Case Report

Primary Spinal Epidural Extraosseous Ewing's Sarcoma with Brachial Plexus Infiltration

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Abstract

The Ewing's sarcoma family of tumors are aggressive malignant small round blue cell tumors of undifferentiated mesenchymal origin. Skeletal Ewing's sarcoma is a common entity that classically involves the diaphysis of the long bones, pelvis, ribs, and sacrum. Extraosseous Ewing's sarcoma (EES) is rare, most commonly presenting as a paravertebral mass lesion. Its manifestation as an anterior epidural mass lesion with extension along brachial plexus is an even rarer phenomenon. A 25-year-old male presented with neck stiffness and progressive weakness of the bilateral upper and lower limbs. Magnetic resonance imaging of the cervical spine revealed an anterior epidural mass lesion compressing the cervical cord and extending along the right brachial plexus, suggesting imaging differentials of EES and lymphoma. The patient underwent laminectomy with gross tumor resection, and histopathology confirmed a diagnosis of EES. EES should be kept in the differential diagnosis of anterior epidural mass lesions in young adults, specifically when the lesion shows extension along multiple neural foramina and nerve plexus.

Keywords: *Ewing's sarcoma family of tumors, Ewing's sarcoma, extraosseous Ewing's sarcoma, peripheral primitive neuroectodermal tumor*

Introduction

The Ewing's sarcoma family of tumors are aggressive tumors comprising osseous Ewing's sarcoma, extraosseous Ewings sarcoma (EES), peripheral primitive neuroectodermal tumor, and Askin tumor.[1] EES is relatively rare constituting approximately 15% compared to osseous Ewing's sarcoma. The most common location for EES is paravertebral soft tissue mass, which can extend to spinal epidural space; however, literature about involvement and extension along the brachial plexus is scarce.[2] Given the rarity of EES and nonspecific imaging features, the diagnosis on imaging alone can be challenging. A final diagnosis of EES is made on histopathological evaluation; however, a high index of suspicion on imaging helps accelerate the diagnostic and metastatic workup as well as planning of further management.

Case Report

A 25-year-old male presented to the emergency facility with complaints of progressive quadriparesis of 5-day duration, with more significant involvement of the bilateral upper limbs. There were no antecedent history of trauma or fever and no bowel/bladder symptoms at presentation. He had complaints of vague neck pain and stiffness for 2-month duration, which used to get relieved on taking over-the-counter analgesics. On examination, the motor power was 1/5 in the bilateral upper limbs and 3/5 in the bilateral lower limbs. The pain and temperature sensation were impaired below the C2 level.

Magnetic resonance imaging (MRI) of the whole spine revealed a relatively well-defined heterogeneous mass in the anterior cervical epidural space extending from C2 to C7 vertebral levels. It caused posterior displacement and compression of the cervical spinal cord [Figure 1a-c]. The mass was noted extending along the widened right C2–3 to C6–7 neural exit foramina infiltrating the root and trunk of ipsilateral brachial plexus. It infiltrated the right-sided vertebral foramina extending from C2 to C5 level encasing the V2 segment of the right vertebral artery without any luminal narrowing [Figure 1d]. Subtle marrow edema of the posterior vertebral

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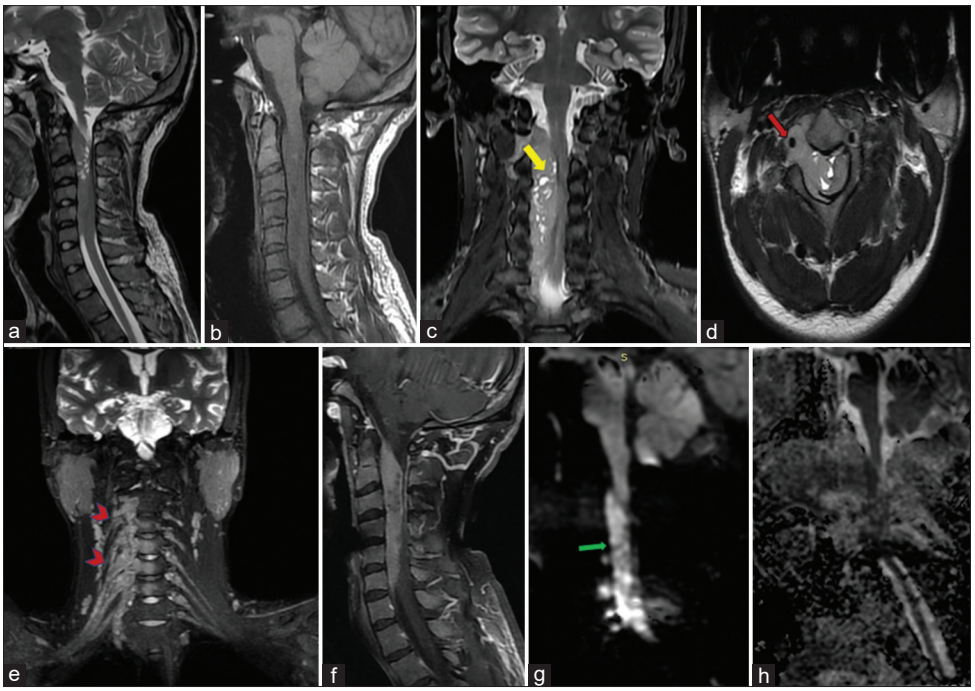

Figure 1: Sag T2 (a), T1 (b), Cor STIR (c), and axial T2 images (d) show T2 hyperintense, T1 isointense mass in the rt anterior epidural space, displacing cord to the left side with internal cystic areas (c, yellow A). Widening of rt C2-3 neural foramen and encasement of right vertebral artery (red A in d). The three-dimensional Cor STIR MIP image (e) showing the infiltration of rt brachial plexus roots and trunk (red ahead). Postcontrast Sag T1 fat saturated image (f) showing moderate enhancement with nonenhancing cystic areas. Note restricted diffusion (green A) on Sag diffusion-weighted images (g), apparent diffusion coefficient map hypointensity (h). Sag-Sagittal, Cor-Coronal, A-Arrow, rt-right

bodies was noted involving C3–C6 vertebral levels [Figure 1e].

The lesion displayed a slight hyperintense signal on T2-weighted and isointense signal on T1-weighted images, with few tiny cystic spaces within it. Sagittal diffusion-weighted images (DWIs) revealed restricted diffusion with low apparent diffusion coefficient values [Figure 1g,h]. Postcontrast images revealed moderate enhancement with few nonenhancing cystic areas [Figure 1f]. No areas of hemorrhage/calcification were seen on gradient recalled echo images. Computed tomography (CT) of the cervical spine was done to look for any periosteal reaction or bony involvement. It revealed the subtle erosion of the C4–C6 vertebral bodies posteriorly. No focal areas of calcification were noted. In view of the imaging findings, the diagnosis of primitive neuroendocrine tumor/EES was made with a differential of lymphoma because of extension along the right brachial plexus. Contrast-enhanced CT of the chest and abdomen was done which did not reveal any metastasis.

The patient underwent surgical decompression with laminectomy of C3–C7 vertebrae and gross total resection of the tumor. Intraoperatively, the tumor was ill-defined soft, suckable, grayish, completely extradural with moderate vascularity being seen pushing and compressing the cervical cord to the left. The cervical cord was nonpulsatile, and right-sided cervical neural exit foramina were widened. There was no obvious bony involvement by the tumor. After tumor decompression, the cord became lax and pulsatile. Intraoperative neuromonitoring showed some improvement in motor evoked potentials in the bilateral lower limbs after tumor decompression. Intraoperative frozen section was suggestive of a high-grade round cell tumor. Postsurgery, the patient continued to have a gradual improvement in motor power in all four limbs. At discharge, the patient had the power of 4/5 in the bilateral lower limbs, 4/5 in right upper limb, and 3/5 in the left upper limb.

Histopathology showed a small round cell tumor composed of monomorphic to mildly pleomorphic round cells, with scant cytoplasm, round hyperchromatic nuclei, and inconspicuous nucleoli [Figure 2a and b]. The tumor was positive for vimentin, CD99 [Figure 2c and d], and Bcl-2 and was negative for leukocyte common antigen, chromogranin [Figure 2e and f], and synaptophysin. Additional markers cytokeratin and myogenin were negative, which ruled out an epithelial and skeletal muscle differentiation. Based on these findings, a diagnosis of EES was made.

Discussion

Extraosseous tumors resembling Ewing's sarcoma were first described in 1969 by Tefft.[3] EESs are rare in comparison with Ewing's sarcoma of the bone, usually reported in the second and third decades of life with equal frequency in both males and females. EES can occur virtually anywhere

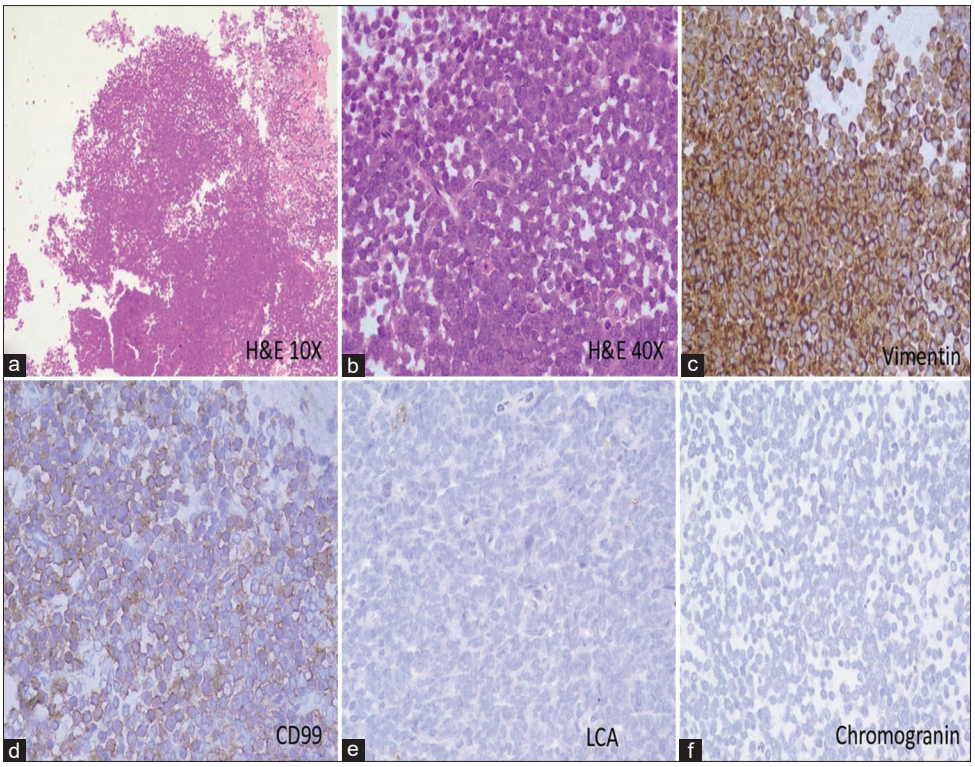

Figure 2: (a) Image (H and E, ×10) showing a low power view of a tumor appearing very cellular and "blue." (b) Image (H and E, ×40) showing a high power view of the tumor, which is composed of monomorphic to mildly pleomorphic round cells with scant cytoplasm, round hyperchromatic nuclei, and inconspicuous nucleoli. The tumor cells are immunopositive for vimentin (c), show membranous positivity for CD99 (d), and are immunonegative for leukocyte common antigen and chromogranin (e and f), respectively

in the body, with most frequent sites of occurrence being the paravertebral region, chest wall, lower extremities, trunk, and pelvis.[4]

EES primarily occurring in the anterior spinal epidural space is a rare occurrence with only a few cases reports in the literature,[5] and extension along the brachial plexus nerve root is even rarer. However, it is more common than the primary intradural EES. In a review of patients with extradural spinal EES/peripheral primitive neuroectodermal tumor, the lumbar region was the most common site followed by thoracic and cervical spine, with sacrum being the least affected site.[6] The tumor generally tends to spread locally, infiltrating deep fascial spaces, muscles, or skeletal structures. The involvement of brachial plexus in our case might be secondary to the spread of the tumor along with the nerve roots.

EES presenting as the primary intradural lesion is extremely rare, and <50 cases have been reported in the literature.[7] They have a male preponderance with a predilection to involve the lumbosacral region followed by cervical and dorsal regions. The extradural and intradural EESs have similar clinical manifestations and prognosis.[7]

The clinical presentation depends on the varying degree of cord compression and the level involved, with the most common presenting symptoms being radicular pain, paresis, and sensory disturbances with or without bowel/bladder symptoms.[8] The median diagnostic delay in the literature was 3 months and is explained by the nonspecific symptoms at onset.

MRI is the modality of choice for imaging in EES. These tumors are reported to display isointense to hyperintense signal on T1 images and hyperintense on T2 images with smaller tumors appearing homogenous and larger tumor appearing heterogeneous secondary to internal areas of hemorrhage and necrosis. Some studies have shown the presence of flow void within the lesion at MRI, which may aid the diagnosis. Calcification is an atypical feature.[9] Lymphadenopathy is not a commonly reported feature. Our case demonstrated a slightly hyperintense T2 signal of the tumor with internal cystic areas and homogenous enhancement, similar to previous reports. The restricted diffusion of the tumor signifies increased cellularity.

Imaging differential diagnosis of EES involving the epidural space included benign and malignant nerve sheath tumors, lymphoma, leukemia, histiocytic diseases, hemangioma, and epidural meningioma. Because of extension along multiple neural foramina, encasement of the V2 segment of the right vertebral artery, infiltration of brachial plexus, and STIR hyperintensity of the adjacent vertebral body, the benign entities were excluded from the differential diagnosis. Lymphoma, EES, and nerve sheath tumors have the propensity to extend along the

neural foramina and brachial plexus. Lymphoma involving epidural space originates from the body of vertebra or paravertebral lymph node and then extends to epidural space. It shows a homogenous hyperintense signal on T2 with restricted diffusion and homogenous enhancement; however, the presence of cystic internal spaces is not described. The primary spinal intradural EES may also extend along the nerve roots and be indistinguishable from nerve sheath tumors.[9] These intradural tumors can also mimic other intraspinal tumors such as meningioma and ependymoma.

Surgical intervention is considered the primary approach in the management of these cases, particularly to relieve cord compression symptoms, as well as for cytoreduction. Inadequate tumor margin or residual tumor is associated with increased tumor recurrence.[10] EES is radiosensitive, but definitive radiotherapy is indicated when only an intralesional resection is possible.[11] Neoadjuvant and adjuvant chemotherapies produce comparable results in patients with localized disease.

EES is aggressive with a high predilection for local recurrence as well as distant metastasis. The prognosis of EES is more favorable compared to the osseous counterpart. Patients with localized disease have an estimated 5-year survival rate of about 70%, however, with a relapse rate of 30%. Patients with metastatic or recurrent disease have a poorer prognosis.[12]

Conclusion

Although primary spinal epidural EES is rare, it should be considered in the differential diagnosis of spinal epidural masses with features, such as extension along multiple neural exit foramina, infiltration along nerves, and encasement of vessels. DWI can be of help in determining the high cellularity of the lesions even in the spinal cord lesions. Gross tumor resection followed by radiotherapy and chemotherapy can alter the disease course and improve the 5-year survival rate.

Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

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