

Atypical Presentation Of Pre-Sacral Malignant Peripheral Nerve Sheath Tumour (MPNST): A Case Report

Mohd Kamel MJ; Kamel MJ; Sahran Y; MZ Norazman; Faisham WI

Department of Orthopaedic, Universiti Sains Malaysia, Kelantan

INTRODUCTION:

Malignant peripheral nerve sheath tumor (MPNST) involving the sacrum is a rare clinical condition. MPNST is high-grade malignant tumors comprising 5-10% of soft tissue sarcomas.¹Symptoms depend on the tumor anatomical location, and whether it compresses neighboring structures, such as the sacral nerve roots or intrapelvic organs.²

REPORT:

We report a case of 30-year-old gentleman who complained of radicular-pain over his left lower limb for 2 years. The pain was increased with walking and decreased by rest and analgesics. He didn't experience back pain, abdominal pain, fever, and no trauma. Power of lower limb was generally grade 4 with positive straight leg rising test. He was treated clinically as prolapsed intervertebral disc (PID) for 2 years. He lost significant weight over the duration. Constipation and frequent micturition getting worse over time. For such, he sought second opinion.

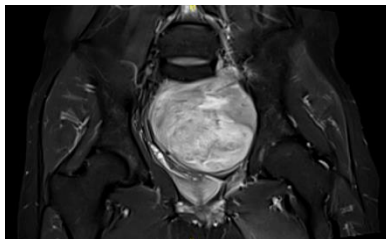


Figure 1

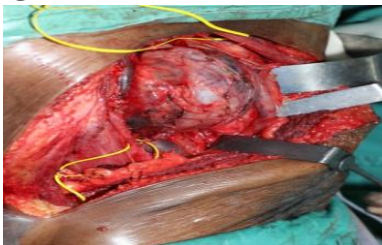


Figure 2

At our centre, MRI of the pelvic was performed and revealed a huge pre-sacral mass (figure 1). The solid mass was heterogeneously enhanced, measured 11x13 x14cm. It caused pressure erosion to the subjacent sacrum. Tru-cut biopsy suggestive of MPNST and patient underwent tumor resection through anterior approach, Left S1,S2,S3 and sciatic nerve embedded inside the tumor (figure 2).

CONCLUSION:

MPNST are highly aggressive neoplasms with five-year survival rate 33-39%. Mean age of presentation is 39.7 years in patient without von Recklinghausen disease (NF).¹ We would like to highlight that even though rare, it may occur in the sacrum or presacral region. Not uncommon for the tumor to grow large in pelvic before being diagnosed. Wide-resection is the mainstay of treatment, but the involvement of surrounding critical structures and the tumors' enormous size limit the extent of resection in the pelvic.¹ Adjuvant radiation therapy is important for local control, and chemotherapy may be of benefit in some patients with metastases and whose tumors cannot be completely removed.

REFERENCES:

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