Egyptian Journal of Radiology and Nuclear Medicine

CASE REPORT Open Access

Imaging findings of juvenile nasopharyngeal angiofibroma invading orbital apex and middle cranial fossa: a case report

Roberto Castellana[1*], Gianluca Fanelli[1], Gianmarco Lunardi[1], Massimo Rosset[1], Stefano Piccolo[2], Irene Ariozzi[1], Mara Oggero[1], Roberto Castellana[3], Pierpaolo Petti[4], Monica Alessi[1] and Massimiliano Natrella[1]

Abstract

Background Juvenile nasopharyngeal angiofibroma (JNA) is an infrequent, highly vascular tumor that predominantly affects male adolescents. Although benign, it rarely may exhibit invasive growth, leading to significant morbidity, as in our case presentation. Diagnosis relies on clinical evaluation and imaging, with MR and CT playing vital roles in confirming the tumor's presence, determining its extent, and aiding in treatment planning. MR shows a mass with signal voids at T2-weighted images and intense contrast enhancement at T1-weighted images, while CT shows bone remodeling and destruction. A biopsy is usually avoided due to bleeding risks.

Case presentation We present a case of a 16-year-old male with nasal obstruction and recurrent epistaxis from the right nostril, headache, and mucopurulent rhinorrhea. Furthermore, he had vision impairment in the right eye for one week associated with proptosis and periorbital swelling. MR and CT imaging evidenced the presence of JNA with extensive involvement in the nasal cavity, nasopharynx, pterygopalatine fossa, and infratemporal fossa. Moreover, the mass invaded the orbital apex and middle cranial fossa contacting the cavernous sinus and internal carotid artery. Preoperative embolization and complete surgical resection were performed. No complications occurred in the postoperative period and histopathological analysis confirmed the diagnosis of JNA. No recurrences were found at 12 months MR follow-up.

Conclusions Early detection and accurate imaging evaluation are essential for the effective management of JNA. Regular post-treatment MR follow-ups are crucial to detect recurrences or assess fibrosis stability. MR and CT continue to be indispensable tools in the diagnosis and follow-up of JNA.

Keywords Case report, Computed tomography, Juvenile nasopharyngeal angiofibroma, Magnetic resonance, Middle cranial fossa, Orbital invasion

Background

Juvenile nasopharyngeal angiofibroma (JNA) is a rare tumor with an incidence of approximately 1:150,000, accounting for about 0.05% of all head and neck tumors [1, 2].

The pathogenesis of JNA is still unclear and hormonal and genetic factors have been suggested to explain the almost exclusive occurrence of JNA in male adolescents with an average age at diagnosis of 14–16 years [3, 4].

*Correspondence:
Roberto Castellana
castellanaroberto@yahoo.it
[1] Local Health Authority of Valle d'Aosta: Azienda Unita Sanitaria Locale Della Valle d'Aosta, Aosta, Italy
[2] Università Degli Studi Di Torino, 10126 Turin, Italy
[3] ASL Bari, Servizio Di Radiologia Di Conversano, 70014 Conversano, Italy
[4] Via Gramsci, 5, 70017 Putignano, Italy

JNA is a histologically benign but highly vascular tumor, with an invasive and destructive growth, that causes significant morbidity. Malignant transformation is extremely rare and associated with recurrent radiotherapy [5].

JNA generally originates in the posterior nasal cavity, near the superior margin of the sphenopalatine foramen, and early extends medially into the area of least resistance—adjacent nasal cavity and nasopharynx—and, laterally, into pterygopalatine fossa, enlarging the sphenopalatine foramen and eroding the bone behind it. JNA may further extend laterally into the infratemporal fossa and cause anterior displacement of the posterior maxillary wall. As JNA grows posteriorly, it may encounter various points of minor resistance, potentially leading to its involvement with critical anatomical structures represented by the internal carotid artery (ICA), accessible through the vidian canal, the cavernous sinus via the foramen rotundum, and the orbital apex through the inferior orbital fissure. Moreover, it may extend from the orbital apex to the middle fossa of the skull through the superior orbital fissure [6–8].

Bone involvement in JNA can happen through two main mechanisms: direct pressure resorption and spread along perforating arteries. The tumor's growth exerts direct pressure on the adjacent bone, leading to the gradual resorption or erosion of the bone tissue. This pressure-induced resorption weakens the bone and allows the tumor to infiltrate further. Moreover, JNA can extend its growth along the perforating arteries that pass through the bone. By utilizing these pathways, the tumor can invade the surrounding bone tissue [6, 7].

Typical clinical presentation includes progressive unilateral nasal obstruction (80–90%) with rhinorrhea and recurrent unilateral epistaxis (45–60%) [1, 6]. Additional symptoms can include headaches and facial pain caused by the obstruction of the paranasal sinuses, as well as unilateral secretory otitis media resulting from compromised Eustachian tube function. In rare and advanced cases, proptosis and alteration of the vision may be also present and indicate the involvement of the orbit [6, 9].

The preoperative diagnosis is primarily based on clinical and imaging characteristics while performing an incisional biopsy carries the risk of significant bleeding [1].

Preoperative imaging with CT and MR plays a crucial role in confirming the diagnosis, determining the extent and stage of the tumor, and aiding in treatment planning. Furthermore, postoperatively, it is utilized to evaluate tumor persistence or recurrence.

Here, we report MR and CT findings of a rare and very large JNA in a 16-year-old male.

The article was written according to CARE (CAse REport) guidelines [10] and the CARE checklist is reported in Table 1.

Case presentation

A 16-year-old male presented to the outpatient department of otorhinolaryngology with the chief complaint of worsening nasal obstruction from the right nostril, headache, and mucopurulent rhinorrhea. His symptoms had begun four months before and he also reported multiple episodes of epistaxis from the right nostril. Furthermore, he had vision impairment in the right eye for one week associated with proptosis and periorbital swelling. The oculomotor function was preserved. His past medical history was not significant. The relevant events in the patient's history are presented in Table 2.

Endoscopic examination revealed a large mass with a smooth surface and clear signs of hypervascularization that obstructed the right nasal cavity and extended posteriorly in the nasopharynx, requiring further assessment with an urgent MR.

A MR of the head, orbits, and maxillofacial regions with contrast medium (gadobutrol) was performed with a 1.5 T MR scan (GE SIGNA) using axial T1-weighted SE, axial and coronal T2-weighted FSE, coronal T2-weighted FSE with fat suppression, axial diffusion-weighted, axial T2-weighted FLAIR and post-contrast 3D T1-weighted CUBE sequences.

A large mass measuring about 9 cm and centered in the right sphenopalatine foramen was evidenced (Fig. 1). The expansile lesion invaded the right nasal cavity, nasopharynx, sphenoid sinuses, ethmoid cells, right pterygopalatine fossa, and right infratemporal fossa, causing prominent bowing of the posterior maxillary wall and flattening of the right maxillary sinus. Moreover, the mass cranially invaded the right orbital apex, contacting the optical nerve, and the right middle cranial fossa, contacting the cavernous sinus and internal carotid artery (Fig. 2).

The lesion was characterized by an intermediate signal at T1-weighted images, an intermediate-to-high heterogeneous signal with serpentine flow voids and degenerative cystic components at T2-weighted images, and intense and heterogeneous contrast enhancement with small intratumoral vessels. No portions of the mass with a high signal at DWI images were evidenced.

Moreover, the right maxillary artery was slightly enlarged in comparison to the left side and the right optic nerve showed no evident alterations (Fig. 3).

A JNA was suspected based on MR findings. To gain a clearer understanding of bone involvement, a non-contrast maxillofacial CT scan was conducted.

Table 1 CARE Checklist

Topic	Item	Checklist item description	Reported on
Title	1	The diagnosis or intervention of primary focus followed by the words "case report	Title
Key words	2	2 to 5 key words that identify diagnoses or interventions in this case report, including "case report"	Keywords
Abstract	3a	Introduction: What is unique about this case and what does it add to the scientific literature?	Abstract, P1
	3b	Main symptoms and/or important clinical findings	Abstract, P2
	3c	The main diagnoses, therapeutic interventions, and outcomes	Abstract, P2-3
	3d	Conclusion—What is the main "take-away" lesson(s) from this case?	Abstract, P4
Introduction	4	One or two paragraphs summarizing why this case is unique (may include references)	Background, P8
Patient Information	5a	De-identified patient specific information	Case present., P1
	5b	Primary concerns and symptoms of the patient	Case present., P1
	5c	Medical, family, and psycho-social history including relevant genetic information	Case present., P1
	5d	Relevant past interventions with outcomes	Case present., P1
Clinical Findings	6	Describe significant physical examination (PE) and important clinical findings	Case present., P1-2
Timeline	7	Historical and current information from this episode of care organized as a timeline	Table 1
Diagnostic Assessment	8a	Diagnostic testing (such as PE, laboratory testing, imaging, surveys)	Case present., P3-7
	8b	Diagnostic challenges (such as access to testing, financial, or cultural)	N/A
	8c	Diagnosis (including other diagnoses considered)	Case present., P6
	8d	Prognosis (such as staging in oncology) where applicable	Discussion, P9
Therapeutic Intervention	9a	Types of therapeutic intervention (such as pharmacologic, surgical, preventive, self-care)	Case present., P9
	9b	Administration of therapeutic intervention (such as dosage, strength, duration)	N/A
	9c	Changes in therapeutic intervention (with rationale)	N/A
Follow-up and Outcomes	10a	Clinician and patient-assessed outcomes (if available)	Case present., P9-10
	10b	Important follow-up diagnostic and other test results	Case present., P10
	10c	Intervention adherence and tolerability (How was this assessed?)	N/A
	10d	Adverse and unanticipated events	Case present., P9-10
Discussion	11a	A scientific discussion of the strengths AND limitations associated with this case report	Discussion, P1-9
	11b	Discussion of the relevant medical literature with references	Discussion, P1-9
	11c	The scientific rationale for any conclusions (including assessment of possible causes)	Conclusions
	11d	The primary "take-away" lessons of this case report (without references) in a one paragraph conclusion	Conclusions
Patient Perspective	12	The patient should share their perspective in one to two paragraphs on the treatment(s) they received	N/A
Informed Consent	13	Did the patient give informed consent? Please provide if requested	Yes

Table 2 Relevant events in the patient's history

2021, July	Worsening nasal obstruction from the right nostril, headache, and mucopurulent rhinorrhea
2021, August	First of multiple episodes of epistaxis from the right nostril
2021, November the 18th	Vision impairment in the right eye associated with proptosis and periorbital swelling
2021, November the 25th	MR of the head, orbits, and maxillofacial regions with contrast medium, followed by a maxillofacial CT
2021, December the 13th	Selective embolization of the mass
2021, December the 14th	Surgery
2021, December the 18th	MR follow-up evidencing a small elongated enhancing nodule localized at the right foramen rotundum
2022, December	MR follow-up showing stable findings, consistent with fibrosis

CT demonstrated diffuse bone remodeling, thinning, and destruction that involved the posterior margin of sphenopalatine foramen extending to the base of the medial pterygoid plate, right maxillary sinus, right nasal cavity, ethmoid sinuses, sphenoid sinuses, and anterior skull base. The right superior and inferior

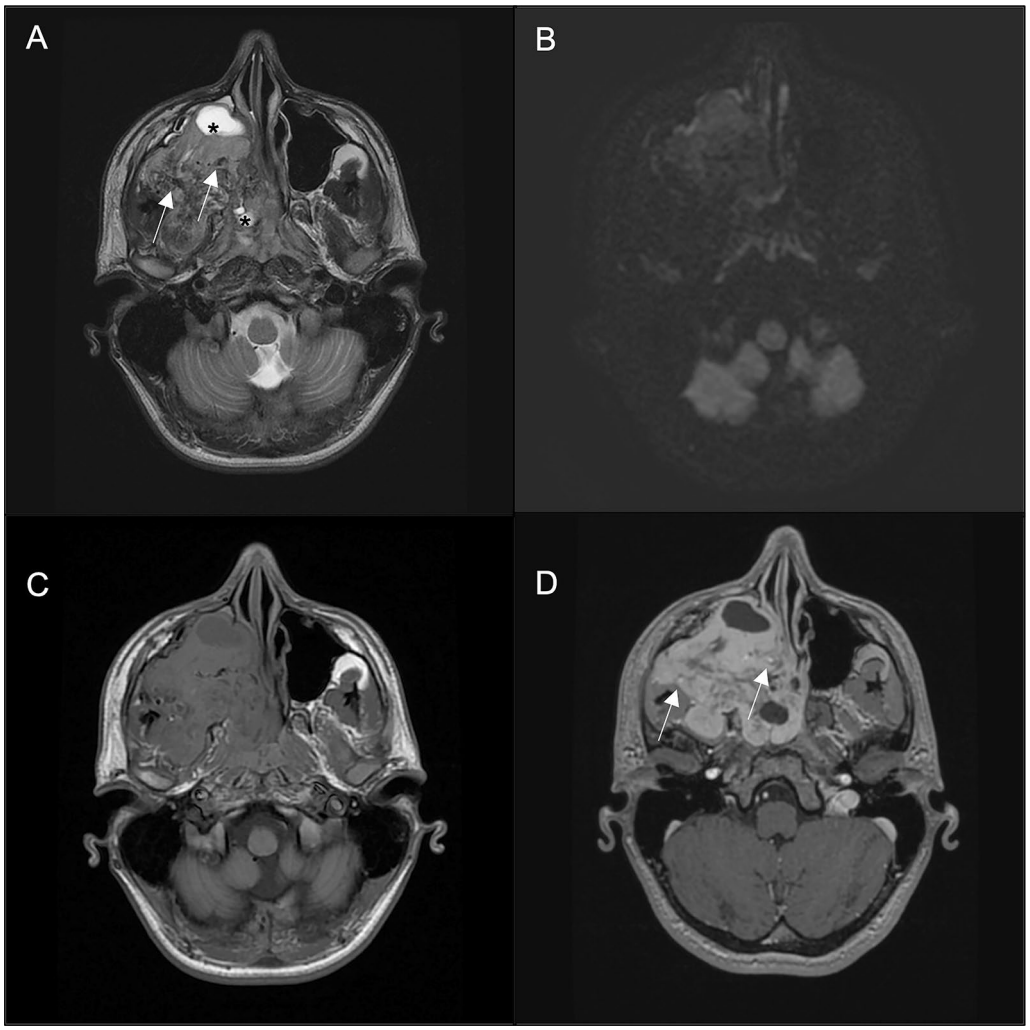

Fig. 1 MR imaging findings of juvenile nasopharyngeal angiofibroma (JNA) in a 16-year-old male with nasal obstruction and recurrent epistaxis from the right nostril and vision impairment from the right eye. At axial T2-weighted images (**A**) JNA shows intermediate-to-high heterogeneous signal with serpentine flow voids (arrows) and degenerative cystic components (asterisks). At axial DWI (**B**), JNA does not present a high signal. At axial non-contrast T1-weighted images (**C**), JNA is predominantly isointense, with intense and heterogeneous enhancement after contrast injection (**D**). Note that some small intratumoral vessels are evidenced after contrast injection (arrow in **D**)

orbital fissures and the right foramen rotundum were enlarged in comparison to the left side. Furthermore, the mass inferiorly extended to the right portion of the hard palate (Fig. 4).

To prevent the risk of significant bleeding, a biopsy was avoided and the patient was urgently transferred to a reference center specialized in JNA treatment. There, preoperative embolization and complete surgical resection were performed. No complications occurred in the postoperative period and histopathological analysis confirmed the diagnosis of JNA.

The post-treatment MR examination, performed 4 days after surgery, evidenced the presence of a small elongated enhancing nodule localized at the right foramen rotundum, this finding was stable at the 6 and 12-month MR follow-ups and, therefore, was consistent with fibrosis.

Discussion

Juvenile nasopharyngeal angiofibroma is typically suspected based on the characteristic symptoms observed in a male adolescent [6]. The first step in the evaluation process usually involves conducting a nasal endoscopy. A sizable, lobulated mass located behind the middle turbinate is usually revealed by this examination [6, 11]. CT and/or MR are crucial for confirming the diagnosis and demonstrating the extension of the mass [6]. CT excels at demonstrating bone erosions while MR is more

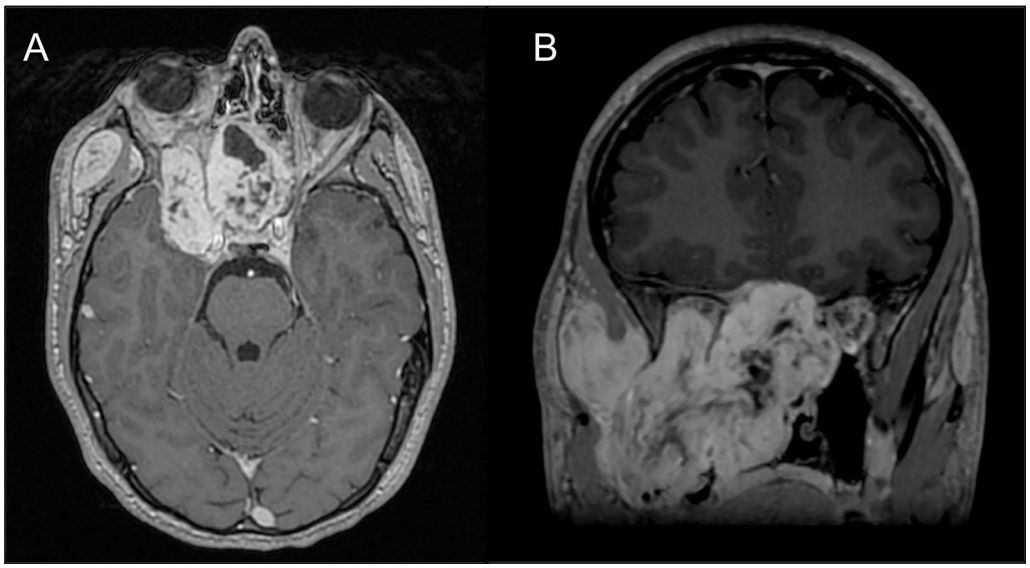

Fig. 2 Axial (**A**) and coronal (**B**) contrast-enhanced T1-weighted MR images show that the juvenile nasopharyngeal angiofibroma invades the right orbital apex, contacting the optical nerve, and the right middle cranial fossa, contacting the cavernous sinus and internal carotid artery

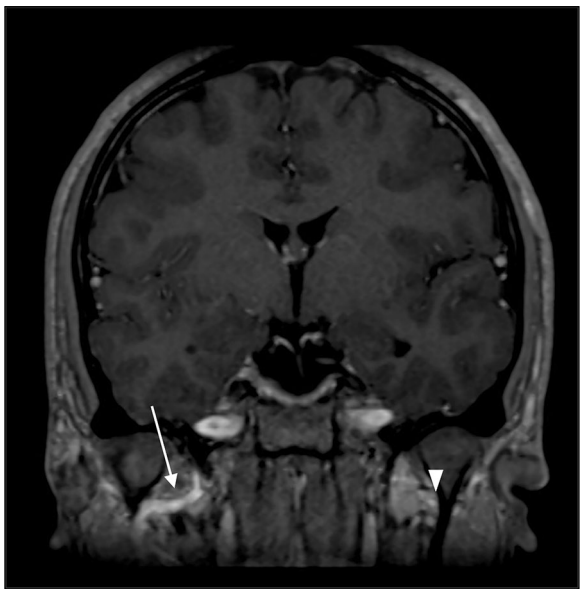

Fig. 3 Coronal contrast-enhanced T1-weighted MR image shows an enlargement of the right maxillary artery (arrow) in comparison to the left side (arrowhead)

advantageous in evaluating soft tissue and intracranial involvement [12].

According to Lloyd et al. [7], three findings on CT and MR imaging should suggest a diagnosis of JNA: (1) a soft tissue mass in the nasopharynx or nasal cavity, (2) a mass in the pterygopalatine fossa, (3) erosion of the posterior osseous margin of the sphenopalatine foramen extending to the base of the medial pterygoid plate.

All of the 72 patients with JNA reported by Lloyd et al. [7] exhibited these findings, which were also observed in our case.

Another characteristic finding of JNA is the Holman-Miller sign, characterized by anterior bowing of the posterior maxillary wall due to the presence of a mass in the pterygomaxillary space [7, 13].

Furthermore, characteristic MR features of JNA include intratumoral signal voids on T2-weighted images and intense tumor enhancement after contrast injection [1, 12, 14].

In our case report, we observed that the tumor did not exhibit restricted diffusion, which aligns with the findings reported in the case series by Aimli et al. [12].

A rare radiologic finding of JNA, that occurred in our patient, is the presence of intratumoral degenerative cystic components.

In some instances, JNA may be diagnosed in an advanced stage, characterized by skull base erosion and invasion into the intracranial and orbital regions. In our patient, this invasion led to vision loss and proptosis. These symptoms respectively occur in about 12.3% and 6.9% of patients with JNA [15].

Patients with advanced stages of the disease usually have a high risk of recurrence, which in most cases is a persisting tumor after incomplete resection [16]. Therefore, post-treatment follow-up is of paramount importance. While the clinical examination remains a fundamental aspect, it cannot be isolated from endoscopy and radiological investigations because many recurrences may not exhibit any symptoms [17]. Imaging

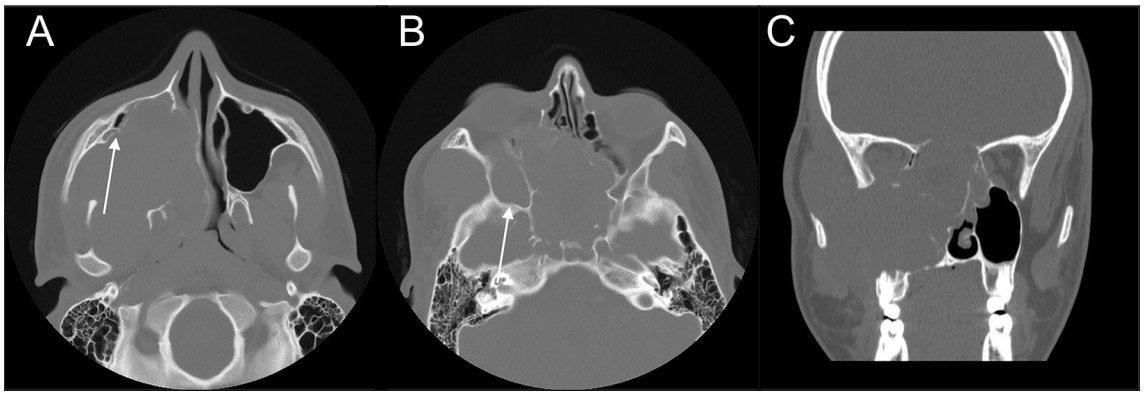

Fig. 4 Axial CT images show bone remodeling, thinning, and destruction that involve the posterior margin of sphenopalatine foramen extending to the base of the medial pterygoid plate, right maxillary sinus, right nasal cavity, ethmoid sinuses, sphenoid sinuses, anterior skull base and right portion of the hard palate. Note the anterior bowing of the posterior wall and flattening of the right maxillary sinus (arrow in **A**). Moreover, enlargement of the right inferior orbital fissure in comparison to the left side is evidenced (arrow in **B**)

follow-up typically relies on MR as the preferred modality to detect recurrences [12], which usually present strongly enhanced nodules after contrast media injection [17, 18]. However, also postoperative fibrosis may show contrast enhancement. The stability of the mass at serial MR examinations is consistent with fibrosis [1, 18]. Recently, the use of prostate-specific membrane antigen (PSMA) PET/CT has been proposed for detecting recurrent or residual JNA exactly by targeting PSMA in the tumor-associated neovasculature and not in scar/fibrosis [17]. Indeed, in the prospective study with 22 patients, Thakar et al. [17] found higher specificity and positive predictive value of PSMAPET/CT (100% and 100%) in comparison to MR (53.3% and 41.67%, respectively).

Conclusions

Juvenile nasopharyngeal angiofibroma is a rare tumor that may present with invasive growth and cause significant morbidity in young patients. The diagnosis heavily relies on the indispensable role of MR and CT imaging, which can reveal the characteristic tumor localization, patterns of spreading, and pronounced vascularization. These imaging modalities are particularly valuable since biopsies are generally avoided in these cases due to the potential risk of severe bleeding.

Abbreviations

CT	Computed tomography
FLAIR	Fluid attenuated inversion recovery
FSE	Fast spin echo
JNA	Juvenile nasopharyngeal angiofibroma
MR	Magnetic resonance
SE	Spin echo

Acknowledgements

Not applicable.

Author contributions

RC was a major contributor in writing all the parts of the case report. SP and GL provided valuable assistance in the writing process. During the writing process, MR, GF and MN contributed with critical feedback, playing a pivotal role in enhancing the report's overall quality and clarity. All authors contributed to the writing and review of the manuscript and approved the final version for submission.

Funding

No funding was obtained for this study.

Availability of data and materials

Not applicable.

Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

Written consent to publish this case was obtained from the father of the patient.

Competing interests

The authors declare that they have no competing interests.

Received: 3 August 2023 Accepted: 14 September 2023
Published online: 22 September 2023

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