MEDICAL SCIENCE

Giant dumbbell shaped malignant peripheral nerve sheath tumour arising from sciatic nerve: A case report

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ABSTRACT

Malignant peripheral nerve sheath tumours in the sciatic nerve are sporadic and are often misdiagnosed. We herein report a case of a 22-year-old woman presenting with low back pain radiating to the right lower limb and right lower limb weakness. CT and MRI scan showed a large complex solid cystic lesion arising from S1 and S2 nerve roots extending from the right presacral space and traversing through the obturator foramina and emerging in the right gluteal region. Surgical resection with a bi-modal (Trans abdominal and gluteal) approach resulted in complete excision of the mass with an appreciable margin. The histopathological report showed a malignant peripheral nerve sheath tumour. Post tumour board discussion patient underwent adjuvant radiation. The patient has been under regular follow-up for six months and is disease-free.

Keywords: Malignant peripheral nerve sheath tumour (MPNST), dumbbell shaped, sciatic nerve, computed tomography (CT), magnetic resonance imaging (MRI), radiation therapy.

1. INTRODUCTION

Malignant peripheral nerve sheath tumour (MPNST) is a rare malignant soft tissue sarcoma of ectomesenchymal origin (Panigrahi et al., 2013). It is extremely rare for MPNST to originate from the sciatic nerve, with only a few cases having been documented to date. MPNST is commonly witnessed in patients belonging to the age group of 20 to 50 years (Weiss and Goldblum, 2001). Management of MPNST should include achieving complete surgical resection (R0 resection) followed by adjuvant radiotherapy and chemotherapy (James et al., 2016). Chances for developing local recurrence (40-65%) and distant metastases (30-60%) after undergoing aggressive treatment are extremely high (Anghileri et al., 2006). In patients with high grade MPNST the overall 5-year survival is poor (20-50%) (Eilber et al., 2004). A positive margin on histopathology increases the risk of mortality by 1.8-fold. Since MPNST are very aggressive tumour's which elicit vague

symptoms such as radicular pain and sensorimotor deficits it is essential to make an early diagnosis.

2. CASE REPORT

A 22-year-old female came to our outpatient clinic complaining of low back pain radiating to the right lower limb for the past eight months and right lower limb weakness for two months. No history of urinary or bowel incontinence. On examination, we were able to demonstrate right foot drop. Contrast-enhanced CT abdomen-pelvis showed a large heterogeneously enhancing well-defined lesion noted in the pelvis extending through the right obturator foramina and invagination along the planes of gluteus musculature and causing splaying of the muscles, measuring approximately 15x10x6cm. The fat planes between the presacral region and the lesion are lost along with the displacement of adjacent pelvic organs. High-resolution chest CT showed no evidence of metastasis. MRI lumbar spine revealed a mixed intensity heterogeneously enhancing solid cystic lesion appearing to arise from the S1 and S2 nerve roots within the neural foramina on the right side extending into the presacral region (Figure 1). Posteroinferiorly, the lesion extends into the gluteal region displacing the gluteus maximus muscle posteriorly. The nerve roots of S1 and S2 appear thickened, causing the widening of S1 and S2 neural foramina on the right side. Medially it is abutting and displacing the sigmoid colon. CT guided biopsy from the lesion suggested a MPNST.

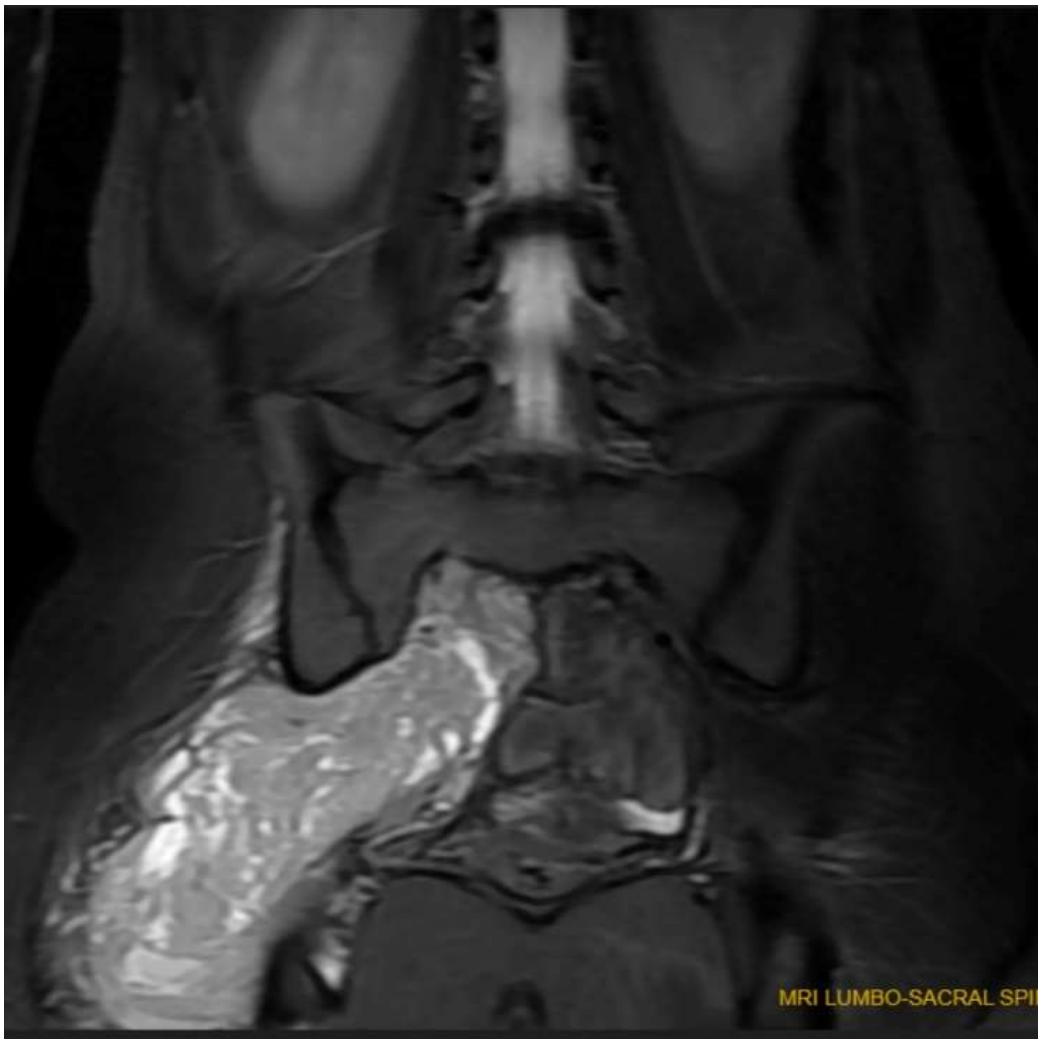

Figure 1 MRI lumbosacral spine showing tumour arising from s1 and s2 nerve roots

The patient's case was presented in a tumour board discussion and she was advised upfront surgery. The patient was planned for resection of tumour with the Trans abdominal and gluteal approach. The patient was initially placed in a supine position and a midline incision was taken over the anterior abdomen. Mass was found to arise from the presacral region and was only confined to the right side (Figure 2). After separating the tumour from the right ureter and dissecting out the mesocolon, the mass was noted to be originating from S1 and S2 nerve roots. The internal iliac vessels were identified, splayed and carefully dissected from the

tumour. The proximal branches of internal iliac vessels were preserved and terminal branches were ligated. Dissection was continued up to obturator foramina. For the next part of the surgery, the patient was shifted to a left lateral position and the most prominent portion of the tumour over the right gluteal region was incised (Figure 3, 4). Once the normal portion of the sciatic nerve was identified on dissection, a small segment was cut and sent for frozen section and the margins were found to be negative. Tumour in its entirety was removed through the gluteal incision (Figure 5). A drain was inserted from the gluteal region, with one limb passed through the obturator foramina into the pelvis and the other limb placed in the subcutaneous plane in the gluteal region. Post-operatively patient was managed with active physiotherapy and a foot drop splint. There was no evidence of bowel/bladder incontinence or additional neurological deficit post-surgery. The histopathological report suggested a low-grade MPNST with margins negative for malignancy (Figure 6). However, as there was suspected margin positivity at the level of foramen patient was advised adjuvant radiotherapy. The patient is currently doing well and has been disease free in the last six months of follow-up post radiation.

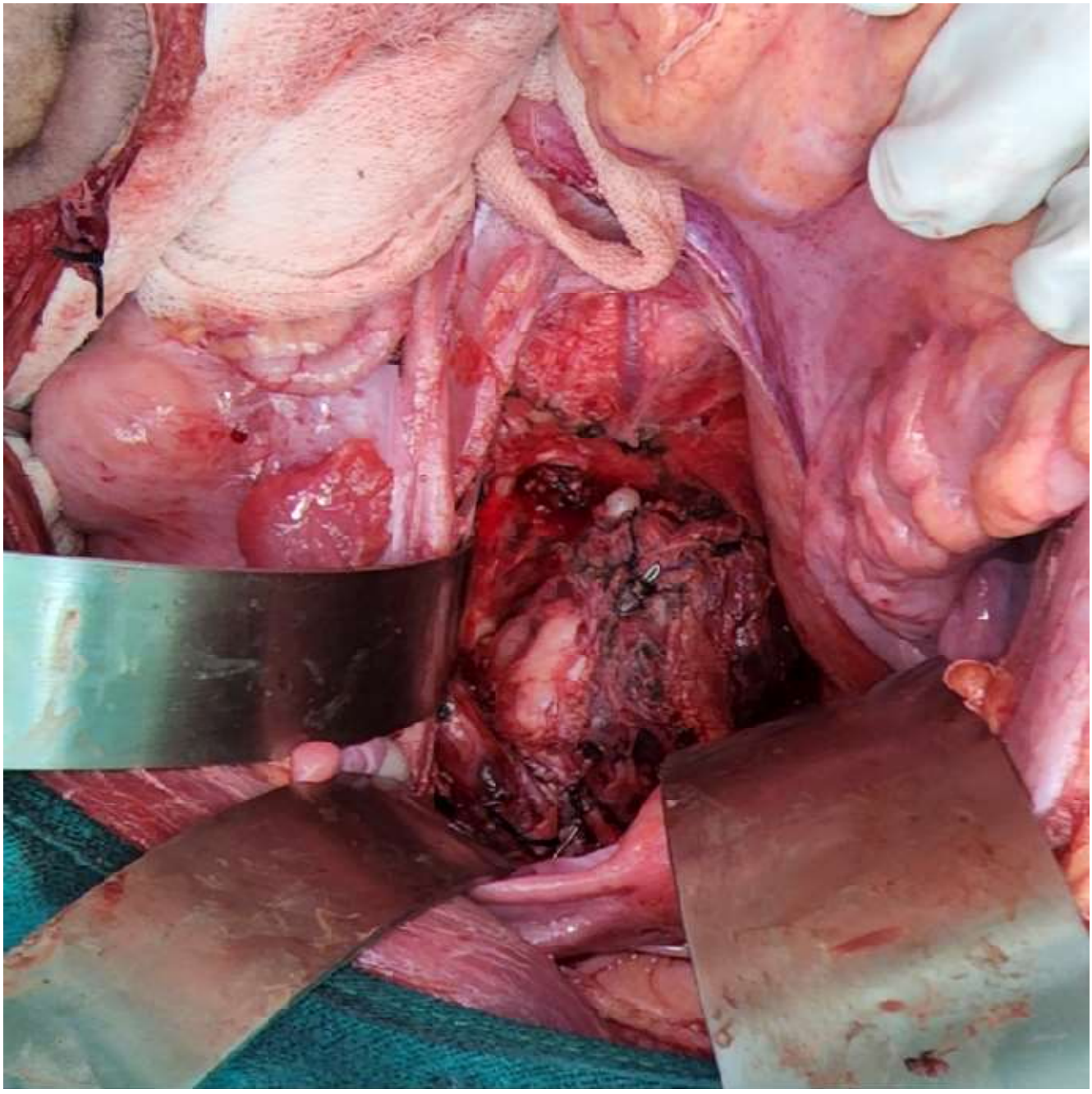

Figure 2 Intraoperative image of tumour in transabdominal approach

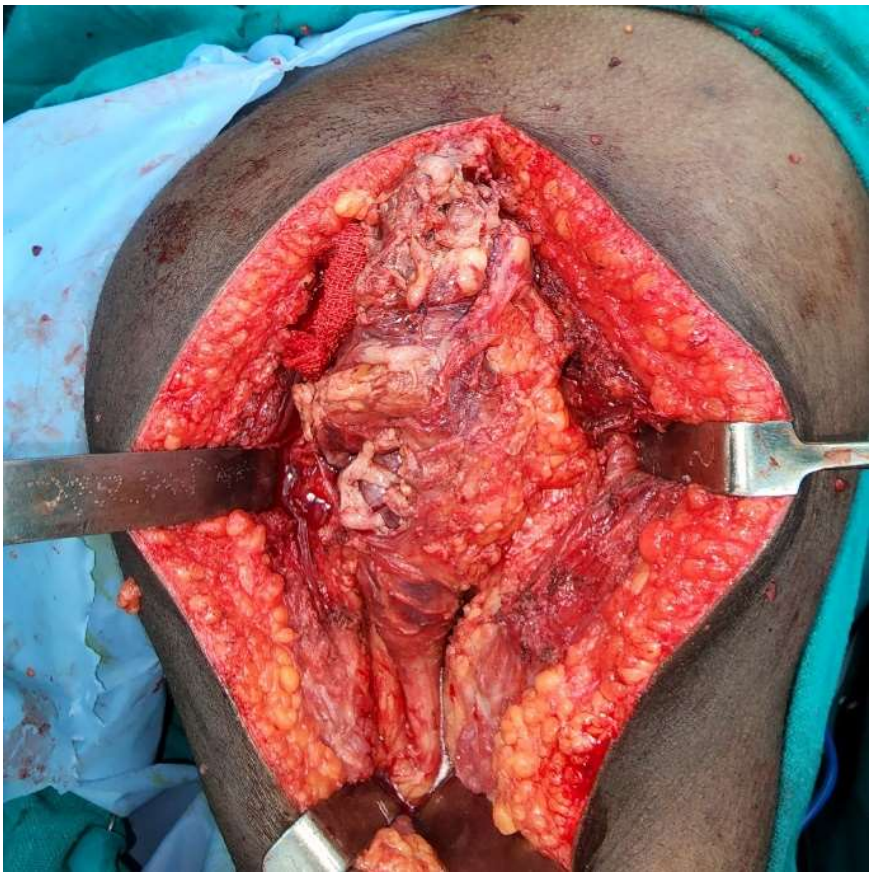

Figure 3 Intraoperative image of tumour in gluteal approach

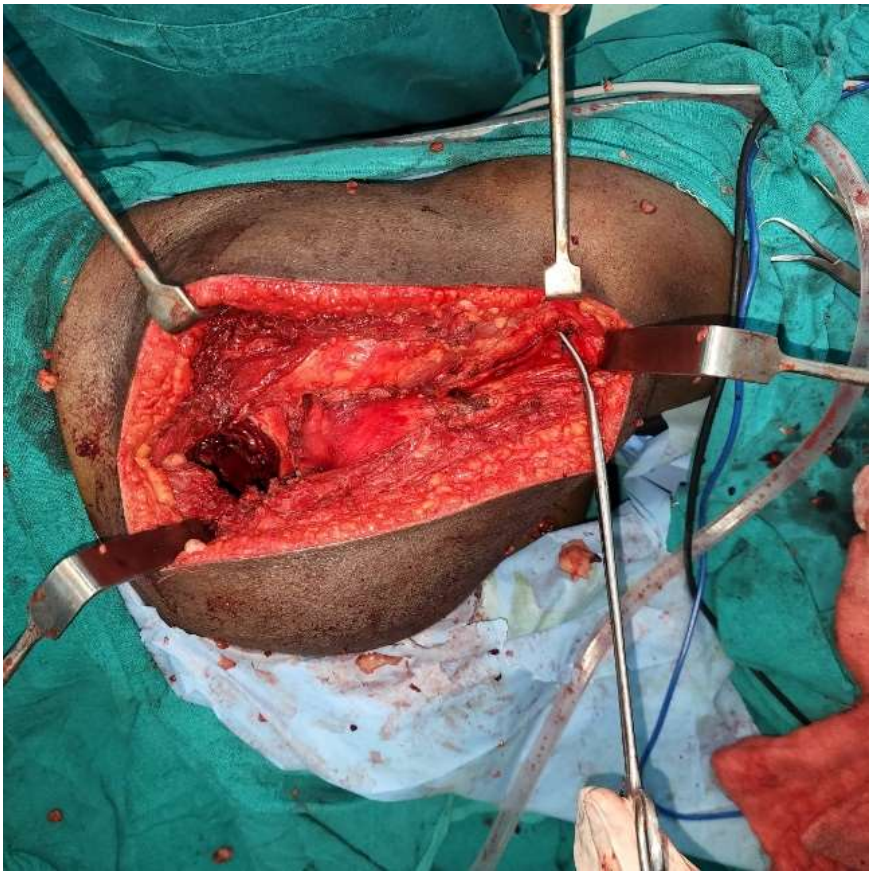

Figure 4 Intraoperative image post excision of tumour

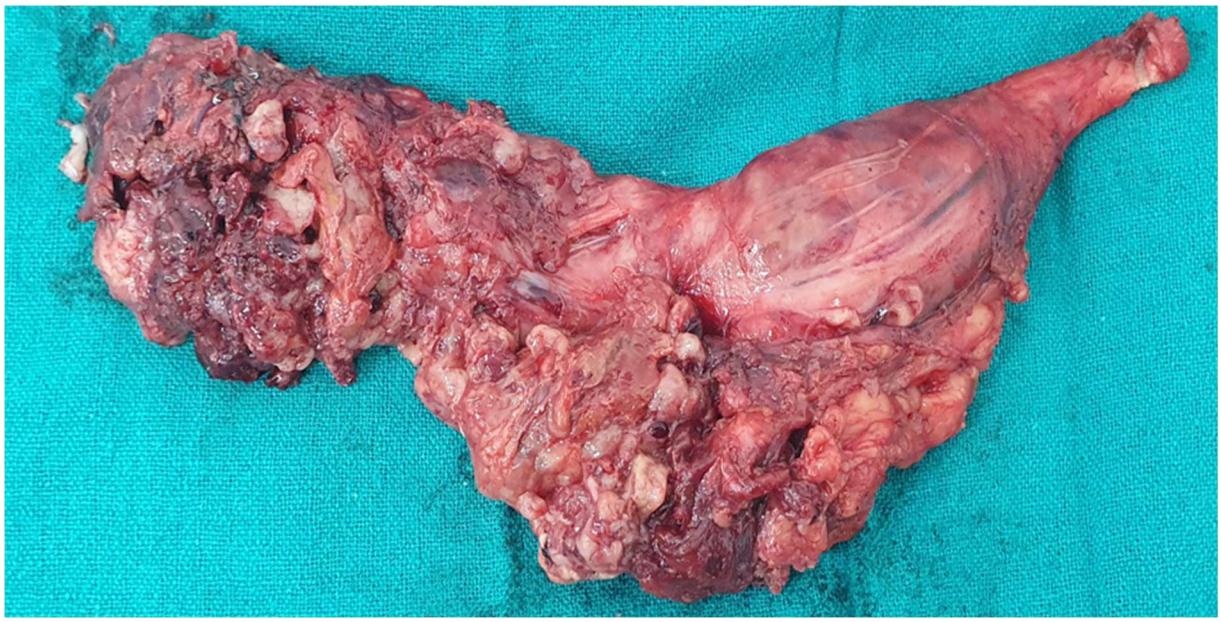

Figure 5 Excised specimen of dumbbell shaped tumour

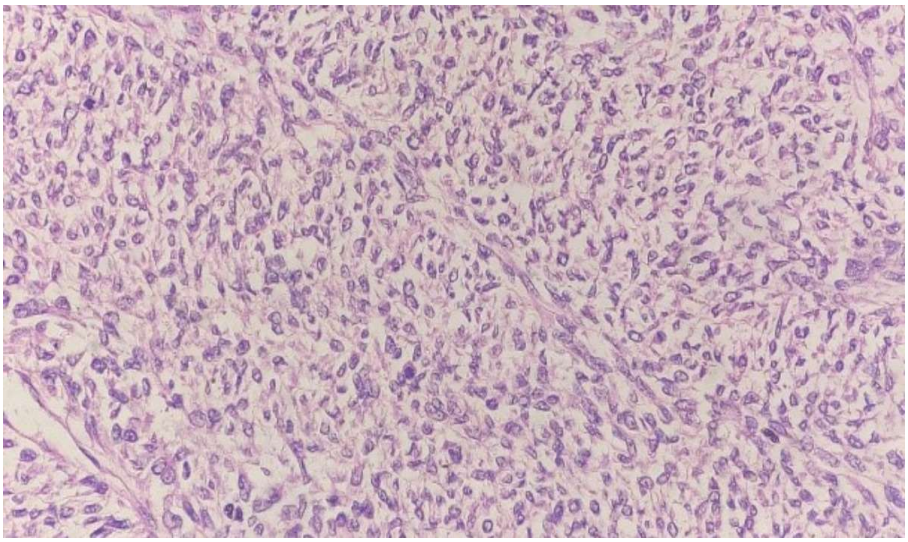

Figure 6 Histopath image showing monomorphic spindle cells arranged in interlacing fascicles, palisades and whorls

3. DISCUSSION

MPNSTs are sporadic tumours of the nervous system that develop from peripheral nerve sheath fibroblasts or Schwann cells (Dabski et al., 1990; Garg et al., 2004). It is estimated that MPNST is present in 0.001% of the population and accounts for 3-10% of all soft tissue sarcomas (Winslow et al., 2015). Patients with NF1 are at a greater risk as 8–13% of plexiform neurofibromas show the potential to transform into MPNST (Winslow et al., 2015; Evans, 2002). Approximately 10% of MPNSTs tend to have a history of previous radiation exposure (Ducatman and Scheithauer, 1983; Foley et al., 1980). MPNST belongs to the subcategory of soft tissue sarcomas as per WHO central nervous system tumours classification (Louis et al., 2016). MPNSTs are highly aggressive soft tissue sarcomas with high local recurrence rates and distant metastases (Cury et al., 2007). Ideally, peripheral nerves can develop MPNSTs anywhere along the deep tissues but are commonly found to involve major nerve trunks. Intra pelvic MPNST is significant since it is difficult to diagnose in clinical settings. Pelvic tumours have a moderate growth rate and are not palpable along the nerve route. Before they cause symptoms, they are typically challenging to diagnose. Motor deficits, paraesthesia, radicular pain and ureteral obstruction make up the most typical clinical manifestations of intrapelvic MPNST. Hematuria and bone pain may indicate invasion of adjacent structures.

MRI is the best diagnostic modality for assessing the tumour and its relation to the nearby structures. It is vital to differentiate MPNST from other benign lesions on MRI as benign tumours classically present with peripheral ring enhancement and a hypo-intense central area on T2–weighted images (White et al., 1990); however, malignant tumours are represented by areas of necrosis, haemorrhage and heterogenous contrast enhancement (Kar et al., 2006).

Histologically, MPNSTs are described to have spindle cells with hyper-chromaticity and a fasciculate appearance. Areas of necrosis, perivascular hyper-cellularity, elongated and wavy nuclei and significant mitotic activity are also frequently seen (Stucky et al., 2012). Immunohistochemistry markers sensitive to MPNST include s-100, vimentin and Neuron-specific enolase (NSE).

The treatment of choice for intra-pelvic MPNST is upfront surgery. The main aim of surgical intervention should be to achieve R0 resection, which may involve sacrificing surrounding structures. It is challenging for a surgeon to completely excise the mass and achieve negative margins without injuring the involved nerve trunk in intra-pelvic MPNSTs. Our patient underwent wide local excision of MPNST; unfortunately, the sciatic nerve could not be salvaged. A combination of transabdominal and gluteal approaches should be considered when pelvic malignancies expand beyond the pelvis through the obturator foramina. As per earlier studies, adjuvant radiation and chemotherapy have shown better survival outcomes (Stucky et al., 2012; Baehring et al., 2003). However, the only surgical procedure likely to have a favourable prognosis is total excision before metastasis.

The prognosis for MPNST patients is typically not good. Poor prognostic markers are high-grade tumours, large size, proximally located tumours, positive resection margins and NF1 (Angelov et al., 1998; Anghileri et al., 2006). The 5-year survival rate is directly related to NF1. Patients without NF1 have a reported 5-year survival rate of up to 50%, whereas patients with NF1 have only 10 (Doorn et al., 1995). These tumours are prone to local recurrence and systemic dissemination, with pulmonary, hepatic and bone being common sites of metastasis.

4. CONCLUSION

MPNST arising from sciatic nerve are extremely rare and henceforth very challenging for a surgeon to diagnose and manage. Vague symptoms and infrequent incidence often lead to misdiagnosis. Only with a high index of suspicion and prompt radio-imaging and biopsy can an early diagnosis be achieved. Aggressive management with upfront surgery (R0 resection) and immuno-histochemical analysis followed by radiotherapy is of utmost importance to prevent recurrence and improve survival.

Author Contributions

Rahul Rajendran has collected information and prepared the manuscript which was reviewed by all authors.

Informed consent

Not applicable.

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Conflict of interest

The authors declare that there is no conflict of interests.

Data and materials availability

All data sets collected during this study are available upon reasonable request from the corresponding author.

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