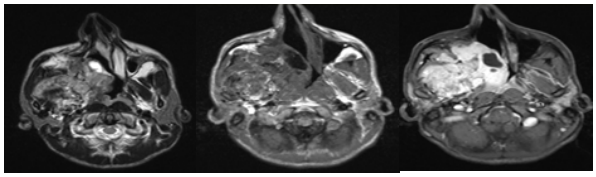
The patients' ages ranged from 9 to 16 years (mean age, 13.5 years), and all patients were male. The primary presenting symptom was nasal obstruction in 2 patients and epistaxis in 2 patients; 6 patient had combined nasal obstruction and epistaxis. The tumor was primarly located on the right side in 7 patients and on the left side in 3 patient. All tumors had lobulated contours.

Tumor extension into the nasopharynx, the nasal cavity, and the pterygopalatine fossa was a common feature in all patients. The infratemporal fossa was involved in five cases, the maxillary sinuses were involved in two, and the sphenoid sinus and the ethmoid air cells were involved in five cases. The cheek region was involved in two patients, and two patients had orbital involvement. The tumor extended into the cavernous sinus in three patients. The Holman-Miller sign was present in three cases. Erosion of the adjacent bones was found in four patients, and all patients demonstrated medullary bone marrow edema. Of the six cases, intratumoral cystic components were observed in four.

MRI signal intensity of the tumors were heterogeneous in all cases. Tumors were largely isointense to muscle on T1-weighted images and hyperintense on T2-weighted images. All lesions had internal signal-void regions, and all exhibited intense enhancement after intravenous (IV) contrast injection. In four patients for whom the diffusion-weighted images were available, diffusion restriction was not an associated feature.



CECT sinus of 13 yr old male showing intensly enhancing mass in nasopharynx with right maxillary sinus, infratemporal fossa and orbital extension



12 year old male(T2, FLAIR, PC images) showing heterogenous S.I. mass in the RT side of the nasopharynx with extension into RT nasal cavity, maxillary sinus, orbit, parapharyngeal, masseter space, infratemporal fossa with intracranial extension

DISCUSSION

Juvenile nasopharyngeal angiofibroma is a locally aggressive, polypoid tumor of the sphenopalatine foramen that originates from the primitive mesenchyme. It is a highly vascular tumor and occurs in adolescent males. Due to its intense vascular supply, diagnostic biopsies are mostly avoided. Thus, the patient history and the clinical and imaging findings play an important role in diagnosis. In this study, the mean age of the patients and their chief complaints on admission were consistent with the literature. In all cases, the diagnosis of JNA was based on CT & MRI findings, and the tumor extension was evaluated with MRI.

The gold-standard treatment of JNA is the surgical resection of the tumor, and surgical options include endoscopic surgery. Endoscopic surgery is advantageous for cosmetic reasons and also has the main advantage of less intraoperative blood loss. However, the use of the endoscopic surgical approach for locally invasive advanced-stage lesions is limited. Preoperative tumor embolizations performed with direct or transarterial injections of the liquid embolic agents are now frequently used to reduce the intraoperative blood loss. Radiotheraphy, alone or combined with surgery, may be used in the treatment of partially resectable or advanced-stage tumors. The reported recurrence rates for JNA are between 20 per cent and 50 per cent, and advanced-stage or large-sized lesions carry the risk of higher recurrence rates. [**1, 2**]

JNA is a histopathologically benign tumor, but it extends into adjacent foramina, fissures, and sinonasal spaces and demonstrates a locally invasive behaviour [**1**] Although it may extend into unexpected locations, it usually follows a predictable spread pattern. Originating from the sphenopalatine foramen, the pterygoid process, and the sphenoid sinus, JNA mostly extends medially and laterally, largely because it encounters less resistant barriers in these directions [**3**]. Medially, the tumor usually invades the nasopharynx, the nasal cavity, and the maxillary and ethmoid sinuses. Laterally, it invades the pterygopalatine fossa and causes an anterior bowing of the posterior wall of the maxillary sinüs (Holmann-Miller sign). Through the pterygomaxillary fissure, the tumor extends into the infratemporal fossa and laterally into the cheek [**1, 3**].

Through the skull base, JNA may extend posteriorly into the pterygoid canal, pterygoid processes, and parapharyngeal space. It may erode the pterygoid processes and reach the greater wing of the sphenoid bone through the involvement of the medullary bone marrow. Also, from the infratemporal fossa, the tumor may extend into the orbita through the inferior orbital fissure. From the orbit, it may extend into the middle cranial fossa and the parasellar region through the superior fissure, where it may encase the internal carotid artery and invade the cavernous sinus. Invasion of the roof the sphenoid sinus and medial extension from the cavernous sinus may result in intracranial involvement as well as the invasion of the roof of the infratemporal fossa, foramen rotundum, or foramen ovale. Rarely, the tumor 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 tumor is frequently limited to the extradural compartment.

The pterygopalatine fossa, nasopharynx, and nasal cavity were invaded in all patients in this study. CT and MRI are the two primary imaging modalities used in the diagnosis and staging of these tumors. CT or MR angiography studies may also be used to detect the feeding vessels [**5**]. CT is superior in demonstrating the bone erosions and the invasion of the sphenoid bone, and MRI is more beneficial for the evaluation of the soft tissue, medullary bone marrow, and intracranial involvement . MRI is the preferred modality for post-treatment follow-up, and recurrences are detected in approximately 25 per cent of the patients . MRI is also 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 sequences [**1**]. However, most of the cases in our series showed isointensity to muscle on T1-weighted images, and one case exhibited high-signal intensity. On T2-weighted images, the tumors were of isohyperintense signal. Intratumoral signal voids and intense enhancement of the tumor after IV contrast injection are characteristic MRI features of JNA, and we observed these findings in all of our patients. The lesions' lateral extensions were appreciated better from axial and coronal images, while the evaluation of the superior extensions of the tumors were easier using the sagittal and coronal images [**1, 5**]. Although the presence of intratumoral degenerative cystic components is a rare radiological finding of JNA, we observed this feature in four out of six patients.

MRI plays an essential role in detecting the intracranial, dural, and intracavernosal extensions of the tumor and demonstrates the relation of the tumor with the 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 sequences are reported to be more sensitive in detecting leptomeningeal spread.

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 branches or from the vertebral and internal carotid artery branches is also possible' The recurrence risk is largely determined by the stage of the tumor and by the surgical approach that is used. Preoperative staging of the tumor is of paramount importance in surgical planning and directly affects the surgical approach and the success of the operation. Thus, the imaging studies are expected to provide accurate information regarding the extent of the lesion and to guide the surgeon.

CONCLUSION

Despite our retrospective study including a relatively low number of subjects, we concluded that MRI accurately delineates the extent of these tumors and aids in staging. The differential diagnosis includes nasopharyngeal carcinoma, lymphoma, rhabdomyosarcoma, sinonasal polyp, and adenoid hypertrophy. DWI and their mean ADC values can be used for distinguishing between benign and malignant tumors. In adolescent males, a nasopharygeal tumor that exhibits intense contrast enhancement with intratumoral signal voids and extends into the pterygopalatine fossa with the destruction of the pterygoid processes strongly supports the diagnosis of a JNA. Also, it may include cystic components.

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