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Case Report

Early recognition and diagnosis of Ewing sarcoma of the cervical spine

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ABSTRACT

Ewing sarcoma of the cervical spine is a rare diagnosis. Early recognition and treatment of this condition improves survival. Reported is a case of a 20-year-old male patient who presented with increasing neck pain. Computed tomography and magnetic resonance imaging showed a mass with a primary differential diagnosis of Ewing sarcoma. As a result of mass effect and spinal cord compression, immediate medical intervention was deemed necessary to prevent neurologic deficits.

Introduction

Ewing sarcoma accounts for one-quarter of all bone tumors in childhood. Its incidence is highest during the second decade of life [3,8]. Ewing sarcoma (ES) occurs most commonly in the pelvis and appendicular skeleton in particular the femur and less commonly in the spine. Vertebral involvement is uncommon and is reported to be less than 5% [3]. Among the tumors appearing in the vertebral column, cervical spine is the least involved. However, the disease involvement in the spine could present with nerve root or spinal cord compression with associated poor prognosis [8]. Thus, a high index of suspicion is required to ensure early diagnosis of the disease in children and young adults who present with back pain. Our case is unique because of the unusual involvement of the cervical spine in a young adult. This would increase awareness for the clinician about cervical spine involvement of the disease process, and the role of imaging studies for early detection and diagnosis.

Case report

A 20-year-old male patient was admitted with a 2-month history of progressively worsening neck pain. Initial cervical spine computed tomography, CT, showed an enhancing right anterior epidural mass (0.9×3.8 cm) associated with spinal canal narrowing and irregularity of the C-3 vertebral body and posterior element (Figs. 1 and 2). Subsequently, cervical spinal magnetic resonance imaging, MRI, performed revealed abnormal, hyperintense signals in the C-5 vertebral body posterior element, and epidural soft tissue component causing spinal cord compression and edema (Figs. 3 and 4). An interventional radiology image guided needle biopsy was then performed,

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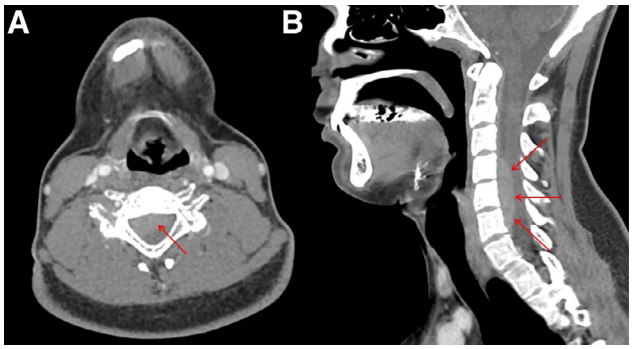

Fig. 1 – Axial and Sagittal CT scan in soft tissue window showing anterior epidural soft tissue mass lesion from C4-C6 level (red arrows) compressing on the spinal cord at this level. (Color version of figure is available online.)
CT, computed tomography.

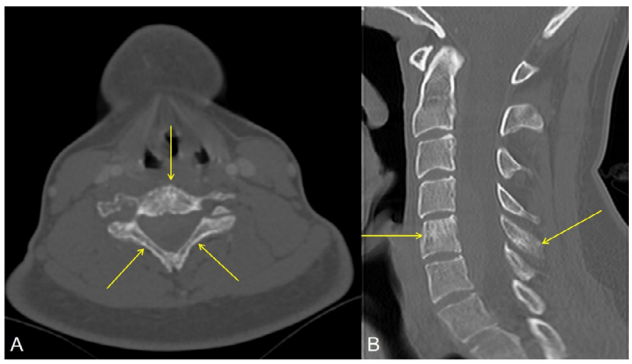

Fig. 2 – Axial and Sagittal CT scan in bone window showing permeative lytic lesion of the C5 vertebral body and posterior elements (yellow arrows) associated with the anterior epidural soft tissue mass at C4 -C6 level. (Color version of figure is available online.)
CT, computed tomography.

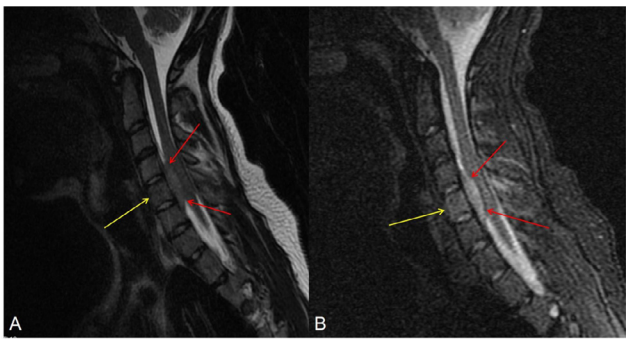

Fig. 3 – MRI Sagittal T2 and STIR images showing T2 hyperintense anterior epidural mass (red arrows) causing spinal cord compression at C4-C6 level. There is associated prominent edema in the involved C5 vertebral body and posterior elements (yellow arrows). (Color version of figure is available online.)
MRI, magnetic resonance imaging.

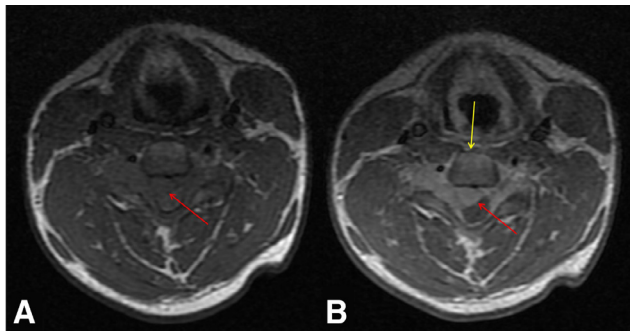

Fig. 4 – MRI Axial T1 pre- and postcontrast images showing T1 hypointense anterior epidural soft tissue mass (red arrow) on image A, with corresponding hyperenhancement of the mass (red arrow) and adjacent vertebral body (yellow arrow) on the postcontrast image B. (Color version of figure is available online.)
MRI, magnetic resonance imaging.

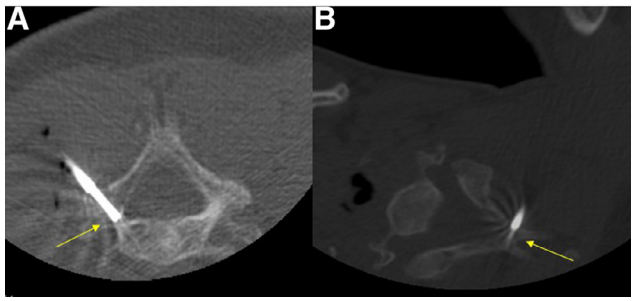

Fig. 5 – Axial CT scan showing a 12-gauge Elson bone biopsy needle advanced from the right paravertebral approach to enter into the right lateral osteolytic part of the lesion (yellow arrow in A) and 13 gauge biopsy needle into the spinous osteolytic part of the lesion in (yellow arrow in B). Three core specimens each were obtained. Aspiration specimen were also obtained using 18-gauge Franseen needle. (Color version of figure is available online.)
CT, computed tomography.

and small round blue cell neoplasm was a concern. Immunostains were positive for CD 99 FLI 1 and negative for synaptophysin and chromogranin. Morphology and immuno profile made it suspicious for ES. The fluorescence in situ hybridization assay performed was inconclusive due to inadequate cells. During the hospitalization course, the patient received 10 mg of IV dexamethasone every 8 hours. Nine days later, another biopsy was performed, also indicative of small round cell sarcoma. The patient was discharged while ES gene and fluorescence in situ hybridization results were still pending, however due to tumor location and new logical symptoms with tingling, ES treatment were commenced, which improved the tingling symptoms. Further CT scans of the abdomen and thorax revealed splenomegaly, but otherwise no metastatic disease. The patient had a bone scan undertaking that showed focal activity in mid cervical spine suspicious for bony involvement. Core bone biopsy using paravertebral approach along with fine needle aspiration of specimen was performed (Fig. 5).

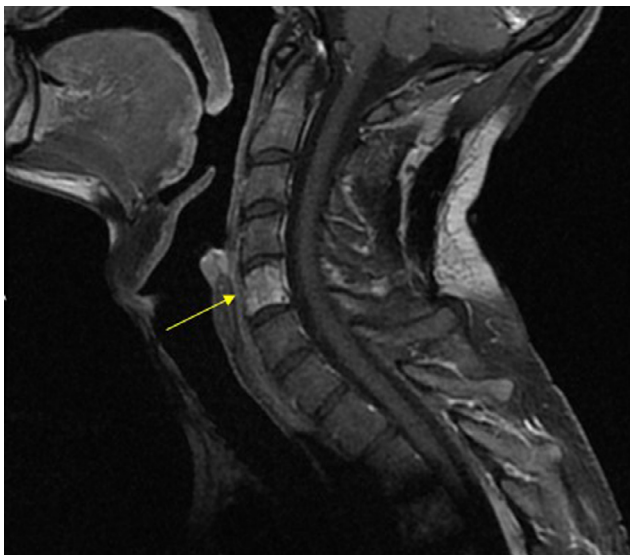

Fig. 6 – MRI of the cervical spine after 4 cycles of chemotherapy showing only residual enhancement of C5 vertebral body and posterior element (yellow arrow) and almost complete resolution of previously seen anterior epidural mass. (Color version of figure is available online.) CT, computed tomography; ES, Ewing sarcoma; ESFT, Ewing's sarcoma family tumor; FISH, fluorescence in situ hybridization; MRI, magnetic resonance imaging; SRBCT, small round blue cell tumor.

The EWSR1 gene break apart rearrangement studies that were performed were positive, supporting and confirming the final diagnosis of ES of the cervical spine arising from the C-5 vertebral body. Given that this is an isolated lesion with no other systemic disease, he was started on pediatric protocol AEWS 1221. The goal is to treat the patient with a total of 4-6 cycles total followed by repeat imaging and then possible resection, and if that is not feasible, then radiation treatment. Repeat imaging of the cervical spine after cycle 4 of chemotherapy showed almost complete resolution of his tumor (Fig. 6).

Discussion

First defined by Dr. James Ewing in 1921, ES (also known as Ewing's sarcoma) is a small round blue cell tumor which, after osteosarcoma, is the next most frequent occurring form of bone cancer in children and young adults [1–5]. ES is characterized by Ewing's sarcoma family tumor containing 1 of 2 chromosomal translocations, with over 90% having a t(11;22) (q24;q12) translocation and the balance expressing a t(21;12)(22;12) translocation [1,3–8]. Annual estimates of cases in the United States range from 200 to 500, and for children and adolescents being 4%-10% of all primary bone malignancies [2,3]. Early detection of ES is crucial as these tumors are aggressive, metastasize early, and tend to recur after resection, thereby presenting with an ominous prognosis upon diagnosis [2,3]. ES is slightly more prevalent in males than in females, with male to female ratios within the approximate range of 1.3-1.5:1 [3]. ES is more so found in younger individuals, most frequently adolescents ranging from ages 10 to 20 years old, of Caucasian background and less frequently reported in individuals of African and Asian ethnicity [4–6]. In the head and neck region, the mean age of occurrence is 10.9 years old [3]. ES is also more often observed occurring first in long bones and the pelvic girdle, with lower occurrences witnessed and clinically reported in the head and neck region, but it is capable of occurring in just about any bone [3,5,6,9]. Treatment for ES includes systemic multidrug chemotherapy (aiding with the suppression of micro metastasis and tumor reduction before surgery), surgery (eg, tumor resection), and radiotherapy (especially for unresectable primaries) [1,3].

The case reported is of particular interest as it illustrates a finding of ES in the cervical spine, a place where ES does not frequently develop. About a mere 3% of diagnosed ES is in the bones of the neck and head (excluding the jaw), with 0.9% of studied cases involving the nonsacral spine [5,9]. This is especially important because of the nature of ES and its characteristically negative prognosis. Our patient presented with worsening neck pain attributed to the compression on the anterior aspect of the spinal cord by the soft tissue component associated with the C5 vertebral body lesion. The location of this tumor also placed the patient at great risk for neurologic deficits if the compression were to continue uninhibited by surgical intervention. In addition, because of these risks associated with ES in this critical location, ES AEWS1221 protocol was followed before laboratory confirmation of the ES diagnosis. Dini et al makes an observation that is also noted in the case presented; ES of the spine presents in patients with local pain, a palpable mass, and neurologic deficit [8]. However, it is important to note that in the presented case, neurologic deficit was not an immediate symptom of the growth, but rather was a function of the growth with resultant compression on the spinal cord.

This case demonstrates the importance for early detection and diagnosis, with radiological imaging being an excellent primary resource for initial detection. Based on one study by Widhe et al of a group of patients who were later diagnosed with ES, only 19% of that sample population was suspected to have a bone tumor, with sciatica and tendonitis being common misdiagnoses in those cases [9]. In the case reported, initial CT scan of the cervical spine was the modality that revealed the ES mass, and it was recognized. In the aforementioned study by Widhe et al, 43% of the radiographic images were incorrectly deemed normal and the cancer was not identified [9]. This shows the importance of close scrutiny of such radiological images. The CT in the presented case also demonstrated significant spinal canal narrowing, and compression of the spinal cord. Demonstration of mass effect as in this case could be an initial indicator for ES [9]. Based on this presentation, this mass lesion could have been interpreted, not just as ES, but also with multiple differential diagnoses including, but not limited to, osteosarcoma, osteomyelitis, pseudo hemangioma, neuroblastoma, malignant lymphoma, aneurysmal bone cyst, and vertebra plana (Langerhans cell histiocytosis) as mentioned by Iacoangeli et al as well as Dini et al in their findings [1,6,8].

CT showed hyperintense signaling representative of the mass in the patient presented. The follow up MRI imaging also

added increased value to making the diagnosis, showing varying intensity in the affected area (hypointense on T1 imaging and hyperintense on T2 and T2 fat-sat images and showing enhancement on postcontrast images). Dini et al had similar success with using CT as a valuable tool for determining the outline, extent of the soft tissue component, and involvement of the vertebral bodies [8]. Thus, just as with Dini et al, MRI study was very useful with early detection of ES, with care taken to avoid MRI misinterpretations as in the case of Goktepe et al [8,9]. Thus, to avoid overlooking small or inconspicuous growths, high field MRI with great anatomic detail and contrast enhanced sequences are strongly recommended [10]. A definite and conclusive diagnosis can then be confirmed via prompt laboratory testing of suspicious cells extracted via interventional radiology image guided needle biopsy.

Summary

Early recognition of ES is essential, especially in critical areas in which it is not often seen such as the spine, due to the poor prognosis of the disease with high rates of malignant metastasis. Imaging studies such as CT and MRI, when carefully performed and examined, can prove to be invaluable tools for early recognition of ES to allow for laboratory confirmation and prompt treatment.

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