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CASE REPORT

Primary Ewing Sarcoma of the Body of Cervical Spine

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ABSTRACT

Primitive neuroectodermal tumors (PNETs) are malignant neoplasm originating from neural crest cells that generally affects children and young adults (4–15 years). Ewing sarcoma family of tumors occurs most commonly in the long bones of the extremity and less commonly in the spine. We present a case of a 21 years old male with acute onset neck and left upper extremity pain which rapidly progressed to spastic paraplegia without bladder bowel involvement. He underwent a C7 corpectomy and titanium cage fixation. Immunohistochemistry clinched the diagnosis of PNET/ Ewing sarcoma. Our case was unique in the primary involvement of the body of cervical vertebra in a young adult which was very similar to Kochs spine.

Keywords: Cervical spine, Ewing sarcoma, Primitive neuroectodermal tumors.

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INTRODUCTION

Primitive neuroectodermal tumors (PNETs) are malignant neoplasm originating from neural crest cells that generally affects children and young adults (4–15 years).[1,2] When present outside the central nervous system, it is called peripheral PNET and is included in the Ewing sarcoma family of tumors.[2,3] Ewing sarcoma family of tumors occurs most commonly in the long bones of the extremity and less commonly in the spine.[4] Amongst the tumors appearing in the vertebral column, cervical spine is the least involved.[5] The involvement in cervical spine is most commonly of the posterior elements with occasional spread into the body.[5] Our case was unique in the primary involvement of the body of cervical vertebra in a young adult which was very similar to Kochs spine.

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CASE REPORT

A 21-year-old male presented to us with pain in the neck extending to the left upper limb of 15 days onset. He had stiffness of both lower limbs with difficulty in walking. There was history of hypertonic spasms involving both lower limbs. However, there were no bowel or bladder disturbances. He had hypoesthesia below the neck. Examination showed a power of grade 5/5 in all four limbs with hypertonia and exaggerated reflexes in both lower limbs.

Magnetic resonance imaging (MRI) showed a lytic lesion involving the body of C7 vertebra with extradural soft tissue compressing the spinal cord. The lesion was isointense on T1W and hypointense on T2W image. There was no evidence of cord signal intensity changes at that level (Fig. 1). Due to patient being symptomatic and radiological obliteration of anterior subarachnoid space at the level of lesion, a decision was taken to surgically decompress the cord by anterior approach followed by corpectomy and fusion.

On the day of surgery, the patient developed bilateral lower limb weakness and had grade zero power in both lower limbs. The surgery as planned, was performed after explaining the prognosis and preoperative neurological status to the relatives. With the patient in supine position, C7 corpectomy using anterior cervical approach was done. The cord was decompressed till the lateral limits. The cord was pulsatile at the end of the surgery. Fusion was performed with expandable titanium cage (Fig. 2).

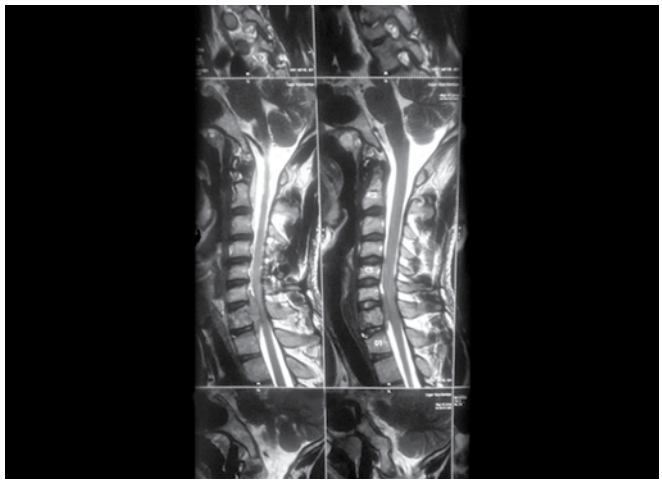

Fig. 1: Preoperative T2-weighted sagittal section of MRI cervical spine reveals lytic lesion in body of C7 vertebra with extradural soft tissue compressing the spinal cord. No evidence of cord signal intensity changes

Intraoperative steroids and citicoline were given. Patient was extubated on table but was not moving both lower limbs. Magnetic resonance imaging was done on postoperative day one showed signal intensity changes in the cervical cord, suggestive of myelomalacia (Fig. 3). He was given hyperbaric oxygen therapy (5 sittings), continued on steroids and given physiotherapy. He progressively improved in his lower limb power and was able to walk on postoperative day 8. A delayed MRI, performed after 2 months of surgery, showed resolution of myelomalacia.

Histopathology showed bony trabeculae with intervening sheets and islands of tumor cells which were small with high nucleocytoplasmic ratio and large vesicular nuclei suggestive of Non-Hodgkins lymphoma. Tissue immunohistochemistry showed malignant small round cell tumor with rosette formation. The tumor cells expressed Mic2, Fli1 and were negative for desmin and AE1/AE3 suggestive of primitive neuroectodermal tumor/Ewing sarcoma (Fig. 4).

Fig. 2: Intraoperative picture depicting the titanium cage placed after C7 corpectomy

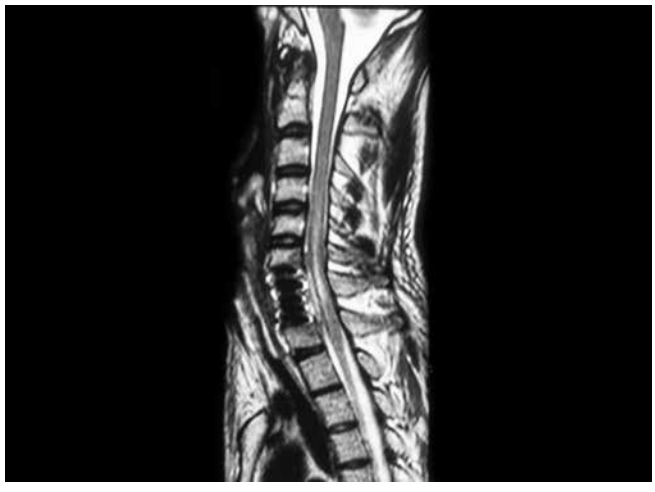

Fig. 3: First postoperative day T2-weighted sagittal section of MRI cervical spine reveals signal intensity changes in the cervical cord, suggestive of myelomalacia

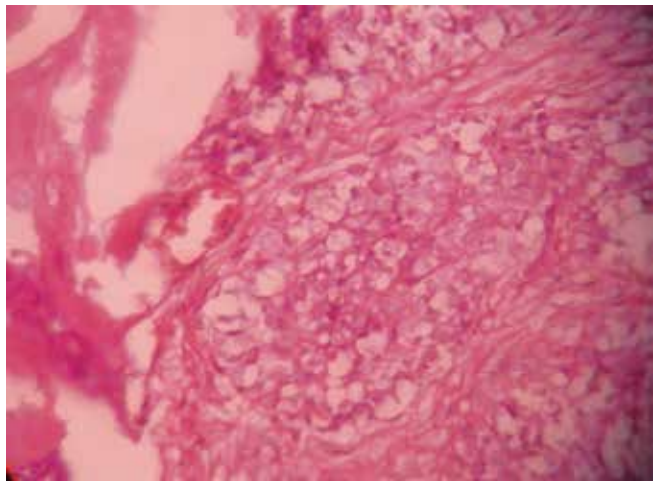

Fig. 4: Slide hematoxylin and eosin histological sections showing the tumor is composed of small round cells with a perivascular nest of cells in a rosette-like pattern. The cells had large vesicular nuclei with scanty cytoplasm (hematoxylin and eosin, 20×)

DISCUSSION

Ewing sarcoma or red marrow tumor, a highly malignant bone tumor, was first described by James Ewing in 1921. It is the second most common malignant tumor in young patients, commoner in males.[4] Annual incidence rate is less than 2 cases per 1,0000,000.[4] They are commonest in growing bones and occur most commonly in the age group of 4 to 15 years and rarely above the age of 30 years.[4]

Primitive neuroectodermal tumors are malignant neoplasm composed of small round cells, of neural crest origin and variable degree of differentiation.[6] Long bones, paravertebral region, chest wall and pelvis are commonest sites.[7] Primary involvement of the vertebral column is rare. In a series of tumors involving the vertebral column reported by Ilaslan et al, cervical spine is the least commonly affected (3.2%).[5] They most commonly affected the posterior columns and only one case in their series had sole involvement of the vertebral body.[5]

Local pain is the commonest first symptom.[4] Even though, there are no pathognomic symptoms or signs, the presentation depends on the affected site and degree of tumor invasion.[6] Systemic manifestations such as fever, anemia, etc. have been described. Diagnostic imaging modalities include: MRI and computed tomography (CT). However, there are no specific radiologic signs.[8] Cytological and histological analyses are not sufficient and have to be supplemented with immunohistochemistry for final diagnosis.[9]

Due to aggressive behavior of the neoplasm and its great potential to metastatize, treatment should be multimodal involving radical surgical excision, radiotherapy and chemotherapy.[2] We need to recognize the entity for early treatment and adjuvant therapy with any. Disease

free survival is approximately 45% at 7 years. Certain factory like tumor volume >100 cm^3, axial location, increased LDH levels, low serum albumin levels, metastasis, older age and neural differentiation have been associated with poorer outcomes.[10]

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