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Case Report

A Fatal Case of Primary Osteogenic Sarcoma of Cervical Spine: A Rare Entity

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ABSTRACT

Primary osteosarcoma of cervical spine is a rare condition with poor prognosis. The clinical, radiological as well as histopathological features of osteosarcoma vary widely, often leading to diagnostic difficulties. The report highlights the rarity of osteosarcoma of the cervical vertebra wherein the radiological and clinical findings suggested an infective pathology and histological findings remained the gold standard for final diagnosis of neoplastic etiology. A 21-year-old female presented with pain in the nape of the neck and tingling numbness in left upper & lower limb. On examination patient had quadriparesis. Radiological investigations were suggestive of infective etiology most likely Koch's causing involvement of C3-C5 vertebrae with prevertebral, paravertebral, parapharyngeal and intraspinal infective soft tissues abscess. She underwent C4 laminectomy with excision of the lesion and C1-C7 fixation with titanium Hartshil. The patient eventually died in the post-operative period. The intraoperative squash diagnosis was reported as a spindle cell tumor with malignant potential. The subsequent neuropathological diagnosis of C4 vertebral tumor on paraffin sections revealed characteristic features of osteogenic sarcoma (osteoblastic variant) showing osteoblastic giant cells and neoplastic growth of atypical spindle shaped cells arranged in sheets and trabeculae laying down mineralized osteoid and permeating the adjoining host bone. Early detection and accurate diagnosis is important for improving not only patient prognosis but also the quality of life. We should always consider this rare entity, particularly in young patients who present with intractable neck pain.

Introduction

Osteogenicsarcoma is one of the most common malignant tumors of bone arising frequently in extremities and only rarely involve spine. Primary vertebral osteogenicsarcoma accounts for only 0.85-3% of all osteogenicsarcoma and 3.6-14.5%⁽¹⁾ of all primary spinal tumors. Most osteogenic sarcoma arising denovo are located in

metaphysial area of long bones particularly lower end of femur, upper end of tibia and upper end of humerus. The mean age of occurrence is 38 years for axial osteogenicsarcoma than its appendicular counterpart which occurs predominantly between 10-25 years of age⁽²⁾. Osteogenicsarcoma is a high grade tumor with poor prognosis. Overall the prognosis of cervical osteogenicsarcoma is worse

Keywords: Primary osteogenicsarcoma, cervicalvertebra, young female, death.

when compared to osteogenicsarcoma at conventional sites. It is a locally aggressive tumor treated with chemotherapy, radiotherapy and surgical resection. Due to rarity of spinal osteogenicsarcomas we report a case of 21 yearsold female who presented with primary osteogenic sarcoma of cervical spine.

Case Summary

A 21 years old female presented with pain in the nape of neck and tingling numbness in left upper and lower limb since 7 months. On examination patient had quadriparesis. Radiological investigations were suggestive of infective etiology most likely Koch's causing involvement of C3-C5 vertebrae with pre vertebral, parapharyngeal and intraspinal soft tissue abscess. [Fig 1&2]. She underwent C4 laminectomy with excision of the lesion. The patient eventually died during postoperative period.



Fig 1: Xray cervical spine showing Osteolytic lesion of C4 vertebra causing cord compression

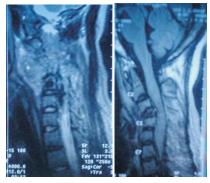
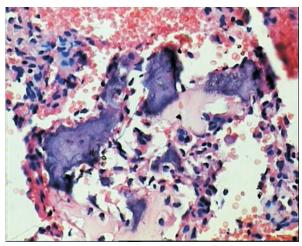


Fig 2 : MRI spine showing collapse of C4 vertebra and intraspinal extension

Pathological Findings

The intraoperative squash diagnosis was reported as spindle celltumor with malignant potential. The subsequent neuropathological examination of C4 vertebral tumor on paraffin sections revealed characteristic features of osteogenic sarcoma showing neoplastic growth of atypical spindle shaped cells arranged in sheets and trabeculae lying down mineralized osteoid and permeating the adjoining host bone. [Fig 3(a, b, c, d)]





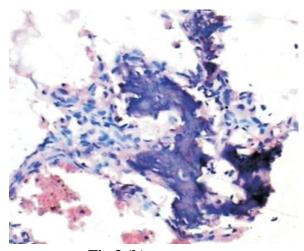


Fig 3 (b)

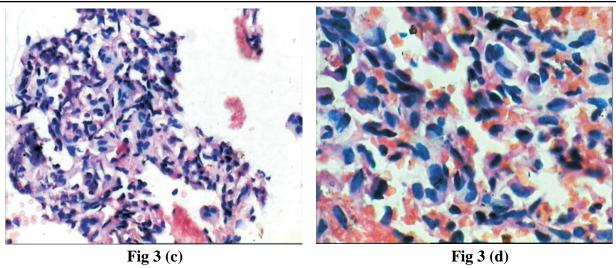


Fig 3 (a,b,c,d) Microphotograph of osteogenic sarcoma showing atypical spindle cells lying down mineralized osteoid and permeation of host bone

DISCUSSION

Primary spinal osteosarcomas are rare, mostly located in the sacral region followed by lumbar and thoracic segment of spine⁽³⁾. Cervical segment being the least common site needs high degree of suspicion for diagnosis. Clinically osteosarcoma almost always present with insidious onset neck pain which progressively become intractable and is associated with neurological deficit in more than two third of patients. Age distribution of primary vertebral osteosarcoma is bimodal. The mean age of occurrence is 4th decade for axial osteosarcoma which is two decade later than its appendicularcounterpart. A second smaller peak in incidence of the tumor occurs in patient older than 50 years (4). Patient presenting with non mechanical back pain along with neurological deterioration should be thoroughly evaluated for primary spinal malignancy. Clinical examinations and radiological investigations are complimentary but histopathological examination remains gold standard in the establishment of diagnosis. Though radiology plays an important role in establishment of diagnosis, however imaging studies alone may be misleading at times. Plain computed radiograph, tomography, magnetic resonance imaging, angiography and dynamic bone scintigraphy are used for diagnosis and evaluation of tumor extent. Plain radiograph and CT shows a mixture of osteoblastic and osteolyticareas. However purely osteolytic (moth

eaten appearance) and pure sclerotic osteosarcoma (ivory, dense cloudlike appearance)are also been reported. The differential diagnosis for a lytic expansile lesion on a cervical spine radiograph in thisage group include aneurysmal bone cyst, giant cell tumor, eosinophilic granuloma, osteoblastoma, fibrous dysplasia and osteosarcoma⁽⁵⁾. So conclusive diagnosis is based on histopathology. Tumor commonly involve spinal canal as a soft tissue mass which makes cross sectional imaging like CT and MRI essential. In many parts of developing world tuberculosis is the most common cause of vertebral body infection with majority of cases aged around 20 years, and it is difficult to differentiate atypical tuberculous lesion of vertebra from neoplastic process radiographically and clinically (as in our case). Osteogenicsarcoma represents a heterogeneous group of tumor with different histologic features. Conventional osteogenicsarcomas are intramedullary and high grade besides this another group of low grade osteogenicsarcomas consisting of low grade intramedullary and various forms of surface osteogenicsarcoma. (6) According to World health organization bone osteosarcoma is curreclassified as follows; conventional, telengectesic, small cell, low grade central, secondary, parosteal, periosteal, high grade surface⁽⁷⁾. Majority of tumor shows osteoid matrix mineralization and few shows lytic lesion. Conventional osteosarcoma is the classic form of

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osteosarcoma and traditionally according to the predominant type of extracellular matrix produced it is further subdivided in to osteoblastic, chondroblastic and fibroblastic subtypes⁽³⁾ The tumor cells are often highly anaplastic with pleomorphic and hyperchromatic nuclei and are spindle shaped. Poor prognosis is due to location and proximity of vital structures.

Conclusion

Early detection and accurate diagnosis is important for improving not only patient prognosis but also quality of life. This rare entity should be considered particularly in young patients who present with intractable neck pain.

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