



## Solitary Fibrous Tumor Developing Two Years After Primary Cranial Synovial Sarcoma: A Rare Case

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**ABSTRACT** Synovial sarcoma is an aggressive and rare malignant soft tissue neoplasm, typically affecting young adults and predominantly arising near major joints. Primary cranial involvement is extremely uncommon. In rare circumstances, patients may develop multiple soft tissue tumors either synchronously or metachronously, raising a challenging differential diagnosis between metastatic disease and a second primary malignancy.

We report the case of a woman diagnosed with a primary high-grade cranial synovial sarcoma, treated with wide surgical resection followed by adjuvant radiotherapy, who remained disease-free for two years. She later presented with multifocal skeletal lesions. Biopsies taken from two distinct bone lesions demonstrated diffuse STAT6 positivity and the absence of SS18 rearrangement by molecular analysis. These features, confirmed by independent pathological reviews and a multidisciplinary tumor board, were consistent with a multifocal solitary fibrous tumor rather than metastatic disease.

This rare case highlights the diagnostic complexity of multifocal skeletal lesions developing after primary cranial synovial sarcoma and emphasizes the importance of thorough pathological, molecular, and multidisciplinary evaluation.

**Keywords:** Synovial sarcoma, solitary fibrous tumor, cranial tumor, multifocal bone lesions.

### INTRODUCTION

Synovial sarcoma accounts for 5-10% of all adult soft tissue sarcomas<sup>1</sup>. The incidence of synovial sarcoma does not show a gender bias. The median age of synovial sarcoma patients is approximately 35 years<sup>2</sup>. Overall 5-year survival rates for patients range from 37% to 81%<sup>2</sup>. Synovial sarcoma can occur anywhere in the body and at any age, but it most commonly occurs in and around the joints. It can also occur in the cranial region, but this has been reported rarely. It has a unique metastatic pattern and, unlike many other sarcomas, metastasizes to lymph nodes, other soft tissues, and bone. The simultaneous or metachronous development of two or more primary soft tissue sarcomas has been reported<sup>3</sup>. These are primary lesions occurring in multiple soft tissue sites without pulmonary or lymphatic involvement. Radiotherapy has been used as a palliative treatment for intracranial lesions. Synovial sarcoma remains a challenge in both

diagnosis and treatment. Future treatment developments are expected to focus on molecular targets, including inhibition of angiogenesis<sup>4</sup>.

Here, we present a rare case of primary cranial synovial sarcoma followed two years later by multifocal skeletal lesions, two of which were histologically consistent with solitary fibrous tumor, raising an important diagnostic and therapeutic dilemma.

### CASE PRESENTATION

In the case we reported, informed consent was obtained from the patient. A woman presented to the neurosurgery outpatient clinic approximately two years ago with complaints of a swelling in her head. An extradural mass was identified posterior to the left parietal region. The mass was resected with wide margins, and the sample was sent for pathology. Histopathological examination revealed a high-grade synovial sarcoma with clear surgical margins. Immunohistochemical

analysis showed positivity for vimentin, Bcl-2, CD99, TLE-1, CD34, focal CD31 and EMA, with a Ki-67 proliferation index of approximately 20%. The tumor was negative for GFAP, desmin, SMA, HMB-45, and Melan-A. Molecular analysis demonstrated SS18 gene rearrangement, supporting the diagnosis of synovial sarcoma. Despite the absence of tumor necrosis, the lesion was classified as high-grade based on high cellularity, elevated mitotic activity (24 mitoses per 10 high-power fields), and the presence of a round cell component.

Postoperatively, the patient received adjuvant cranial radiotherapy. Adjuvant radiotherapy of 6000 cGy in 30 fractions was applied to the left parieto-occipital region using IMRT technique and helical tomotherapy. Systemic staging at the time of diagnosis revealed no evidence of extracranial disease. The patient remained clinically and radiologically disease-free for approximately two years.

Two years later, the patient presented with a one-month history of progressive left elbow pain. Physical examination revealed painful and limited elbow flexion–extension as well as severely restricted pronation and supination. Neurovascular examination was normal.

A preoperative brain magnetic resonance image taken 2 years ago revealed a lesion with a lobulated contour measuring approximately 31 x 30 millimeters in the posterior bony structure of the left parietal posterior, destroying both the inner and outer tables and demonstrating heterogeneous enhancement on the post-contrast series (Figure 1).

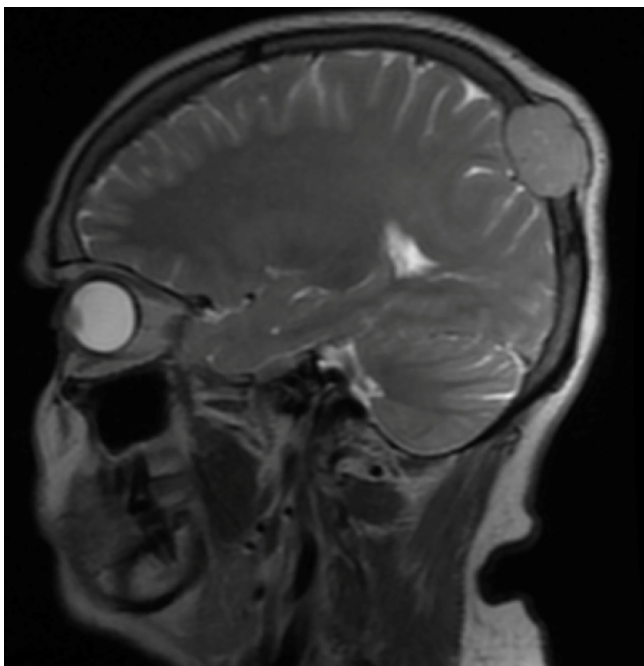


Fig. 1 — Preoperative brain magnetic resonance imaging.

A postoperative brain magnetic resonance image taken 2 years ago revealed a postoperative defective appearance measuring approximately 40 x 18 x 44 millimeters in the posterior bony structure of the left parietal posterior (Figure 2).

An elbow radiograph revealed a lytic mass in the radial neck, expanding the bone and causing a pathological fracture (Figure 3).

Contrast-enhanced elbow MRI was performed. A 2x2 cm lesion was observed in the proximal radius, with an area of intense intramedullary peripheral edema causing cortical expansion and cortical thinning and destruction. Post-contrast images revealed a homogeneous contrast area (Figure 4).

PET CT imaging revealed an appearance consistent with a craniectomy defect in the left parietal bone. No residual pathological FDG uptake consistent with recurrence was observed in the surgical site. Skeletal system, increased FDG uptake with an SUV max of 8.3 was observed in lytic bone lesions in the left proximal radius, left half of the sacrum, left pubis, and left femoral neck, indicating malignancy (Figure 5).

To ensure diagnostic accuracy and address the multifocal nature of the skeletal involvement, sequential biopsies were obtained from two distinct anatomical sites: the left superior pubic ramus and the sacrum. These specimens were evaluated independently by two separate pathology centers, with additional confirmation provided by an external reference laboratory. Immunohistochemical staining demonstrated diffuse nuclear STAT6 positivity, focal positivity for TLE-1 and CD34, and negativity for S100 and Bcl-2 (Figure 6).

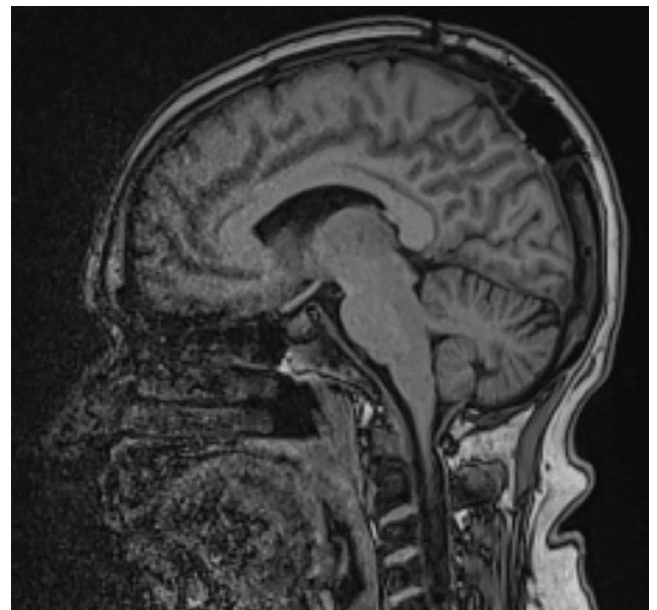


Fig. 2 — Postoperative brain magnetic resonance imaging.



Fig. 3 — Image of a mass in the radial neck that is lytic in character, expands the bone and causes pathological fracture.



Fig. 4 — This is the image of a 2x2 cm lesion that causes thinning and destruction of the cortex, with an intramedullary peripheral dense edema area causing expansion in the cortex in the proximal radius.

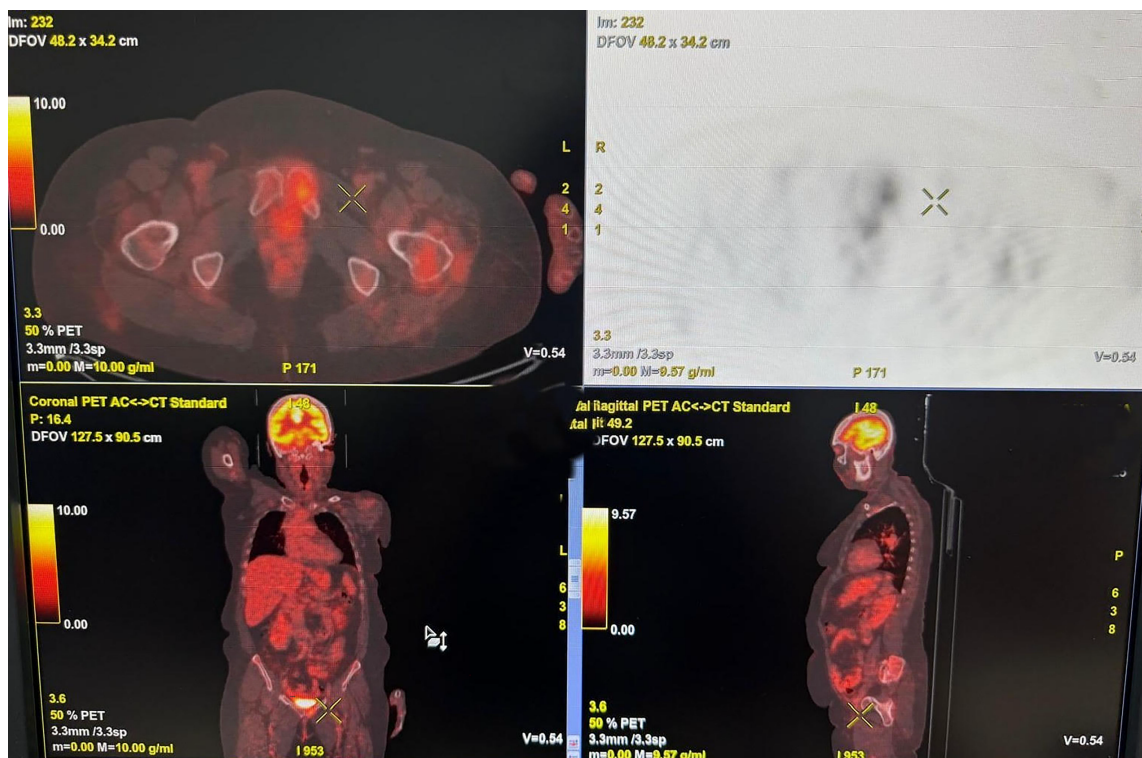


Fig. 5 — PET CT imaging revealed an appearance consistent with a craniectomy defect in the left parietal bone. No residual pathological FDG uptake consistent with recurrence was observed in the surgical site. Skeletal system, increased FDG uptake with an SUV max of 8.3 was observed in lytic bone lesions in the left proximal radius, left half of the sacrum, left pubis, and left femoral neck, indicating malignancy.



*Fig. 6 — Sequential biopsies were obtained from the left superior pubic ramus and the sacrum. Histopathological examination revealed features consistent with solitary fibrous tumor. Immunohistochemical staining demonstrated diffuse nuclear STAT6 positivity, focal positivity for TLE-1 and CD34, and negativity for S100 and Bcl-2.*

To further differentiate these lesions from the primary malignancy, molecular analysis was performed at an external center, which confirmed the absence of SS18 rearrangement in the skeletal lesions. Based on the findings of STAT6 positivity and the absence of SS18 rearrangement, the lesions were interpreted as a second primary solitary fibrous tumor.

Considering the patient's poor general condition (ECOG performance status 3), multifocal skeletal involvement, a multidisciplinary tumor board recommended non-surgical management. The patient was treated with immobilization using a long-arm splint, palliative radiotherapy, and systemic therapy with pazopanib. As palliative radiotherapy, 300 cGy fractional dose was applied in 10 fractions to left sacrum + left pubic bone + left femoral head using IMRT technique and helical tomotherapy, for a total of 3000 cGy palliative radiotherapy. No orthopedic surgical intervention was performed.

## DISCUSSION

Synovial sarcoma is a clinically rare malignant tumor. This malignancy is generally considered a soft tissue malignant tumor originating from joints, synovium, and tendon sheath synovium and is typically seen in the extremities of young adults. However, it can occur anywhere in the body, including the heart, kidney, throat, and tongue, and case reports are increasing<sup>5-8</sup>.

The case we report represents a synovial sarcoma that primarily originates in the cranial region. This location is quite rare for synovial sarcoma.

The clinical symptoms of primary intracranial synovial sarcoma are nonspecific. Diagnostic findings depend on the tumor's location, size, and complications. Clinical symptoms of primary intracranial synovial sarcoma include headache, nausea, vomiting, limb hemiplegia, and speech impairment<sup>9,10</sup>. In our case, the initial symptom was a palpable mass in the head. Notably, two years after the initial diagnosis, the patient presented with elbow pain, and subsequent scans revealed multifocal skeletal lytic lesions. The emergence of multifocal bone lesions in our patient two years after the initial treatment raised a critical diagnostic question: metastatic synovial sarcoma or a second primary tumor?

Biopsies from two different skeletal lesions showed features consistent with a solitary fibrous tumor. The diagnostic certainty was established through both immunohistochemical and molecular evidence; the lesions demonstrated diffuse nuclear STAT6 positivity while simultaneously showing an absence of SS18 rearrangement. This combination effectively rules out the possibility of metastatic synovial sarcoma and confirms the diagnosis of a second primary multifocal solitary fibrous tumor. Furthermore, the diagnosis and the subsequent management plan were validated by a multidisciplinary tumor board consisting of orthopedic

surgery, pathology, medical oncology, and radiation oncology specialists. While the inability to obtain tissue samples from every single focus is considered a limitation, the consistency of the STAT6 positivity across two separate biopsy sites, the exclusion of SS18 rearrangement, and the multidisciplinary consensus provide a high level of diagnostic confidence.

The most common site of metastasis for synovial sarcoma is the lung. This is followed by lymph node involvement. Synovial sarcoma rarely metastasizes to the skeleton and, when it does occur, most commonly involves the long bones<sup>2</sup>. Synchronous sarcomas are primary soft tissue sarcomas that occur in multiple soft tissue sites, usually without pulmonary or lymphatic involvement. It is often difficult to distinguish between synchronous and metachronous manifestations of second primary and soft tissue sarcoma metastasis<sup>2</sup>. As seen in the case we report, the patient's primary synovial sarcoma originated from the cranium, and multifocal lytic lesions that appeared 2 years later on the axial skeleton and extremities were considered to be the second primary. The pathology report supports the finding that it is a solitary fibrous tumor.

## CONCLUSION

This rare case highlights the diagnostic complexity of multifocal skeletal lesions developing after primary cranial synovial sarcoma. The integration of multi-site biopsies, independent pathological reviews confirming STAT6 positivity alongside the absence of SS18 rearrangement, and a multidisciplinary tumor board evaluation involving orthopedists, pathologists, medical oncologists, and radiation oncologists was essential in successfully distinguishing this second primary tumor from metastatic disease.

*Ethical Approval:* No ethical approval is required. Informed consent was obtained.

*Consent for publication:* Informed consent was obtained from the patient.

*Availability of data and materials:* The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request

*Conflict Of Interest:* The authors declare no conflict of interest regarding this article.

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