EWING'S SARCOMA FAMILY OF TUMOUR IN ADULTS: A RARE CASE OF POSTERIOR MEDIASTINAL EWING'S SARCOMA WITH INTRASPINAL EXTENSION CAUSING SPINAL CORD COMPRESSION

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Introduction: Ewing's sarcoma family of tumours (ESFTs) are rare, small round cell tumour which originate from neuroectoderm. They are highly malignant tumours and may arise from bone or extraosseous soft tissue. Extraosseous Ewing's sarcomas (EESs) are relatively rare member of ESFTs and usually found in chest wall (trunk) and extremities. EESs are reported to be associated with poorer prognosis as compared with osseous Ewing's sarcoma.

Discussion: The patient is a 40 year-old lady who was previously healthy and presented to emergency department with chronic recurrent epigastric pain associated with left pleuritic chest pain without neurological deficit of bilateral lower limbs. Magnetic resonance imaging of thoracolumbar spine showed large lobulated left posterior mediastinal mass which appears iso-intense in T1W, heterogenous in T2W and enhanced post contrast. The mass is seen from the level of T1 till T6 vertebrae with intraspinal extension through the minimally widened left T5/T6 and T6/T7 neural foramina causing spinal cord displacement and compression. Histopathologic examination suggestive of Ewing's sarcoma / primitive neuroectodermal tumour. Unfortunately, she developed acute neurology with complete thoracic spinal cord injury AIS A with sensory level T4. She was referred to our centre for multidisciplinary management with conclusion of neoadjuvant chemotherapy before tumour debulking surgery or posterior instrumentation. Neoadjuvant chemotherapy was commenced. However, on day 3 of first cycle chemotherapy, patient developed neutropenic sepsis and unfortunately deceased.

Conclusion: EESs are aggressive tumours with high incidence of local recurrence and systemic metastasis, therefore poorer outcome. Early diagnosis with high degree of suspicious is essential due to unspecific presentation at disease onset. Principles of management of EES have been extrapolated from the experienced of treating primary osseous Ewing's sarcoma. Multimodality treatment consisting of adequate surgical resection, aggressive chemotherapy (both neoadjuvant and adjuvant) and adjuvant radiotherapy if indicated are recommended treatment for patient with extraosseous Ewing's sarcoma.