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# A Rare Case of a Primary Spinal Solitary Fibrous Tumor/Hemangiopericytoma in a 9-Month-Old Patient

## Authors' Contribution:

Study Design A  
Data Collection B  
Statistical Analysis C  
Data Interpretation D  
Manuscript Preparation E  
Literature Search F  
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**Conflict of interest:** None declared

**Patient:** Female, 9-month-old  
**Final Diagnosis:** Hemangiopericytoma • solitary fibrous tumor  
**Symptoms:** Lower extremity weakness • mass in lumbar region • movement disorder  
**Medication:** —  
**Clinical Procedure:** Biopsy • computed tomography • magnetic resonance image  
**Specialty:** Pathology • Radiology

**Objective:** Rare disease

**Background:** Solitary fibrous tumors (SFTs)/hemangiopericytomas (HPCs) are mesenchymal tumors commonly found in middle-aged patients, usually localized to thoracic pleurae. Spinal tumor involvement is rarely seen, and its imaging findings are largely inconsistent because of the rarity of these cases. We present a case report of a 9-month-old girl with a rare intraspinal tumor with histologic evidence of SFT/HPC, but no STAT6 nuclear immunoreactivity.

**Case Report:** A 9-month-old girl, born at term with good prenatal care, presented to the emergency room with regression of developmental milestones. The patient was in good health until 2 months, when she developed decreased spontaneous leg movements. Physical exam revealed diffuse muscular atrophy, with no deep tendon reflexes, sensation, or spontaneous movements of the lower extremities. Computed tomography and magnetic resonance imaging showed a heterogeneous irregular mass filling the lumbosacral spinal canal, extending through the neural foramina to the prevertebral/perivertebral and presacral regions. The tumor was biopsied and referred to the National Institutes of Health for consultation and the diagnosis of SFT/HPC was confirmed on the basis of its histologic features, despite the fact that the tumor was negative for STAT6 immunoreactivity.

**Conclusions:** Although the tumor histology was consistent with SFT/HPC, it was negative for STAT6 nuclear immunoreactivity, which is unusual and may exclude the diagnosis. To our knowledge, this is the youngest patient to present with a spinal SFT with these features.

**MeSH Keywords:** Hemangiopericytoma • Solitary Fibrous Tumors • STAT6 Transcription Factor

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## Background

Solitary fibrous tumors (SFTs) and hemangiopericytomas (HPCs) are primary mesenchymal neoplasms that were first described in 1931 and 1942, respectively [1,2]. SFTs are commonly found in middle-aged patients and are generally localized to thoracic pleural sites [3]. The current literature is limited for cases in the pediatric population, specifically for cases of spinal involvement [4]. Also, imaging findings including contrast-enhanced magnetic resonance imaging (MRI) and computed tomography (CT) scans of spinal SFTs are inconsistent, as they have not been widely studied because of the rarity of these cases. We report a case of a 9-month-old girl with a rare lumbosacral intraspinal tumor with histologic features consistent with an SFT/HPC, but with a discordant immunohistochemical profile. We believe this case to be unique given that, to our knowledge, this is the youngest patient to present with a spinal SFT.

## Case Report

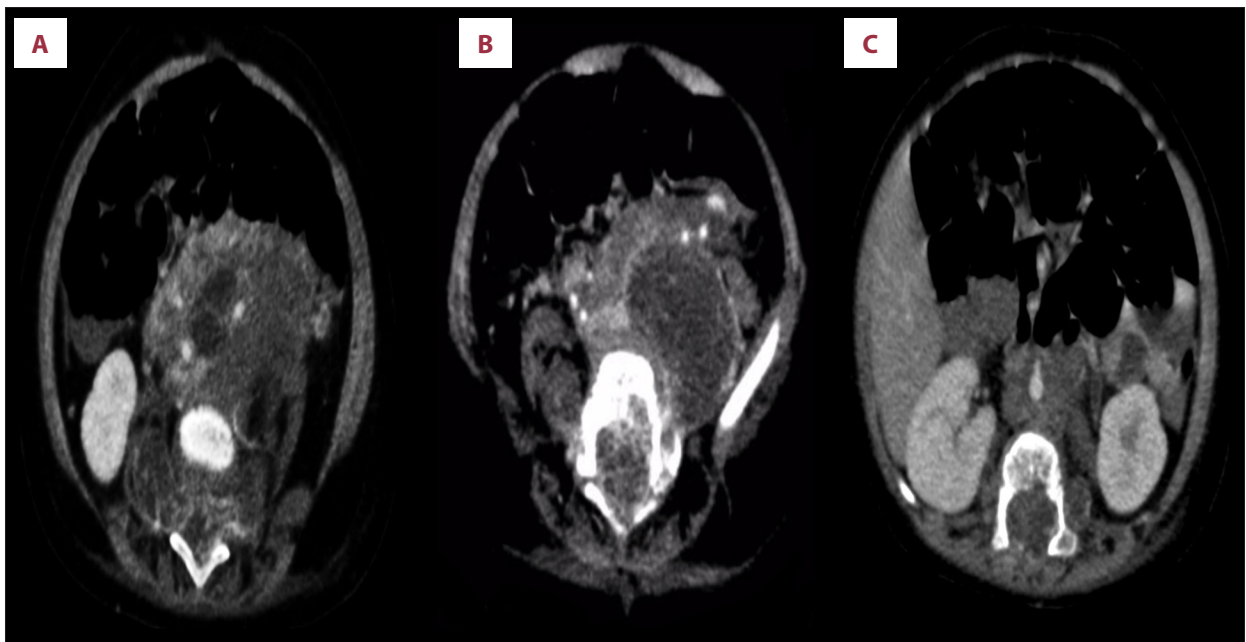
A 9-month-old girl, born at 39 weeks gestational age to a 22-year-old mother (1 pregnancy, 1 birth, no miscarriages or abortions) with good prenatal care, presented to the emergency room with regression of developmental milestones. The patient was in good health (active, alert, accomplishing

milestones) until approximately 2 months before admission when she developed decreased spontaneous leg movements. The patient's parents reported that about 1 week before admission, the patient completely stopped moving her lower extremities, and could no longer sit up or roll over. They also reported frequent wet diapers and irritability. They denied loss of movement in her head, neck, or upper extremities. A neurological exam revealed diffuse muscular atrophy, along with absent deep tendon reflexes, sensation, and spontaneous movements of the lower extremities. The patient also had a decreased anal sphincter tone and an absent anal wink reflex. The patient's trunk and upper extremities had adequate muscle tone, positive deep tendon reflexes, and no loss of sensation or spontaneous movements. These physical exam findings suggested a spinal cord etiology.

Additional data regarding peripartum examinations (blood, urine, and cerebrospinal fluid) were not available.

## Imaging findings

A contrast-enhanced multidetector CT of the abdomen and pelvis revealed a heterogeneously enhancing lumbosacral infiltrative mass showing irregularly shaped borders, regions of central hypoattenuation, and no calcified components. The mass bulk measured approximately 4.2×6.4×10.6 cm and filled the



**Figure 1.** (A) Contrast-enhanced axial computed tomography shows a heterogeneously enhancing infiltrative lumbosacral mass showing irregularly shaped borders, regions of central hypoattenuation, and lack of calcified components. The mass is filling and expanding the spinal canal extending to the retroperitoneum via the bilateral neural foramina, with encasement of the inferior mesenteric artery. (B) Encasement of the bilateral common and proximal external iliac arteries is also noted. (C) Encasement of the infrarenal abdominal aorta is better appreciated at this more cranial level. The distal ureters were also encased by the mass (not shown).

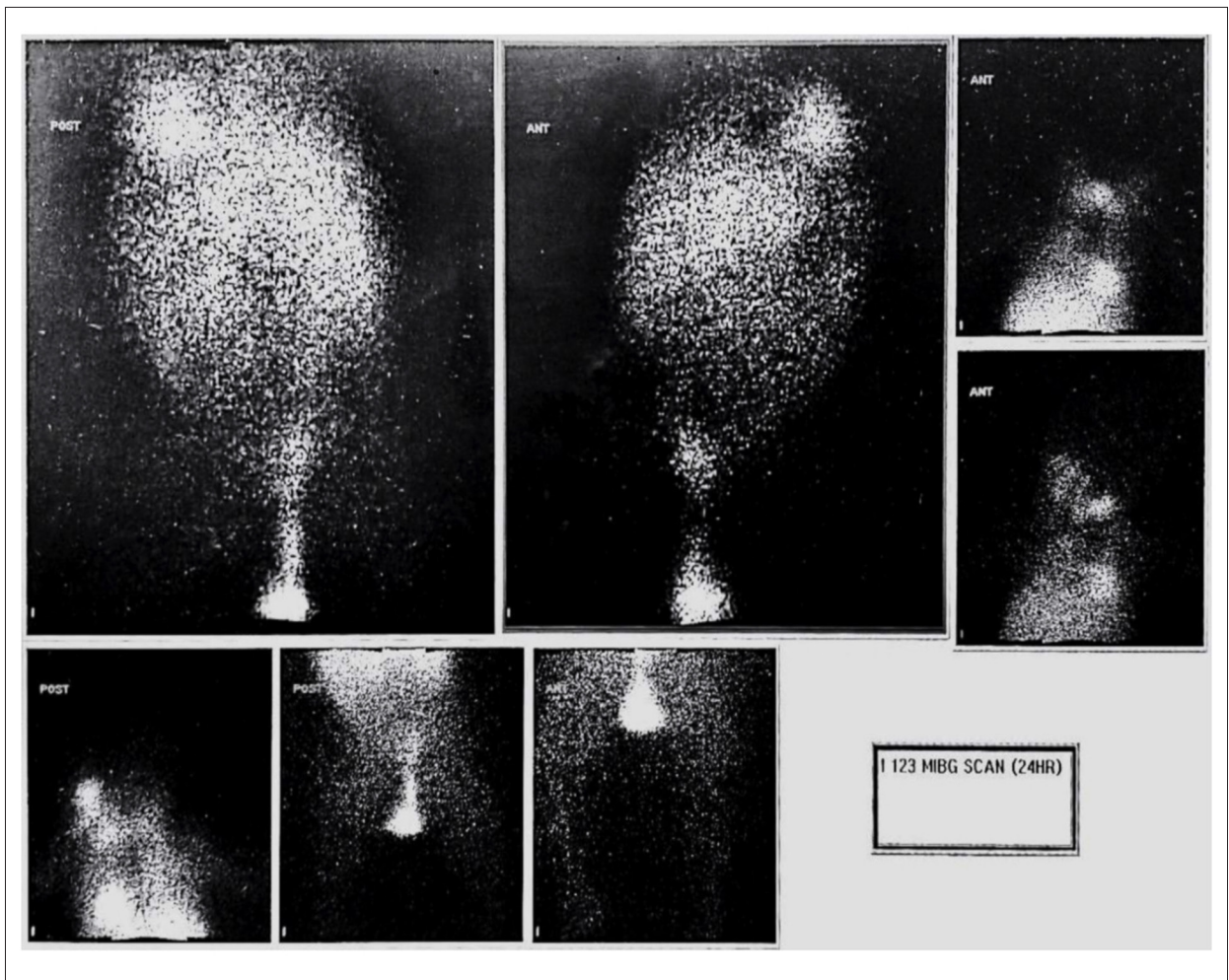


**Figure 2.** (A) Sagittal T2-weighted sequence shows a heterogeneously hyperintense mass with multifocal internal hypointensities filling the entire spinal canal of the lumbar and upper sacral region. At its superior aspect, the mass abuts and superiorly displaces the conus medullaris. The cauda equina nerve roots are not identified and in view that this mass completely fills the lumbosacral spinal canal, are likely compressed or involved. There is subsequent spinal canal expansion with areas of scalloping. (B, C) Sagittal T2-weighted sequences demonstrate that the mass extends bilaterally through expanded lumbosacral neural foramina to the prevertebral/perivertebral and presacral regions with subsequent displacement of presacral vessels and bilateral psoas muscles. (D) Coronal T2-weighted sequence shows bilateral retroperitoneal involvement extending just inferior to the inferior poles of the bilateral kidneys. (E, F) Sagittal T1-weighted postcontrast fat-sat sequences also show that the mass has a heterogeneous enhancement pattern, which also involves various posterior vertebral bodies at the inferior lumbosacral region, as evidenced by contiguous vertebral body enhancement.

lumbosacral spinal canal and multiple bilateral neural foramina with bone expansion and remodeling of the lumbar and upper sacral vertebral bodies. It also infiltrated the retroperitoneal compartment and bilateral psoas muscles, with anterior displacement of the infrarenal abdominal aorta, inferior mesenteric artery, both proximal external iliac arteries, and left internal iliac artery (Figure 1).

A lumbosacral spinal MRI demonstrated a large T1 hypointensity and T2 mixed-signal intensity (hyperintense with internal areas of T2 hypointensity) heterogeneously enhancing mass filling the entire spinal canal of the lumbar and upper

sacral regions. At its superior aspect, the mass abuts and superiorly displaces the conus medullaris. Cauda equina nerve roots are not identified and in view that this mass completely fills the lumbosacral spinal canal, they are likely compressed or involved. There is subsequent spinal canal expansion with areas of bone scalloping. At the inferior lumbosacral region, the enhancing mass is seen involving various posterior vertebral bodies (as evidenced by continuous vertebral body T2 hyperintensity and enhancement). The mass extends bilaterally through expanded lumbosacral neural foramina to the prevertebral/perivertebral and presacral soft tissues. There was subsequent mass effect and displacement of presacral vessels



**Figure 3.** Posterior and anterior whole-body metaiodobenzylguanidine (MIBG) scintigraphy was performed using 370 MBq of iodine-123. Multiple spot images of the entire body were collected 24 h after radionuclide administration, showing normal physiologic MIBG uptake in brown fat, nasal mucosa, and salivary glands, heart, liver, and bowel, with excretion activity in the bladder.

and of the bilateral psoas muscles. Bilateral retroperitoneal involvement is seen extending just inferior to the inferior poles of the bilateral kidneys. The urinary bladder is displaced anteriorly and appears elongated because of extrinsic compression. There appears to be some degree of free intraperitoneal fluid (Figure 2).

On the basis of these imaging findings and taking into account the lesion's location and patient demographics, our initial differential diagnosis included neuroblastoma, chordoma, chondrosarcoma, Ewing's sarcoma, and other sarcomatous lesions.

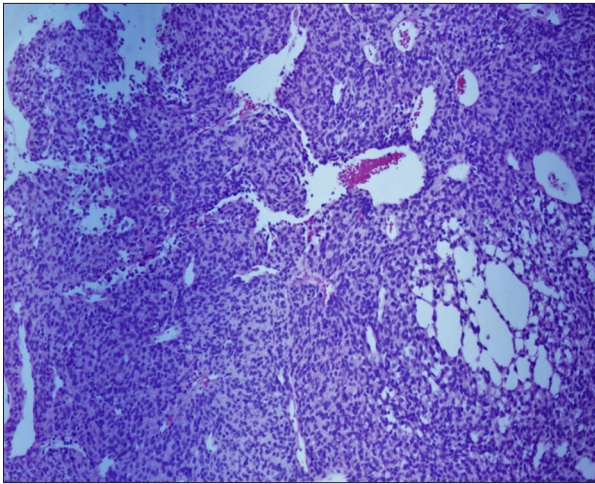
A whole-body metaiodobenzylguanidine (MIBG) scintigraphy was requested by the pediatric oncologist. Posterior and anterior whole-body MIBG scintigraphy using 370 MBq of iodine-123 demonstrated normal physiologic MIBG uptake in brown fat, nasal mucosa, and salivary glands, heart, liver, and

bowel, with excretion activity in the bladder, excluding a diagnosis of neuroblastoma (Figure 3).

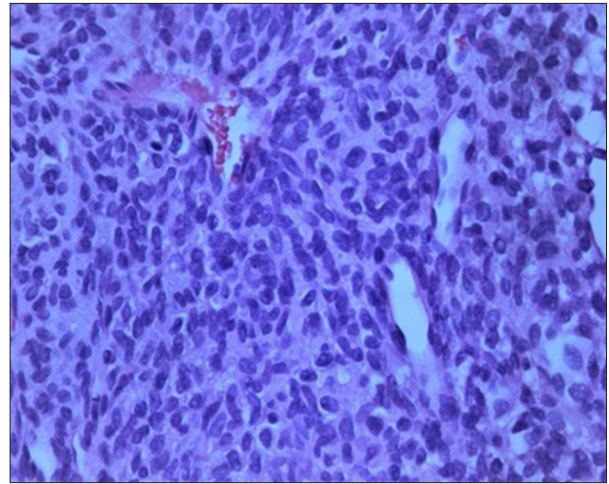
### Pathologic findings

An epidural/intervertebral disc biopsy was taken and referred to our pathology service. Upon histologic evaluation (hematoxylin-eosin stain, original magnification  $\times 100$ ) we observed monotonous sheets of spindle cells with abundant dilated "staghorn" vessels (Figure 4). Higher magnification (hematoxylin-eosin stain, original magnification  $\times 400$ ) showed monotonous spindle cells, with minimal cytologic atypia and no mitosis (Figure 5).

On immunohistochemistry evaluation, the tumor stained positive for CD56, CD99, S100, and vimentin, whereas it was negative for CD34, Bcl-2, synaptophysin, chromogranin, desmin,



**Figure 4.** The tumor is composed of monotonous sheets of spindle cells with abundant dilated “staghorn” vessels (hematoxylin and eosin,  $\times 100$ ).



**Figure 5.** Higher magnification showed monotonous spindle cells with minimal cytologic atypia and no mitosis (hematoxylin and eosin,  $\times 400$ ).

actin, and epithelial membrane antigen (EMA). Phosphohistone H3 and Ki67 staining protocols were consistent with a low proliferative index, reporting 3 mitoses in 10/HPF and low (up to 10% of cells in some areas) proliferation, respectively. The diagnostic impression was of a spindle cell lesion with features of SFT/HPC with inconclusive immunohistochemistry.

Given that our institution does not perform the STAT6 immunoreactivity protocol, the case was referred to the National Institutes of Health (NIH) for consultation. NIH evaluation confirmed the diagnosis of SFT/HPC on the basis of histologic features, despite having a negative STAT6 nuclear immunoreactivity.

After the diagnosis was received and discussed with pediatric oncologists, the patient was transferred to another hospital center in the U.S. mainland for specialized management and treatment options. Hence, further management and follow-up information is not available.

## Discussion

SFTs and HPCs are rare mesenchymal tumors that were initially described as separate entities; however, because of similar pathologic characteristics they were recently merged under a single classification [1,2]. Even though these tumors can arise in any part of the body, they are uncommonly found in the central nervous system (CNS), located above the tentorium, around the pineal gland, or extending from the spinal cord meninges. In fact, patients with spinal SFTs/HPCs are commonly middle-aged and complain of motor and sensory symptoms unique to the tumor location [3]. In the majority of the previously reported cases the tumor arises from the thoracic spine, involving the intradural extramedullary compartments [4].

Spinal SFTs/HPCs rarely appear on imaging studies in the current literature. When this tumor is suspected, CT may aid in diagnosis, but MRI with intravenous gadolinium contrast is more commonly used [5]. On imaging, SFT/HPC of the CNS can be found as an oval or irregular heterogeneous mass extending from the dural planes with well- or ill-defined margins on CT or MRI [6]. When infiltration of adjacent structures such as vertebrae or soft tissues occurs, the tumor margins may be ill defined and could be indicative of malignant variants of the disease. Regions of patchy hypoattenuation are likely caused by ischemia from inadequate blood supply to the tumor center. When a spinal lesion with these features is identified, it should be differentiated from common spinal lesions including meningiomas or nerve sheath tumors such as schwannomas. A schwannoma usually presents as a well-defined dumbbell-shaped spinal lesion, typically heterogeneously hyperintense on T2-weighted images with areas of cystic degeneration and contrast enhancement. Meningiomas commonly appear isointense relative to the spinal cord on both T1- and T2-weighted images, with a homogeneous enhancement pattern [7].

Particularly in the pediatric population, a neuroblastoma should be considered, as it is the most common extracranial solid tumor in childhood and the most frequently diagnosed neoplasm during infancy. In the majority of cases, a neuroblastoma occurs within the abdomen, but may present along the paraspinal region, extending into the neural foramina, causing compression of nerve roots and of the spinal cord [8].

Before the World Health Organization (WHO)'s revision of SFT/HPC, some studies attempted to characterize the imaging features of SFT and HPC separately, with conflicting results demonstrating a wide range of tumoral MR signal intensities and enhancement patterns [9]. Current unified WHO guidelines

state that SFT/HPC demonstrate isointensity on T1-weighted MRI and high or mixed signal intensity on T2-weighted MRI. However, in our case we observed a heterogeneous signal on T1-weighted images, as well as a mixed and predominantly hyperintense signal on the T2-weighted images.

Diagnostic confirmation of CNS SFT/HPC is currently achieved through the combination of histologic and immunohistochemical analysis. On histomorphologic assessment, these tumors are highly vascular and characteristically display a prominent irregular branching pattern known as staghorn vessels. In addition, these tumors may present with varying degrees of cellularity of spindle to ovoid tumor cells and scant cytoplasm within a collagenous stroma. Although the majority of SFTs exhibit a benign histology and behavior, atypical malignant forms can occur. The malignant criteria should be advocated by a high mitotic count, increased cellularity, hemorrhage or necrosis, pleomorphic nuclei, and foci of dedifferentiation [10].

Immunohistochemistry with existing markers has been of variable value in making a definitive diagnosis. Beside the suggestive histomorphologic features, SFT/HPC tumors have been formerly diagnosed by the frequent coexpression of CD34, Bcl-2, and CD99 [11]. Generally, the most characteristic immunohistochemical marker is CD34, present in 95% of cases; however, CD34 reactivity can be absent or only focally present in some atypical cases of SFTs or expressed in other benign mesenchymal tumors or sarcomas [12].

The other markers commonly identified yet variably expressed in SFTs include CD99, Bcl-2, nuclear  $\beta$ -catenin, and EMA. All these markers are nonspecific and could be positive in other soft-tissue tumors that may mimic SFTs [12]. SFTs may also rarely stain for S100, cytokeratin or desmin and differential diagnosis in such cases may include a wide variety of spindle cell tumors primarily of fibroblastic/myofibroblastic or peripheral nerve sheath lineage. In contrast, immunohistochemical nuclear expression for the upregulated STAT6 protein, which results from a gene fusion event on chromosome 12, has been previously recognized in many studies as a highly sensitive and specific immunohistochemical marker for SFT/HPC, being useful to distinguish this tumor type from histologic mimics [11,12].

Currently, SFTs/HPCs are classified in a three-tier grading system that aids in determining treatment algorithms. In this classification, grade I tumors are highly collagenous and have areas alternating between hypocellularity and hypercellularity. Grade II tumors are characterized by high cellularity, reticular fibers, and scarce collagenous structures. Grade III tumors have similar histologic characteristics to grade II, but demonstrate anaplasia, with  $>5$  mitoses per 10 high-power fields [13].

In our case, histologic evaluation of the specimen revealed a highly cellular tumor with monotonous sheets of spindle cells, a low amount of collagen, and thin-walled branching vessels in a staghorn pattern. These histologic findings were consistent with a grade II SFT/HPC.

On immunohistochemistry analysis our specimen was CD99 positive. However, it was S100 positive and negative for both CD34 and Bcl-2 markers, which is an uncommon immunoprofile for SFT/HPC [14]. Furthermore, our specimen was also negative for STAT6 nuclear protein immunoreactivity, which is remarkably unusual for SFT/HPC tumors. Although a negative nuclear protein immunoreactivity is sufficient to exclude the diagnosis, it may be lost in cases of dedifferentiated SFTs [15].

In a recent study validating the STAT6 immunoreactivity by Tan et al. [10], the majority of STAT6-negative tumors suspected to represent SFTs were reclassified, with alternative diagnoses supported by additional immunohistochemical or molecular studies. In their study, reclassification of the STAT6-negative tumors revealed some common mimics of SFT/HPC that they recommend be considered in the differential diagnosis in the pediatric population. These were fibroblastic or myofibroblastic tumors, nerve sheath/neural tumors, sex cord stromal tumors, and poorly differentiated small round blue cell tumors. Histologically, each of these tumors demonstrated focal to diffuse areas comprised of spindle cell proliferations with variably collagenous backgrounds and focal HPC-like features. In fact, they concluded that SFT is an overly diagnosed tumor in the pediatric population and encouraged the use of STAT6 nuclear stain as confirmatory for its diagnosis.

A review by Geramizadeh et al. [15] analyzes the role of immunohistochemistry in the diagnosis of SFTs, states that a double-negative CD34 and Bcl-2 tumor (as in our case) makes the diagnosis of SFT highly unlikely, and that to call a tumor SFT, a CD34 marker should be positive. They considered that the only exception would be in malignant and dedifferentiated cases, pertinent to the pathologic analysis in our case that demonstrated minimal cytologic atypia without reaching the criteria for a malignant tumor.

## Conclusions

This case makes it clear that correlating the radiological characteristics of a tumor with its pathologic findings and biological behavior still remains a challenge. First of all, our case is particularly rare in that the histologic characteristics showed features classically attributed to SFTs/HPCs, which was unexpected for the patient's age group. In addition, the cytologic analysis showed a completely discordant immunohistochemical profile that is of a debatable diagnosis in the current literature.

To our knowledge, this is the youngest patient to present with a spinal tumor with these pathologic features.

The pathologic challenges encountered in this case report suggest that future studies are still needed to clarify the biological nature of the immunohistochemical marker of these tumors. Even though a rare presentation of a STAT6-negative SFT/HPC was the final diagnosis in this case, additional studies should

be undertaken to establish unified imaging features of SFT/HPC to develop more specific radiologic diagnostic criteria that could potentially correlate with pathology.

### Conflicts of interest

None.

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