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Case Report

The role of diagnostic and interventional radiology in juvenile nasopharyngeal angiofibroma: A case report and literature review

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ABSTRACT

Juvenile Nasopharyngeal Angiofibroma is a rare and locally aggressive benign tumor. It is characterized by high vascularization leading to spontaneous bleeding or massive hemorrhage. We reported a case of 13 years old boy with a swollen at the right face, nasal congestion, recurrent nose bleeding, hyposmia, headache, breathing difficulty, and a nasopharyngeal mass at CT scan. Angiography examination showed a highly vascularized mass which was successfully occluded through preoperative embolization procedures. It is suggested that imaging modalities is not only playing an important role in diagnostic process, but also as an adjunct treatment approach in patient with Juvenile Nasopharyngeal Angiofibroma.

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Introduction

Juvenile Nasopharyngeal Angiofibroma (JNA) is a rare benign tumor that accounts for 0.05%–0.5% of all the head and neck tumors [1,2]. This tumor mostly affected men from aged 7 to 29 years old [1]. The JNA is located at the posterior nasopharynx and sphenopalatine foramen [3]. The characteristics of this tumor are locally aggressive and highly vascularized resulting in prone to bleeding [1–6]. The manifestations of this tumor are unilateral swelling of the face, nasal congestion and hyposmia [1–6].

As for the diagnostic process, imaging modalities are more preferred than biopsy owing to the high bleeding risk. CT scan is one of the imaging modalities that commonly used in JNA

cases to evaluate the tumor and bone destruction caused by the aggressiveness of this tumor [1–6]. A surgery remains the definitive treatment of JNA, but nowadays interventional radiology procedures play an important role in the treatment process. Here, we report a patient with JNA tumor and the role of imaging modalities from the diagnostic process until the treatment.

Case illustration

A 13 year old boy came to the clinic in March 2014 and complained about the swelling at his right face that was getting bigger for the past 4 months. The patient also experienced

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Fig. 1 – Clinical presentation of the patient.



Fig. 2 – The CT of paranasal sinus in Feb 2014.

nasal congestion, recurrent nose bleeding for around 100 cc, hyposmia, proptosis, diplopia, headache, and breathing difficulty. No other abnormalities were found in both physical and laboratory examinations (Fig. 1).

The patient underwent a paranasal sinus CT scan. Based on the examination, there were soft tissue tumor at the right nasal cavity, right sphenoid sinus, posterior ethmoid sinus, nasopharynx and pterygopalatine fossa sized $5 \times 5 \times 4.5$ cm as well as the destruction of inferior right sphenoid sinus wall. It was corresponded well with the JNA (Fig. 2). A biopsy procedure was then carefully performed to avoid the bleeding, and the result demonstrated a connective tissue stroma which consists of spindle cells with vascular patterns; as such was confirmed the diagnosis of JNA.

Further carotid angiography examination showed a highly vascularized tumor at the right internal maxillary artery (Fig. 3A) and branches of external right carotid artery (Fig. 3B). The patient then underwent preoperative embolization as an adjunct therapy at 24 hours before the surgery to reduce the risk of bleeding. The procedure was conducted at the right internal maxillary artery (Fig. 4), right pharyngeal artery (Fig. 5) and right facial artery (Fig. 6). The embolization was accomplished successfully and then the treatment was fol-

lowed by mass extirpation, maxillectomy and reconstruction surgery.

Discussion

The patient in this case was a 13 year old boy, which was well corresponded with a study of Parikh et al that reported JNA affecting a man within the range of age 7 to 29 years old [1]. The findings such as a recurrent nose bleeding, hyposmia, breathing difficulty, proptosis and diplopia are considered as complications of extensive tumor growth [2–4]. Imaging modalities are necessary to evaluate these manifestations.

Based on the study conducted by Gupta et al [2], a CT scan was a reliable tool to diagnose JNA because it can evaluate the bone destruction, widening between foramens and fissures in cranium. For further evaluation of tumor extension, MRI is more preferred than CT because of its ability to explore the soft tissues in intracranial region [5]. A mass in both nasopharyngeal region and pterygopalatine fossa, and the destruction of inferior right sphenoid sinus wall are suggested to be strongly associated with the complications of this case.

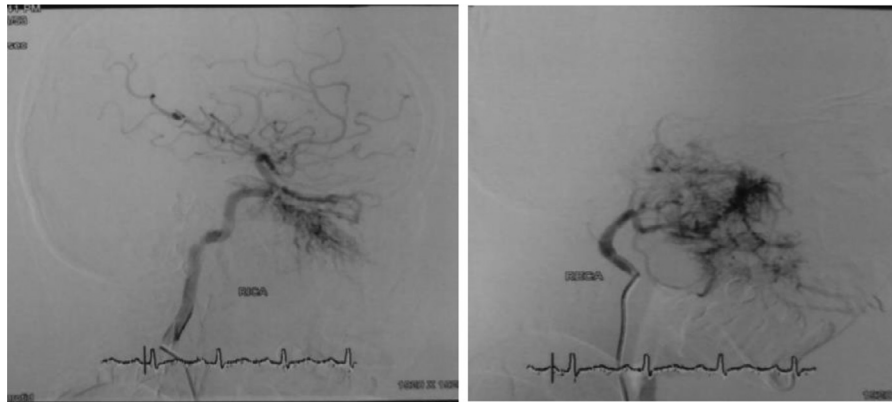


Fig. 3 – Tumor vascularization (a) branches of right internal carotid artery (left) and (b) branches of right external carotid artery (right).



Fig. 4 – The angiography of right internal maxillary artery before (left) and after (right) embolization.

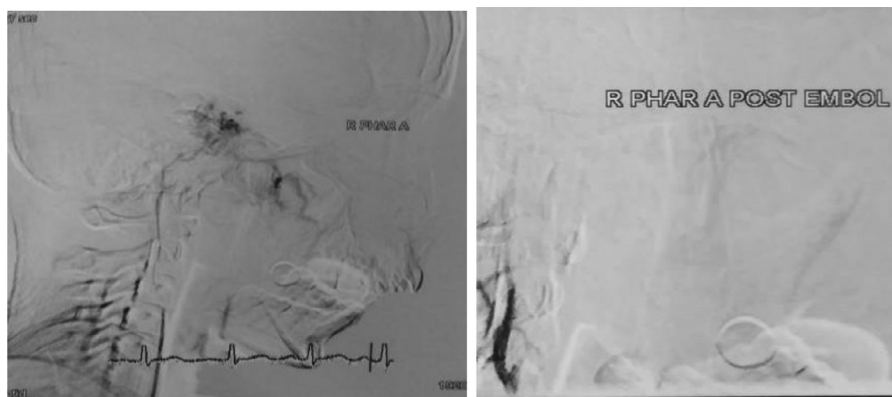


Fig. 5 – The angiography of right pharyngeal artery before (left) and after (right) embolization.

These findings are also similar with the reports conducted by Gupta et al [2] and Atalar et al [4].

Further examination using carotid angiography showed feeding arteries from the branches of Internal Carotid Artery (Internal Maxillary Artery) and External Carotid Artery (Facial and Pharyngeal Arteries). This condition is aligned with the studies reported by Parikh et al [1], Ballah et al [3], Atalar et al [4] and Sirakov et al [6] suggesting that branches of ex-

ternal and internal carotid arteries can become the feeding arteries.

Preoperative embolization in this case was performed within 24 hours prior to the surgery. Sirakov et al also reported that this procedure should be performed 24-72 hours before undergoing the surgery in order to occlude the arteries [6]. Preoperative embolization is proven to be safe as well as effective to avoid the fatal intraoperative hemorrhage, shorten

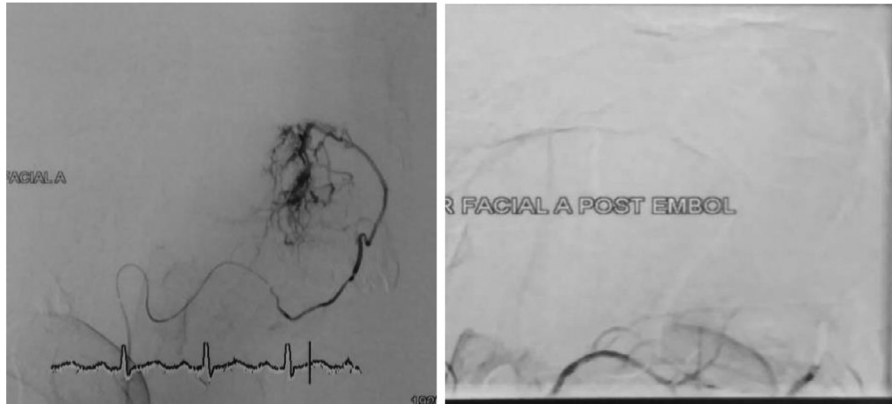


Fig. 6 – Angiography of right facial artery before (left) and after (right) embolization.

the surgery durations, and widen the surgeon's field of view. As a result, the tumor can be resected perfectly [1].

This study requires some improvements. We strongly recommend performing the post resection histopathology examination and postsurgical CT scan to confirm the final diagnosis.

Conclusion

The CT scan offers a substantial benefit in determining the mass and bone destruction caused by the JNA tumor. Moreover, angiography is also essential to evaluate the feeding arteries of the tumor. According to these findings combined with preoperative embolization as an adjunct therapy, we suggest that imaging modalities can be crucial during both diagnostic and treatment processes in a patient with JNA tumor.

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