CASE REPORT

Spinal epidural extraskeletal Ewing sarcoma

Abstract A rare case of extraskeletal Ewing sarcoma, arising primarily in the spinal epidural space is reported. An 18-year-old male presented with a 2-month history of right shoulder pain progressing to complete paraplegia and urinary retention over the course of 2 days. Magnetic resonance imaging demonstrated an extradural mass extending from the C6 to T1 level. Histopathologic examination confirmed the diagnosis. The literature is reviewed and radiological differential diagnosis of this rare neoplasm is briefly discussed.

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MOTS CLÉS Sarcome d’Ewing ; Rachis ; Tumeur épidurale ; Imagerie de résonance magnétique

Introduction

Ewing sarcoma of bone is the most frequent malignant bone tumor in the first 10 years of life and the second after osteosarcoma in the second decade of life. It was first described in 1921 by James Ewing [7]. Although the most common place for Ewing sarcoma is the bone, it may also arise from soft tissues. Ewing sarcomas arising from soft tissues are referred as extraskeletal Ewing sarcoma (EES). Since it was first described by Tefft et al. [31] in 1969, a number of cases have been reported to arise in various locations such as small intestine, vagina, kidney, skin, larynx, esophagus, paravertebral region, or epidural space as well [6,8,17,22,26,32]. However, we encountered only 31 cases in which EES arose primarily from the epidural space of the spine during the review of the literature. We present the magnetic resonance (MR) imaging findings of EES located in the spinal epidural space. The literature on spinal epidural EES is reviewed and imaging variability of this rare neoplasm is discussed.

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Case report

An 18-year-old male presented with a 2-month history of right-sided-neck and right shoulder pain progressing to complete paraplegia and urinary retention over the course of 2 days. Laboratory studies including erythrocyte sedimentation rate and complete blood count were within normal limits. MR imaging revealed a large, right extradural mass extending from the C6 to T1 level and displacing the thecal sac to the left. The mass was hypo-intense to cord on T1-weighted images and hyperintense on T2-weighted images, and showed heterogeneous, moderate enhancement on contrast material enhanced T1-weighted images (Fig. 1A,B,C,D). Axial MR images revealed the extension of the mass to the right neural foramen at the C7-T1 level and neural foraminal widening was detected at this level (Fig. 1E). Scalloping was detected at the posterior aspect of the C7 vertebral body on sagittal images. No other bony changes were seen. A few foci of high signal representing myelomalacia were detected within the spinal cord on T2-weighted images.

The patient underwent complete resection of the mass with laminectomy between C5-T1. Microscopically, the tumor was made up of generally solid sheets and infiltrating cords. Tumor cells showed uniform small round nuclei containing fine chromatin, scanty eosinophilic cytoplasm, and indistinct cytoplasmic membrane (Fig. 2A,B). There was a periodic acid-Schiff (PAS)-positive and diastase negative staining within tumor cells. CD99 was expressed focally. Other immunohistochemical markers including keratin, chromogranin A, synaptophysin, GFAP, and LCA were negative.

The patient received chemotherapy with cyclophosphamide, vincristine and adriamycin. Present follow-up is 13 months and the patient is without clinical and radiological evidence of recurrent or metastatic disease.

Discussion

Involvement of the spinal epidural space is an unusual and rare presentation of EES. Besides the current case, only 31 cases of EES arising in the spinal epidural space have been reported in the literature (Table 1). The mean age was 19.1 years (range, 4-47 years) at presentation, with 50% of the patients being between 10 and 20 years old, in the 32 cases of spinal epidural EES including our case. Our patient’s presentation at an age of 18 years is just below the mean age. There was a predominance in occurrence in male patients (21/32, 65.6%). The male-female ratio is 1.5:1 in ESB, however this ratio was calculated at 1.9:1 in spinal epidural EES.
The lumbar epidural space has been the most commonly reported site in 41% of the cases followed by the thoracic epidural space. With the case herein presented, only 7 cases of cervical epidural EES (22% of patients) have been reported to date [27,13,19]. Rare cases of EES involving primarily the central nervous system have been reported in the literature. Pekala et al. reported two cases of pathologically proved intracranial EES and they concluded that as the treatment and prognosis were quite different from each other, the differentiation of EES and central primitive neuroectodermal tumor was crucial [21]. The most common clinical presentation of EES arising primarily in the spinal
epidural space is back pain with or without radicular pain in all patients, paresis of one or more limbs, sensory disturbances, and bladder or bowel dysfunction. All these symptoms are related to the compression of the spinal cord, nerve roots, and/or cauda equina as seen in the other spinal epidural disorders. The most important clinical differential diagnosis may be the intervertebral disc herniation [9,23]. The mean diagnostic delay is 5.7 months (range 4 days to 18 months) and this delay can be explained by the nonspecific symptoms at presentation [26]. Physicians can fail to diagnose the disorder at an early stage because of the rarity of this neoplasm.

The most useful imaging studies for diagnosis are computerized tomography (CT) and MR imaging. CT may reveal a well-defined hypodense mass displacing the thecal sac on nonenhanced scans. Calcification has not been reported in any case. On contrast-enhanced CT scans, the tumors may exhibit heterogeneous contrast enhancement. Central, nonenhancing areas representing necrosis within the tumor can also be seen. CT may also demonstrate bony changes. CT myelography has been previously used for diagnosis. It may demonstrate displacement of the thecal sac, and neural foraminal widening [13]. MR imaging is useful for determining the extent of the tumor because of its high soft tissue resolution. The tumor is usually hypo-isointense to the cord on T1-weighted images and hyperintense on T2-weighted images, as in the case herein presented. Mild to moderate and homogeneous or heterogeneous enhancement can be seen after the administration of contrast material.

Radiological differential diagnosis of spinal epidural EES is broad and includes a number of disorders such as hematoma, abscess, hemangioma, meningioma, lipoma, nerve sheath tumors, dermoid and epidermoid tumors, and various epidural metastases. Heterogenous contrast enhancement of the mass excluded the possibility of cystic lesions such as abscess and hematoma in our case. Also, the lack of fat and bony changes except the vertebral scalloping excluded the possibility of vertebral tumors, dermoid and epidermoid tumors, and lipoma. The lack of intradural component excluded the possibility of meningioma. In fact, benign epidural neoplasms were not considered in our case. Leukemia was not considered because of the normal white blood cell count. Finally, we established our radiological differential diagnosis as malignant nerve sheath tumor, malignant mesenchymal tumor and lymphoma on the basis of the MR imaging findings. Because of the rare occurrence, spinal epidural EES was not considered before the histopathologic examination.

Although clinical and radiological findings are important in early diagnosis of primary spinal epidural EES, the definitive diagnosis is made on the basis of histopathologic examinations. EES has been recognized as being histologically indistinguishable from Ewing sarcoma of the bone and other round cell tumors. The histopathologic diagnosis of the small round cell malignant tumors is based on immunohistochemical and ultrastructural analysis. The classical histopathologic findings are sheets, lobules, and occasional rosettes of round cells with irregular nuclei and sparse cytoplasm [13]. After diagnosis, a laminectomy with tumor resection should be done. However, complete resection is not possible in some tumors, which exhibit infiltration into the adjacent structures. A partial tumor resection should be done in such cases. The combination of surgery, chemotherapy, and radiotherapy offers the best chance for a long-term disease free survival [15].

In conclusion, primary spinal epidural EES is a rare neoplasm that is difficult to diagnose. Many imaging techniques are available, however MR imaging seems to be more effective in the overall assessment. Although rare, the possibility of EES should be kept in mind in assessing the spinal epidural lesions.

References


