Case of Intradural Extramedullary Ewing Sarcoma in an Adult

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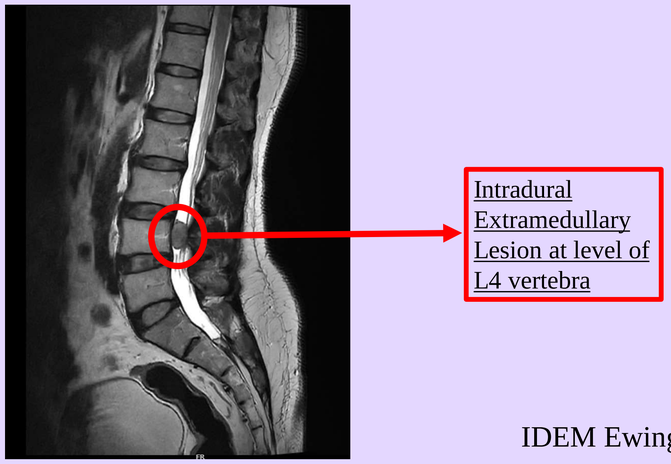
INTRODUCTION

- Ewing Sarcoma (EWS) is a malignant tumour characterised by primitive round cells lacking distinct differentiation. It accounts for less than 5% of all soft tissue sarcomas. Ewing sarcoma has a peak incidence at 10-15 years. Less than 1% of cases are seen in patients greater than 40 years of age.
- EWS are predominantly skeletal in origin. Extra-skeletal EWS, such as spinal EWS, are rare, with Intra Dural Extra Medullary (IDEM) EWS lesions, such as in this case, being even rarer.
- Hybrid gene EWS-FLI1, due to t(11;22)(q24;q12) gene rearrangement, is seen in 80% of cases. The EWS-FLI1 fusion protein acts as an aberrant transcription factor which could play a part in the pathogenesis of Ewing sarcoma.

CASE REPORT

A 53-year-old male presented with complaints of lower back ache since 3 months, with bilateral lower limb paraesthesias more in the right lower limb. There is no history of fever, trauma, or neck pain. MRI spine showed well defined Intradural Extramedullary (IDEM) lesion about 2 x 2.2 x 1.2 cm at the level of L4 vertebra with homogenous contrast enhancement and extension along the exiting nerve roots.



MANAGEMENT

- Patient underwent L4, with partial L3, laminectomy with durotomy, and excision of the lesion. Postoperatively patient had good neurological improvement in both lower limbs, more on the right side.
- Biopsy of IDEM lesion was suggestive of Ewing Sarcoma. Next Generation Sequencing (NGS) was positive for EWSR-FLI1, confirming the diagnosis of WHO CNS Grade 4 Ewing Sarcoma.

AMP Classification	CDS variant details	Interpretation	Treatment Recommendations	Treatment Response
***EWSR1/FLI1* (FUSION)** Total Read depth - 33x				
Tier II	NA	Oncogenic	NA	Diagnostic

- 4 cycles of IE regimen chemotherapy followed by PETCT imaging showed no evidence of locoregional or distant metastasis. Chemotherapy continued with VAC/IE regimen

DISCUSSION

- This case details an unusual scenario of Ewing Sarcoma of spinal origin (IDEM) arising in an adult of 53 years of age.
- EWS patients over the age of 40 have been observed to have a lower chance of survival. This could be linked to late stage of malignancy at diagnosis in older patients, as well as a higher incidence of comorbidities, leading to increased risk of treatment-related toxicity or poor performance of treatment.
- IDEM Ewing Sarcoma is associated with a high recurrence and metastasis rate. Lumbo-sacral region is the most common location. Patients most commonly present with pain in the lower back and motor disturbances in the lower limbs, as in this case.
- Surgical intervention is the primary treatment in Spinal EWS, with complete resection leading to increased survival.

CONCLUSION

IDEM Ewing Sarcoma is an unusual variant of the soft tissue malignancy, requiring a multidisciplinary approach. Diagnosis is done based on immunohistochemistry and molecular analysis of excised lesion. Subsequent chemotherapy is planned based on metastatic presence (and extent of) or absence on PETCT imaging