INTRODUCTION
Malignant peripheral nerve sheath tumours (MPNSTs) are sarcomas which originate from peripheral nerves or from cells associated with the nerve sheath, such as Schwann cells. World Health Organization (WHO) combined the term MPNST replacing previous heterogeneous and often confusing terminologies, such as; malignant Schwannoma, malignant neurilemmoma, and neurofibrosarcoma for tumours of neurogenic origin and similar biological behaviour. They can occur either spontaneously or in association with neurofibromatosis-1 (NF1).

However, this case reports MPNST which started from left inguinal region and then eventually spread to omentum and presented like omental cake.

CASE REPORT
A 21-year-old unmarried male presented with a history of mass in his left inguinal region for the last 4 months. He was a non-smoker with no previous history of any type of radiation exposure. This mass was painless and gradually progressing in size. It was not associated with fever, testicular or scrotal swelling. Both testes were normally located in the scrotum. His lower limbs were also normal with no evidence of any mass or infection on it. There was no history of abdominal pain, distention, nausea, vomiting, diarrhea or constipation.

He consulted a surgeon who excised this mass on May 19, 2009 and histopathology report stated a malignant peripheral nerve sheath tumour. Resection margins were positive. Mitotic activity was 34/10 HPF (high grade). Tumour size was 9 x 5.5 x 4.5 cm (pT2). S-100 was positive while CD34 was focal positive. ASMA and CK-AE1/AE3 were negative. No pre-operative CT scan or any tumour markers for germ cell tumour were done. Figure 1 shows the status of residual mass after resection. After surgery, he was referred to our Institute for further management. Just after 2 weeks of surgery, he developed marked abdominal distension. Post-operative CT scan revealed marked omental caking (Figure 1) and ascites with residual mass in left spermatic cord. Multiple enlarged lymph nodes were also reported in the left inguinal region; the largest was 3.6 x 3.0 cm in size. His alpha fetoprotein, LDH, β-hCG and CEA levels were within normal limit. Hepatitis profile for “B” and “C” viruses, was also negative. CT scan of chest revealed no evidence of any pulmonary metastases. He was advised for tru-cut biopsy of omentum which revealed malignant peripheral nerve sheath tumour. S-100 was positive while CD34, ASMA and CK-AE1/AE3 were negative. Disease was labeled in stage-IV as it was pT2, Nx, M1 (omental metastases).

Omental Caking: A Rare Manifestation of Malignant Peripheral Nerve Sheath Tumour
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ABSTRACT
Malignant peripheral nerve sheath tumour (MPNST) is a very rare tumour with an incidence of one per 100,000 and constitutes between 3 to 10% of all soft tissue sarcomas. Most of the sarcoma involve the extremities and retroperitoneal regions. However, this case presented with mass in left inguinal region and then spread rapidly to omentum, assuming the appearance of an omental cake. Mass responded well to chemotherapy comprising of Ifosfamide and Doxorubicin.

Key words: Omentum. Soft tissue sarcoma. Omental cake. Inguinal region. Malignant peripheral nerve sheath tumour.
attendant. Chemotherapy was started after their permission. He received two cycles of Ifosfamide and Doxorubicin with MESNA protection (AIM protocol). Ifosfamide 1500 mg/m² day 1-4 and Doxorubicin 20 mg/m² day 1-3. Cycle was repeated after every 3 weeks. Interim analysis after two cycles of chemotherapy revealed good clinical response. Omental caking resolved marked and ascitic fluid was cleared completely (Figure 2). He is being continued with the same chemotherapy protocol and will complete six cycles.

DISCUSSION

The etiology of MPNST’s is unknown but there is a higher incidence in patients with a history of radiation exposure.2,3 Up to 50% of MPNSTs occur in patients with NF1 demonstrating the tendency for this tumour to arise from a pre-existing neurofibroma.4 MPNSTs generally occur in adulthood, typically between the ages of 20 and 50 years of age. Approximately 10-20% of cases have been reported to occur in first 2 decades of life. These tumours are biologically aggressive in nature.5 Surgery is the mainstay of treatment. The goal of operation is to achieve complete surgical excision with negative margins which could not be achieved in this case. This offers the best outcome with respect to both local recurrence and distant metastases. They are usually deep-seated and often involve the proximal upper and lower extremities as well as the trunk.

MPNST is a very rare tumour with an incidence of one per 100,000 and constitutes between 3-10% of all soft tissue sarcomas.6 MPNSTs are generally considered chemotherapy and radiotherapy-resistant tumours and however, this patient responded very well to chemotherapy. The local recurrence rate for MPNSTs has historically been reported to range from 40-65% and the distant recurrence rate has similarly been reported to range from 40-68%.7 Five-years survival has been reported to range from 16-52%. Longer survival has been correlated with complete surgical excision, small tumour size (< 5 cm), and the presence of a low grade component.

MPNST presentation with omental caking remains unique for this type of tumour. Omental caking is usually seen in ovarian tumours but not in nerve sheath tumours. Hence this case report adds another novel differential to the list of causes of omental cake appearance.

REFERENCES