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(CASE REPORT)

Radiotherapy in advanced recurrent juvenile nasopharyngeal angiofibroma: A case report

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Abstract

Juvenile nasopharyngeal angiofibroma (JNA) is a rare, highly vascular benign tumor characterized by locally aggressive behavior. While surgery remains the standard treatment, management of recurrent or advanced disease with skull base extension remains challenging.

We report the case of a 15-year-old male presenting with recurrent JNA initially treated surgically. The patient subsequently developed recurrence managed with embolization followed by incomplete resection. Further imaging demonstrated locoregional progression with skull base and intracranial extension. Given the extent of disease and limited surgical options, definitive radiotherapy was delivered to a total dose of 50 Gy. Treatment was well tolerated, with no significant acute or late toxicity. At follow-up, the patient showed complete clinical response and progressive radiological regression with no evidence of disease progression.

Radiotherapy represents an effective and well-tolerated treatment option for recurrent or unresectable JNA, particularly in advanced stages when surgical management is not feasible.

Keywords: Juvenile Nasopharyngeal Angiofibroma; Radiotherapy; Recurrent Disease; Skull Base Extension; Embolization; Intracranial Extension

1. Introduction

Juvenile nasopharyngeal angiofibroma (JNA) is a rare, highly vascular tumor that predominantly affects adolescent males, typically during the second decade of life [1,2]. Despite its benign histological nature, it demonstrates locally aggressive behavior with progressive extension into adjacent structures such as the nasal cavity, paranasal sinuses, pterygopalatine fossa, infratemporal fossa, and skull base [2,3].

Clinically, JNA most commonly presents with recurrent epistaxis and progressive nasal obstruction. Advanced disease may be associated with facial deformity or neurological symptoms due to skull base or intracranial extension [3]. Owing to its marked vascularity, biopsy is contraindicated, and diagnosis relies primarily on imaging. Computed tomography (CT) and magnetic resonance imaging (MRI) are essential for assessing tumor extent and guiding management, while angiography plays a key role in identifying feeding vessels and facilitating embolization [4,5].

Surgical resection remains the standard treatment, with endoscopic approaches increasingly favored for early and intermediate stages [2,6]. However, management of advanced or recurrent JNA remains challenging, particularly in

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cases with skull base or intracranial extension where complete surgical excision may not be feasible or is associated with significant morbidity [3,7]. In such settings, radiotherapy represents an effective therapeutic option, providing high rates of local control with acceptable toxicity, especially with modern techniques such as intensity-modulated radiotherapy (IMRT) [7–9].

We report a case of recurrent, locally advanced JNA with intracranial extension treated with definitive radiotherapy, highlighting the role of this modality in achieving durable disease control when surgical management is limited.

2. Case Presentation

A 15-year-old male with no significant past medical or family history was referred to our department for management of a recurrent juvenile nasopharyngeal angiofibroma.

The disease initially presented in 2019, at the age of 10 years, with recurrent epistaxis associated with progressive unilateral nasal obstruction and hyponasal speech. There were no associated symptoms such as rhinorrhea, facial pain, hearing impairment, or history of craniofacial trauma. Endoscopic evaluation revealed a vascularized mass occupying the left nasal cavity, for which the patient underwent surgical resection. Histopathological examination confirmed juvenile nasopharyngeal angiofibroma.

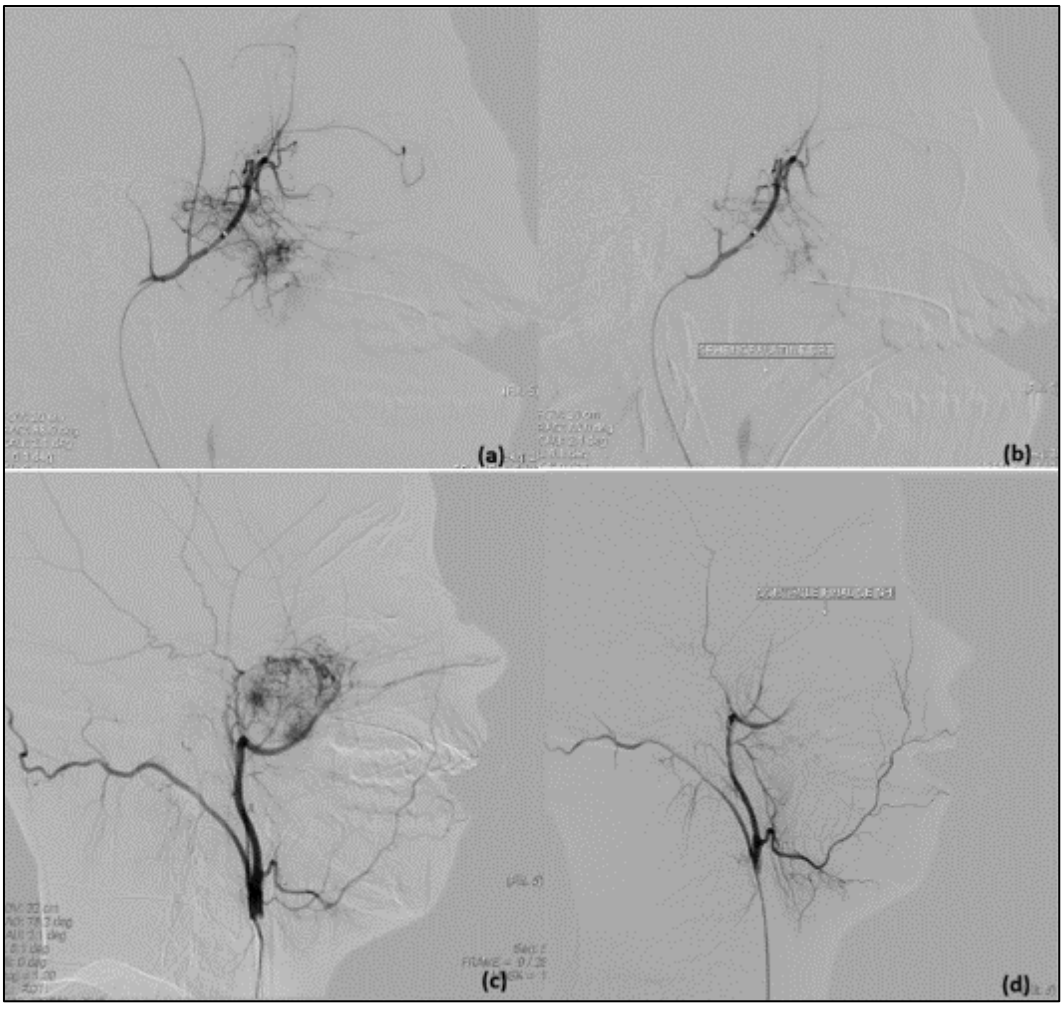

Figure 1 Angiography illustrating the vascularization of a juvenile nasopharyngeal angiofibroma. (a) Selective angiography demonstrating a hypervascular tumor blush. (b) Angiography and embolization of the sphenopalatine artery. (c) Lateral projection showing tumor staining with irregular vascularity. (d) Control angiography following embolization demonstrating a reduction in tumor vascularity

In 2023, the patient developed recurrent epistaxis and nasal obstruction. Imaging confirmed tumor recurrence, and he underwent preoperative embolization followed by a second surgical procedure. Resection was incomplete due to tumor extension, and histopathological analysis again confirmed the diagnosis of JNA.

Follow-up magnetic resonance imaging demonstrated further locoregional progression of the lesion, prompting referral to our institution for definitive management.

At presentation, the patient was in good general condition (ECOG performance status 0–1). Laboratory evaluation revealed mild anemia (hemoglobin 10.5 g/dL), attributed to chronic blood loss, for which iron supplementation was initiated. There was no history of blood transfusion.

Clinical examination showed a mass filling the left nasal cavity on anterior rhinoscopy, while the contralateral nasal cavity was unremarkable. Otologic and oropharyngeal examinations were normal, and no cervical lymphadenopathy was detected.

Magnetic resonance imaging revealed a highly vascular mass centered in the left nasopharynx, causing obstruction of the nasal cavity and choana. The tumor extended laterally into the pterygopalatine and infratemporal fossae, superiorly toward the skull base, and involved the medial orbital wall. Additional findings included invasion of the clivus, extension into the maxillary sinus, infiltration of the medial pterygoid muscle, involvement of the superior orbital fissure, and intracranial extension. These features were consistent with advanced recurrent disease classified as Radkowski stage IIIb.

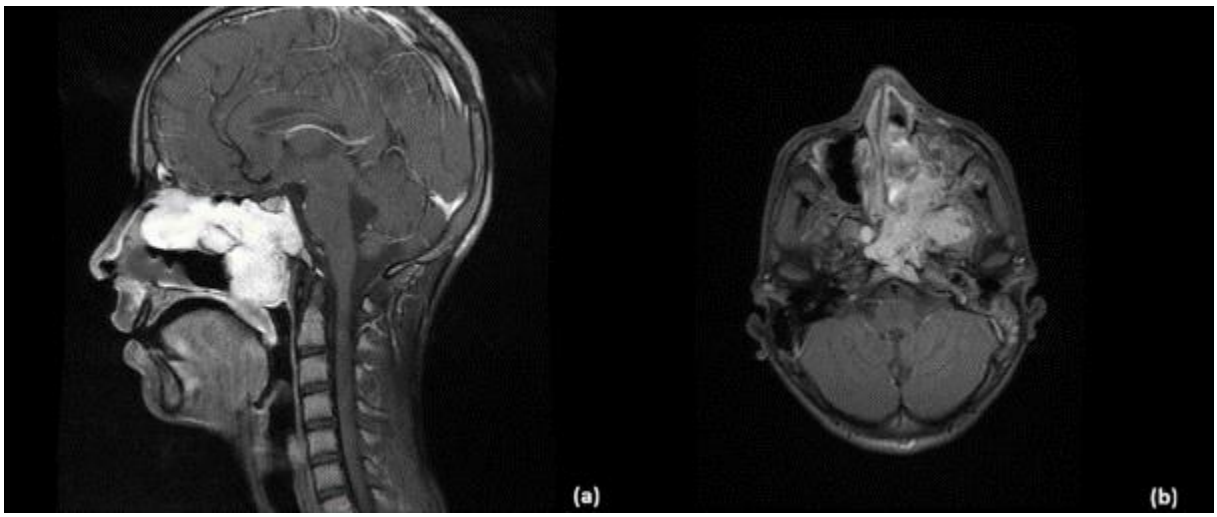

Figure 2 Magnetic resonance imaging of a juvenile nasopharyngeal angiofibroma. (a) Sagittal contrast-enhanced T1-weighted image showing a large enhancing mass centered in the nasopharynx with extension toward the skull base. (b) Axial contrast-enhanced T1-weighted image demonstrating a heterogeneous enhancing lesion involving the nasal cavity and adjacent structures

Histopathological examination confirmed a benign angiofibroma without evidence of malignant transformation.

Given the extent of disease, characterized by skull base and intracranial involvement, surgical management was deemed unsuitable. The case was discussed in a multidisciplinary tumor board, and the decision was made to proceed with definitive radiotherapy.

The patient underwent CT-based simulation in the supine position using thermoplastic immobilization, with MRI fusion for target delineation. The gross tumor volume (GTV) was defined according to MRI findings, and a 5 mm margin was applied to generate the planning target volume (PTV), without a separate clinical target volume. Intensity-modulated radiation therapy (IMRT) was delivered to a total dose of 50 Gy in 25 fractions. Dose constraints to organs at risk, including the optic pathways, brainstem, and temporal lobes, were respected.

Treatment was well tolerated, with no clinically significant acute toxicities observed.

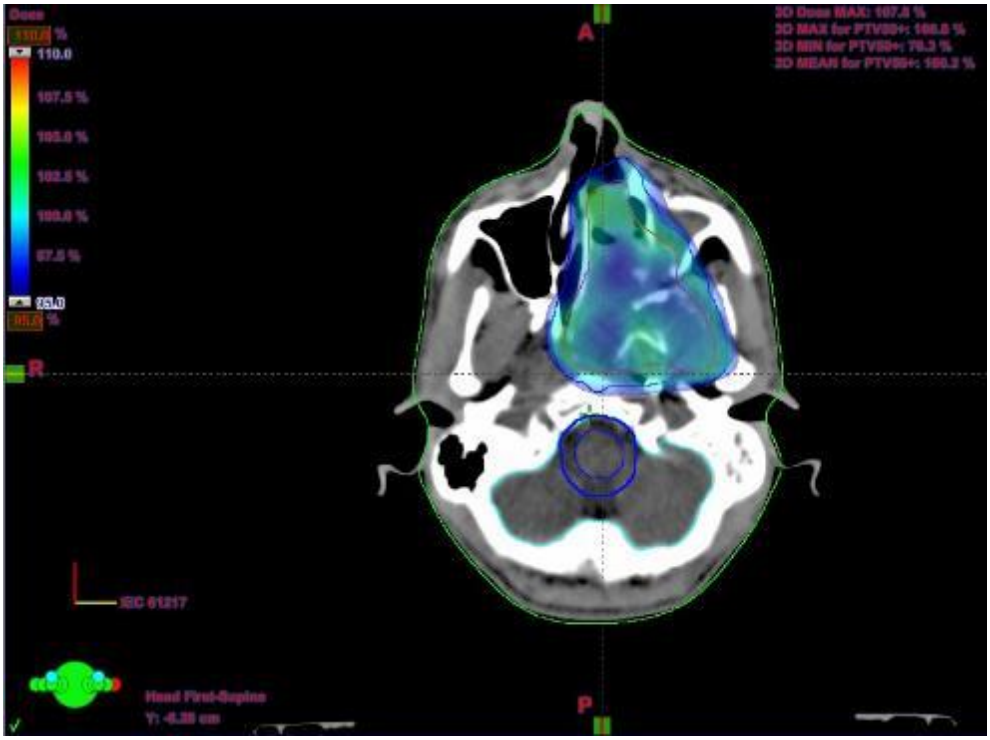

Figure 3 Axial CT-based treatment planning image illustrating dose distribution of intensity-modulated radiotherapy. The planning target volume is covered by the prescribed isodose, with sparing of adjacent organs at risk

At 18 months of follow-up, the patient remains in good clinical condition, with complete resolution of epistaxis and nasal obstruction. Serial MRI evaluations demonstrated progressive tumor regression with sustained radiological stability. No evidence of local progression been observed. Furthermore, no late radiation-induced complications, including optic neuropathy, temporal lobe necrosis, endocrine dysfunction, or ocular toxicity, have been identified to date.

3. Discussion

Juvenile nasopharyngeal angiofibroma (JNA) is a benign but locally aggressive tumor for which surgery remains the standard treatment. However, management becomes particularly challenging in recurrent or advanced disease, especially in the presence of skull base or intracranial extension, where complete resection may be difficult and associated with increased morbidity [10]. In such situations, repeated surgical interventions may fail to achieve durable local control.

In the present case, disease progression occurred despite prior surgery and embolization, with incomplete resection followed by further radiological progression. This clinical course is consistent with previously reported patterns in which radiotherapy is indicated following failure of surgical management or when further resection is not feasible [11].

Radiotherapy has demonstrated high efficacy in advanced JNA, with reported local control rates ranging from approximately 80% to 100% in unresectable or recurrent cases [12,14]. It allows treatment of tumor extensions that are difficult to access surgically, particularly at the skull base. Our findings are consistent with these data, with sustained clinical and radiological control observed at 18 months.

Dose selection remains heterogeneous across studies. Most series report effective tumor control with doses ranging from 30 to 45 Gy, while higher doses around 50 Gy are frequently employed in cases of bulky, progressive, or recurrent disease [14,15]. Rare reports have described further dose escalation up to 60 Gy in selected aggressive cases, with satisfactory tumor control, although such approaches remain exceptional and are not routinely recommended. In this context, the use of 50 Gy in our patient appears appropriate given the locally advanced recurrent presentation and intracranial extension.

Tumor response following radiotherapy is typically gradual, and persistent radiological abnormalities in early follow-up do not necessarily indicate treatment failure. Previous studies have shown that radiological regression may continue

for several months after treatment [15]. In our case, serial MRI demonstrated progressive tumor reduction associated with complete clinical resolution of symptoms, consistent with the expected post-radiotherapy evolution.

The use of intensity-modulated radiotherapy (IMRT) is particularly advantageous in JNA due to the complex tumor geometry and proximity to critical structures. IMRT enables conformal dose distribution with improved sparing of adjacent organs at risk, including the optic pathways, brainstem, and temporal lobes [13]. This is especially important in young patients, in whom long-term toxicity must be minimized. In our case, IMRT allowed adequate target coverage with no significant acute or late toxicity observed during follow-up.

Within the evolution of radiotherapy techniques, proton therapy has emerged as a promising modality for the management of advanced JNA. By providing improved dose distribution and reduced exit dose, proton therapy may further decrease irradiation of surrounding normal tissues. Recent series have reported excellent local control with minimal toxicity, along with a potential reduction in long-term risks such as neurocognitive impairment and secondary malignancies [16]. However, limited availability currently restricts its widespread use, and IMRT remains a highly effective and accessible alternative in most clinical settings.

Overall, this case supports the role of definitive radiotherapy in recurrent and advanced JNA, demonstrating that modern techniques such as IMRT can achieve durable local control with minimal toxicity, even after failure of prior surgical management.

4. Conclusion

Definitive radiotherapy represents an effective therapeutic option for recurrent or unresectable juvenile nasopharyngeal angiofibroma, particularly in advanced stages with locoregional extension. It provides durable local control with acceptable toxicity and should be considered when surgical management is not feasible or has failed.

Compliance with ethical standards

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Disclosure of conflict of interest

All authors declare that there are no conflicts of interest regarding the publication of this manuscript. The authors declare no competing financial or non-financial interests.

Statement of informed consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. According to institutional policy, ethical committee approval is not required for single case reports.

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