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Case Report

The US/ MRI/PET-CT Imaging Findings of a Malignant Triton Tumor of the Shoulder in a NF-1 (Von Recklinghausen Disease) Patient

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Abstract

We report a malignant triton tumor case in a 48-year-old woman with neurofibromatosis-1 who presented with a rapid growth over the shoulder. She was previously diagnosed of schwannoma by a needle aspiration cytology. CT showed a soft tissue tumor.

MRI showed a T2WI high intensity tumor with central more hyper intense area. And the interruption of capsule was also observed. The diffusion weighted image (ADC map) showed a peripherally dominant restricted diffusion and the FDG-PET showed highly uptake in viable peripheral area. Histopathologically the diagnosis of MTT was made with the positive immunostaining of S-100, myogenin and high MIB-1 index (30%).

Keywords: Malignant Triton Tumor (MTT); neurofibromatosis-1; Rhabdomyoblastic differentiation; Interruption of outer capsule; Restricted diffusion

Introduction

Malignant Peripheral Nerve Sheath Tumor (MPNST) accounts for about 5-10% of all soft tissue sarcomas [1]. Patients with neurofibromatosis-1 develop sarcomas usually after 10 to 20 years and multiple MPNST occur in some cases [2]. In cases of NF-1 patients, it usually occurs at a slightly earlier age and tends to be larger than those with sporadic cases. Classically it arises as a large fusiform or eccentric mass in a major peripheral nerve. It may demonstrate (bone or cartilage, skeletal or rhabdomyoblastic (malignant triton tumor), histiocytoid, glandular differentiations. Malignant Triton Tumor (MTT) constitutes about 5% of all MPNST [3]. It is commonly seen in the head, neck, extremities and the trunk [1,4]. The extremely rare incidence of this tumor in the shoulder or axillar region has prompted us to report this case. This neoplasm was first described in 1938 by Mason and Martin who suggest that the neural elements induced the differentiation to the skeletal muscle. It can occur in sporadic form or over a setting of (NF-1). When MTT develops over NF-1, the diagnosis can be confirmed based on morphologic histological grounds supported by an immunostains such as the S-100 protein. Desmin, myogeninare are also immunochemically positive for rhabdomyoblasts focally or diffusely [4]. MTT is an essentially aggressive tumor, and the 5-year survival rate has been reported to be 5-15%. However, modern treatment such as complete surgical resection with adjuvant radio-, chemo-therapy has improved the prognosis [5].

Case Presentation

A 59-year-old woman who was diagnosed as NF-1(Von Recklinghausen disease) tenderness in her left shoulder. On ultrasound sonography, a heterogeneous low echoic mass measuring 27 x 21x 34 mm was revealed between infraspinatus and teres minor displacing the long head of the triceps. The echotexture was low with deep attenuation. And the doppler mode showed peripheral slightly high echoic signals suggesting vascurality.US guided biopsy showed a schwannoma. Immunochemically S-100 protein was diffusely positive. 3 months later, MRI revealed the solid and cystic mas measuring 74 x 36 x 42 mm with small nodules contiguous to the main tumor along the subscapular and scapular circumflex neurovascular bundle .The diffusion weighted image (DWI: b factor=1000) of shoulder tumor showed heterogeneous high signal (mean ADC=0.913 x 10-3mm2/s). Eleven months later, continuous enlargement of the tumor was observed with an interruption of outer capsule (Figure 1) on T2WI. On fat suppressive T2WI, a heterogeneous high signal tumor with central hyperintensity area was observed (Figure 2). The DWI showed peripherally dominant hyper signal. The corresponding ADC map suggested relatively high cellularity (mean ADC=1.017 x 10-3mm2/s) in the peripheral portion of the tumor. F18-Positron Emission Tomography-Computed Tomography (PET-CT) demonstrated a peripherally dominant high signal like a dougnat without distant metastasis. The maximum Standard-Uptake-Value (SUV) was 11.8.One month later, the tumor grew up to 110 x 101 x 94 mm in size, Then, total resection of the tumor was performed finally, and the pathological examination revealed that the tumor was a malignant triton tumor type of MPNST (Figure 3) .The cut surface of the tumor revealed massive central necrosis.

The Ki67 index was 30%.In MPNST, its 20 percent level was used as a discrimination point from the benign schwannoma [6-24] .Round cells with eosinophilic cytoplasm were found. The immunochistochemical stains of myogenin and desmin, were focally positive consistent with rhabdoid differentiation. And negative for AE1/3, HMB45, MelanA, CD34, bcl-2 c.

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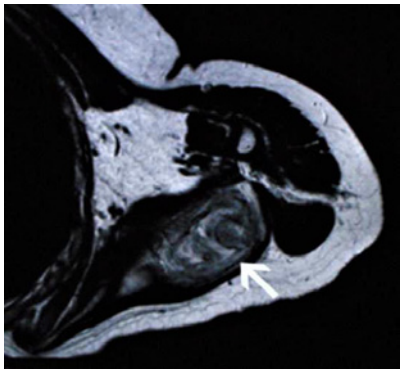
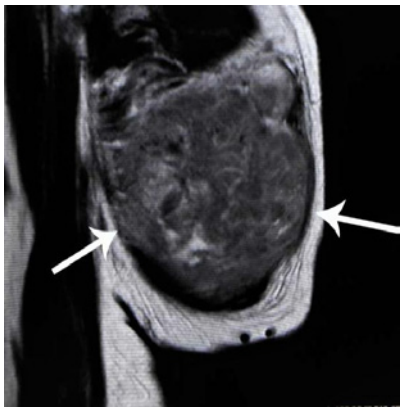


Figure 1: The axial T2-weighted MR image showed heterogeneously high–low signal intensity (arrow).



Figurre 2: On axial T2WI, rapid growing of the heterogeneously high intensity tumor was noted .There was a interruption of the outer capsule (arrows).

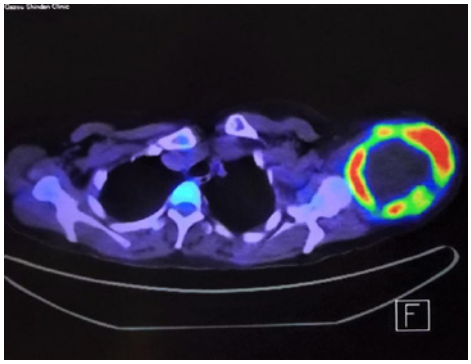


Figure 3: F18- PETCT demonstrated a ring uptake of FDG like a dougnat.

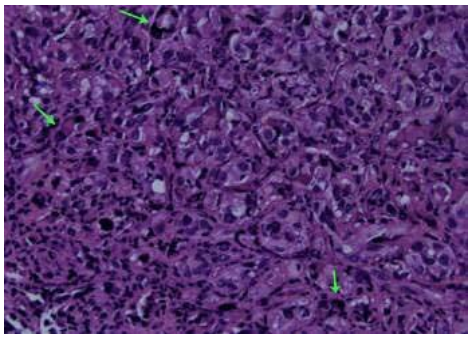


Figure 4: Round cells with eosinophilic cytoplasm morphologically consistent with rhabdoid differentiation are identified

Discussion

Schwannoma is a slow growing, well-circumscribed and encapsulated round or ovoid tumor. A spindle-shaped cells with large nuclei and with nuclear palisading arranged in interlacing bundles known as Verocay bodies. On the other hands, MPNST are composed of spindle cells. MPNST shows extensive pleomorphism, simulating a indistinct margin areas of hemorrhage and necrosis. MTT is a malignant peripheral nerve sheath tumor with rhabdomyosarcomatous differentiation. Pierre Masson firstly reported the MTT, then Woodruff and Perino, et al. identified 84 cases [12]. The age ranged from newborn to 75 years old (mean 34 years), and males and females are equally represented. They reported that its local recurrence was more than 40%, metastasis 30-60%, and a 5 year survival rate was 15-34%.Such tumors show focal positivity for S-100 protein in 50-90% of cases, suggesting a nerve sheath origin. Rhabdomyoblasts are positive for immunostains such as desmin, myogenin and myo-D1[1-4]. MTT arises in two principal forms: sporadic or in association with NF-1. Slightly more than half of the cases of MTT have been reported to occur in NF-1 patients [1,3,10]. Usually MTT with NF-1 showed marked male predominance with more predilections for younger age groups compared with the sporadic forms [17].

A 18F-FDGPET is a noninvasive whole-body imaging technique and is widely used to evaluate tumor staging and detection of recurrence, and for monitoring treatment response. Schwannoma generally shows relatively high FDG uptake [21]. The prognosis of MTT depends on the location, grade and completeness of surgical margins. In a study it was observed that MTT in association with NF-1 has a poor prognosis compared to sporadic form [10]. There is a breakpoint in 11p15, considered a region of myogenic differentiation. This gene is probably responsible for rhabdomyoblastic differentiation. Amplification of c-myc oncogene is probably responsible for its aggressive biologic behavior [17]. The infiltrative tumor border on CT or MR suggests malignancy including other high grade sarcoma such as MFH (undifferentiated pleomorphic sarcoma). But, a malignant neoplasm may have a smooth, non-infiltrating margin at times. Bone erosion may occur with both benign and malignant neoplasms, but is more irregular with malignant cases.

Preoperative radiological diagnosis is important to avoid over-treatment and unnecessary extensive procedures, but it is very difficult because diagnostic imaging findings for MTT has not been well established. The lesion was isoechoic to the muscle, with smooth and lobulated margins, apparently showing an expansive rather than an infiltrative behavior. (Power) Doppler evaluation displayed a rich vascularization of the mass. MRI showed iso signal on T1-weighted images and rather inhomogeneous high signal intensity with sporadic scattered low signal intensity areas on STIR and T2-weighted images (Figure 4). Post-contrast T1-weighted images with fat suppression demonstrated inhomogeneous enhancement.

MRI findings suggesting malignant transformation include large

size (>5cm), peripheral enhancement pattern, perilesional edema, intratumoral cystic change, and heterogeneity on T2WI [23]. Li, et al. [8] also reported that imaging features suggestive of malignancy may include larger size lack of contiguity with adjacent specific nerves, and an infiltrative margin. Intratumoral cystic change commonly occurs as a result of hemorrhage or necrosis in schwannoma, but rarely in neurofibroma [11].This feature can assist in the differentiation of neurofibroma from MPNST, in which malignant transition may result in the occurrence of necrosis or hemorrhage. The malignant transformation of the tumor from a preexisting benign neurofibroma might cause necrosis or hemorrhage of the tumor, which would be high signal on T1WI and heterogeneously low signal on MR T2WI images.

Matsumine et al. [24] analyzed 19 cases of malignant peripheral nerve sheath tumor.

In which,they found the important findings indicating the aggressiveness of MPNST was a perilesional edema.Ill-defined margins may reflect invasion of the tumor into the marginal tissue contiguous to the tumor. MPNST including MTT may occur as solitary or multiple enlarged masses associated with major nerve trunks such as the brachial plexus, sacral plexus, and sciatic nerve and may be asymptomatic or present with various sensory and motor symptoms, including projected pain or compressing and infiltrating surrounding issues and structures [25,26]. About 40–50% of benign peripheral schwannomas may have central enhancement on CT corresponding to central zone of tightly packed cellular components (Antoni A) surrounded by hypocellular myxoid material (Antoni B) corresponding to target sign on MRI T-2 weighted images (central low and peripheral high signal intensity). The central enhancement (target sign) is rarely seen in MPNST. In large tumor, there is central necrosis or degeneration, with peripheral tumor enhancement. The borders in MTT are frequently irregular and infiltrative borders invading adjacent structures or destroy adjacent bones. On imaging, the differentiation features between benign and malignant nerve sheath tumor are size greater than 5 cm, rapid growth, prominent enhancement, infiltrative margins, and marked heterogeneity with necrosis, peripherally avid uptake of F18-FDG on PET [27].

Survival of MTT is associated with complete tumor resection. The overall 5-year survival rate is affected by the patient's age, size, location and margins affecting survival. Longer survival is associated with early diagnosis (mainly due to the improved imaging), complete surgical resection and neoadjuvant therapy. Most large soft tissue tumors of the shoulder region, presenting in the elderly are malignant such as high grade (undifferentiated) sarcoma, leiomyosarcoma, fibrosarcoma, liposarcoma, or myxofibrosarcoma. The MR appearance is not tumor specific. Large volume, liquefaction and cellular components are features on MR shared by these sarcomas. One of the discriminating point of MRI from a myogenic tumor such as leiomyosarcoma was that no apparent low signal intensity component on T2-weighted image [28]. However, the differentiation from other soft tissue sarcoma such as fibrosarcoma, myxoid liposarcoma, or myxofibrosarcoma assumed to be difficult except the fact that this case occurred from the schwannoma in NF-1 patient.

Although, DWI in the diagnosis of sarcoma is not completely established, the utility of DWI was studied in some literatures [29]. In our case, DWI signal of the tumor changed from diffuse high to peripheral high signal intensity reflecting central necrosis. The corresponding ADC map suggested relatively high cellularity (mean ADC=0.913-1.017 x 10-3mm2/s). Regarding MTT, the ADC value have not been reported before, so its high cellular nature was disclosed with this study.

As for FDG-PET, the SUV max of the tumor was reported as 11.8 which reflects MPNST including MTT [30]. Broski SM et al. described that all lesions with SUV max>8 were malignant in their study of both benign and malignant peripheral nerve sheath tumor and stated that SUV max cutoff of 6.1 yielded 90.0% sensitivity and 78.3% specificity for MPNSTs [31]. However, benign schwannoma showed high uptake of FDG at times [32,33].

Conclusion

We report a Malignant Triton Tumor (MTT) case in a 48-year-old woman with NF-1 who presented with a rapid growth. MRI demonstrated a T2WI heterogeneously high intensity tumor with interruption of capsule.

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