Primary epidural liposarcoma of the cervical spine: Technical case report and review of the literature

Hamid Borghei-Razavi, Khairi-Mohamed Daabak, Sahar Bakhti, Uta Schick

**Abstract**

Liposarcoma is the most common soft tissue sarcoma in adults. These tumors have a high incidence of osseous metastases, with a propensity to the spine; however, primary spinal involvement is very rare. A 56-year-old female patient presented with a 4 month history of cervical pain, including radiation to both upper limbs, without radicular distribution. Magnetic resonance imaging (MRI) showed an epidural lesion with gadolinium enhancement and bilateral extension into the intervertebral neural foramina (C5–C7), with spreading on the right side of the tumor into paravertebral tissue. The histopathological diagnosis was myxoid liposarcoma. To our knowledge it is the first case of primary myxoid liposarcoma of the cervical spine in the literature. Although rare, our case demonstrates that liposarcoma should be considered in the differential diagnosis of cervical tumors.

**Introduction**

Liposarcoma is the most common soft tissue sarcoma in adults. It is classified into five distinct histological types: well-differentiated, dedifferentiated, myxoid, round cell, and pleomorphic [1].

Myxoid liposarcoma is the second most common subtype, accounting for 30%–40% of cases, and can frequently metastasize to extrapulmonary locations [1–3]. These tumors have a high incidence of osseous metastases, with a propensity to the spine [4]. However, primary spinal involvement is rare (one case with first manifestation involving the thoracic spine, two cases with lumbar involvement, and one case of intradural involvement).

In this article, we present for the first time a patient with primary liposarcoma of the cervical spine as the primary tumor origin.

**Case presentation**

**History and physical examination**

A 56-year-old female patient presented with a 4 month history of cervical pain, including radiation to both upper limbs, without radicular distribution. There was no clinical evidence of cervical cord compression. The patient was in good general condition, and had no other symptoms and no weight loss. Her medical history was unremarkable.

**Neurological examination on admission revealed apraxia and triceps paresis on both sides. Two stages of surgery were performed using an anterior and posterior approach.**

**Imaging findings**

Magnetic resonance imaging (MRI) showed an epidural lesion with gadolinium enhancement and bilateral extension into the intervertebral neural foramina (C5–C7), with spreading on the right side of the tumor into paravertebral tissue. MRI also showed vertebral body infiltration and edema at C5–C7 level. The intervertebral disks were intact at all levels (Fig. 1). A preoperative CT scan showed evidence of bone destruction at C5–C7 and CT angiography showed vertebral artery displacement on the right side (Fig. 2). Preoperative clinical and imaging examinations (including abdominal CT scan, thorax CT scan, and bone scan) showed no evidence of tumors in other organs and confirmed the primary origin of the tumor.

**Surgical intervention and histopathology**

In the first step, the patient underwent incisional biopsy using an anterior approach. Histopathological examination showed a small, round cell malignancy consistent with myxoid sarcoma (Fig. 3). Immunohistochemistry indicated a positive reaction for CD34 (particularly in vessel networks), whereas other markers for muscles and epithelial differentiation, as well as S100 protein, were absent.

CD45 was rarely positive. The Ki67/MIB1 proliferation index was 6%. The histopathological diagnosis was myxoid liposarcoma (MLS). Biopsy tissue was sent for cytogenetic analyses, which subsequently confirmed the t(5,6) (q13;p11) translocation, typical of myxoid tumors.
postoperative physical examination, bone scan, and chest and abdominopelvic CT scans showed no metastasis. The patient was prepared for the next surgery in two stages. During the first stage, she underwent C6 and C7 vertebrectomy, discetomy of C5/6, C6/7, and C7/T1 with tumor resection, as well as paravertebral and intraspinal tumor and bilateral neurolysis of the nerve roots C5, C6, C7 and C8 via the anterior approach. The removed tumor was yellow and avascular in color (Fig. 4). Anterior reconstruction was performed using an expandable cage (Fig. 5, left).

The second stage of surgery was performed using a posterior approach 4 days after resection of the tumor from the neural foramina C5 and C5 neurolysis, as well as fusion surgery from C5 to T1 (Fig. 5, left).

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<tr>
<th>Author</th>
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<td>Hamlat et al. [9]</td>
<td>Primary liposarcoma of the thoracic spine</td>
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<td>Lamjati et al. [7]</td>
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To our knowledge our case is the first case of primary cervical liposarcoma.

Fig. 1. T2 and T1-weighted magnetic resonance images after gadolinium infusion show an epidural mass at C5–C7 with pre and paravertebral involvement and compression effect on cervical cord. (Upper left: sagittal T2, upper right: sagittal T1 with contrast, lower left: coronal T1 with contrast, lower rights: axial T2).
right). Postoperatively, cervicobrachialgia was improved, but other neurological symptoms, including motor weakness, were similar to the preoperative state without any new neurological defects. Because of extensive tumor invasion, complete tumor resection was not possible.

One month after surgery, she was referred to a radiation oncologist. The patient underwent palliative radiotherapy. Six months post-radiation, MRI control revealed no demonstrable residual tumor (Fig. 6).

**Technical note**

During the transpedicular screw placement at C3–C7, it would be very difficult in most cases because of the midline incision limitation to achieve the correct angle shown by the navigation system. We suggest a very small incision for each screw, approximately 2–3 cm, beside the midline incision to perform transmuscular screw placement. With such an incision, it would be easier to achieve a proper angle for transpedicular screw placement (Fig. 7). This method has been described for lumbar transpedicular screw fixation. However, in transpedicular cervical fusion surgery, it has not yet been described in the literature.

**Discussion**

Liposarcoma is the most common soft tissue tumor and accounts for approximately 20% of all mesenchymal malignancies [1,7,8]. Liposarcoma usually originates from primitive mesenchymal cells, rather than mature adipose tissue [8–10]. The peak incidence of liposarcoma occurs between 40 and 60 years of age, and commonly has an indolent course [1,11]. The diagnostic criteria have been well established and five histological types are described: well-differentiated, dedifferentiated, myxoid, round cell, and pleomorphic [1,7,10].

Pleomorphic liposarcoma is a high-grade tumor with the highest prevalence between the ages of 60 and 70 years, and it affects both sexes equally. It is the rarest form of liposarcoma [9,10,12]. Myxoid liposarcoma is the second most common subtype, occurring more frequently in younger patients during the fourth and fifth decades of life [1,4,11]. Myxoid liposarcomas are usually located on the extremities, particularly the thighs. In contrast to other types of liposarcomas that generally metastasize to the lungs, myxoid liposarcomas have a tendency to metastasize to extrapulmonary sites, such as soft tissue, retroperitoneum, mediastinum, chest wall, peritoneal surface, and heart, which may be due to the abundance of fat tissue at these sites [1–4]. Skeletal metastasis has recently been reported as the most common site of metastasis in myxoid liposarcoma [4,11].
Myxoid liposarcoma has few or no mitotic figures and is characterized by proliferating small signet-ring lipoblasts in different stages of differentiation, a prominent anastomosing capillary network, and a mucoid matrix rich in hyaluronidase-sensitive acid mucopolysaccharides [1,13]. There is a distinctive pattern of capillary-sized vasculature, described as a “chicken wire” or “crows’ feet” pattern [1].

Cytogenetically, myxoid liposarcoma is characterized by the reciprocal translocation t[5,6] (q13;p 11), which results in the FUS-DDIT3 fusion, and less commonly, the t(12;22)(q13;q12) translocation that results in the EWS-DDIT3 fusion [13].

Primary tumors of the spine are relatively infrequent lesions compared with metastatic disease, multiple myeloma, and lymphoma [5]. Their presentation is variable and insidious before the signs of spinal cord compression are detected, at which point the diagnosis becomes obvious. Therefore, MRI is one of the most valuable modalities for anatomic evaluation of spinal tumors [5,9].

Noble and colleagues [14] showed that negative bone scans do not rule out bone metastasis and MRI provides the most sensitive technique for the diagnosis of bone metastases in myxoid liposarcoma. In another study, Schwab et al. [15] revealed that bone scans and positron emission tomography (PET) scans lack sufficient sensitivity to detect spinal metastasis from MLS. Screening with whole-spine MRI may lead to the earlier detection of spinal metastasis.

Schwab et al. [16] demonstrated that FDG-PET may also lack the sensitivity needed to detect these lesions. They advocated total spine MRI for the screening of skeletal metastases. Schwab et al. [4] reported that more than half (56%) of the total metastatic sites in MLS represent skeletal metastases, 70% in the absence of pulmonary spread, and a high incidence of metastasis to the spine.

There are several case reports regarding primary spinal involvement of MLS in the literature (Table 1). Hamlat et al. reported a primary liposarcoma of the thoracic spine with an extension into the epidural space without meningeal involvement [9].

Cho et al. [1] reported intradural involvement of multicentric MLS. Lmejjati et al. [7] and Barra de Moraes et al. [12] and in two separate studies reported primary liposarcoma of the lumbar spine.

In another study, Ogose et al. [2] showed metastasis of MLS to the thoracic epidural space without bone involvement.

Our case represents a primary myxoid liposarcoma of the cervical spine due to the histological features consistent with this diagnosis. The lesion developed primarily within the vertebrae. The preoperative and postoperative examinations produced no evidence that the tumor was a metastatic presentation from an original tumor elsewhere. The cervical spine is an unusual location, even for metastasis [6,17]. To our knowledge, this is the first case of primary myxoid liposarcoma of the cervical spine reported in the literature.

Schwartz and co-workers [18] defined criteria for differentiating true primary liposarcoma of bone from metastasis and involvement from surrounding tissue as follows: “the tumor should be a histological proved liposarcoma which arose within bone, and metastasis has been ruled out”. In addition, when bone invasion occurs from soft tissue liposarcoma, deep penetration is unusual. Most metastases are predominantly located on one side of the midline with frequent pre-vertebral soft tissue extension [19].

Complete resection is the treatment of choice. Because prognosis seems to be poor, chemotherapy and radiotherapy are indicated both in the neoadjuvant or adjuvant settings [20]. Therapeutically, surgical resection and radiotherapy are usually sufficient for local control [1].
Treatment with chemotherapy alone was significantly associated with metastasis [11]. Because MLS has been regarded as a radiosensitive tumor, early detection and radiotherapy with or without surgery can hopefully improve patient quality of life and survival [2].

The specific “crows’ feet” vascular pattern is one of the most striking features of MLS compared to other sarcoma classes, and is pathognomonic of these subclasses. Vreeze et al. [21] suggested preoperative radiotherapy, arguing that its effect the specific vascularization of MLS, explains the radiosensitivity.

Our patient had a destructive bone lesion in the cervical spine. Histopathology of the lesion showed myxoid liposarcoma. Common sites for this type of liposarcoma were surveyed and no other lesions were found. Therefore, the cervical spine lesion was non-metastatic.

The most common site for the origin of liposarcoma is primitive mesenchymal cells, rather than mature adipose tissue. However, in our case, we propose that it originated from mature epidural adipose tissue.

References