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Malignant peripheral nerve sheath tumor with extensive osteosarcomatous and chondrosarcomatous differentiation: A case report

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ABSTRACT

BACKGROUND: Malignant peripheral nerve sheath tumor is an uncommon tumor of the peripheral nerves. The commonest presenting symptom is soft tissue mass and pain with local neurological findings. Imaging modalities are unhelpful in making a reliable diagnosis. Treatment is radical resection with adequate clear resection margins. Radiotherapy improves the local control, but the prognosis remains poor especially in those with divergent differentiation.

SUMMARY: A 23-year-old man with no history of neurofibromatosis presented with a swelling on the back which has been gradually increasing in size and causing him discomfort. The tumor was surgically excised and the histopathological examination revealed malignant peripheral nerve sheath tumor with extensive osseous and cartilaginous differentiation. He developed pulmonary metastases one year after the surgical resection. Pulmonary metastatectomy was therefore performed and the histopathology of the metastatectomy specimen revealed metastatic malignant peripheral nerve sheath tumor, but without any osseous or cartilaginous differentiation. He remained well with no recurrence or metastases at 9-month follow-up.

CONCLUSION: Malignant peripheral nerve sheath tumor is a malignant tumor that behaves aggressively despite adequate radical resection. This case also illustrates extensive osseous and cartilaginous divergent differentiation of the primary tumor which was surprisingly absent in the metastatic lesions. This finding warrants further research.

1. Introduction

Malignant peripheral nerve sheath tumor (MPNST) is an uncommon soft tissue sarcoma that arises from the peripheral nerves. It may originate from normal nerves or pre-existing plexiform neurofibromas and perineuriomas. The tumor is sporadic in the majority of the cases but it is associated with neurofibromatosis type 1 (NF1) in 20%–50% of the cases especially in younger patients [1,2]. MPNST commonly affects the trunk and extremities and less often the head and neck area [3]. The tumor is well known for its aggressiveness and tendency to metastasize to distant organs especially lung and bones. The local control of the tumor is best achieved by radical surgical resection and adjuvant radiotherapy. However, this does not prevent early development of distant metastases.

We report herein a case of MPNST of the trunk with bone and cartilage differentiation in a young man who had no clinical features of neurofibromatosis type 1 (NF-1) and who developed pulmonary metastases one year after surgical resection and adjuvant radiotherapy. Of interest is that the metastatic lesions in this case – unlike the primary tumor – did not exhibit any divergent osseous or cartilaginous differentiation.

2. Case report

A 23-year-old male presented to the surgical clinic with a swelling over the left side of his back. The swelling was noticed several years earlier, but was small and painless. He was examined at another facility and was advised to observe. The swelling became larger, harder and uncomfortable over the recent months and was therefore referred as a case of 'back lipoma'. There was no significant past surgical or medical history and he denied any family history of neurofibromatosis. Examination revealed a hard oval swelling (7 × 4 cm) just to the left of the upper lumbar spine which was partially mobile. Routine blood tests were within normal range

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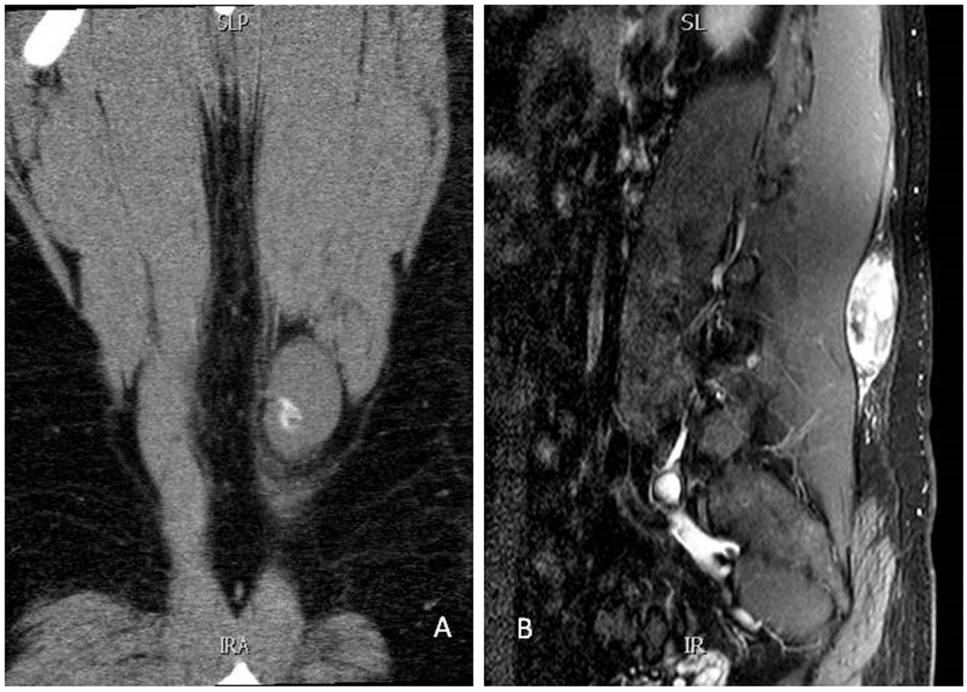

Fig. 1. Magnetic resonance imaging coronal view (panel A) and sagittal view (panel B) showing clearly the lesion with some calcification.

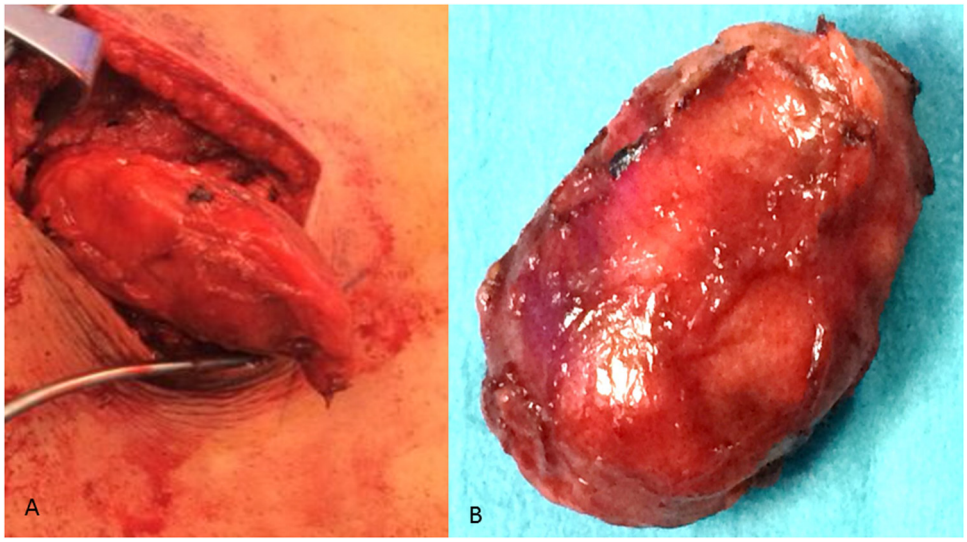

Fig. 2. An operative view showing the lesion as it was dissected from the surrounding structure (Panel A). Panel B showing the excised mass which looked well encapsulated with a smooth surface.

and review of the computed tomography (CT) and magnetic resonance imaging (MRI) scans which were done outside confirmed the presence of a soft tissue mass which was unlikely to be a lipoma (Fig. 1). Fine needle aspiration cytology (FNAC) was offered, but refused by the patient and hence surgical resection of what appeared to be an encanpsulated mass (Fig. 2) was carried out.

Pathological examination revealed a soft tissue mass measuring $7 \times 4 \times 2.5$ cm, with soft myxoid areas and firm hard areas. Histologically, the tumor exhibited areas of malignant spindly mesenchymal cells with brisk mitotic activity within loose myxoid stroma and foci of tumor necrosis. In addition, areas with marked osteosarcomatous and chondrosarcomatous differentiation were identified (Fig. 3A–D). However, no rhabdomyosarcomatous or glandular differentiation was noted. Immunohistochemical stains show focal nuclear staining of the malignant spindly cells for S100 protein but they were negative for smooth muscle actin, desmin and myogenin. These findings confirm the tumor as MPNST. Based on the pathological findings, CT scan of the chest, abdomen and pelvis was performed and this excluded presence of distant metastases. Also, neurosfibromatosis type I (NF1) gene sequencing was ordered at a referral laboratory and found to be negative.

The case was discussed in the multidisciplinary tumor board and adjuvant radiotherapy was recommended. After finishing all radiotherapy sessions, he remained well with no evidence of recurrence or metastases at 6- and 9-month follow-ups. However, at 12-month follow-up a body combined ^{18}F-fluorodeoxyglucose–positron-emission tomography and CT (FDG–PET–CT) scan showed a right lung metastatic nodule with avid FDG activity (SUV_{max} 6.8) (Fig. 4). Another smaller nodule was also noted in the anterior aspect of the left upper lobe. The patient underwent right mini-thoracotomy