CASE REPORT

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Sporadic Malignant Triton Tumor of Shoulder: a Case Report

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ABSTRACT

Background: Malignant triton tumors (MTT) are subtype of malignant peripheral nerve sheath tumor (MPNST) which develop from Schwan cells of peripheral nerves or within neurofibromas, and shows rhabdomyoblastic differentiation. It is a rare soft tissue tumor with poor prognosis. **Objective:** We report a case of Malignant Triton Tumor (MTT) arising in the right shoulder in a 46 year old male patient presented to our Musculoskeletal Oncology Clinic at Royal Rehabilitation center at King Hussein Medical Center during June 2018. **Case presentation:** The patient was complaining of an 8 months long progressive right shoulder pain and swelling at the posterior lateral area of the shoulder. As accurate diagnosis is crucial in such case, investigations that included x-rays and magnetic resonance imaging (MRI) demonstrated an soft tissue tumor involving the right shoulder area leading to the differential diagnosis of aggressive soft tissue tumor which laid down the plan of an open incisional biopsy to be reported histopathological as a case of Malignant Triton Tumor which is a very rare and aggressive sarcoma originates from the peripheral nerve sheaths as it is subtype of malignant peripheral nerve sheath tumors after which excision of the entire tumor with safety margin was performed and referred for adjuvant chemotherapy. **Conclusion:** The treatment of choice is radical tumor excision with wide margins followed by chemotherapy and /or radiotherapy to improve the 5 years survival rates.

Keywords: Malignant Triton Tumor, peripheral nerve sheath tumors.

1. BACKGROUND

Malignant triton tumors (MTT) are subtype of malignant peripheral nerve sheath tumor (MPNST) which develop from Schwan cells of peripheral nerves or within neurofibromas, and shows rhabdomyoblastic differentiation. It is a rare soft tissue tumor with poor prognosis. It can be found in association with neurofibromatosis type 1 (NF-1) or sporadically, accounting for 5-10% of soft tissue sarcoma (1-3). MPNST occurs in about 2-5% of patients with NF-1 (4-5).

MTT was first introduced by Masson and Martin in 1932 as a very rare tumor and has more aggressive clinical course and worse prognosis than MPNST without rhabdomyoblastic differentiation (6, 7). It was found the 5 year survival rate is 5-15% (9, 10) compared to 50-60% for MPNST (8); however, studies suggest that complete tumor excision with adjuvant chemotherapy improve survival rate (2-4). MTT are developed from Schwan cells of peripheral nerves, usually close to the trunk, in the head and neck or in the upper and lower limbs (12) and it is usually seen in patients younger than 35 years of age.

2. OBJECTIVE

We report a case of Malignant Triton Tumor (MTT) arising in the right shoulder in a 46 year old male patient presented to our Musculoskeletal Oncology Clinic at Royal Rehabilitation center at King Hussein Medical Center during June 2018.

3. CASE PRESENTATION

A46 year old male patient was presented to our Musculoskeletal Oncology Clinic at Royal Rehabilitation Center at King Hussein Medical Center complaining of progressive right shoulder pain over 8 months duration, aggravated by any movement and associated with a swelling which increase in size with time. There was no history of trauma to the same area, and no positive

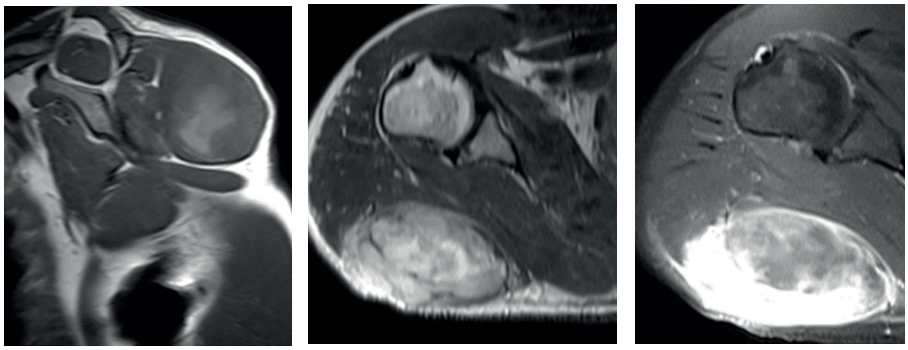

Figure 1 (a, b, c). MRI images demonstrate mixed signal intensity on T1 and high signal intensity on T2 with heterogeneous enhancement in fat sat post contrast imaging

family history for neurofibromatosis1 nor 2. On physical examination, we found a soft tissue mass in the posterior aspect of right shoulder, tender to deep palpation and firm. As part of work up, x- ray examination was done and showed increase soft tissue intensity.

For further evaluation, magnetic resonance imaging with intravenous contrast preform and reveled a relatively well defined lobulated superficial located large soft tissue tumor about 7 x 6 x 5 cm in the posterior aspect of the right shoulder with mixed signal intensity on T1 and high signal intensity on T2 with heterogeneous enhancement in fat sat post contrast imaging (Figures 1a, 1b, 1, c).

Based on the above findings, we proceed with Open inscional biopsy under general anesthesia which revealed spindle cells and prominent mitotic activity with rhabdomyoblastic differentiation which is characteristic for MTT. To finish work up, chest abdomen pelvic CT scan was done and did not show any evidence of distant metastasis.

Later, patient underwent excisional tumor resection with safety margin with nearest margin 30 mm, microscopic examination revealed multi lobulated tumor composed predominantly of hypercellular areas containing uniform spindle cells arranged in intersecting fascicles with perivascular accentuation of the tumor cells , extensive areas of geographic necrosis and prominent mitotic activity noted (18MF/10HPF) with areas of rhabdomyoblastic differentiation evidenced by Desmin and Caldesmin immunostain and Immunohistochemical studies showed CK,PGP9.5,cd68,S-100 and SMA positive (Figures 2a, 2b, 2c)..

The patient was referred to the Hematoncology department and started adjuvant chemotherapy. He continued to follow up in our clinic with no evidence of recurrence of the tumor nor distant metastasis after 5 years of his treatment.

4. DISCUSSION

MTT is a very rare and aggressive sarcoma originates from the peripheral nerve sheaths so it is subtype of MPNSTs develop from Schwan cells of peripheral nerves or within neurofibromas (6), and shows rhabdomyoblastic differentiation, so called mosaic tumor because it has two components muscular and neurogenic. Masson and Martin in 1932 showed the malignancy of MTT with Von Recklinghausen disease (NF-1) (5, 6).

MTTs found in association with NF 1 in two thirds of the cases or sporadically, the MTTs with NF-1 are mainly found in

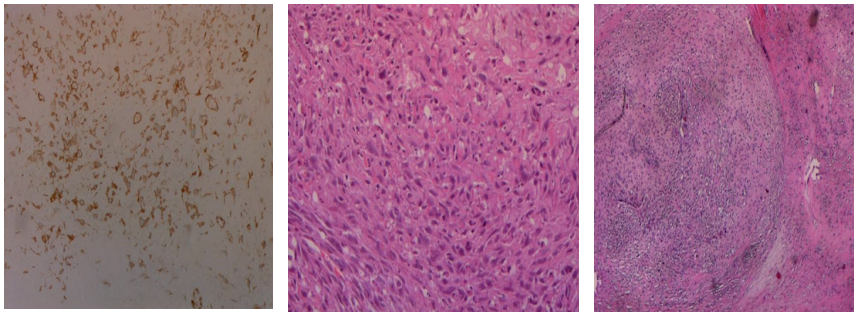

Figure 2 (a, b, c). Histopathology slides showing the rhabdomyobalstic differenitiation and Desmin and Caldesmin immunostain

males of young age groups while the sporadic forms are found in females of older age groups (9, 10). Our reported case is sporadic and our patient age within the average age groups but not meet the gender predominance (14, 15).

The clinical presentation of the MTTs is painless or painful soft tissue mass firm increase in size with time arise in different sites including head and neck , trunk, upper and lower limbs, mediastinum and retroperitoneum. Usually arise from medium to large nerves (2, 15).

The diagnosis of MTTs is made by histopathology after surgical excision and immunohistochemical studies which confirm the presence of rhabdomyoblastic differentiation by Desmin and Caldesmin immunostain and S-100 protein suggesting a nerve sheath origin which is found in our case.

MTTs have a worse prognosis than the MPNSTs as the 5 year survival rate of MTTs 5-15% in contrast to MPNSTs where is 50-60% (13). So the treatment guidelines for the MTTs to improve the survival rate is excision of the tumor with wide safety margin followed by adjuvant chemotherapy and some authors add radiotherapy (10). The location of the tumor affects the prognosis as reported in the literature as the head and neck and in the upper and lower limbs have better prognosis than tumors found in the trunk, buttock and retroperitonium (16).

5. CONCLUSION

MTTs a rare sub type tumor of MPNSTs which is very rare and aggressive sarcoma originates from the peripheral nerve sheaths with worse prognosis and more aggressive behavior and mainly histopathological diag-

nosis. The treatment of choice is radical tumor excision with wide margins followed by chemotherapy and /or radiotherapy to improve the 5 years survival rates.

- **Data Access Statement**: Data supporting this study's findings are available upon reasonable request.
- **Ethics Statement**: This study was conducted in accordance with ethical standards and approved by the relevant Institutional Review Board.
- **Author Contributions Statement**: The authors contributed equally to the research, writing, and review of this manuscript. Final proofreading was made by the first author.
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