



Case Report

Pre-pubertal juvenile nasopharyngeal angiofibroma with intracranial extension

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Abstract

Juvenile nasopharyngeal angiofibroma (JNA) is a rare benign vascular tumor predominant in adolescent and adult young male. It represents only 0.05 to 0.5% of all head and neck neoplasms. Clinical presentations of JNA are dominated by recurrent and unprovoked epistaxis, and nasal obstruction. More rarely other unusual clinical manifestations can be observed, especially in forms invading oropharyngeal structures and skull base. Headache and exophthalmia are exceptional. The JNA occurring in the pre-pubertal period can pose a diagnostic challenge for clinicians because of their often-atypical clinical presentations, and the absence of conventional radiological features at this age. We report an original observation of JNA with endocranial extension in pre-pubertal boy revealed by persistent headache and unilateral exophthalmia.

Keywords: Juvenile nasopharyngeal angiofibroma, Endocranial extension, Headache, Exophthalmia, Pre-pubertal period.

Introduction

Juvenile nasopharyngeal angiofibroma (JNA), or juvenile angiofibroma according to the new World Health Organization terminology and classification of head and neck tumors¹ is a rare benign vascular tumor predominant in adolescent and adult young male. [2-5]

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It represents only 0.05 to 0.5% of all head and neck neoplasms. [4-6] This tumor is characterized by significant local aggression [3-5] and a high rate of morbidity and mortality, mainly due to the existence of endocranial extension.²

The JNA occurring in the pre-pubertal period can pose a diagnostic challenge for clinicians because of their often-atypical clinical presentations, and the absence of conventional radiological features at this age⁷; it can be fatal.⁸

We report an original observation of JNA with endocranial extension in pre-pubertal boy revealed by persistent headache and unilateral exophthalmia.

Case report

10-year-old boy, with no medical history, was referred to us by his family doctor for persistent and resistant to analgesic symptomatic treatment headaches. These headaches had been associated for two weeks with a progressive right exophthalmia.

Somatic examination was normal except for painless moderate right exophthalmia. Specialized neurological examination and blood pressure profile were correct as well as visual acuity and fundus of both eyes. Nor was there any abnormality in the examination of the thyroid compartment or cervical lymphadenopathy. The basic biologic parameters were within normal limits. The ear nose throat examination with nasal endoscopy showed a whitish tumor of the right nasal fossa, polylobulated, traversed by numerous blood vessels, and which reached the level of the head of the inferior turbinate. The craniofacial computed tomography (CT) showed a lobulated and no contrast-enhanced tumor located in the right nasal fossa, and extending to right ethmoid cells, right maxillary sinus, right infra-temporal fossa, and cavernous sinus (Figure 1). Craniofacial magnetic resonance imaging (MRI) confirmed these findings by showing a heterogeneous lobulated tumor of the right nasal fossa, with isosignal on T1-weighted sequences (Figure 2), heterogeneous hypersignal on T2-weighted sequences (Figure 3), and strongly enhancement after gadolinium injection (Figure 4). This tumor extended to the right ethmoid cells, right maxillary sinus, right infra-temporal fossa, and cavernous sinuses. The angiographic sequences (angio-MR) objectified the richness of the peri-tumor vascularization (Figure 5). The surgery was refused by the parents; the child was then treated with external radiotherapy.

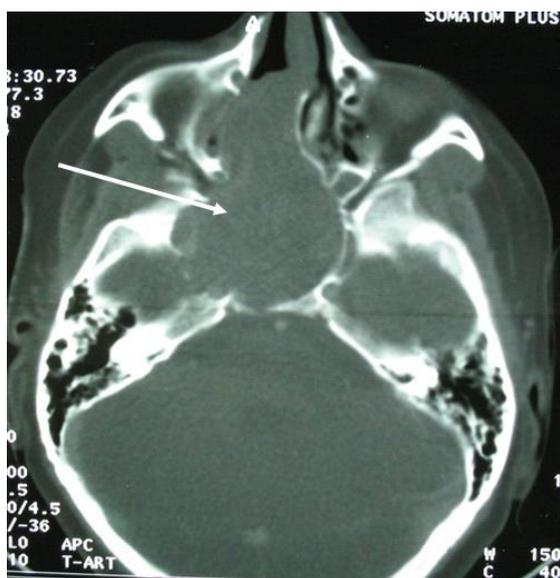


Figure 1. Axial craniofacial CT in bone window: naso-ethmoidal expansive process with extension to the right infra-temporal fossa.



Figure 2. Axial T1-weighted craniofacial MRI without injection: isointense right nasopharyngeal mass with endocranial extension.

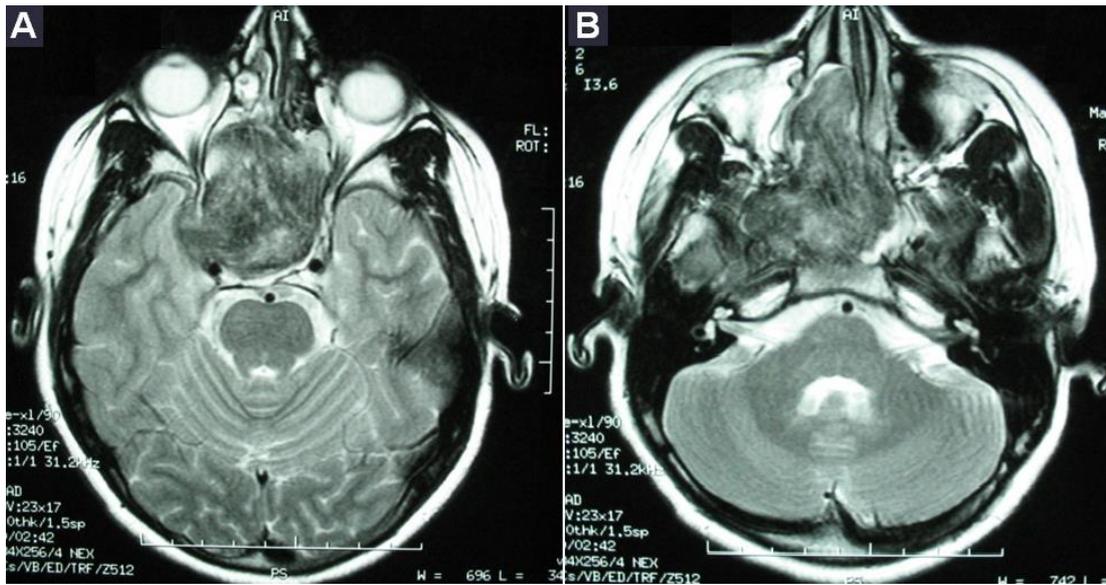


Figure 3. Axial T2-weighted craniofacial MRI sections passing through the nasopharynx (A) and the base of the skull (B): heterogeneously hyperintense mass extending to the right cavernous sinus with moderate right exophthalmia.



Figure 4. T1-weighted craniofacial MRI with gadolinium injection in axial (A), sagittal (B) and coronal (C) planes: heterogeneously and strongly enhanced right nasopharyngeal mass with extension to supra-sellar region and right infra-temporal fossa.

Discussion

Clinical presentations of JNA are dominated by recurrent and unprovoked epistaxis, and nasal obstruction. [1-8] More rarely other unusual clinical manifestations can be observed, especially in forms invading oropharyngeal structures and skull base [9,10] Headache and exophthalmia are exceptional. [7,9,10]

Exophthalmos is the witness of an orbital extension of the tumor. [9,10] The headaches are seen mainly in the evolved forms of the tumor with endocranial extension. This complication of the tumor occurs particularly in adolescents; indeed, in the series of Mattei TA et al, 20 cases among the 67 adolescents treated surgically for JNA, had an intracranial extension (29.85%).¹¹



Figure 5. Angio-MR showing the hypervascular character of the mass.

The diagnosis can be made based on the clinical and very specific radiological aspects (CT, MRI, and angio-MR) without the need for biopsy. [12-14] Surgical excision is the treatment of choice for these tumors; preoperative embolization reduces the risk of intraoperative bleeding [2,5,11,15,16]. Traditional or endoscopic surgery for JNA with intracranial extension represents a real challenge for ENT and neurosurgeons' practitioners [2,6,11,16].

External radiotherapy may be a therapeutic alternative for advanced forms of JNA found to be inoperable, even with significant intracranial extension, with satisfactory results. ⁸

Postoperative recurrences are relatively common.³ Their frequency is estimated at 20% ¹ justifying that regular radiological monitoring is recommended.¹⁶

Conclusion

Although exceptional and benign, the JNA deserves to be well known because of the potential risk of morbidity and mortality. Endocranial extension is a major factor for both therapeutic choice and prognosis. Imaging, in particular MRI and MR-angiography, greatly facilitated the non-invasive diagnosis of this tumor.

Our observation is characterized by its prepubertal character and the unusual revealing symptoms.

Author contributions

NR, SY, MG, SB took care of the patient and made the literature research. SB, MG drafted the manuscript and supervised the manuscript. The final version has been read and approved by all authors.

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Conflict of interest

All authors declare that they have no conflict of interest.

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