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Case Report

Malignant peripheral nerve sheath tumor of anterior abdominal wall-A rare presentation

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ABSTRACT

Malignant peripheral nerve sheath tumor is highly malignant tumors that occur most commonly in head and neck as well as extremities. Very rarely it seen in the retroperitoneum. When associated with neurofibromatosis, they are having a poor outcome. We present a case malignant peripheral nerve sheath tumor of anterior abdominal wall. The location and rapid increase in size of the swelling permitted early diagnosis and treatment.

1. Introduction

Malignant peripheral nerve sheath tumor are common in extremities as well as head and neck. Soft tissue sarcomas include more than 70 histologic subtypes. Historically, the most common subtypes in adults (excluding Kaposi's sarcoma) were malignant fibrous histiocytoma (28%), liposarcoma (15%), leiomyosarcoma (12%), synovial sarcoma (10%), and malignant peripheral nerve sheath tumor (6%). The overall 5-year survival rate for patients with all stages of soft tissue sarcoma is 50% to 60%. Of the patients who die of sarcoma, most succumb to lung metastasis, which 80% of the time occurs within 2 to 3 years after initial diagnosis.

2. Case Report

A 27 yr. old gentleman presented with swelling in right side of the abdomen for 10yrs. The swelling was small but gradually increased in size. He noticed a sudden increase in size since 1month. A painless swelling initially now experiencing dull pain. He had no of fever. He gives history of weight loss.

He is a known case of Neurofibromatosis-1 with brain stem glioma, caufe au lait spots and multiple swelling in the body. He is hypothyroid on thyroxine supplement.

10x7 cm swelling in the anterior abdominal wall involving right hypochondrium, right lumbar and extending into right iliac region. Purely parietal non tender warm immobile with variable consistency.

MRI of the abdomen revealed a large well circumscribed lobulated solid cystic lesion in the right lumbar region in the superficial inter muscular plane (anterior abdominal wall) measuring 6.8x8.2x9.8cm. The mass is heterogeneously isointense on T1W with few areas of hyperintensity and heterogeneously hyperintense on T2W. It demonstrated heterogenous post contrast enhancement with peripheral enhancing soft tissue rim and non-enhancing areas likely representing areas of necrosis. No evidence of infiltration into the parietal peritoneum. Core needle biopsy showed spindle cell neoplasm.

Wide excision with skin, muscles up to the parietal peritoneum was done. The anterior abdominal wall was strengthened with polypropylene mesh and wound closed by creation of V-Y flap.

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Histopathological examination showed dermis with an infiltrating neoplasm composed of spindle cells with moderate cytoplasm and elongated nuclei in bundles and sheets. Focally the spindle cells show marked atypia with increased mitosis(>20/10HPF). Large areas of necrosis seen with ectactic vessels. Hypo and hypercellular areas with macrocystic changes. Periphery shows collections of small mature lymphocytes and muscle bundles.

2.1. Immune histochemistry

Cytokeratin (AE1/AE 3) negative, bcl2 negative, S100 negative, Ckit negative, SMA negative, CD34 negative, TLE1 negative, Desmin negative, Myogenin negative.

A diagnosis of Spindle cell sarcoma -high grade compatible with Malignant Peripheral nerve sheath tumor was made.

CT thorax, FNAC was done for other swellings in the neck, axilla and a lesion in retroperitoneum which did not suggest any malignant change.

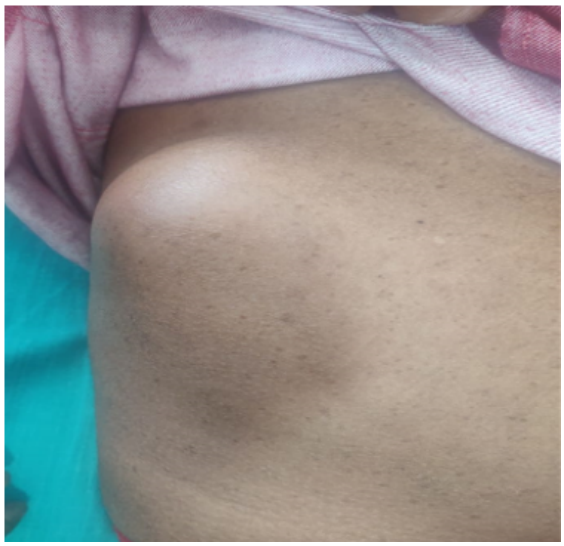

Fig. 1: Picture showing swelling in the anterior abdominal wall

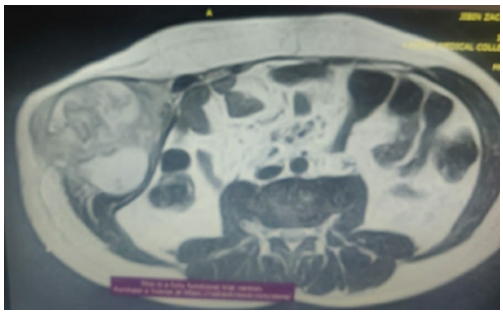

Fig. 2: MRI showing the tumor

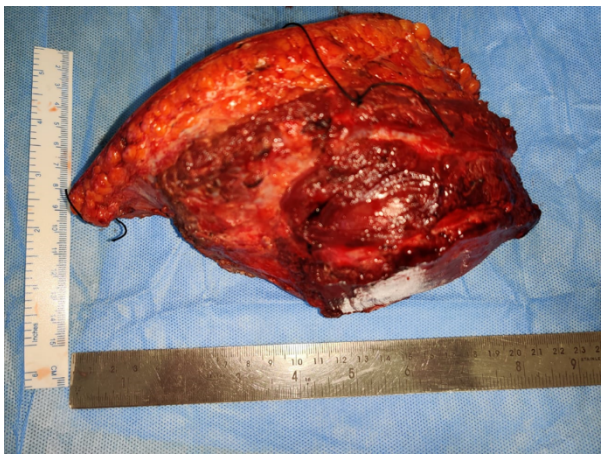

Fig. 3: Excised specimen

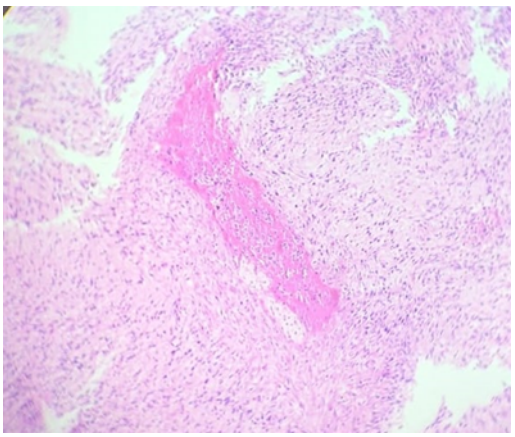

Fig. 4: Histopathology slide showing central necrosis

3. Discussion

MPNSTs previously named as malignant schwannomas as it develops from Schwann cells. In 1978, WHO grouped malignant schwannomas, malignant nerve sheath tumors, neurosarcoma and neurofibrosarcomas as MPNSTs.[1]

The diagnosis of MPNSTs requires specific histologic features. The features helping in differentiating MPNST from neurofibromas: nuclear atypia, hypercellularity and mitotic activity.[2] Even though both neurofibromas and MPNSTs comprise spindle cells the neurofibromas have more collagen fibers than MPNSTs, which serves as another point of distinction between them.[3] A 5-year survival rate for patients with MPNST without NF1 is as high as 50% but it drops to as low as 10% for MPNST patients with NF1.

S100 protein has been reported as an important single marker for diagnosing MPNST. S100 protein is often negative in high-grade MPNST.[4] S100 protein is weakly positive in 50% of MPNSTs, but it shows diffuse and a strong positive reaction in neurofibromas. Therefore, when S100 protein is diffuse and demonstrates a strong reaction,

MPNST is excluded.[2] In our case S100 was negative

Complete surgical resection of MPNSTs is required for cure. Local and distant recurrence are seen even with complete surgical resection. Radiation and chemotherapy are used as adjuvant in treatment of tumors.[4,5] Our patient underwent radiotherapy due to high grade nature of the tumor and to prevent local recurrence. This case is reported due to its location which is anterior abdominal wall, a rare one.

4. Source of Funding

None.

5. Conflict of Interest

The authors declare no conflict of interest.

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