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An undiagnosed chronic back pain may come out with severe morbidity in young patient, needs to be evaluated adequately: A case report and review of literature

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Abstract
Introduction: there are various possible etiology for chronic back pain and malignancy is one of them. Extraskeletal Ewing's Sarcoma usually involves sacrum while dorsolumbar junction involvement is uncommon. We are reporting a case of Extraskeletal Ewing's Sarcoma with rare presentation related to the age, site of lesion and unusual extension.
Case presentation: 35 year old male presented to casualty with urinary retention and paraplegia of sudden onset with history of chronic back pain of 6 months. We investigated him and diagnosed extra skeletal Ewing's Sarcoma of paravertebral origin along D12-L1 spine with intaspinal intadural extension. Patient was given palliative chemo-radiotherapy.
Conclusion: Chronic back pain of unknown etiology should be evaluated properly and while making diagnosis possible malignant etiology should be kept in mind even in young patient to prevent undue morbidity and mortality.

Keywords: chronic back pain, extraskeletal ewing's sarcoma, ewing's sarcoma

Introduction
Back pain is a common reason for patients to seek medical advice. Outpatient department (OPD) has plenty of cases with back pain and due to common belief of their benign etiology these are not often taken seriously. Back pain may be due to muscular cramps, axial spine pain, disc degeneration, fracture of vertebra and tumors [1]. Most of the symptoms are related to benign pathology but malignancy is of great concern for morbidity and mortality. Making early diagnosis is also necessary to detect hidden malignant pathological process. Consultation of quacks and bone setters by this group of patients may make the picture worse resulting delayed presentation with severe complications. Here we are presenting a case of undiagnosed neglected case of back pain that landed into complete paraplegia with bladder and bowel involvement. Later when diagnosis was made it came out to be extra skeletal Ewing's sarcoma of paravertebral origin which is a rare finding. Extra-skeletal Ewing's sarcoma (EES) involving the central nervous system is not a common entity. EES is a rare soft tissue neoplasm histologically similar to skeletal Ewing's sarcoma found predominantly in 10 to 30 years age group with aggressive course. Involvement of epidural space by these EES is more uncommon [2].

Case Report
35 year old male presented to casualty with complain of urinary retention and sudden onset of weakness in bilateral lower limbs for which uretheral catheterization was done. On examination both lower limbs were flaccid and deep tendon reflexes were absent bilaterally. There were silent plantar reflexes, while power was 0/5 for hip flexors, knee extensors and ankle dorsiflexors. Sensory loss was up to mid-thigh region which progressed gradually upto level of inguinal ligament (groins).

There was no sensory and motor deficit on upper limb and higher mental function including all cranial nerves were normal. Patient had history of dull back pain located on left paravertebral region since 6 months which was insidious in onset and progressed gradually. Initially there was mild pain on nights which relieved after analgesics; later on it became constant and severe in intensity. There was radiation of pain to left flank and abdomen. Pain was not associated with any swelling. Patient consulted a bonesetter for that he has given massage and assurance.There has history of weight loss and loss of appetite but no history of evening rise of fever or chronic cough was there and there were no close contacts of tuberculosis in family.

On x ray and ultrasound pleural effusion was found, and 300 ml of hemorrhagic fluid was drain. On contrast enhanced computed tomography (CECT) of chest a poorly marginated heterogeneously enhancing soft tissue mass lesion in left paravertebral region at D12-L1 level was there measuring approximately 7.7/7.0/6.6 cms in dimensions. Medially mass was extending into left neural foramina from D12- L1 level. FNAC revealed atypical round cell with scant to moderate vacuolated cytoplasm suggestive of malignant round cell tumor. On biopsy extensive necrosis with viable focus of malignant round cell tumor were found. Tumor cells were strongly and diffusely positive for NKX2.2 while negative for LCA, Desmin and CK. Histopathology confirmed Ewing's sarcoma. Patient counselled regarding prognosis and sequelae of surgical intervention but he didn't give consent for surgery. Pt has been given a course of 3 cycles of chemotherapy with Vincristine, Actinomycin D, Cyclophosphamide and MRI was repeated. On post chemotherapy MRI, mass was found to be extending from D8 to L1 with involvement of D10, D11, and D12 costovertebral junction with intraspinal epidural component of soft tissue extending from D10 to D12 vertebral level compressing and displacing the lower dorsal cord. Tumor was unresponsive to chemo-radiotherapy and no improvement in sensory or motor function was there.

Discussion

Ewing's sarcoma involves the metaphyseal plates of long bones in childhood. In spine non-sacral involvements are extremely rare. EES has predilection for the soft tissues of the trunk like paravertebral and intercostal regions [2, 3]. It presents equally in both sexes of around 20 years age in contrast to skeletal Ewing's sarcoma which has male prediction of around 10 years. The most common presentation is painful bony swelling and most common frequent primary sites are femur, pelvis, humerus and fibula [2-4]. As with Ewing's sarcoma EES frequently metastasize into lungs and bone with poor prognosis [4].

Diagnosing Ewing sarcoma often requires immunohistochemical and molecular markers such as CD99 and FLI-1; these are sensitive but not specific. Recently, NKX2-2 has been reported as a diagnostically useful immuno-histochemical marker for Ewing's sarcoma [5-8].

The presence of initial benign musculoskeletal symptoms often leads to a delay in diagnosis or being misdiagnosed and treated for disc disease as in our patient who was treated with analgesics until he presented to emergency with urinary retention and paraparesis [9]. Symptoms included back pain or radiculopathy which may be due to muscular cramps, nonspecific axial spine pain, disc degeneration, fracture of vertebra, tumor and infection [1, 5]. A tumor in retroperitoneal organs (kidney, urinary tract, pancreas) or uterus, ovary in a female patient, abdominal aneurysm, and metastasis of gastrointestinal cancer to the lumbar spine can be source of back pain [10, 11].

For undiagnosed chronic back pain an important point while making diagnosis is to differentiate serious pathology (malignancy, infection). Physician should consult to specialist if Red flag sign are present, which are possible indicators of serious spinal pathology: chest pain, fever and unexplained weight loss, bladder or bowel dysfunction, history of malignancy, associated medical co-morbidities, deteriorating neurological deficit, disturbed gait, saddle anaesthesia, and age of onset <20 years or >55 years [10, 11]. When low back pain is present regardless of rest or movement, it should be considered malignancy or diseases of internal organs. When pain is present in a lower limb in addition to low back pain, there is a high possibility of radicular pain. History of weight loss, loss of appetite, evening rise of fever and steroid intake suggests infective etiology. History of injury and presence of motor palsy suggest fracture of vertebra while pain without occurrence of nerve root symptoms is defined as nonspecific low back pain [10, 11]. Locally examine for any tenderness and swelling over back. Positive Straight leg raising test, bowstring sign, lassegue test indicate towards nerve root compression. Radiographs, CT, MRI and bone scintigraphy of spine helps in ruling out tumor, infection, and vertebral fracture [10]. For suspicious tumor or infection blood tests (C-reactive protein, ESR, alkaline phosphatase, and tumor markers) are useful.

In case of EES the mean diagnostic delay calculated from the previous cases is 4.5 months. The symptoms commonly include back and/or radicular pain in all patients, paresis in about 70%, sensory disturbances in 35%, and to lesser extent bladder and bowel dysfunction in about 12% of patients. Distant metastases occurred in nearly 40% of the cases. Lung, spine, and brain were the most frequent sites of metastasis [12-15].

Neurological deficit is most important determinant for deciding treatment of disease. It is advisable to confirm diagnosis by needle biopsy and that subjected to a neoadjuvant chemotherapy regimen, which not only help shrink the primary tumor, thereby increasing chances of total excision, but also take care of micrometastasis and give an idea about responsiveness of the tumor to adjuvant therapy [3, 17]. This is followed by surgery or radiotherapy or both. Surgical intervention to relieve cord compression symptoms, as well as for cytoreductive purposes is considered the primary approach in the management. There is also consensus about use of chemotherapy and local radiotherapy. Vincristine, actinomycin, cyclophosphamide and or iphosphamide (VAC/IE) regimen is preferred for localized disease with radiotherapy (45 Gy in 25 fractions). Regimens such as VAdriaC (vincristine, adriamycin, and cyclophosphamide) are used to treat metastatic disease. There is also evidence that neoadjuvant therapy may be of limited use in spinal epidural EES. Patient treated with chemo-radiotherapy after surgery have better 1 year survival than patient treated with surgery, chemotherapy and radiotherapy alone [15-18]. In our patient chemo-radiotherapy was not found to be very effective in reducing tumor mass. However patient did not give consent for surgery.

Conclusion:- We advise the physicians to keep a broad vision while managing cases of undiagnosed chronic back pain and not to consider it benign until proven otherwise. A prompt intervention is required to keep neurological damage to a minimum, and a correct combination of surgery, chemotherapy, and radiotherapy is required for better long-

term patient outcome in case of EES and malignancies.

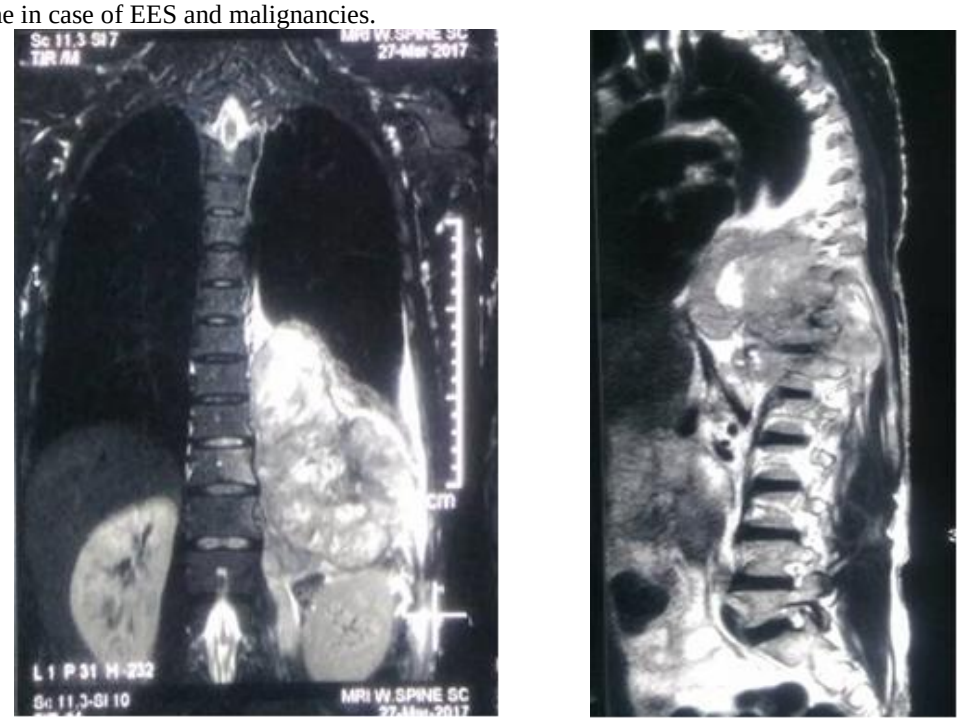

Image 1: Post chemotherapy MRI showing alterd signal intensity lesion in left paravertebral region extending from D8-L1 level with involvement of left D10, D11, D12 costovertebral junction

Image 2

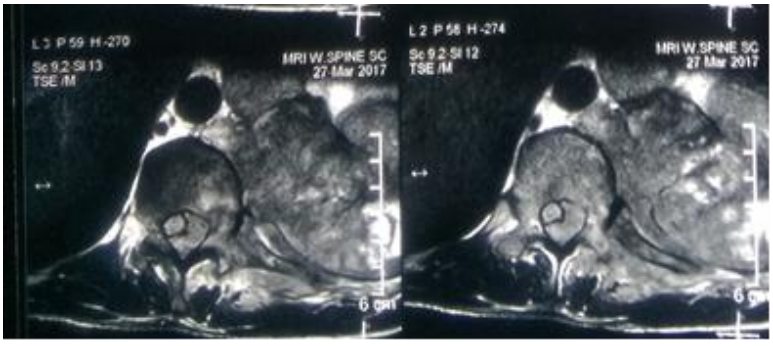

Image 3

Image 2 and 3 post chemotherapy MRI showing large inttradural component of soft tissue extending from D10 to D12 vertebral level, compressing and displacing the lower dorsal cord with signal alteration in the cord at D10-D11 and D11-D12 vertebral level.

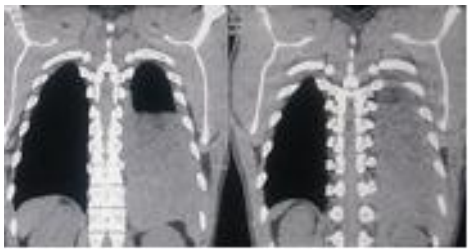

Image 4: Pre-chemotherapy CT showing heterogeneously enhancing soft tissue mass lesion in left paravertebral region at D12- L1 level. Fluid density is seen adjacent to the mass lesion with few interspersed hyperdensity possibly pleural effusion

Consent: Informed consent was obtained from the patient for the procedure and possibility of publication.

Conflict of interest: The authors declare no conflict of interest.

Author's contributions: Study concept and design: PKP, PKS, LM, AB; acquisition of data: PKP, PKS, RR, AB; analysis and interpretation of data: PKP, PKS; drafting of the manuscript: PKP, PKS, LM, AB; critical revision of the manuscript for important intellectual content: PKS, PKP. All authors read and approved the final manuscript.

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