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Synovial sarcoma of cervicodorsal spine: A case report

Wali Umer Shah
Pakistan Institute of Medical Sciences (PIMS), Islamabad.

Syeda Dania Shujaat
University of Oklahoma Health Sciences Center, Oklahoma, USA

Naseeb Ullah
Aga Khan University

Salman Mansoor
Shifa International Hospital, Islamabad.

Synovial sarcoma of cervicodorsal spine: A case report

Wali Umer Shah,[1] Syeda Dania Shujaat,[2] Naseeb Ullah,[3] Salman Mansoor[4]

Abstract

Synovial sarcoma is a soft-tissue sarcoma. Its involvement of the spine is extremely rare. We report a 40 year old male who presented with shoulder pain and progressive weakness in all four limbs for six months with a visible, slowly growing bulge in his upper back. On examination he had quadriparesis and diffuse sensory deficit. MRI of the cervical spine showed a large soft tissue mass, iso to hypointense, extending into the neural canal, compressing the cord. The mass had a few internal areas of contrast enhancement with extension into the right paraspinal regions involving the vertebral bodies. Sub-periosteal spine dissection was done. Tumour was primarily extradural, involving and extending from paraspinal soft tissues to the posterior arches, laminae and spinous processes of the verteberae, with their destruction. Gross radical removal of the visible mass was done, followed by three cycles of radiation therapy. Excisional biopsy showed synovial sarcoma TYPE II. In conclusion, synovial sarcomas should be kept in the differentials of a mass arising in spinal axis.

Keywords: Sarcoma, Synovial, Quadriplegia, Granulomatous disease.

Introduction

Synovial sarcoma is an uncommon neoplastic entity accounting for 7-10% of all soft-tissue sarcomas.[1] Its involvement of the spine is extremely rare as most of synovial sarcomas involve extremities of the body near large joints.[2,3] Among the 3 % head and neck region involvement, majority of tumours are in hypopharynx.[4,5] The reporting of synovial sarcoma of the spine is limited to case reports so far in the literature.[7,9] so the management remains undecided. We report a rare case of primary synovial sarcoma of the cervicodorsal spine here.

Case Report

We report a case of a 40 years old male who presented to us in June 2014 in outpatient of Pakistan Institute of Medical Sciences (PIMS) Islamabad with complaints of shoulder pain bilaterally radiating to upper limbs and progressive weakness in all four limbs for the past six months. According to the family there was a slowly growing bulge visible in his upper back region. Three months prior to presenting to us he had an excisional surgery for the lump in the dorsal region of the back (Further details of the surgery were not available). His biopsy at that time was suggestive of granulomatous disease and he was taking anti tuberculous agents since then, with no significant improvement. The mass reappeared at the same site and the patients now developed urinary incontinence as well.

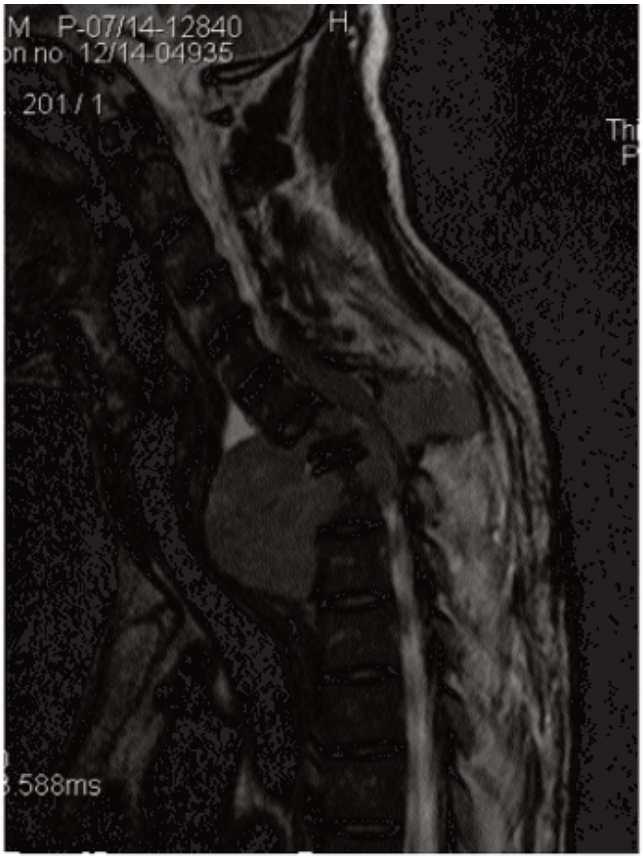


Figure-1: T 1 weighted sagittal view showing a large soft tissue mass, iso to hypo intense. Mass is extending into the neural canal, compressing and probably involving the cord also.

[1]Pakistan Institute of Medical Sciences (PIMS), Islamabad, [2]University of Oklahoma Health Sciences Center, Oklahoma, USA, [3]Aga Khan University Hospital, Karachi, [4]Shifa International Hospital, Islamabad.
Correspondence: Naseeb Ullah. Email: kakar441@gmail.com

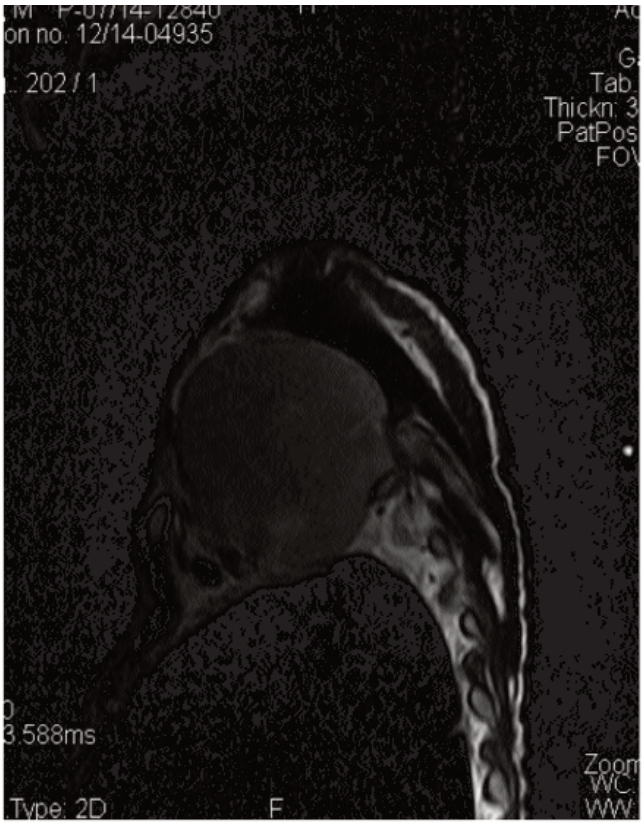


Figure-2: T 1 weighted image with contrast in the same sagittal view showing few internal areas of enhancement in the mass.

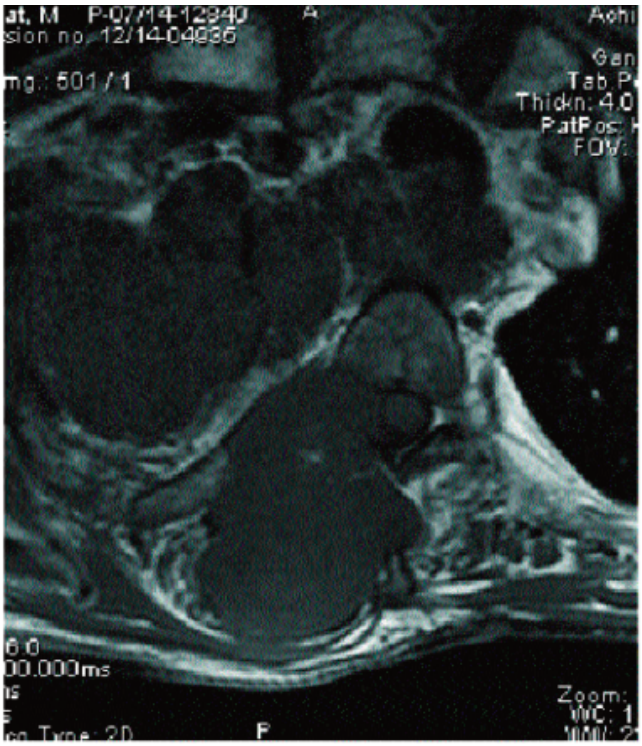


Figure-3: Axial T 1 weighted image showing the mass is extending to the right Para spinal regions and involving the vertebral bodies.

On examination patient was alert and obeying commands. Speech and cranial nerves examination was normal. He had quadriparesis with lower limbs power 0/5 bilaterally and upper limbs 2/5 on right side and 4/5 on left side. There was generalized wasting of muscles of upper and lower limbs bilaterally. There was a sensory loss to pinprick sensation diffusely up to the dermatomal level of T4.

As patient was worsening despite a previous surgery and anti tuberculous medications and the mass had reappeared, we decided for reimaging of the region (Figure 1, 2 & 3 with description). Radiologist suggested biopsy for classifying this apparently malignant mass in his detailed report. Surgery for maximum possible excision and histopathology of the mass was planned. Spine was explored from C5 to D5 with midline posterior approach, sub-periosteal dissection was done. It was a large tumour primarily extradural, involving and extending from Para spinal soft tissues to the posterior arches, laminae and spinous processes of the vertebrae, with their destruction. It was also compressing the cord. Superiorly it was reaching the retropharyngeal space, inferiorly it was reaching the lower border of fourth thoracic vertebra, and anteriorly it was touching and compressing the mediastinal structure as oesophagus. Gross radical removal was done for the whole visible mass in its entirety, including the part in the mediastinum. Mass was grayish white, rubbery to feel and highly vascular. It was 14x12x4 centimeters in size, weighing 550 grams. Complete mass was sent to the histopathological laboratory.

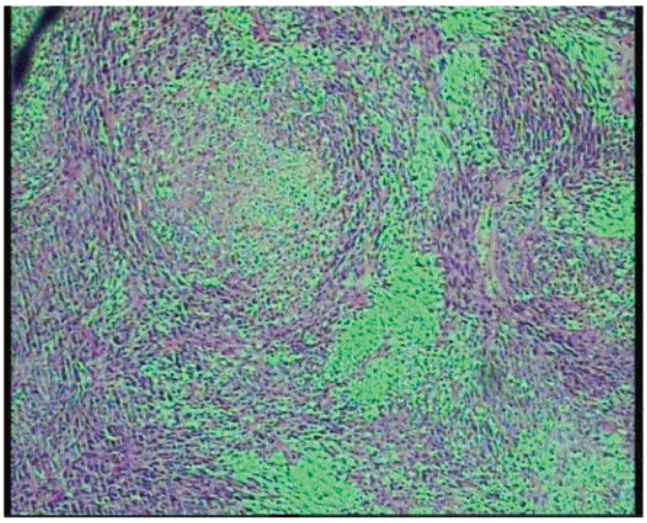
Histopathology

On microscopic examination it was a cellular tumour with fascicular pattern of growth, composed of monomorphic population of spindle cells showing elongated nuclei and mitoses. Hypo cellular areas, foci of necrosis along with infiltration by mast cells was also appreciated. Background contained vessels with a haemangiopericytoma like pattern [Slide photos 1, 2 & 3).

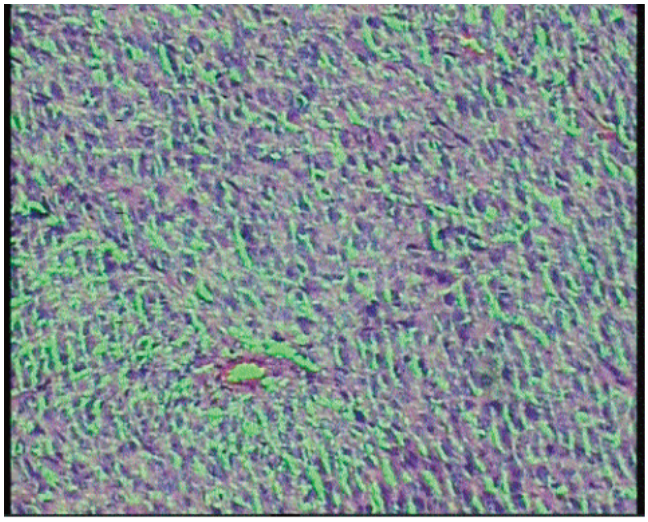
On immunohistochemical analysis it was positive for CD99 and Vimentin [Slide photos 4 & 5).

S-100 C protein, CD68 and EMA were negative [Slide photos 3, 6 & 7). On these grounds it was diagnosed as a synovial sarcoma TYPE II.

After surgery and biopsy result we consulted the oncology center for further intervention, and it was



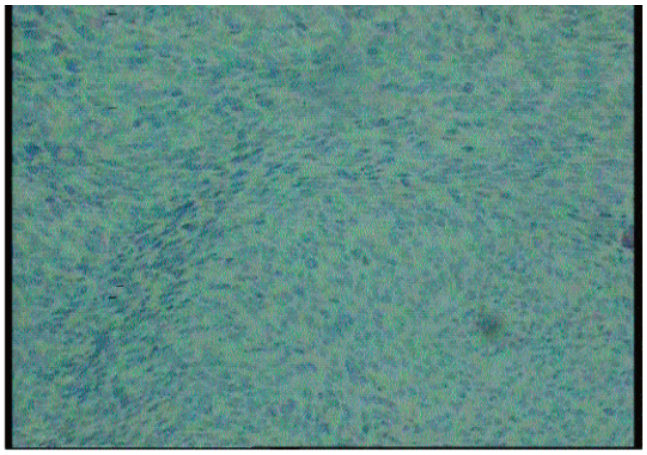
Slide photo-1: Low power view of cellular tumour arranged in fascicles of spindle cells. A focus of necrosis and hypo cellular areas can also be appreciated.



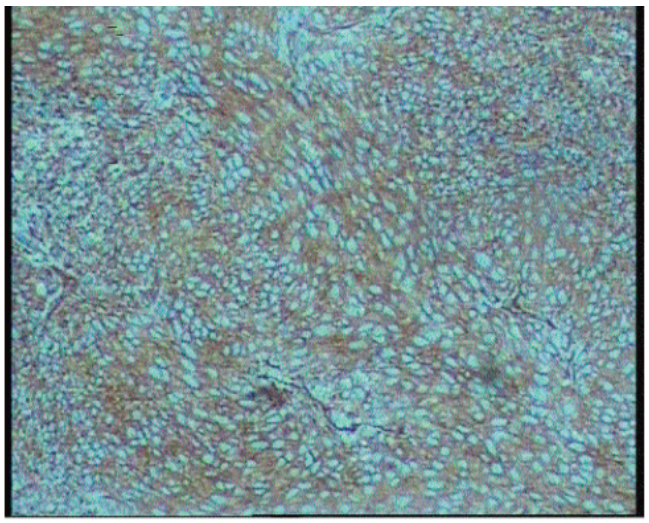
Slide photo-2: High power view of monomorphic spindle cells with elongated nuclei showing scattered mitotic figures. A dilated and ectatic (haemangiopericytoma like) blood can also be seen in the center.

decided to give him three cycles of radiotherapy in "palliative setting". During this period patient made a good post operative recovery. He was pain free started to walk with a single person support but, bladder was still incontinent and he was catheterized.

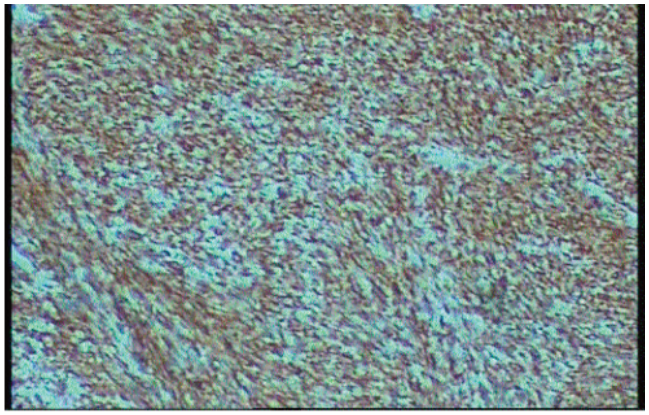
As the patient was clinically improving we planned for imaging after radiation and settling of surgical changes (edema, stitches removal). At this stage, his family requested for earlier discharge and they would come back for follow up. He was discharged with advice of regular physiotherapy and follow up imaging of the spine.



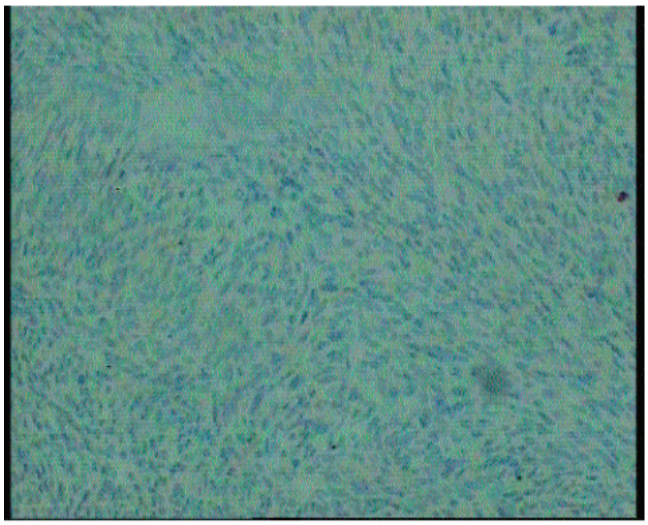
Slide photo-3: Tumour cells demonstrating negative expression for EMA immunohistochemical stain.



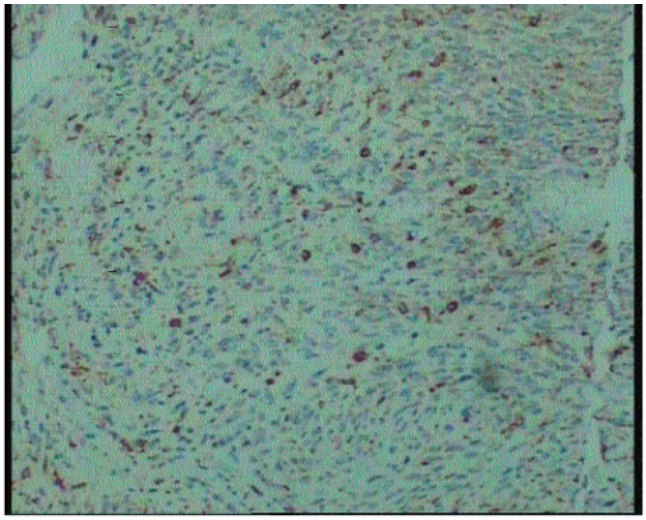
Slide photo-4: Tumour cells demonstrating strong and diffuse positive expression for CD99 immunohistochemical stain.



Slide photo-5: Tumour cells demonstrating strong and diffuse positive expression for Vimentin immunohistochemical stain.



Slide photo-6: Tumour cells demonstrating negative expression for S100 immunohistochemical stain.



Slide photo-7: Tumour cells demonstrating negative expression for CD68 immunohistochemical stain.

But he did not come for follow up after discharge. Their residence was far from any tertiary care hospital, so his family was approached who revealed that the patient did well for the first two years after surgery but then developed fever and diarrhoea and died within days while after being admitted at a local facility which was not a tertiary care center as family was financially weak. All these sad flaws in management were because of poor economic conditions and lack of communication with our centre.

Discussion

Synovial sarcoma of the soft tissue are named so not because of synovial origin but because of the microscopic appearance of their degenerated mesenchymal cells that is very similar to synovial tissue.[6]

Their appearance near large joints usually present with local pain, swelling and walking abnormality which may initially be misinterpreted as bursitis, lipoma or myosarcoma. They involve the head and neck region in only 3% of the cases,[4,5] with features of throat pain, dysphagia or dyspnoea, with involvement of the spine limited to only case reports so far in the literature.[7-9] Not surprisingly, in case of the spinal involvement, features of spinal cord compression, neurogenic pain and radiculopathy, are encountered which add to the neurological differential diagnoses of the rarity list.

Imaging can be proceeded with either C.T scan or MRI but MRI appears superior in assessment of soft tissue involvement details. The tumour is highly malignant and engulfs soft tissues, bones, ligaments, nerve roots and spinal cord.

Synovial sarcomas usually vary in size between 2 and 9 cm[10,11] but our patient had 14x12x4 cm which has not been reported yet. One study showed that 5.8% of cases have distant metastases at diagnosis, mainly to lung. Metastasis was very much associated with primary tumour size: 32 times more in tumours of size > 5 cm than for tumours <5 cm.[12]

Histopathology gives the diagnosis of choice. Synovial sarcoma has two main subtypes based on histological appearance of cells: the biphasic form reveals both epithelial and spindle cells and the monophasic type reveal spindle cells only (our case).[7]

Treatment usually applied for these lesions is surgical excision followed by radiation and/or chemotherapy. Prognosis is favourable unless the metastatic lesion appear in other tissues like lungs[8] and liver[9] in which case mortality is high. Role of chemotherapy is controversial.[13]

Synovial sarcomas near large joints need total amputation and limited excision results in a high recurrence (60-90%) within two years of surgery.[14] There is a possibility that our patient had a recurrence or metastasis in other tissues which may have resulted in his death after two years of treatment, because the was young and had no other major illnesses. Due to his affordability and education status we could unfortunately not ensure a regular follow up and screening for possible recurrence or metastasis, as expected in these cases.

Conclusion

As with all rarities, although down the list, synovial

sarcomas should be kept in differentials of a mass arising in spinal axis as the high index of suspicion and early diagnosis benefits the patient prognosis, morbidity and mortality.

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Conflict of Interest: None.

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