Acute flaccid paralysis: a rare presentation of Ewing sarcoma in the spine

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Introduction and background

-Ewing sarcoma is a primary bone sarcoma.
-Most common malignant bone tumor in children <10 years old.[1]
-Median age of presentation is 15 years old.
-Presenting symptoms:

- Palpable mass or regional pain
- Paresthesias with generally reported progression over weeks to months
- Symptom duration median of 2-9 months

-Primary Ewing Sarcoma of the spine is extremely rare representing only 8% of cases.[2]
-Rare cases of associated cauda equina syndrome have been reported in the literature with reported evolution of symptoms related solely to tumor over the course of days.[3-5]

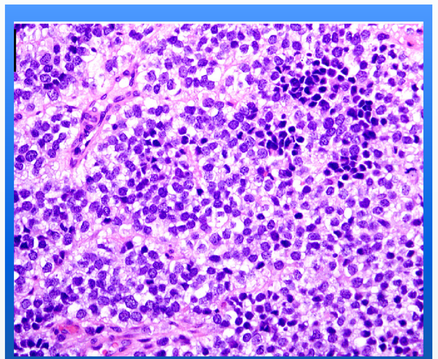

Figure 1: Ewing Sarcoma: Characteristic sheets of small round cells with scant cytoplasm.[6]

-Metastases detectable in 25% of patients, most common in lung.
-Most consistently associated with t(11;22)(q24;q12) chromosomal translocations.[2]
-Histology demonstrates small round cell tumors as seen in Figure 1.

Case presentation

15 year old previously healthy male presented with acute flaccid paralysis

-Preceding 3 weeks of low back pain → workup per PCP included negative spine x-ray.
-Patient described sudden loss of sensation and strength from waist down over several minutes with associated inability to void.
-No other systemic symptoms, including fever or pain.
-Examination: 1+ patellar/achilles reflexes bilaterally with no sensation from ASIS inferiorly. Negative Babinski reflexes. Neurologically intact in upper extremities with appropriate mentation.

Workup and diagnosis

-Initial differential diagnosis:

- Osteomyelitis
- Epidural abscess
- Localized spinal infarction or hemorrhage
- Guillain-Barre syndrome
- Spinal cord tumor, including osteosarcoma
- Acute disseminated encephalomyelitis

-Outside hospital labs: normal BMP, PT/INR, magnesium, and phosphorus.

-MRI brain w/ and w/o contrast normal.

-MRI spine demonstrated tumor extending posteriorly from T12-L2 with osseous, dorsal epidural, and paraspinous soft tissue involvement with thecal sac impingement; see figure 2.

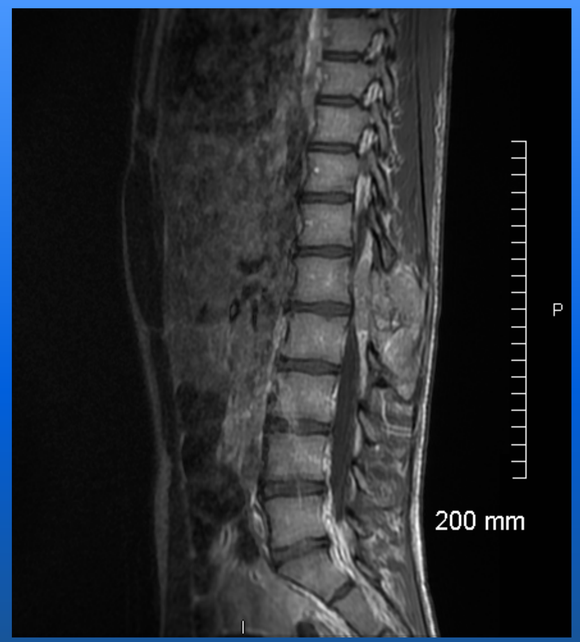

Figure 2: Tumor extending from T12-L2 with soft tissue involvement. Thecal sac impingement, resulting in cauda equina syndrome.

Patient was emergently taken to the OR per neurosurgery where he underwent T12-L2 laminectomy and removal of surrounding paraspinal musculature.

Intraoperative findings:

-Small round blue cell tumor, highly malignant and vascular, suspicious for Ewing sarcoma vs neuroblastoma.
-Pathology confirmed EWSR1 gene rearrangement at 22q12.

Concluding points

1) Importance of a broad differential in rapidly evolving acute flaccid paralysis:

- Initial leading differentials were focused on generally acute pathology (infectious vs autoimmune vs infarction/hemorrhage)
- Ewing sarcoma and other primary bone malignancies should be considered in acute neurologic presentations.

2) Ewing sarcoma variability in presentation:

- Other reported cases describe days to week-long progression of symptoms when Ewing sarcoma presents in the spine (already a rare primary location).
- This case is unique in the rapid evolution of acute flaccid paralysis over span of few minutes.

References

[1]Prusakowski, MK, Cannone, D. Pediatric Oncologic Emergencies. Emergency Medicine Clinics of North America. 2014. 32(3): 527-548.
[2]Cotterill SJ, et al. Prognostic Factors in Ewing's Tumor of Bone: Analysis of 975 Patients From the European Intergroup Cooperative Ewing's Sarcoma Study Group. Journal of Clinical Oncology. 2000. 18(17): 3108-3114.
[3]Gopalakrishnan CV, et al. Primary Ewing's sarcoma of the spine presenting as acute paraplegia. J Pediatr Neurosci. 2012. 7(1): 64-66.
[4]Kannan, KK et al. Unusual Presentation of a Primary Ewing's Sarcoma of the Spine with Paraplegia: A Case Report. J Clin Diagn Res. 2015. 9(3): RD01-RD03.
[5]Electricwala, AJ, Electricwala JT, Primary Ewing's Sarcoma of the Spine in a Two-Year-Old Boy. Case Reports in Orthopedics. 2016. 1–4.
[6]Image adapted from Histopathology. 2017. 71(5): 786-794.