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Ewing's sarcoma of the mobile spine. Three unusual observations

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Abstract

Background. Ewing's sarcoma is a bony highly malignant tumour, it occurs most frequently in the second decade of life. Ewing's sarcoma is a rare affection, located usually in the pelvis, the femur, the humerus, the ribs, the mandible and clavicle, other location are rare especially in the spine. We report three cases of spinal Ewing's sarcoma, two primary spine locations and one on young adult with unusual clinical presentation.

Cases presentation

Case 1. The first patient is a girl of 14 years old without past medical history. She presented initially two months before consultation a neck pain followed days after by a weakness of the left upper limb; the spine imaging performed objectified a destructive process of C2 with a spinal cord compression. The patient was operated beneficiating of a spinal cord decompression and a subtotal removal of the tumour. The pathologist's results were in favour of Ewing's sarcoma and the patient was oriented to oncology.

Case 2. The second patient is a man of 31 years old operated five years before he consulted for shoulder Ewing's sarcoma followed by chemotherapy and radiotherapy, he presented two months before consultation a cauda equina syndrome. Spine MRI objectified a double location of an epidural tumour at T3-T4 and S1-S2 levels. The patient was operated beneficiating of subtotal removal of the tumour. The laboratory exam results were in favour of Ewing's sarcoma and the patient was oriented to oncology.

Case 3. The third patient is a 6 years old boy who presented a 1 month history of low back pain followed by a rapidly deteriorating weakness of both lower limbs over a weak. On examination there was bilateral spastic paraplegia, hypoesthesia below the level of Th10 and a urinary retention. The MRI imaging revealed a lesion on the levels Th8, Th9 and Th10 vertebras involving the body, pedicle, lamina, and the transverse process on the left side with an epidural invasion compressing the spinal cord. The tumour was radically removed. Pathology report was in favour of Ewing's sarcoma. Two weeks after surgery the patient was able to walk. He was referred for adjuvant systemic chemotherapy.

Conclusion. Ewing's sarcoma is rare malignant tumour. The location in the spine exposes the patient to more complications because of the neurostructures compression. The surgical total removal followed by radio and chemotherapy is the only option with the best prognostic and guaranties an acceptable life quality.

Keywords
Ewing's sarcoma,
spinal cord compression,
child malignancy

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Introduction

Ewing's sarcoma is a bony highly malignant tumour, it occurs most frequently in the second decade of life (1,3,4,5,7,8,10). Ewing's sarcoma is a rare affection (4), located usually in the pelvis, the femur, the humerus, the ribs, the mandible and clavicle (7), other location are rare especially in the spine(4,8). We report three cases of spinal Ewing's sarcoma two primary spine locations and one on young adult with unusual clinical presentation.

Case presentation

Case 1

The first patient is a girl of 14 years old without past medical history. She presented initially two months before she consults a neck pain followed days after by a weakness of the left upper limb; the spine CT and MRI performed objectified a destructive process of C2 with a spinal cord compression (Figure 1). The patient was operated beneficiating of a spinal cord decompression and a subtotal removal of the tumour, the lesion is considered to cause a spinal instability so an occiptocevical fixation was put. The pathologist's results were in favour of Ewing's sarcoma and the patient was oriented to oncology for adjuvant treatment.

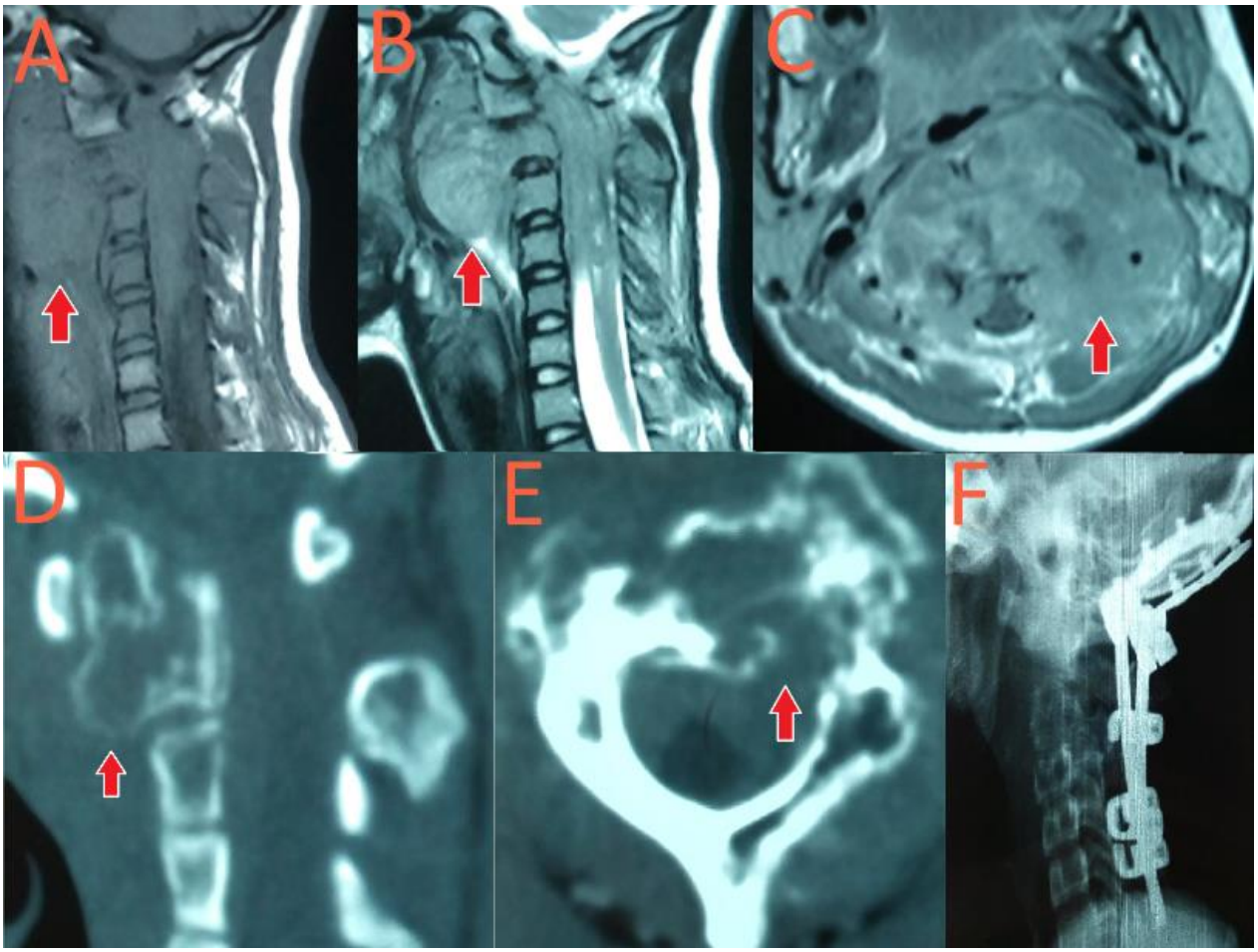

Figure 1 Patient 1 imaging. **A**: sagittal T1 WI sequence cervical MRI; **B**: sagittal T2 WI sequence; **C**: axial T2 WI sequence; **D**: sagittal bone window CT; **E**: axial bone window CT; **F**: post-operative x rays.

Case 2

The second patient is a man of 31 years old operated five years before for shoulder Ewing's sarcoma followed by chemotherapy and radiotherapy, he presented two months before consultation a cauda equina syndrome. Spine CT and MRI objectified a double location of an epidural tumour at T3-T4 and S1-S2 levels (Figure 2). The patient was operated beneficiating of subtotal removal of the both lesions through Th3 and Th4 then S1 and S2 lamenectomy. The laboratory exam results were in favor of Ewing's sarcoma and the patient was oriented to oncology.

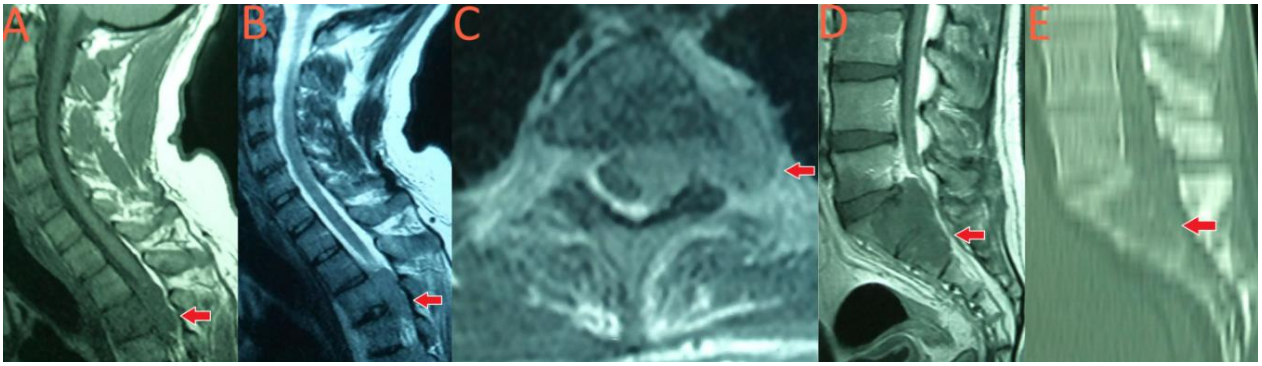

Figure 2. Patient 2 imaging. **A**: sagittal cervical T1 WI MRI; **B**: sagittal cervical T2 WI MRI; **C**: axial cervical T2 WI MRI; **D**: sagittal lumbosacral T2 WI MRI; **E**: sagittal lumbosacral bone window CT.

Case 3

The third patient is a 6 years old boy who presented a 1 month history of low back pain followed by a rapidly deteriorating weakness of both lower limbs over a weak. On examination there was bilateral spastic paraplegia, hypoesthesia below the level of Th10 and a urinary retention. The MRI imaging revealed a lesion on the levels Th8, Th9 and Th10 vertebras involving the body, pedicle, lamina, and the transverse process on the left side with an epidural invasion compressing the spinal cord (Figure 3). The tumour was radically removed. Pathology report was in favour of Ewing's sarcoma. Two weeks after surgery the patient was able to walk. He was referred for adjuvant systemic chemotherapy.

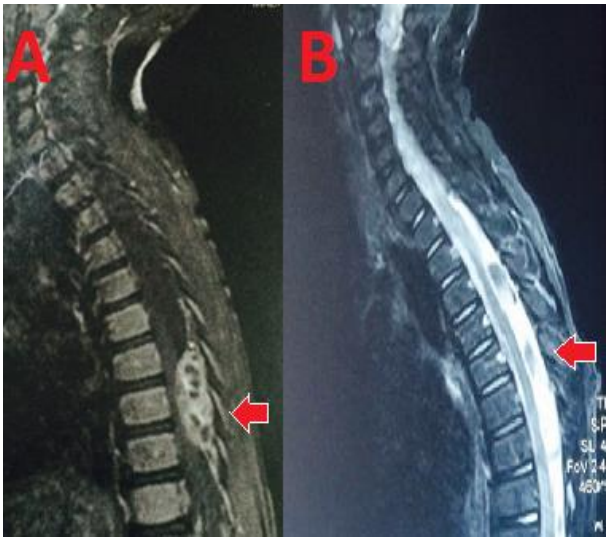

Figure 3. Patient 3 spine MRI. **A**: preoperative T1 injected sequence; **B**: postoperative T2 WI.

Discussion

Ewing Sarcoma (ES) was first described by James Ewing in 1921(2,6). It is a small round blue neoplastic cells (1,2,3,4,5,6,7,8,9) with clear to lightly eosinophilic cytoplasm, evenly dispersed chromatin, and indistinct nucleoli (3,7). ES is found in bones or in the surrounding soft tissues (1,3,4,5,6,7,8). ES is usually diagnosed in the second decade of life (1,3,4,5,7,8,10), the occurrence in adulthood is sporadic (1,6,3,8). Ewing's sarcoma present 4% of paediatric malignant tumours (4), It arises mostly in the long bones in 47% of cases, pelvis in 19%, and on ribs in 12% of cases (7), vertebral location is seen in less than 6% of cases (4,8), and mostly located in sacrococcygeal region (5,6,7), its presence on non-sacral spine is found in only 0.9% of all cases (7) with extremely rare involvement of the cervical spine (5,7). The primary location on the spine is unusual (4,6,7). Being a primary bone tumour, extra osseous locations are rare in a review between 1969 and 2015 only 119 cases of extra osseous ES was reported among them 76 cases of epidural ES (3). The clinic is summarized in the triad of local pain, palpable mass and neurologic deficit depending on the site of compression (1,7). Radiographic exams in spinal ES are not specific (3,6,8). Mostly it is a lytic lesion, sclerotic changes are rarely seen (7,8,9) especially in primary lesions(8). Spinal ES is located on posterior elements in 70% of cases and on the body in 30% of cases which could lead to vertebral collapse (8), extraossous location is limited to some reported cases (3,8). Plain radiography shows the lytic lesions tardively (7,9) usually after the neurological signs became obvious (9). CT is very helpful to assess the amount of destruction of the vertebral body and posterior element (7,9). Soft tissue lesions are better

outlined by MRI, ES have hypo or isointense signal on T1 weighted imaging and a hyperintense signal on T2 weighted imaging with heterogeneous enhancement after gadolinium injection (3,8). There is no standard management described for the spinal ES and no difference between child and adult treatment strategy (1,6). Usually it associates surgery chemotherapy and radiotherapy. The surgery assures biopsy, the local control of the tumour, spinal cord decompression and spinal stabilization. Because of the intimate relationship with neurological structures some surgeons prefer intralesional excision or debulking of the tumour in order to prevent long term morbidity (4) but many studies suggest that gross total resection is associated with better outcome (1,2,3,4,6,9). Tumours excised with 2 cm of normal tissue are considered to have safe surgical margins (2) otherwise radiotherapy is highly indicated (1,2,3,4,5,6,7,9). Radiation doses could not exceed 45 Gy (3,4,9). Radiotherapy could be used alone to assure the local control of the tumour (2,3,4,6), but with high risk of deterioration and less overall survival (2,3,4). When surgery and radiotherapy assure the local control of the tumour, chemotherapy is indicated to eradicate systemic micro metastasis (1,2,3,4,5,9). With patients who have no neurological deterioration and If the diagnosis is made before by needle biopsy a neoadjuvant chemotherapy could be proposed in order to shrink the tumour which could facilitate its removal (1,2,6,7,9). The classical chemotherapy protocol of ES is VACA (vincristine, actinomycin, cyclophosphamide and doxorubicin) (3,9) many other drugs have been added like ifosfamide and or etoposide (VAC/IE) which improved the outcome (1,2,3). Prognosis is poor in the spinal ES compared to other locations (1,4), other factors are related to a bad prognosis like the presence of metastasis (1,5,7) the adult age (3,7) and extra osseous location (3). In the other hand tumours located in the sacrum are associated with a poor prognosis due to the late stage diagnosis (4) and those located in cervical spine due to the difficulties of the total resection (7). The local control of the tumour with en bloc resection is associated with the best survival outcome (1,3,4,6,9). Overall median survival in spinal ES is 26 months, the survival rates for 5-year is at 41% and for 10-year 34% (4,10).

Conclusion

Ewing's sarcoma is rare malignant tumor. The location in the spine exposes the patient to more complications because of the neurostructures compression. The surgical total removal followed by radio and chemotherapy is the only option with the best prognostic and guaranties an acceptable life quality.

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