

## Management of Metastatic Spine and Hip Alveolar Soft Part Sarcoma: Case Report and Review of Literature

### Abstract

Alveolar soft part sarcoma (ASPS) is a rare soft tissue tumor. Primary or metastatic involvement of the spine is unusual in ASPS. In most case, it is refractory to chemotherapy and radiation. Surgical resection is the most effective intervention. We report the case of a 38-year-old female having ASPS along with metastatic spine and hip involvement treated surgically as a single-stage operation, which is the first of its kind approach to our knowledge. We present the case of a 38-year-old female with simultaneous L4 pathological fracture with symptomatic lumbar canal stenosis without focal neurology and pathological fracture of neck of femur of left hip, secondary to metastatic ASPS. Since both conditions were contributing equally to her disability and demanded early intervention, they were treated simultaneously with intralesional excision of the tumor and posterior stabilization of the spine and left hip proximal femur resection and replaced it with proximal femur endoprosthesis as a single-stage operation. Postoperatively, she had significant relief of radiculopathy and left hip pain. She was mobilized out of bed on the postoperative day 1 and was discharged from hospital on the postoperative day 6. She was given chemotherapy drug sunitib postoperative. At her last follow-up, 20 months' postoperative, she was asymptomatic and was independent in terms of activities of daily living. Metastatic ASPS of the spine and hip is a rare clinical entity. Simultaneous surgical treatment of the spine and hip pathology is technically demanding. If the conditions demands, as in our case, both of them can be managed safely in a single-stage with good midterm outcome.

**Keywords:** Alveolar soft part sarcoma, hip, metastasis, spine, surgery

### Introduction

Alveolar soft part sarcoma (ASPS) is a rare soft tissue malignancy, which accounts for about 1% of all soft tissue sarcomas.<sup>[1,2]</sup> This lesion usually occurs in adolescents and adults between 15 and 35 years of age, with slight preponderance in females.<sup>[3,4]</sup> The tumour typically occurs in the deep soft tissues, most often in the buttock and thigh, with a smaller number of cases at other soft-tissue locations such as the arm, chest and retroperitoneal tissues.<sup>[4-6]</sup> ASPS is usually chemoresistant and radio-resistant.<sup>[7]</sup> *En bloc* resection of involved vertebra offers the best chance of cure when tumor is contained.<sup>[8,9]</sup> Endoprosthetic replacement is safer method than osteosynthesis in the proximal femur metastatic fracture.. Endoprostheses are not dependent on healing of the fracture, which is often poor in patients with cancer because of systemic and local factors.<sup>[10]</sup> Metastasis to lung, brain, and bone is common in ASPS.<sup>[11,12]</sup>

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The patient was informed about the data of her clinical condition being used for publication and gave consent for the same.

### Case Report

A 38-year-old female presented with low back pain radiating to bilateral buttocks and both lower limbs in the distribution of L4 and L5 dermatome. She walked independently with a walker. There was no bowel and bladder dysfunction. On examination, the patient had a diffuse tenderness in the lumbosacral spine. On neurological examination, she had normal sensations and motor power in the lower limbs. Deep tendon reflexes were normal. No upper motor neuron signs were noted. Babinski's sign was negative. Magnetic resonance imaging (MRI) [Figures 1-3] showed L4 pathological fracture with soft tissue causing severe lumbar canal stenosis and compression of thecal sac along with nerve roots. The patient was referred

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for computed tomography (CT)-guided biopsy that was inconclusive (showing few stromal/spindle cells). Hence, it was decided to proceed with open biopsy along with intralesional debulking of the tumor and stabilization of the spine. The patient suffered an unprovoked hip fracture which was recognized by sudden increase in localized pain around the left hip. It was confirmed by X-ray of the pelvis with both hips [Figure 4].

Scheduled surgery was deferred and trucut biopsy from the left proximal femur was planned. It showed metastatic clear cell carcinoma consistent with renal tissue. Positron emission tomography (PET)-CT scan revealed increased uptake in the rectum, suggesting possibility of colo-rectal carcinoma, but not renal growth or lesion. This caused a diagnostic dilemma. Colonoscopy was performed which did not show any lesion or growth from colon or rectum again adding to confusion.

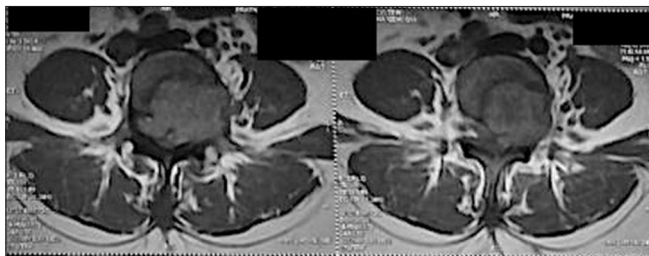
Revised Tokuhashi score<sup>[13]</sup> is useful to determine the prognosis of metastatic spine tumor. Scores range from 0 to 15. Her score was 9 which suggests her prognosis for survival more than or equal to 6 months. Spinal instability neoplastic score (SINS)<sup>[14]</sup> is useful to determine the instability in spine neoplasia. Scores range from 0 to 18. Scores from 7 or more surgery are indicated. In our case, her SINS was 7 which suggests spine to be potentially unstable.

After all the failed attempts to achieve a diagnosis, it was decided to go ahead with the open biopsy along with intralesional excision of the tumor and stabilization of the spine. To get a good functional outcome, the hip lesion had to be tackled as well. Since the patient was young and fit, we decided to go ahead with the hip surgery in the same anesthesia. We got a wide excision of the proximal femoral mass and reconstructed with a megaprosthesis. The patient lost about 1.5 l of blood in total for which she was transfused postoperatively. The total time under anesthesia was 8 h. The patient was mobilized bedside the next day, and the patient was discharged on the postoperative day 6. At the time of discharge, she was walking independently on a walker managing well with nonopioid analgesics. There were no complications during the postoperative period in the hospital. Histopathological study of the sample from the spine and hip was reported as ASPS [Figure 5].

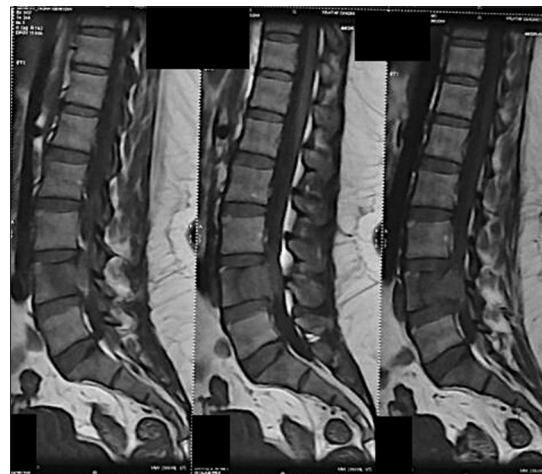
The primary site of the tumor remained unknown. During one of the postoperative visits, we noticed a swelling in the right calf which was firm in consistency and nonpulsatile



**Figure 1:** Preoperative sagittal magnetic resonance imaging (T2-weighted) of LS spine showing L4 pathological fracture with soft tissue encroaching spinal canal



**Figure 3:** Preoperative axial magnetic resonance imaging (T1-weighted) of L4 pathological fracture with soft tissue showing encroachment of lesion into spinal canal



**Figure 2:** Preoperative sagittal magnetic resonance imaging (T1-weighted) of lumbosacral spine showing L4 pathological fracture with soft tissue encroaching spinal canal



**Figure 4:** Preoperative left proximal hip fracture

which was missed initially. The patient reported it as being present for several years which she had ignored as it was asymptomatic. Primary lesion over soleus was missed on PET scan as screening was done from head to knee as per the radiologist protocol.

We did ultrasonography of the left calf swelling that was considered lipoma, till date. There was evidence of lobulated hypoechoic mass arising from soleus muscle belly of about 4.4 cm × 4.4 cm × 10 cm in size [Figure 6]. Fine-needle aspiration cytology was done and it confirmed ASPS [Figure 7]. As her primary tumor was asymptomatic and she was on chemotherapy, it was not intervened surgically. Her postoperative X-ray of the spine and pelvis both hips were showing proper position of implants [Figures 8 and 9].

At her last follow-up, 20 months' postoperative, she was asymptomatic and was independent in terms of activities of daily living. Her follow-up X-rays were not showing any loosening of implants. However, imaging of choice for soft tissue sarcoma is MRI to see any recurrence.

### Discussion

The majority of patients of ASPS have metastasis at the time of diagnosis.<sup>[6]</sup> Five-year survival rate in metastatic

ASPS is 20%.<sup>[8,9]</sup> In our case, primary site (right calf) of ASPS remained indolent as long as 10 years and presented lately due to metastasis of the spine and hip, secondary to ASPS. Surgical stabilization and decompression are strongly recommended for patients with radiation-resistant tumors in the setting of high-grade spinal cord compression.<sup>[15]</sup> Her tumor was not contained in the body of the L4 vertebrae, so *en bloc* resection was not possible. Diffuse soft tissue component arising from the body of L4 was encroaching spinal canal causing severe compression. Wide surgical excision with posterior fixation was done one level above and below the lesion.

Surgical intervention for metastatic pathological hip fracture was imperative to decrease her pain and early mobilization. Surgical intervention varies from intramedullary nailing, open reduction, and internal fixation to prosthetic replacement. Chances of nonunion are higher in internal fixation devices in metastatic proximal femur fracture and the reoperation rate is higher.<sup>[10]</sup>

Proximal femur resection and prosthetic reconstruction are preferred in patients with extensive bone destruction, in patients with pathologic fractures, in tumors resistant

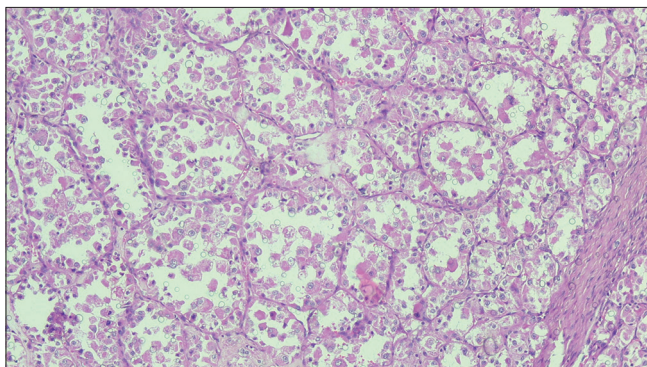


Figure 5: Histopathology showing round or polygonal tumor cells with abundant eosinophilic cytoplasm arranged in the alveolar pattern

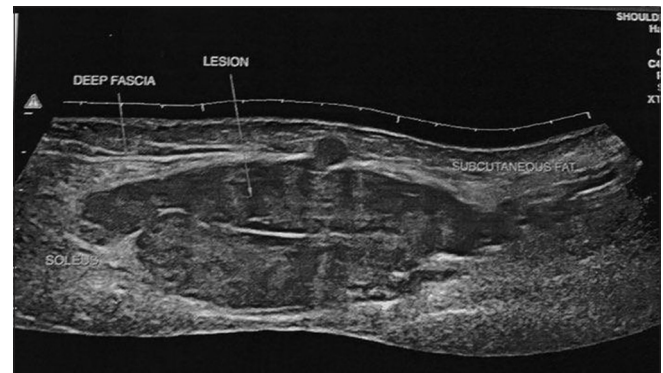


Figure 6: Ultrasound of the right calf showing hypoechoic lesion in soleus of approximate 4.4 cm × 4.4 cm × 10 cm

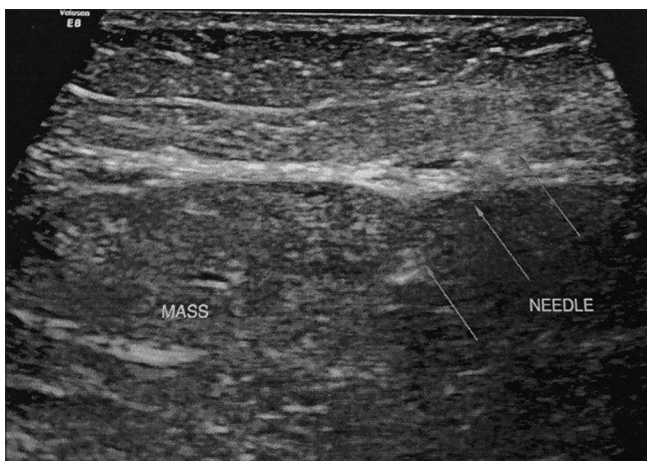


Figure 7: Fine-needle aspiration cytology procedure under ultrasound guidance from the right soleus

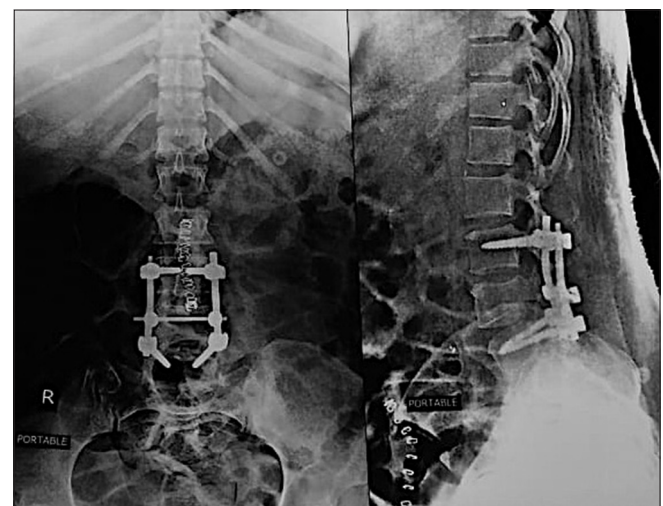


Figure 8: Postoperative X-ray of the lumbosacral spine anteroposterior and lateral view

to radiation therapy, and in patients with more proximal metastatic lesions.<sup>[10]</sup> Endoprostheses replace bone whereas osteosynthetic implants are at best load-sharing and will ultimately fail if the bone does not heal. Thus endoprosthesis is better than osteosynthesis.

Stereotactic body radiotherapy (SBRT) or stereotactic radiosurgery (SRS) with intensity-modulated radiation therapy and an image guidance technique<sup>[16]</sup> has emerged as a new treatment option for spinal metastasis. It is one of the potential treatment options for patients with unresectable or metastatic sarcoma. SBRT offers a noninvasive and convenient fractionation schedule, minimizing treatment burden and chemotherapy delays. Strict immobilization and advanced imaging techniques allow for delivery of highly conformal dose distributions and large dose per fractionation, thereby increasing the biological effective doses beyond conventional fractionation. SBRT is a theoretically attractive local control option for patients with metastatic or locally advanced unresectable soft tissue or bone sarcomas and offers the possibility of increased efficacy. SBRT is convenient and minimizes delays in systemic therapy. In contrast to conventional radiotherapy, spine SRS delivers highly conformal radiotherapy in single- or hypo-fractionated regimens and may offer superior local control of spinal sarcomas.<sup>[17-19]</sup> Indeed, 1-year local control of sarcomas following SRS has been reported to range from 70% to 90% with single-fraction doses of 22–24 Gy. However, patients with rapidly progressive neurologic deficits or emergent spinal instability are not treated with SRS and require surgical intervention.

In our case, proximal femur resection and replacing it with proximal femoral endoprosthesis for pathological hip fracture and intralesional surgical excision with posterior stabilization for metastatic spine tumor were performed in single setting. It helped in early mobilization of the patient [Figure 10] and reduced her overall length of hospital stay.

Her tumor cells expressed transcription factor E3 (TFE3) on immunohistochemistry which confirmed ASPS [Figure 11]. An antibody directed against the C-terminus of TFE3 has emerged as a highly sensitive and specific marker of the tumor.<sup>[2,20,21]</sup> Alveolar soft part sarcoma locus-TFE3 fusion protein is a new target for novel chemotherapy and antiangiogenic therapy.<sup>[2,20]</sup> Drugs such as sunitinib have shown partial response.<sup>[7,22]</sup>

She was started on sunitinib after consultation with an oncophysician.

In summary, ASPS poses a diagnostic and therapeutic dilemma. A high index of suspicion is needed for early diagnosis and treatment. Aggressive surgical resection of the symptomatic metastasis is recommended. Although chemo-resistant, sunitinib has some role



Figure 9: Postoperative X-ray of the pelvis with both hips



Figure 10: Postoperative day 1, she was mobilized with walker and brace

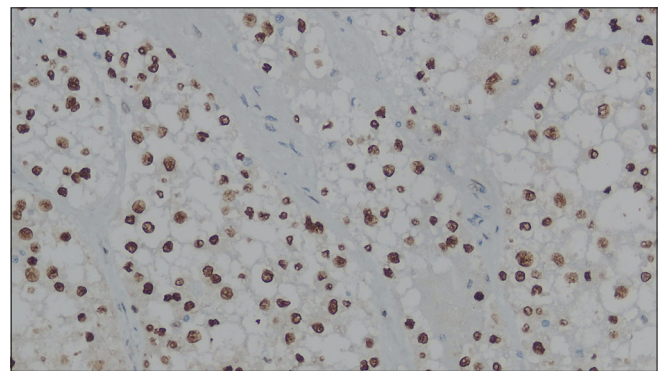


Figure 11: Immunohistochemistry showing nuclear transcription factor E3 expression

in disease control. Single-sitting hip and spine surgery, in a carefully selected patient, is safe with less morbidity, good outcome, and reduced length of stay in hospital. Research should be encouraged for new drug trials for better metastatic and local disease control in ASPS.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

## Conflicts of interest

There are no conflicts of interest.

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