Endometrial stromal sarcoma metastasis to the lumbar spine and sphenoid bone

Mary L. Huang,1 Robert L. DeBernardo,2 Mark Rodgers,3 David J. Hart1
1Department of Neurological Surgery, Case Western Reserve University School of Medicine, University Hospitals Case Medical Center, Cleveland, Ohio; 2Department of Obstetrics and Gynecology, Case Western Reserve University School of Medicine, University Hospitals Case Medical Center, Cleveland, Ohio; 3Department of Pathology, Case Western Reserve University School of Medicine, University Hospitals Case Medical Center, Cleveland, Ohio, USA

Abstract

ESS is typically associated with metastasis to the abdomen, pelvis, and lung. We found three case reports of ESS metastasis to the bone (two to the thoracic spine, and one to the parietal bone). Our objective is to review the literature on endometrial stromal sarcoma (ESS) spinal and intracranial metastases, and report the first case of ESS metastatic to the lumbar paraspinal region and sphenoid bone. A 53-year-old female with ESS status-post radiation, chemotherapy, and pelvic exenteration surgery presented with right hip weakness, back pain, and radicular leg pain that were explained by chemotherapy-induced neuropathy, radiation-induced lumbosacralplexopathy, and femoral nerve and obturator nerve injury during pelvic exenteration surgery. During routine positron emission tomography, we found metastasis to the L3 lumbar spinal region. L3 laminectomy and subtotal resection of the mass was performed with tumor residual in the neuroforamina and pedicles. One month later, an MRI performed for persistent headaches revealed a large lesion in the sphenoid bone that was biopsied transphenoidally with the same diagnosis, but no further surgery was performed. She is intolerant of chemotherapy and currently undergoing whole brain radiation. Delay in the diagnosis and management of lumbar paraspinal and sphenoid bone metastasis of ESS likely occurred because of the uniqueness of the location and aggressiveness of ESS metastasis. Health care providers should be aware of potentially aggressive metastasis of ESS to bone, in particular the unusual locations of the lumbar paraspinal region and sphenoid bone.

Materials and Methods

This case report was developed from direct patient clinical encounters; events that took place in the operating room; review of medical records; review of radiographic studies; and pathology slides. An interdisciplinary group from neurosurgery, gynecologic oncology, and pathology reviewed, discussed, and developed this case report.

Results

A 53-year-old Caucasian female with ESS was referred to our neurosurgery outpatient clinic in March 2010 after a routine PET scan two months earlier revealed increased uptake in the L3 spinous process (Figure 1). MRI of the lumbar spine in March 2010 revealed a 3-centimeter heterogeneously enhancing mass involving the lamina, spinous process, right pedicle, and right transverse process of L3, with expansion into the paraspinous muscles and without involvement of the dorsal fascia or epidural space (Figure 2).

The patient initially presented to her primary care physician three years earlier with symptoms of urinary incontinence, and a workup revealed uterine fibroids. She underwent supracervical hysterectomy. The pathologist identified multiple leiomyomas within the myometrium, some with myxoid change. One year later, the patient developed worsening urinary incontinence. She was found to have a large pelvic mass. Core biopsy showed a myxoid spindle cell lesion with reactivity against desmin and weak reactivity against muscle-specific actin. Immunohistochemistry against CD10 was non-reactive. The features were interpreted as suggestive of leiomyosarcoma. Biopsies of additional intra-abdominal lesions were morphologically similar, but these tumors failed to react against muscle-specific actin, smooth muscle actin and CD10. Therefore a diagnosis of primary endometrial stromal sarcoma was favored.

The patient underwent preoperative radiation therapy followed by extensive resection of the intraabdominal tumor. Discovery of metastatic disease led to an end colostomy and
palliative chemotherapy with taxotere and ifos/Adria. Because of intolerable side effects (including leukocytosis, alopecia, skin changes, fatigue, and nausea) and local progression of disease, she was switched to hormonal therapy with megestrol, then an aromatase inhibitor. Her disease was stable for a year on hormonal therapy, then the patient underwent pelvic exenteration (hemicolecotomy in October 2008; cystectomy, proctectomy, and partial vaginectomy in July 2009). Evaluation of the resected colon, urinary bladder and vagina again showed a spindle cell tumor morphologically similar to that seen in the patient’s previous biopsies and resection, but also small areas of high-grade sarcoma. This specimen showed reactivity with anti-CD10 antibody and no reactivity against desmin, actin, myoD or S100 protein. The findings were interpreted as consistent with endometrial stromal sarcoma, predominately low grade, with focal areas with high grade features.

The patient’s paraspinal mass was unusually aggressive. Two months after pelvic exenteration surgery, the patient reported hip flexion and abduction weakness. Diagnostic considerations included chemotherapy-induced neuropathy, radiation-induced lumbosacral plexopathy, and iatrogenic femoral nerve injury. Her obturator nerve was resected during the surgery. Electromyogram showed right obturator neuropathy, which was believed to explain her right hip flexion and abduction weakness.

At the time of the routine PET scan that revealed the lumbar mass (January 2010), she had no back pain. One month later, she developed dull aching lower back pain, with occasional stabbing on the right, and paresthesias bilaterally to the feet. Physical examination revealed no neurologic deficits and no palpable mass in the lumbar region. A MRI of the cervical and thoracic spine showed a small, rounded, hypointensity of the C4 vertebral body but was otherwise normal. A bone scan showed suspected metastases in the bilateral femoral diaphyses, left humeral head, right seventh rib, and mid-lumbar spine. The original PET scan from January 2010 also showed mild enlargement of a right middle lobe lung mass and other bony metastases.

Despite the presence of other bony and soft-tissue metastases, we decided to decompress the lumbar mass because of the patient’s good performance status, intolerance to chemotherapy, the large size of the lesion that correlated with the imaging and limited her daily activities, and the expectation of a minimally morbid surgery given the predominantly dorsal location of the lesion. The goals of surgery were to improve her hip and back pain and debulk the tumor to facilitate adjuvant treatment.

Intraoperatively, the tumor protruded through the lumbar fascia. A clear plane was evident between the tumor and paraspinous muscles in some areas, but roughly 60% of the tumor was blended with the surrounding muscles and the L3 spinous process was partially eroded. After L3 laminectomy, we found significant epidural tumor extension with adherence to the dura. Epidural tumor in the central canal was resected, but tumor remained in the lateral recess traveling out the neural foramen on the right at L2–3 and in the right L3 pedicle (Figure 3). Based on preoperative imaging and

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**Figure 1.** PET/CT showing increased uptake and erosion of the spinous process in the dorsal L3 region.

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**Figure 2.** Preoperative lumbar MRI images. (A) Sagittal T1 with gadolinium, (B) Axial T1 with gadolinium.
the patient’s known multiple metastases, we
had not planned for gross total resection. We
did not perform fusion and stabilization
because the procedure is associated with a
higher complication rate, longer recovery time,
and felt that it would not affect overall survival.
Further, because of her planned radiation ther-
apy, she had a higher risk of non-union. Total
volume of resected tumor submitted to pathol-
yogy was 181.2 cm³ compared with preoperative
volume estimated by MRI to be 27 cm³. This
change represented a 5.7-fold increase in total
tumor volume over a 6-week period. The
pathology report identified a malignant myxoid
spindle cell lesion with predominately low
grade features and focal high grade morpholo-
gy which was consistent with origin from the
patient’s previously diagnosed sarcoma
(Figure 3). The patient then received palliative
radiation to the lumbar paraspinal region. She
was offered systemic chemotherapy, but
refused because of her previous experience
with the chemotherapy-related side effects.
During workup for a persistent headache
one month later, MRI of the head revealed an
avidly enhancing expansile mass in the body of
the sphenoid bone, with soft tissue extension
into the sphenoid sinus and posterior ethmoid
air cells. Abnormal marrow replacement sur-
rounded the area in the lesser wing of the
sphenoid bone and clivus (Figure 4). A stereo-
tactic-assisted transsphenoidal biopsy was per-
formed without complication. The lesion was
diagnosed as a myxoid sarcoma with
histopathological features similar to those
seen in the preceding biopsy and resection
specimens (Figure 5). The tumor represented
had features of a low histologic grade. The
patient is currently receiving whole-brain radi-
ation. No chemotherapy was administered due
to her history of intolerance.

Discussion
This case represents a significant and
unusual presentation for several reasons: i)
ESS rarely metastasizes to bone; this is the
first reported case of ESS metastasis to the
lumbar spine and sphenoid bone; ii) contrary
to most reported cases of ESS, this tumor grew
very aggressively; iii) there was a delay in
diagnosis due to the broad differential of hip
weakness, back pain, radicular leg pain, and
headache in patients receiving treatment for
ESS; iv) these rare tumors pose significant
challenges to pathologic diagnosis; v) limited
clinical information about spinal ESS meta-
tasis complicates treatment decision-making.

Metastasis of ESS typically occurs in the
pelvic, abdominal, and lung regions. This case
study demonstrates the ability to ESS to metas-
tasize aggressively to bone. Adding to the
paucity of case studies of ESS bone metastasis
(2 to the thoracic region, 1 to the parietal
bone), we see here for the first time that ESS
can also spread to the lumbar spine and spheno-
noid bone. On bone scan, this patient also had lesions in the bilateral femoral diaphyses, left humeral head, and right seventh rib. MRI showed a lesion in the C4 vertebra. Although no biopsy was performed in these other regions, we have high clinical suspicion that these areas may also have ESS metastasis. This patient was unique because her mass was in the lumbar region without vertebral body involvement. The tumor extended into the surrounding soft tissues and epidural space, which has not been previously reported.

It is unclear how long the lesion had been growing and whether the dramatic expansion of the mass in the 6 weeks prior to surgery represented a transformation of a previously indolent lesion. Throughout 2009 while being treated for various abdominopelvic issues, including a suspected presacral abscess, the patient had multiple CT scans of her abdomen and pelvis. At the time, no lumbar spine lesion was identified, though abdominal CT is not ideal for examining lumbar spine pathology. However, in retrospect we found erosion of the L3 spinous process and a 1.5-centimeter soft tissue mass that was missed in the original August 2009 report (Figure 6). In a minimally symptomatic to asymptomatic patient, it is difficult to determine the appropriate treatment. In this case, the decision to operate despite the presence of other metastases was based on the low-grade nature of the ESS tumor, the size of the lesion, and the patient's desire for a minimally morbid surgery. We did not realize how aggressive this tumor was pre-operatively.

In conclusion, delays in diagnosis and management in this case report likely occurred because of the uniqueness of the locations and aggressiveness of ESS metastasis. It is our hope that this report will increase awareness among health care providers of the potentially aggressive metastasis of ESS to the bone, in particular to the craniospinal axis.

Figure 5. Pathology specimen from lumbar tumor resection, H&E stain, 200x magnification.

Figure 6. Abdominal axial CT scan images with bone windowing showing subtle, small mass lesion with erosion of the L3 spinous process.

References