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## Case Report

# Malignant primary intraosseus synovial sarcoma – a rare case report<sup>☆</sup>

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## ABSTRACT

Primary intraosseous synovial sarcoma is an extremely rare malignancy that occurs primarily in young adults. We present a case of a primary intraosseous synovial sarcoma of the right distal ulna in a 19-year-old female. It has a propensity to mimic other radiologic and pathologic diagnosis. Histopathology after a surgical excisional biopsy with a wide margin plus adjunct radio and chemotherapy are necessary to improve prognosis.

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## Introduction

Synovial sarcoma is a rare, highly malignant tumor that occurs mainly in adolescents and young adults between the ages of 15–30 years with peak prevalence in the third decade; there is a slight male preponderance. There are four commonly recognized subtypes which include biphasic, monophasic

fibrous, monophasic epithelial and poorly differentiated tumors [1–3].

Primary intraosseous sarcoma is extremely rare and only 39 cases have been reported in peer reviewed journals in English literature. Only thirteen (13) of these have radiologic support with molecular and/or cytogenetic confirmation of the diagnosis. The total cases reported included 26 monophasic and 11

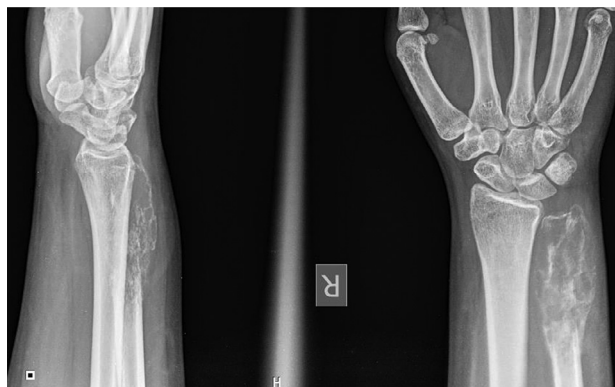
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**Fig. 1 – Lateral and anteroposterior radiographs of the right distal forearm showing a lytic, expansile mass within the distal ulna and adjacent soft tissue swelling.**

biphasic histologic subtypes affecting 18 males and 19 females with no data available for two cases [4]. We present a case of primary intraosseous synovial sarcoma in the distal ulna bone of a 19-year-old female with radiologic imaging and histologically confirmed monophasic fibrous type lesion with immunocytochemistry markers.

### Clinical findings

A 19-year-old female presented in the outpatient clinic with insidious right wrist pain and swelling of 9 months duration with no preceding history of trauma. On examination there was a swollen and painful ulna side of the right wrist joint with pulses and sensation intact. No limitation of the range of movement was seen.

An x ray of the forearm and wrist was requested. It shows an expansile, mixed lytic lesion with a broad zone of transition in the right distal ulna bone (Fig. 1). A provisional diagnosis of a giant cell tumor was made. Fibrous dysplasia and Ewing's sarcoma were listed as possible differential diagnosis.

An MRI of the right forearm done shows a distal ulna intraosseous  $7.07 \times 2.63 \times 1.92$  cm lobulated, iso to hypointense, heterogenous mass to muscle on T1 W. Heterogenous, mixed intensity signal showing a triple signal pattern (hyper, intermediate and hypointense signal patterns to fat) on T2W images. Avid contrast enhancement and areas of cystic necrosis were seen on post contrast T1W images (Figs. 2–4).

A tru-cut biopsy of the distal right ulna done showed a connective tissue lesion composed of spindle-shaped cells arranged in a whorled growth pattern with occasional large cells with ganglion like appearance and a diagnosis of nodular fasciitis was made.

A subsequent excisional biopsy of the distal right ulna bone was done (Fig. 5) with a wide margin of resection and the lesion was sent for histology of the resected bone tissue. Microscopy showed a mesenchymal tumor arranged as vague herringbone and cart wheel appearance. The cells were composed of spindle cells with abundant

cytoplasm and fusiform nuclei. There was marked pleomorphism. The tumor cells had cigar butt shaped nuclei with a coarse granular focal pattern. Numerous mitotic figures and areas of necrosis were also seen. The tumor was seen to destroy bone and adjacent subcutaneous fat. Immunohistochemistry showed a CD 99, Vimentin positive lesion, and negative to desmin, S100, NSE and smooth muscle actin (Figure 6).

She presented a year later with local recurrence of the lesion after surgery and follow up chemotherapy. A follow up right forearm CT scan showed an aggressive soft tissue mass in the region of the previously excised distal ulna (Figs. 6, 7 and 8). The mass was fungating and ulcerating with contact bleeding. A workup to exclude distant metastasis including a chest CT showing no pulmonary metastasis and correction of anemia were done. She then had an above elbow amputation of the right forearm (Fig. 9) with follow up adjuvant radiotherapy and chemotherapy scheduled when stable.

### Discussion

Synovial sarcoma is a rare malignancy [1] with primary intraosseous origin subtype extremely rare. It is commonly found in adolescents and young adults; our case was 19 years of age at presentation. In a large study of 3228 synovial sarcoma patients, the male to female ratio was found to be 52.1% (1707 males) to 47.1% (1521 females) [5]. However, reported primary intraosseous synovial sarcomas subtypes including our study have a male to female ratio of 47.4% (18 males) to 52.6% (20 females). This shows a slight reversal of the male to female ratio with females more commonly affected in the primary intraosseous subtype.

Monophasic synovial sarcoma is prevalent in adolescents and young adults with tumor recurrence being common after excision of the primary tumor [6] which is precisely what we see in our patient as well falling within the age bracket and having local tumor recurrence in place of distant metastasis which usually affects the lungs [7].

The initial diagnosis of a giant cell tumor on plain radiography is plausible as intraosseous osteosarcoma is a mimicker of giant cell tumors in the limbs [8]. MRI is the imaging modality of choice for the diagnosis and initial staging of synovial sarcoma [9]. Typical findings including a mass larger than 5 cm, usually heterogenous, well delineated with a triple signal pattern in which areas of hyper-, iso-, and hypointensities to fat on T2-weighted imaging [10] were seen in this patient. The most reliable confirmation is histology of the excised tumor.

The tru-cut biopsy [11] showed a nodular fasciitis, it has been reported that nodular fasciitis is often misdiagnosed as a soft tissue sarcoma [12] and it is the most important differential diagnosis of this condition. Plaza et al [13] reported that two thirds of their cases had been misdiagnosed as sarcoma. A different study showed a diagnostic dilemma when clinical findings suggestive of nodular fasciitis which turned out to be a peripheral nerve sheath tumor [14]. This case highlights the diagnostic dilemma encountered and the benefit of excisional biopsy and immunohistochemistry [15] in aiding our diagno-

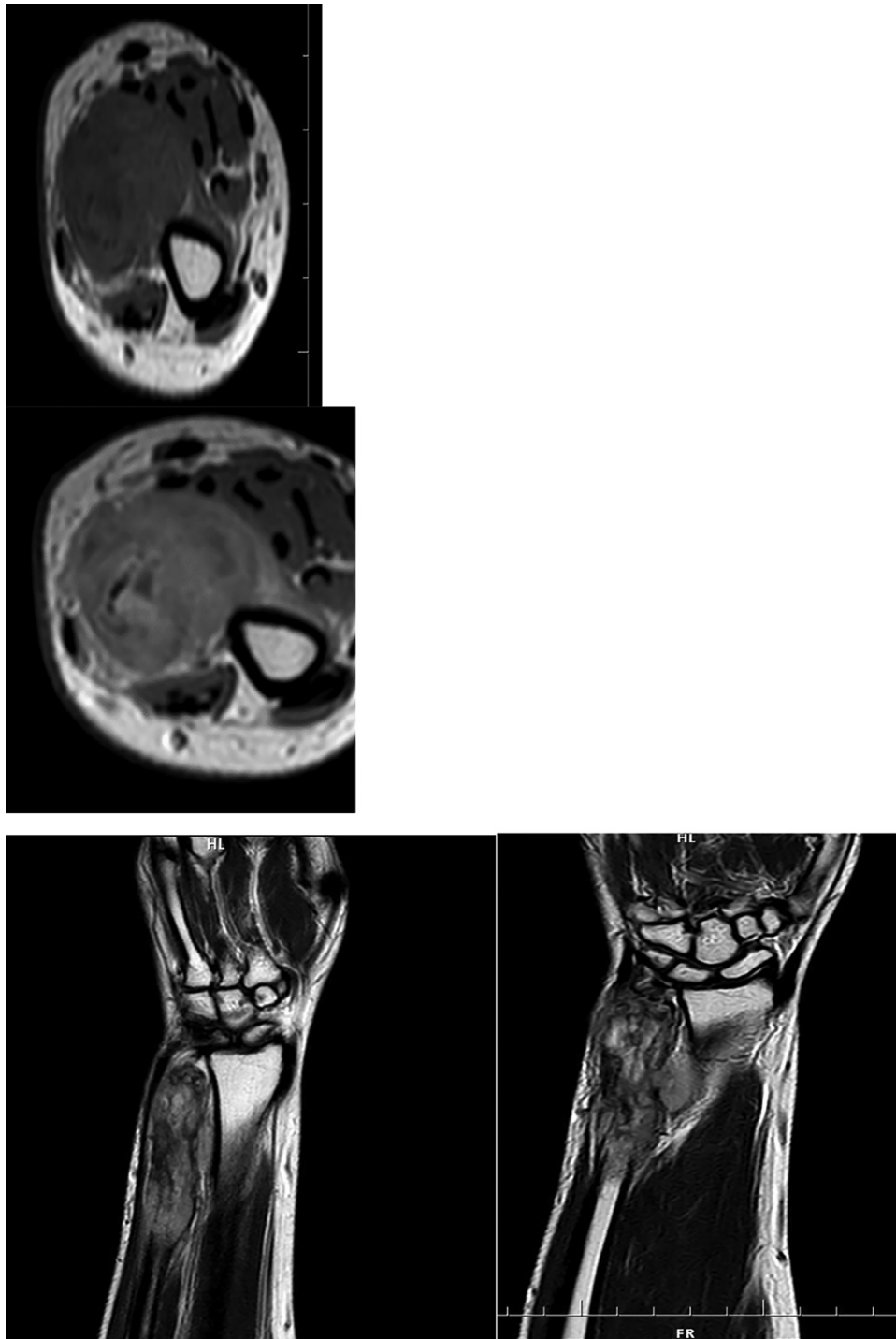
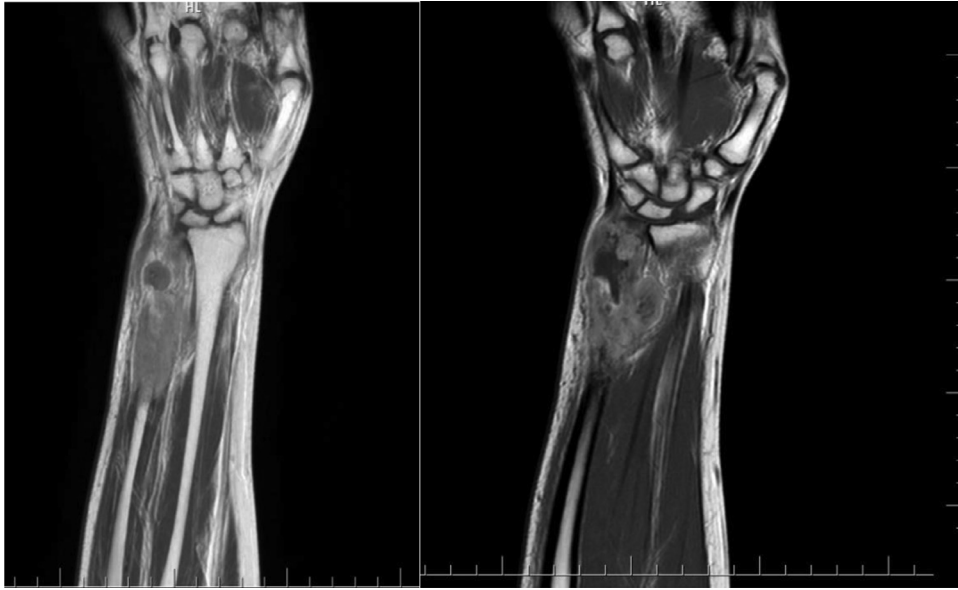


Fig. 2 – T1W pre- and postcontrast axial Images showing right distal ulna lesion with avid enhancement post contrast.



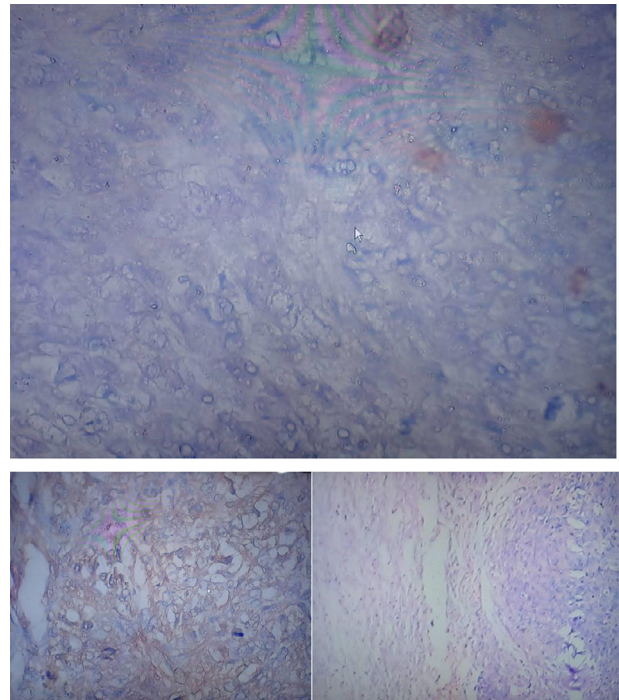
**Fig. 3 – T2W coronal (a) and sagittal (b) images showing mixed intensity, heterogenous lobulated mass in right distal ulna bone.**



**Fig. 4 – T1W post contrast coronal images showing enhancement with necrosis.**



**Fig. 5 – Gross specimen of excised synovial sarcoma of the distal ulna.**



**Fig. 6 – Histological slides showing features in keeping with synovial sarcoma.**

sis. Imaging with nuclear medicine techniques plays an important role in diagnosis, treatment planning, and follow up of sarcomas [16]. It is, however, not available within the resource poor environment we operate, albeit would have contributed to improve overall patient outcome.

Wide surgical resection is the standard of care for management of this lesion with adjuvant radiation therapy for deeper lesions and chemotherapy showing some benefits in certain populations although it remains controversial [5,17]. We performed below elbow amputation and in conjunction

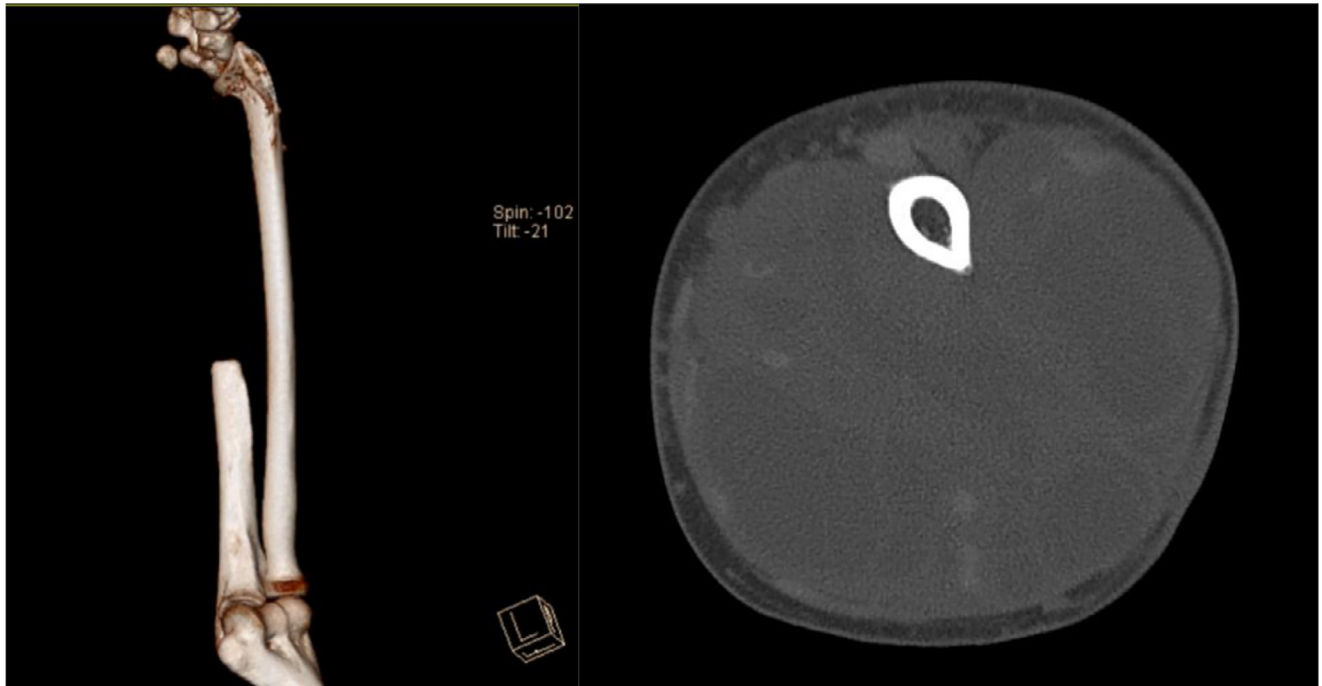


Figure 7 – Volume rendered and axial CT image of right forearm showing excised distal ulna with huge soft tissue mass in the region of the previously excised tumor

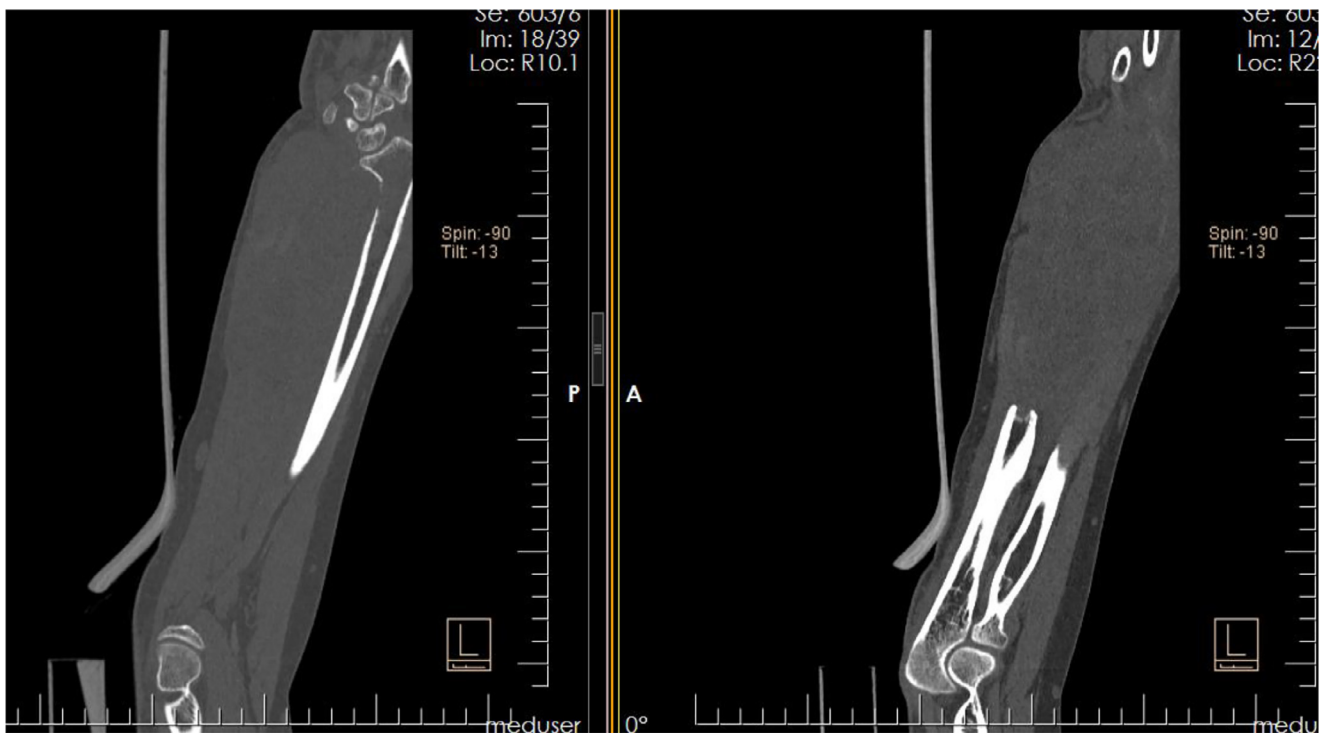


Fig. 8 – MPR reformatted CT image showing extent of recurrent synovial sarcoma involving the soft tissue in the resected tumor bed.



**Fig. 9 – Gross specimen of excised recurrent sarcoma in the right distal ulna region.**

with adjuvant radio- and chemotherapy, we therefore expect a marked increase in life expectancy.

## Conclusion

Primary osseous synovial sarcoma is a rare malignant lesion with only 39 prior reported cases in the English literature. We present a 19-year-old female with recurrent synovial sarcoma of the distal right ulna and have thus demonstrated the importance of imaging, histology with immunocytochemistry in the diagnosis and management of synovial sarcoma.

## Patient consent

The authors obtained written informed consent from the patient for submission of this manuscript for publication.

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