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Case Report

Pediatic intracranial synovial sarcoma: a case report

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Abstract
Object To investigate the MR imaging characteristics of intracranial synovial sarcoma by reviewing this case and literature, aiming to improve preoperative diagnostic accuracy for radiologists and neurosurgeons and guide appropriate treatment planning.
Material and Methods The clinical and MR images of 1 case with SS in the brain by pathology were retrospectively analyzed, and the causes of misdiagnosis were analyzed Combined with relevant literature.
Results MR showed a solid cystic mass in the right frontal lobe, isointense on T1WI, heterogeneous signal on T2WI, Elevated signal on DWI, obvious uneven enhancement on T1WI, and "triple signal sign" and "cobblestone sign" appearance.
Conclusion SS in the brain is very rare. MR has certain imaging features, perfect MR examination is helpful for differential diagnosis.

Keywords Synovial sarcoma · Primary intracranial tumor · Pediatric patient · Triple-signal sign · MRI

1 Introduction

Synovial sarcoma (SS) is a malignant soft tissue tumor of unknown origin. According to the 2020 WHO classification of soft tissue tumors, it is classified as an "undifferentiated tumor" [1]. It accounts for 5–10% of soft tissue sarcomas and is more common in young adults, with a slight male predominance [2]. Here, we present the case of a 7-year-old male diagnosed with intracranial synovial sarcoma, confirmed by postoperative histopathological examination. Magnetic resonance imaging (MRI) revealed a solid cystic mass in the right frontal lobe preoperatively, and the initial diagnosis was a pediatric astrocytoma with hemorrhage.

SS is primarily located around the joints of the limbs and rarely originates from the cranium. Few cases of intracranial SS have been reported in the dura mater [3, 4], particularly in the frontal, parietal, and temporal lobes [5]. They rarely occur in the occipital lobe, cerebellum, sellar region, or ventricles [6–8]. In this case, the tumor was located in the frontal lobe, which is an uncommon intracranial location for SS. The aim of this article is to summarize the MRI findings of a rare case, thereby broadening the diagnostic perspectives for radiologists and neurosurgeons and facilitating preoperative treatment planning.

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2 Case presentation

The patient was a 7-year-old male who had intermittent headaches for five days, accompanied by nausea and vomiting. The laboratory test results were unremarkable. MRI revealed a cystic-solid tumor in the right frontal lobe, which appeared as a low-to-high mixed signal on both T1-weighted and T2-weighted images demonstrating "triple signal sign" and "liquid–liquid interface." There was compression of the basal ganglia around the mass, and edema was seen in the external capsule (Fig. 1A, B). Diffusion-weighted imaging (DWI) showed that the solid nodule was hyperintense and the cystic fluid was hypointense within the mass (Fig. 1C). Diffusion-tensor imaging (DTI) revealed compression of the nerve fibers surrounding the mass, resulting in the reduction or disappearance of focal signals (Fig. 1D). Axial, coronal, and sagittal contrast-enhanced T1-weighted images (T1WI) showed intense enhancement of both the solid nodules and cystic wall of the mass, a characteristic feature often referred to as the "cobblestone sign"(Fig. 1E, F, G). Postoperative magnetic resonance imaging (MRI) revealed that the tumor was completely excised (Fig. 1H). Considering the patient's age, clinical manifestations, and imaging characteristics, the radiologist initially diagnosed the tumor as a hemorrhagic pediatric astrocytoma. On March 10, 2023, a neurosurgeon performed a craniotomy through the frontal region to access the tumor in the right frontal lobe. Histopathological examination revealed a synovial sarcoma. Under the microscope, the tissue exhibited a rather disordered cell arrangement and diverse morphology with bidirectional differentiation characteristics, encompassing both spindle and epithelial cell phenotypes. Some cells were relatively large, with deeply stained and irregular nuclei, and an increased nucleocytoplasmic ratio. Numerous mitotic figures were observed, and the interstitial area was rich in blood vessels (Fig. 2A). Immunohistochemical staining showed negative results for GFAP, while positive results were observed for Bcl-2, CD99, and Vimentin (Fig. 2B–E). Fluorescence in situ hybridization (FISH) analysis, using a dual-color break-apart probe targeting the SS18 gene on 18q11.2, suggests the presence of a t(X;18) translocation

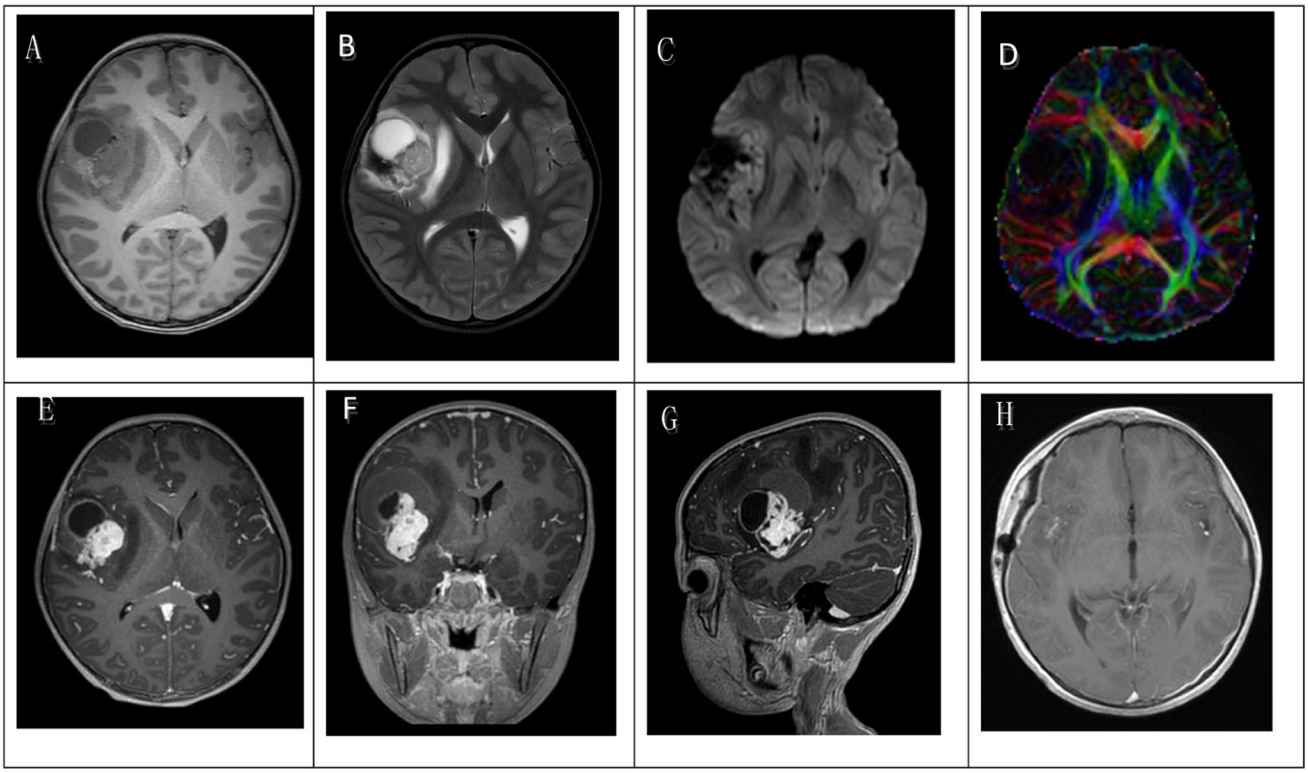

Fig. 1 **A**, **B** Axial T1WI and T2WI reveal synovial sarcoma in the right frontal lobe, which appears as low-to-high signal intensity with the presence of "triple signal sign" and an "liquid–liquid interface."The mass causes significant space-occupying effects and surrounding edema in the external capsule. **C** DWI shows diffusion restriction of solid nodules within the mass with hypointensity. **D** DTI shows displacement and disruption of the nerve fibers surrounding the mass. **E**, **F**, **G** Axial, coronal, and sagittal contrast-enhanced T1WI showing intense enhancement of solid nodules within the mass, commonly referred to as the "cobblestone sign."The multiple cystic structures of the mass had intact cyst wall, and the enhancement was variable. **H** Postoperative axial contrast-enhanced T1WI showed that the tumor had been removed

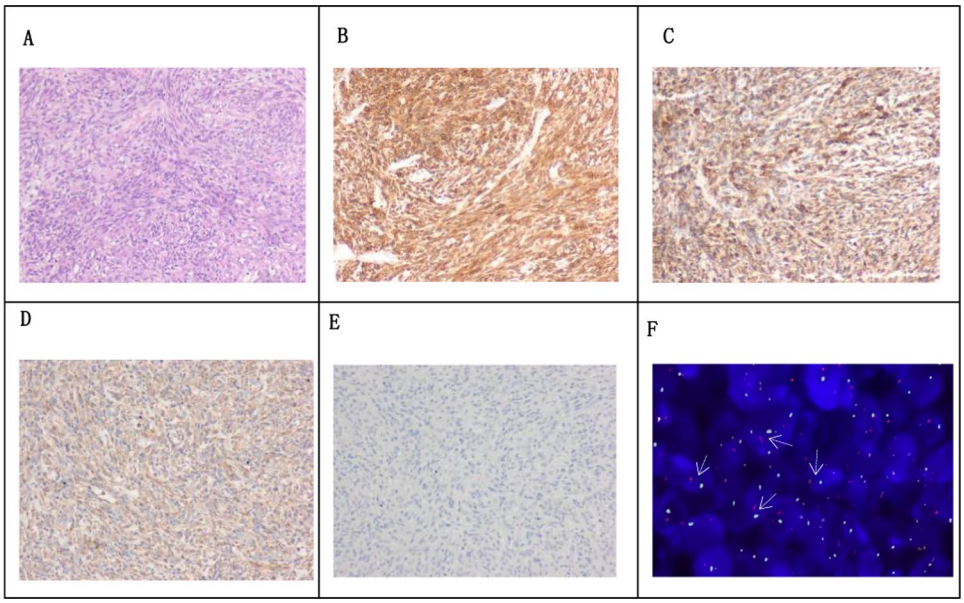

Fig. 2 Histomorphologic, immunohistochemical, and molecular profile of synovial sarcoma in the right frontal lobe. **A** Microscopy shows bidirectional differentiation characteristics encompassing both spindle and epithelial cell phenotypes. **B**–**E** Immunohistochemical staining for Bcl-2+, CD99+, Vimentin+, GFAP– [Original magnification: (**A**) 200×; (**B**–**E**)100×]. **F** Fluorescence in situ hybridization (FISH) with a break-apart probe for the SS18 gene shows scattered, separated red and green signals per nucleus in tumor cells (↑), indicating the presence of a t(X;18) translocation

in 50% of tumor cells(Fig. 2F). One week post-surgery, the patient's physical condition had significantly improved. He subsequently underwent standard adjuvant radiotherapy and has not experienced tumor recurrence to date.

3 Discussion

The clinical manifestations of intracranial SS lack specificity and primarily depend on the location, size, and complications of the tumor. It can present with headaches, nausea, vomiting, seizures, hemiplegia, and language disorders [9]. In this case, the patient experienced headaches accompanied by nausea and vomiting, which are common clinical manifestations of increased intracranial pressure caused by typical intracranial tumors.

Diagnosis of intracranial SS relies on histopathology, immunohistochemistry, and cytogenetics. Histologically, SS can be classified into biphasic, monophasic spindle cells, monophasic epithelial cells, and poorly differentiated types[10]. The first two types were more common. In this case, histological sections showed a biphasic cell type. Immunohistochemistry typically shows the expression of Vimentin and Bcl-2, and often CD56, CD99, and TLE-1 [11]. These are relatively stable immunohistochemical markers for diagnosing intracranial SS, and are consistent with the immunohistochemical results in this case. t(X;18)(p11.2;q11.2) translocation is a cytogenetic marker of SS, and > 90% of SS cases exhibit this molecular feature [12]. This translocation leads to the expression of different SS18::SSX oncogenic fusion proteins, which drive sarcomagenesis. Subtypes include SS18::SSX1 and SS18::SSX2 and less commonly SS18::SSX4[13]. In this case, genetic testing confirmed the presence of the t(X;18)(p11.2;q11.2) translocation (Fig. 2F), which is consistent with the molecular characteristics of synovial sarcoma. The t(X; 18) (p11.2; q11.2) translocation serves as a critical molecular marker that differentiates synovial sarcoma from histological mimics, such as malignant peripheral nerve sheath tumors [14]. This marker is especially significant in central nervous system (CNS) lesions, where limited biopsy material may hinder routine histopathological diagnosis [15]. Our findings underscore the WHO classification's requirement for molecular confirmation in suspected cases of synovial sarcoma [16].

Intracranial SS has limited imaging specificity, which can be summarized as follows: Zhang et al. [1] reported the tumor often has a large volume, irregular morphology, and is predominantly cystic-solid with clear borders [5]. They also noted that the tumors are often accompanied by peritumoral edema and calcification in the surrounding areas. Approximately 12.5% of intracranial SS cases show significant bleeding on imaging. The imaging features of our case are consistent with these findings, except for the absence of calcification; Jones et al. [2] explained that SS exhibits mixed signals on the T2WI sequence, and when it appears as low-to-high signal intensity with the presence of the "triple signal sign" and "liquid–liquid interface," it is a characteristic imaging feature of intracranial SS [17]. In this retrospective study, our present case had typical imaging features mentioned in the literature (Fig. 1A, B). Abdelkrim et al. elaborated that low signal intensity represents remote hemorrhage, equal signal intensity represents tumor tissue, and high signal

intensity represents fresh hemorrhage or cystic necrosis [18]. Sarkar et al. [3] reported that, in T2WI and T1WI-enhanced sequences, the tumor may exhibit nodules with slightly high signal intensity and grid-like low signal intensity, known as the "cobblestone sign," which is an important radiological indicator for diagnosing intracranial SS [19]. There is the special sign in our case (Fig. 1E, F, G). In retrospect, the imaging in this case showed that the tumor was located in the right frontal lobe, which is an uncommon location in SS. The imaging features were typical of the "triple signal sign" and "cobblestone sign." Owing to the lack of in-depth understanding of the imaging features of intracranial SS at the time, the preoperative diagnosis was mistakenly made as pediatric astrocytoma. Through this case report and a comprehensive review of relevant literature, the imaging characteristics of synovial sarcoma were summarized to provide insights for preoperative diagnosis.

Primary intracranial SS in children should be differentiated from pediatric-type, diffuse high-grade gliomas. Diffuse hemispheric gliomas (H3G34-mutant) and diffuse pediatric-type high-grade gliomas (H3-wildtype and IDH-wildtype) are mostly located on the surface of the brain and histologically exhibit bidirectional differentiation between glioblastoma and embryonal tumors [20]. Pediatric-type diffuse high-grade gliomas can present as solid, cystic-solid, or solid lesions with hemorrhage. Imaging signals may exhibit a combination of features resembling SS; however, they typically lack the presence of a liquid–liquid interface and low signal intensity septations within the tumor. Diffuse midline gliomas (H3 K27-altered) in children are located near the midline of the brain, such as the thalamus and brainstem, and mostly present as solid lesions with a less frequent occurrence of cystic necrosis or hemorrhage [21]. Their locations can be used to distinguish them from SS.

The mainstay of treatment for synovial sarcoma remains surgical excision with the addition of radiotherapy and/or chemotherapy based on patient and tumor characteristics [22]. Where neoadjuvant chemotherapy is widely used in the pediatric population, the use of neoadjuvant chemotherapy is controversial in adults. Radiation therapy (RT) is used either in the preoperative or postoperative setting and is associated with a better prognosis [23]. The European Pediatric Soft Tissue Sarcoma Study Group reports a 90% 5-year survival [24]. Age and tumor size are the two most critical factors influencing the prognosis of SS. Generally, younger age and smaller tumor size correlate with a more favorable prognosis [25, 26]. The patient in our case received standard fractionation radiotherapy after surgery (prescribed dose 50 Gy in 25 fractions). Currently, the patient's follow-up MRI examination reveals no evidence of tumor recurrence, and there are no clinical signs of functional area impairment.

In conclusion, we report a rare case of SS located in the right frontal lobe that exhibited typical imaging features of the "triple signal sign" and "cobblestone sign." In this study, we retrospectively analyzed the MRI features of SS with the aim of enhancing the accuracy of preoperative diagnosis and providing a valuable reference for neurosurgeons in formulating treatment plans.

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Data availability All data generated or analysed during this study are included in this published article.

Declarations

Ethics approval and consent to participate. Informed consent was obtained from the patient's legal guardians. The study received approval from the Ethics Committee of Guiyang Second People's Hospital (Approval Number: 2024 Ethical Review No. 17).

Consent for publication The patient's legal guardian has provided written informed consent authorizing the disclosure of the patient's personal information and associated imaging data.

The statement This manuscript does not report data generation or analysis.

Competing interests The authors declare no competing interests.

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