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Case Report: Role of MRI, CT, and 18F-FDG PET/CT in staging and response assessment of pediatric synovial sarcoma

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Synovial sarcoma is a rare and aggressive soft tissue malignancy that predominantly affects adolescents and young adults. Early diagnosis can be challenging due to nonspecific clinical presentation and the heterogeneous imaging appearance of these tumors. Multimodal imaging plays a central role in disease detection, characterization, staging, and treatment planning, and is increasingly used for treatment response assessment. We report the case of a 16-year-old girl presenting with progressive pain and functional limitation of the right shoulder, ultimately diagnosed with a giant synovial sarcoma arising in an uncommon anatomical location. The diagnostic work-up included a comprehensive imaging approach integrating ultrasound, magnetic resonance imaging (MRI), computed tomography (CT), and whole-body ^18F-FDG PET/CT. This multimodal strategy allowed accurate evaluation of tumor extent, assessment of possible bone involvement, and detection of regional lymph node disease. Following three cycles of combination chemotherapy with ifosfamide and doxorubicin, follow-up imaging demonstrated a reduction in tumor metabolic activity on ^18F-FDG PET/CT, consistent with an early metabolic response to therapy, despite the persistence of a relatively stable tumor volume on anatomical imaging. This case underscores the importance of an integrated imaging strategy in the management of pediatric soft tissue sarcomas and highlights the clinical value of combining complementary imaging techniques in rare and complex presentations. In particular, it illustrates how the coordinated use of multiple diagnostic modalities can optimize staging accuracy and support individualized treatment planning in pediatric patients with uncommon and large soft tissue tumors.

KEYWORDS
CT, MRI, PET, staging, synovial sarcoma

1 Introduction

Soft tissue sarcomas represent a relatively small proportion of pediatric malignancies but are associated with significant morbidity and mortality due to their aggressive biological behavior and metastatic potential (1, 2). Among these tumors, synovial sarcoma (SS) is one of the most common non-rhabdomyosarcoma soft tissue sarcomas in adolescents and young adults (3, 4). Tumors arising in the shoulder region account

for approximately 6% of all SS. The disease usually does not arise from inside the joint but from the surrounding soft tissues. The tumor often progresses slowly, taking months to years before patients present themselves, potentially resulting in a delay in diagnosis (5).

Imaging plays a central role in the management of these tumors (6). Magnetic resonance imaging (MRI) is considered the reference modality for local staging due to its excellent soft tissue contrast and ability to assess tumor extent and relationship with surrounding structures. However, MRI findings are not pathognomonic, and additional imaging techniques are often required to fully characterize the disease. Metabolic imaging with 18F-FDG PET/CT has increasingly been integrated into the diagnostic pathway of pediatric sarcomas, particularly for detecting nodal and distant metastases and guiding treatment planning (7–9). In this context, we describe the case of an adolescent patient presenting with a SS arising in the shoulder region, an uncommon anatomical location for this tumor type, and characterized by a considerable tumor burden at the time of diagnosis. The diagnostic work-up included a comprehensive imaging evaluation integrating different modalities, each contributing specific information to the assessment of local tumor extent, regional disease involvement and therapy response. The availability of multiple imaging techniques within the same clinical pathway allowed a thorough characterization of the lesion and supported clinical decision-making.

2 Case description

A previously healthy 16-year-old girl presented with progressive pain in the right shoulder associated with increasing difficulty in arm movement. The symptoms had been present for several months and had gradually worsened over time. There was no history of trauma, previous medical conditions, or systemic disease.

Due to the persistence of pain and its impact on daily activities and sports participation, the patient underwent clinical evaluation. Physical examination revealed localized bulging of the right shoulder, reduced range of motion, and tenderness on palpation of the periarticular region. No systemic symptoms, such as fever or weight loss, were reported. At the time of presentation, the patient had not undergone any previous surgical or oncological interventions.

2.1 Diagnostic assessment

Given the persistence and progression of symptoms, plain radiography and ultrasound of the shoulder were performed as the initial imaging assessments. The shoulder radiograph revealed a soft tissue mass of increased density adjacent to the joint, without significant calcification or osseous involvement. Subsequent ultrasound examination demonstrated a large heterogeneous soft tissue mass involving the periarticular region and extending into adjacent muscular compartments. The lesion appeared poorly defined, with marked internal heterogeneity, findings that raised suspicion for an underlying malignant soft tissue tumor.

MRI of the right shoulder was performed using a 1.5-T scanner (Ingenia, Philips Healthcare, The Netherlands) with standard musculoskeletal protocol including T1-weighted, T2-weighted, STIR, without administration of intravenous contrast medium. The exam demonstrated a large soft tissue mass measuring approximately 18 cm in maximum diameter (Figure 1). The lesion showed a multilobulated configuration with marked internal heterogeneity and areas of variable signal intensity on T2-weighted sequences characterised by both cystic and solid components, with no enhancement in the areas containing cysts. The presence of fluid-fluid levels were noted together with the characteristic "triple sign," consisting of the coexistence of low, intermediate, and high signal intensity regions. These features suggested the diagnosis of SS. The tumor involved several muscles of the shoulder girdle, including the deltoid, infraspinatus, and subscapularis muscles, and extended into surrounding soft tissues. The evaluation of possible bone involvement was initially challenging due to periosteal reaction and inflammatory changes. For this reason, contrast-enhanced computed tomography (CT) was performed and confirmed the absence of direct invasion of the humeral head. Moreover, contrast-enhanced CT provided important information on the enhancement pattern of the lesion, contributing to the characterization of tumor heterogeneity.

To complete staging, a whole-body ^18F-FDG PET/CT, was performed three weeks after MRI. It was acquired 60 min after tracer injection according to institutional pediatric oncology protocol. The examination revealed a highly heterogeneous pattern of the highest metabolic activity (SUV max 12.3) and the mean activity (SUV mean 1.5, SUV peak 7.7) in the primary tumor, with areas of intense uptake corresponding to viable tumor tissue and regions of absent uptake corresponding to necrosis or fluid components. A key diagnostic finding was the identification of metabolically active lymph nodes in the ipsilateral axilla. This information had direct implications for disease staging and therapeutic planning.

Histopathological examination of the excisional biopsy confirmed the diagnosis of monophasic poorly differentiated SS (Figure 2). Molecular testing demonstrated the presence of the characteristic chromosomal translocation t(X;18)(p11;q11) involving the SS18 gene.

Following multidisciplinary discussion, the patient started multimodal therapy consisting of combination chemotherapy with Ifosfamide and Doxorubicin, according to the EpSSG NRSTS 2005 protocol (10). After completion of three cycles of chemotherapy, which were well tolerated, with no relevant treatment-related complications, a follow-up imaging evaluation was performed to assess treatment response. Contrast-enhanced computed tomography demonstrated persistent tumor volume without clear evidence of disease progression. A whole-body ^18F-FDG PET/CT and a contrast-enhanced CT were subsequently scheduled for re-staging of the disease. The examination demonstrated a clear reduction in metabolic activity of the primary tumor compared with baseline, consistent with an early metabolic response to therapy. SUVmax decreased from 12.3 at baseline to 6.7 at follow-up and SUV peak from 7.7 to 2.6 (Figure 3). While contrast-enhanced CT demonstrated

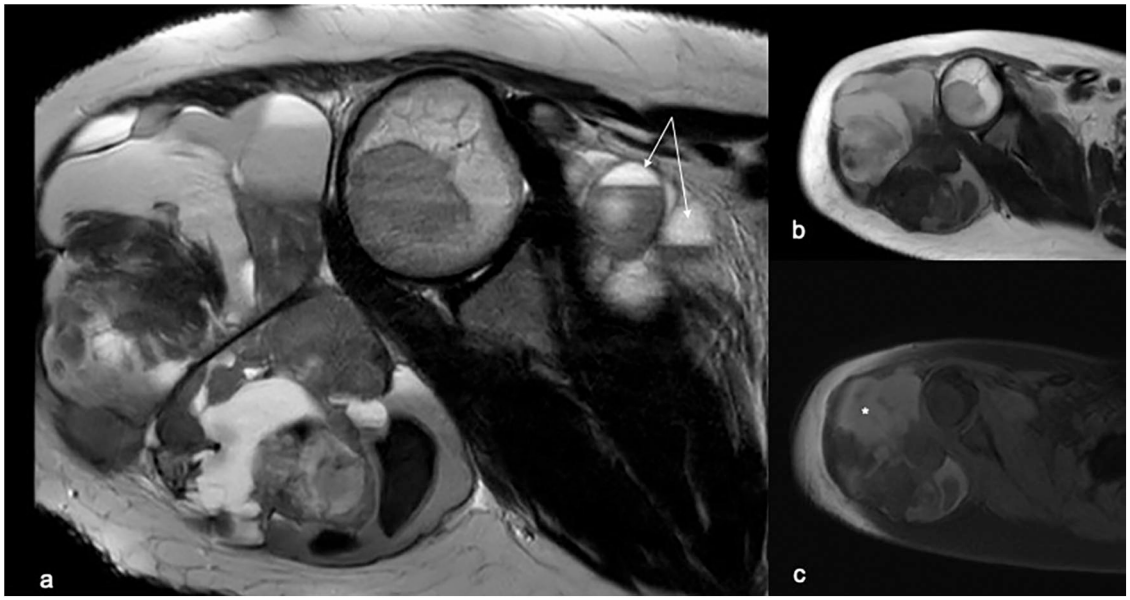

FIGURE 1
MRI characteristics of synovial sarcoma of the right shoulder. Axial T2-w **(a)**, T1w **(b)** and T1w + fat saturation **(c)** showing a large multilobulated soft tissue mass involving the muscles of the shoulder girdle. The lesion demonstrates marked internal heterogeneity with areas of low, intermediate, and high signal intensity on T2w images, consistent with the so-called "triple sign," reflecting the coexistence of solid tumor tissue, necrosis, hemorrhage, and cystic components. Fluid-fluid levels were noted (arrows in "a"). Hemorrhage was confirmed on T1w + fat saturation images (asterisk in "c"). MRI allowed precise assessment of tumor extent and its relationship with adjacent anatomical structures.

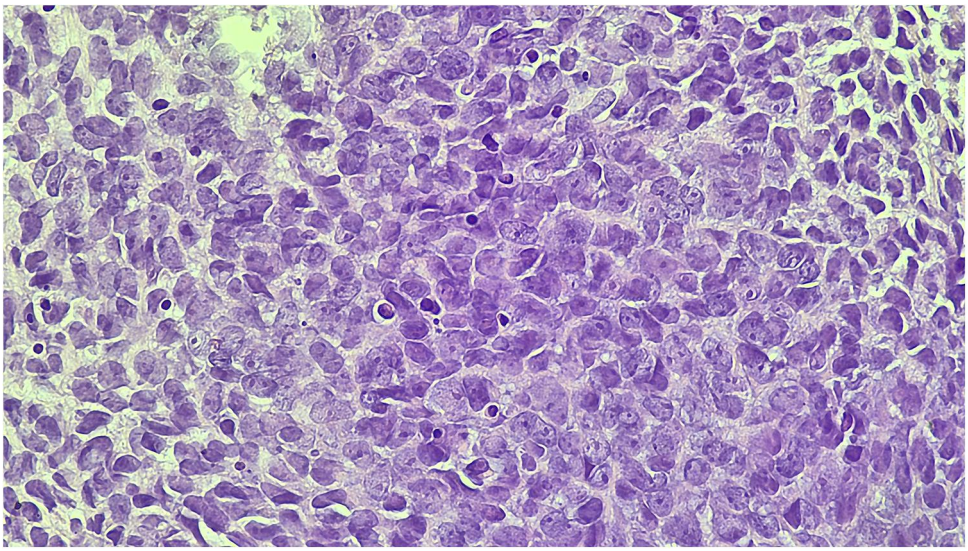

FIGURE 2
Histopathological features of synovial sarcoma. Hematoxylin and eosin (H&E) staining at ×40 showing marked cytological atypia, numerous apoptotic figures, a low mitotic index, and focal areas of necrosis, consistent with monophasic poorly differentiated synovial sarcoma.

changes in the enhancement pattern of the solid tumor component without significant variation in overall tumor volume (Figure 4). These structural changes corresponded to the reduction in metabolic activity observed on follow-up ^18F-FDG PET/CT, suggesting an early biological response to therapy preceding measurable tumor shrinkage. Additionally, multiple areas of symmetrical intense FDG uptake were observed bilaterally in the neck area, the posterior mediastinum, bilateral costover-tebral margins, and bilateral suprarenal fossa, extending to perinephric fat which anatomically correlated to fat at CT, consistent with functioning brown fat.

3 Discussion

SS is a rare but aggressive soft tissue malignancy that requires accurate staging and careful therapeutic planning, particularly in pediatric and adolescent patients, in whom treatment decisions

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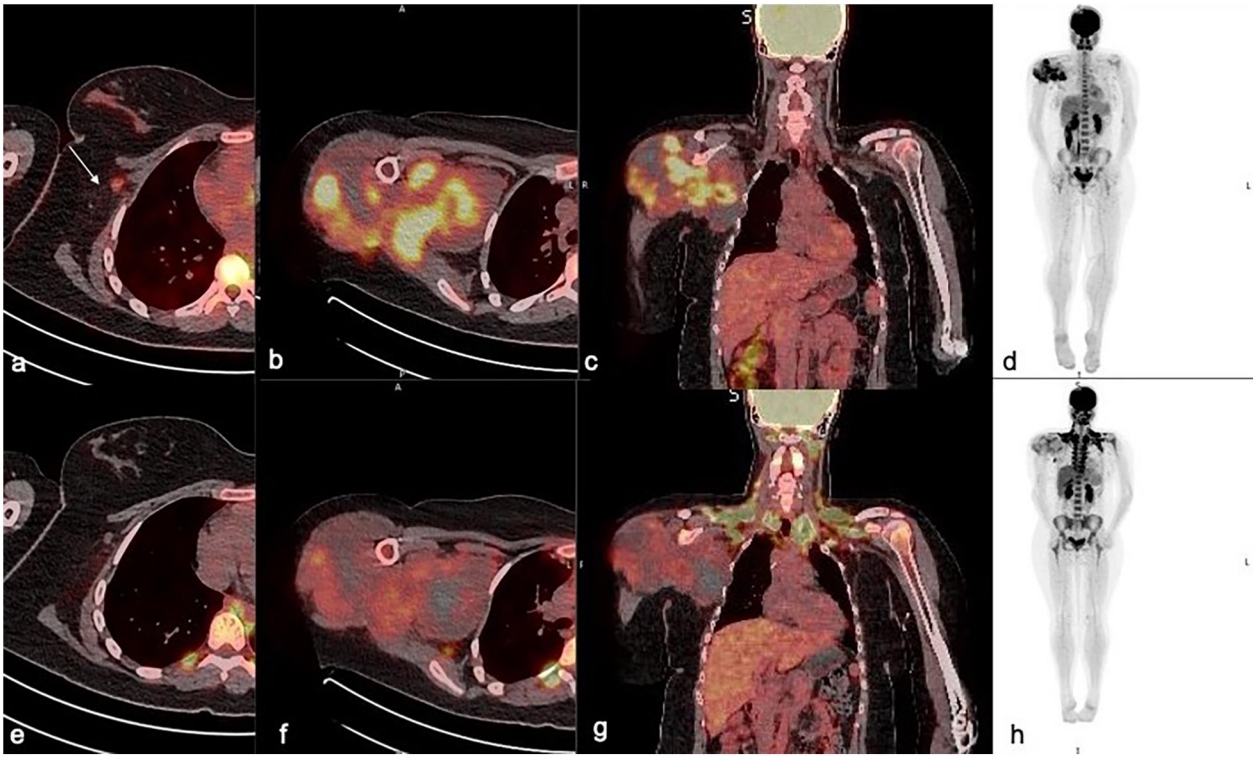

FIGURE 3
Comparison between Pre **(a–d)** and post-treatment **(e–h)** whole-body ^18F-FDG PET/CT. Intense heterogeneous metabolic activity within the primary tumor in the right shoulder region **(b–d)** with a pathologic axillary node (arrow in "a"). Post-treatment ^18F-FDG PET/CT performed after three cycles of chemotherapy demonstrating a global reduction in metabolic activity of the lesion, consistent with a favorable metabolic response to therapy. Symmetric FDG uptake in typical anatomical locations corresponding to fat on CT, consistent with physiologic brown fat, observed on the post-treatment examination **(g,h)**.

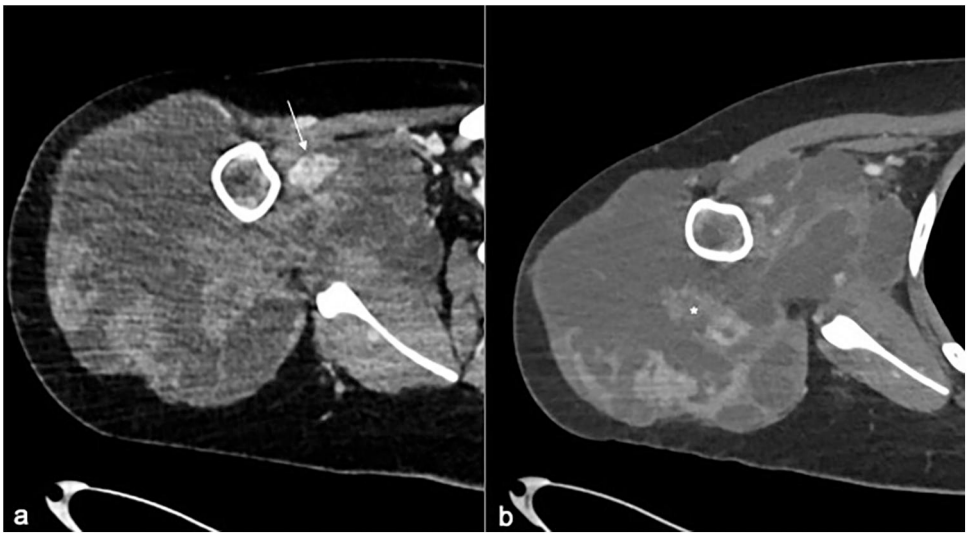

FIGURE 4
Pre-treatment contrast-enhanced computed tomography (CT) of the right shoulder **(a)** confirming the presence of a large soft tissue lesion with heterogeneous enhancement pattern, without bone involvement. CT imaging excluded direct invasion of the humeral head and allowed better evaluation of cortical bone integrity and lesion extent. Post-treatment **(b)** exam demonstrated changes in the enhancement pattern (arrow in "a") of the solid tumor component without significant variation in overall tumor volume.

must balance oncologic control with long-term functional outcomes. SS of the shoulder accounts for 6% of all SS (6) and imaging plays a decisive role not only in diagnosis but also in guiding multidisciplinary clinical decision-making and tailoring therapeutic strategies (11). One of the main strengths of the present case lies in the clear demonstration of the complementary diagnostic value of MRI and PET/CT.

Morphological imaging represents a cornerstone in the differential diagnosis of SS among pediatric soft tissue tumors. In this case, although other malignant entities were considered, the overall imaging pattern was highly consistent with SS. MRI provided detailed anatomical information regarding tumor morphology, local extension, and the relationship between the lesion and surrounding structures, including muscles, periarticular tissues, and bone. This information is essential for surgical planning and for defining target volumes in radiotherapy (12). From a radiological perspective, SS are typically described as heterogeneous masses with high signal intensity on T2-weighted images, reflecting the coexistence of necrotic, cystic, hemorrhagic, and fibrotic components within the tumor. The so-called "triple sign," characterized by the presence of low-, intermediate-, and high-signal intensity areas on T2-weighted sequences, represents a well-recognized imaging feature that emphasizes tumor heterogeneity (6, 13). However, although present in our case, this sign is being reported in approximately 57% of SS in published series (14). Tumor heterogeneity is more frequently observed in lesions larger than 5 cm, and is often associated with complex internal architecture, including fluid–fluid levels, giving the typical "*bowl of grape* " appearance and considered a specific imaging feature of SS, internal septations, calcifications, and mixed cystic and solid components. Nevertheless, smaller lesions (<5 cm) may present differently, often appearing homogeneous on T2-weighted images and demonstrating an ovoid, nodular, or fascicular configuration, typically with homogeneous contrast enhancement and without the characteristic "triple sign" (15). Lastly, a multilobulated tumor morphology and tumor size greater than 5 cm has consistently been associated with worse clinical outcomes in patients with soft tissue sarcomas, reflecting a higher likelihood of local invasion, metastatic spread, and treatment complexity (16).

Contrast-enhanced imaging plays an important role in highlighting tumor heterogeneity in SS, as most lesions demonstrate heterogeneous enhancement with reduced or absent enhancement in areas of cystic necrosis or internal septations. In the present case, while contrast-enhanced CT initially contributed to the assessment of tumor heterogeneity, it proved particularly useful during follow-up, together with PET/CT, by providing information on treatment response, with changes in the enhancement pattern of the solid component despite stable tumor volume. This observation underscores the importance of contrast-enhanced imaging in detecting early treatment responses that may not yet be reflected by changes in tumor size, especially in clinical settings where PET imaging is not available or accessible.

On the other hand, PET/CT proved particularly valuable in identifying regional lymph node involvement that was not clearly detected on morphological imaging, confirming its well-recognized higher sensitivity in detecting nodal and distant disease in pediatric sarcomas (17). Functional imaging enables the identification of metabolically active disease even in the absence of significant anatomical changes, thereby improving staging accuracy and supporting more precise risk stratification (18).

Semiquantitative metabolic parameters, including standardized uptake values (SUVmax, SUVmean, and SUVpeak), are commonly used to characterize tumor biology and monitor response to therapy. Among these, SUVpeak, reflecting metabolic activity within a small, standardized tumor volume, may provide a more robust and reproducible measure than single-voxel metrics in heterogeneous lesions. Although considerable variability exists in reported thresholds due to differences in scanners and acquisition protocols, in synovial sarcoma, a baseline SUVmax above approximately 5 has been associated with higher histological grade and worse clinical outcomes, supporting its role as a clinically relevant imaging biomarker for risk stratification (19). In addition, metabolic imaging with ^18F-FDG PET/CT can help identify metabolically active tumor regions within heterogeneous masses, which are more likely to correspond to areas of higher cellularity and biological aggressiveness. These regions may represent optimal targets for biopsy and may support biologically guided radiotherapy planning, including potential dose escalation to high-risk tumor subregions while sparing surrounding normal tissues, thereby improving local tumor control and reducing treatment-related toxicity (12).

Beyond staging, PET imaging also plays a clinically relevant role in the assessment of treatment response. In the present case, follow-up imaging performed after three cycles of chemotherapy demonstrated a clear reduction in tumor metabolic activity on PET/CT despite the persistence of a relatively stable tumor volume. This finding suggests an early biological response to therapy that would not have been captured by size-based criteria alone. While tumor size remains a widely used parameter for response evaluation, changes in metabolic activity may represent earlier and more sensitive indicators of therapeutic effectiveness. This finding highlights the ability of functional imaging to detect early treatment response before morphological tumor shrinkage becomes evident, a phenomenon that has been reported in high-grade soft tissue sarcomas and supports the role of PET/CT in early response assessment and treatment monitoring (9). Additionally, in the follow-up exam, we detected symmetrical areas of intense FDG uptake consistent with metabolically active brown fat. Although typically considered a physiological finding, recent evidence suggests that brown fat activation on FDG PET/CT may reflect systemic metabolic status and has been associated with improved survival outcomes in oncologic patients (20). In the present case, this finding was interpreted as a physiological phenomenon; however, its potential prognostic implications deserve further investigation, particularly in pediatric and adolescent populations.

In the present case, the multimodal imaging evaluation, enabled a more comprehensive characterization of tumor heterogeneity and nodal disease, allowing a comprehensive

assessment of the lesion from both anatomical and functional perspectives, supporting multidisciplinary treatment planning and monitoring and reinforcing the value of an integrated imaging approach in complex pediatric sarcomas.

The main limitation of this report is the relatively short duration of follow-up, which does not allow assessment of long-term outcomes. Nevertheless, individual case reports remain valuable in the context of rare pediatric malignancies, where clinical experience is limited and each well-documented case contributes to improving diagnostic awareness and management strategies.

4 Take-away lessons

- Multimodal imaging is essential for accurate staging of pediatric synovial sarcoma
- MRI remains the reference modality for local tumor assessment
- PET/CT provides additional value in detecting nodal metastases, tumor heterogeneity and early treatment response
- Early integration of functional imaging may improve treatment planning

5 Patient perspective

The patient and her family reported initial concern due to the persistence of shoulder pain and the progressive limitation of daily activities. After the diagnosis was established, they expressed appreciation for the coordinated multidisciplinary approach and the clear communication provided by the clinical team. At the time of the most recent follow-up, the patient reported good tolerance of therapy and maintained a positive outlook regarding treatment.

Data availability statement

Data will be available upon request to the corresponding author.

Ethics statement

Ethical approval was not required for the study involving human samples in accordance with the local legislation and institutional requirements because [reason ethics approval was not required]. Written informed consent for participation in this study was provided by the participants' legal guardians/next of kin. Written informed consent was obtained from the individual (s), and minor(s)' legal guardian/next of kin, for the publication of any potentially identifiable images or data included in this article. Written informed consent was obtained from the patient's parents for the publication of this case report.

Author contributions

LP: Project administration, Writing – original draft, Investigation, Data curation, Funding acquisition, Conceptualization, Validation, Writing – review & editing. LiM: Investigation, Resources, Writing – review & editing. LMe: Writing – review & editing, Investigation, Visualization. CC: Supervision, Writing – review & editing, Visualization, Validation. DD: Conceptualization, Supervision, Validation, Writing – review & editing. PF: Project administration, Writing – review & editing, Writing – original draft, Funding acquisition, Data curation, Conceptualization, Visualization.

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Conflict of interest

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