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MALIGNANT TRITON TUMOR IN THE LEFT UPPER ARM OF A 22 YEARS OLD MALE

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ABSTRACT

Malignant peripheral nerve sheath tumors (MPNST) constitute a heterogeneous group of malignant tumors. Malignant triton tumor (MTT) is a rare variant of MPNST with rhabdomyo-sarcomatous differentiation. Among the two types, more prevalence is found for MTT associated with neurofibromatosis type 1 (NF1) with male predominance. In patients without NF1, female predominance with tumors commonly located on the trunk has reported. In this article, we present a case of MTT in the left upper arm of a 22- year- old male. A complete surgical resection and adjuvant radiotherapy were done and no evidence of recurrence found during the follow-up for last three years.

INTRODUCTION

Malignant peripheral nerve sheath tumors (MPNST) constitute a heterogeneous group of malignant tumors. About 5-10% of soft tissue sarcomas are comprised of MPNST. Malignant triton tumor (MTT) is a rare variant constituting 5% of MPNST with rhabdomyo-sarcomatous differentiation [1]. They usually arise from Schwann cell or nearby cells with perineural differentiation. MTT are usually of two types, sporadic and neurofibromatosis type 1 (NF1) associated form. Among the reported cases, about two-thirds of MTT arise in patients are associated with NF1 [2]. We present a case of MTT in the left upper arm of a 22- year- old male.

CASE REPORT

A 24-years- old male working as goldsmith presented with swelling below left elbow. The small swelling was first noticed 8 months back and a rapid increase in size was found since the past two months. Ulceration and bleeding occurred during last one month associated with mild restriction of elbow joint movement. No history of pain, fever or trauma or any other swellings or pigmented lesions in the same limb or rest of the body. He was on symptomatic treatment for the same. His Karnofsky performance status was 90%. There was no pallor, icterus, cyanosis, clubbing, lymphadenopathy or edema and vital signs were stable. Local examination found attitude of the limb was normal. There was a swelling of approximate size 15x10x3cm over upper medial aspect of left forearm, just below the elbow joint. It was a hemispherical swelling with central ulceration and bleeding. The surface of the swelling was irregular with two nodules, and well defined margin with the surrounding skin normal. There was no dilated vein, visible pulsations, distal odema, muscle wasting and other swelling in that limb. On palpation, there was no local rise of temperature or tenderness and the hemispherical swelling was of 14x10x4cm in dimension which was 12 cm proximal to wrist joint and 5 cm distal to elbow crease (figure 1A). The surface of the swelling was irregular, nodular with ill defined edges. The swelling was non-fluctuent, non-

transilluminent, irreducible, non-pulsatile and had variable consistency. Skin involvement was in the form of ulceration, with protruding granulation tissue from the floor of the ulcer which bleeds on touch. Base of the ulcer was indurated. The swelling becomes immobile when the wrist flexed against resistance. There was no regional lymphadenopathy. Movements of the elbow joint were normal except for the terminal 10 degrees of flexion. There was no distal neurovascular deficit. Systemic examination found normal. Routine blood investigations and chest X-ray were normal. Magnetic resonance imaging (MRI) scan indicates heterogeneous mass lesion involving the medial compartment of elbow with infiltration of the skin and subcutaneous tissue and adjacent muscles (figure 2). Provisional diagnosis of soft tissue sarcoma (T2b N0 M0) was made. Surgical tumor resection was done with wide local excision of the tumor with 1-2 cm margin clearance (figure 1B), preserving the ulnar nerve followed by reconstruction with ipsilateral Lattisimus dorsi flap. Histopathology of the core biopsy showed malignant spindle cell neoplasm with extensive rhabdomyoblastic differentiation (figure 3). Further, immunohistopathology showed strong positivity of desmin, focal positivity S-100 and SMA, Mib-1 labeling index > 85% and negative for CD34 and CK. Diagnosis was made as MPNST with rhabdomyoblastic differentiation ie. MTT. Adjuvant radiotherapy to the tumor bed was given. During the follow-up for last three years no evidence of recurrence found.

Fig 1. A) Swelling over upper medial aspect of left forearm and B) Surgically removed tumor

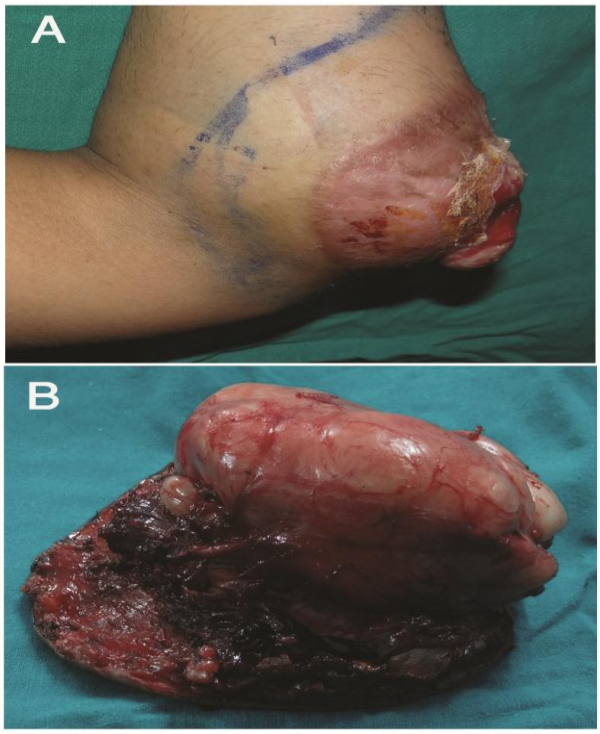


Fig 2. Magnetic resonance imaging of heterogeneous mass lesion involving the medial compartment of elbow

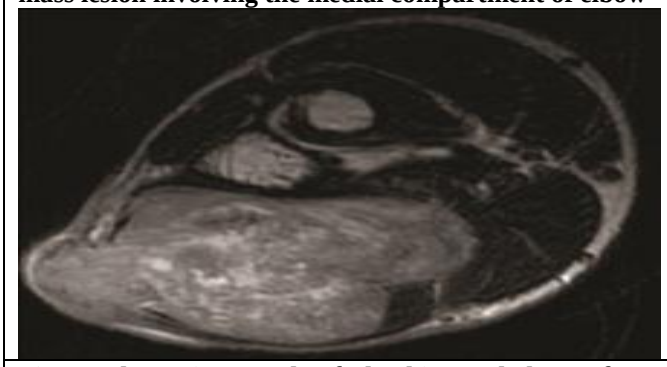
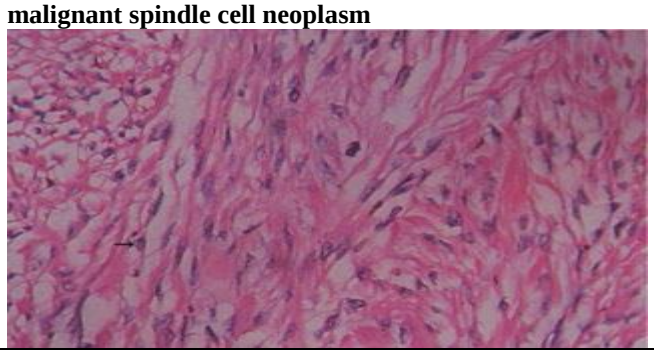


Fig 3. Photomicrograph of the histopathology of core biopsy specimen (H&E, x20). Arrow head indicates malignant spindle cell neoplasm



DISCUSSION

MTT is relatively rare soft tissue sarcomas. The unusual name "triton" was first suggested by Woodruff *et al*, with reference to the observation of supernumerary limbs containing bone and muscle growing the backs of triton salamanders, after the implantation of the sciatic nerve into the soft tissues of the back. About 5% of all MPNSTs are constituted by MTT. The most commonly found locations are on head, neck, extremities and trunk. The rare sites are buttock, mediastinum and retroperitoneum. In our patient, the site was left upper arm.

According to Woodruff et al. about 57% of MTT patient had NF1 [2]. A male (20 - 30 years of age) predominance was noticed in MTT patients with NF1 and presence of tumors most commonly found in the head and neck region. In patients without NF1, female (>50 years of age predominance [3] with tumors commonly located on the trunk [4, 5]. In our case, the gender was a male with the tumor located on trunk.

Histopathological criteria to diagnose the MTT suggested that neoplasm is connected to peripheral nerves or occurs in patients with NF1 or most of the neoplasm consists of Schwann cells or rhabdomyoblasts [6]. The biopsy specimen from our patient meets the criteria of neoplasm contains rhabdomyoblasts, which was further confirmed by IHC. Findings in IHC, such as presence of

strong positivity of desmin, focal positivity S-100 and SMA, Mib-1 labeling index > 85% and negative for CD34 that made the diagnosis as MTT. Further evaluation by genetic analysis could not do in this case as we have no infrastucture

The radiologic features obtained in the MRI are also helpful for the diagnosis of MTT. Since the same features may also be seen in them, MRI did not find as reliable to differentiate MTT from other benign nerve sheath tumors [7, 8]. Several studies demonstrated that fluorodeoxyglucose- Positron Emission Tomography was sensitive and specific tool for distinguishing MPNST including MTT from benign peripheral nerve sheath tumors. The MRI examination of this case showed heterogeneous mass lesion involving the medial compartment of elbow with infiltration of the skin and subcutaneous tissue and adjacent muscles.

The survival rates of patients with MTT were found to be relatively lower than those of patients with MPNST. The reported crude 2 and 5 year survival rates of patients with MTT are 15% and 11%, versus 57% and 39% for MPNST, respectively [9,10]. Complete surgical resection with a wide margin and adjuvant radiotherapy are generally accepted for the treatment of MTT. Systemic chemotherapy was excluded in our patient as there was no evidence of metastatic disease.

This case report concluded that early diagnosis of MTT with complete surgical resection and adjuvant radiotherapy will be beneficial for the better prognosis.

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DECLARATION OF INTEREST STATEMENT

The authors declare that they have no significant competing financial, professional or personal interests that might have influenced the performance or presentation of the work described in this manuscript.

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