Case Report

A Case of Retroperitoneal Malignant Triton Tumor in a Nigerian Boy

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Abstract

Malignant peripheral nerve sheath tumor is a rare tumor occurring in 5%–10% of all malignant soft tissues sarcomas and triton tumor arising from neurofibromatosis type 1 (NF-1) is even rarer with associated high rate of mortality. No case of triton tumor has been reported in Nigeria to the best of our knowledge. We seek to report a case of lately detected retroperitoneal triton tumor presenting in a 12-year-old Nigerian child who was brought with bilateral lower limb weaknesses, weight loss, and a right lumbar mass. There were multiple café au lait spots on the body. Abdominal computerized tomographic scan revealed a huge right retroperitoneal mass crossing the midline, compressing adjacent structures with multilevel intraspinal extensions. Core needle biopsy performed and both histology and immunohistochemical studies confirmed the diagnosis, but patient demised in the course of care. The aim is to heighten suspicion of this extremely very rare malignant tumor in children with NF-1.

Keywords: *Nerve sheath tumors, neurofibromatosis, retroperitoneal tumors*

Introduction

Malignant peripheral nerve sheath tumors (MPNSTs) are rare tumors and constitute about 5%–10% of all soft tissue sarcomas. Malignant triton tumor (MTT), a variant of MPNST with rhabdomyoblastic differentiation is extremely rare with high rate of mortality and are found in only 5% of all MPNST.[1-3] It was first described by Masson[4] as rhabdomyosarcomas in patients with neurofibromatosis type 1 (NF-1). MTT has been found to occur in patients with NF-1 in slightly more than 50% of the cases, whereas the rest are sporadic.[1-3,5] It runs an aggressive cause with high mortality, especially those on a background of NF-1.[3]

Common sites of MTT include head, neck, extremities, and trunk whereas those occurring in the retroperitoneum, viscera, mediastinum, and intracranium are quite rare.[6] Diagnosis is confirmed by histology with immunohistochemical studies for desmin, vimentin, actin, myoglobulin, and S-100 protein.[7,8] The main stay of treatment is complete excision where possible in addition to adjuvant radiotherapy and chemotherapy.[3,5] There are <10 reported cases of retroperitoneal MTT in literature out of which only one was in a child. None has been reported in Nigeria to the best of our knowledge.[7] The rarity of this condition and its aggressive biological behavior and mortality has prompted us to report this case.

Case Report

A 12-year-old boy was brought to the children emergency room of Nnamdi Azikiwe University Teaching Hospital, Nnewi, Nigeria with the complaints of bilateral thigh pains of 6 weeks and inability to walk of 2 weeks duration. There was associated the loss of sensations from the hips down to the feet, significant weight loss, and loss of urinary and anal sphincteric functions. There was no cough and both parents and patient never observed rashes nor pigmented patches on the skin.

When examined, he was found to be chronically ill-looking, severely emaciated with multiple café au lait spots on the trunk and limbs [Figure 1] and subcutaneous nodules on the upper limbs. He had a lumbar swelling

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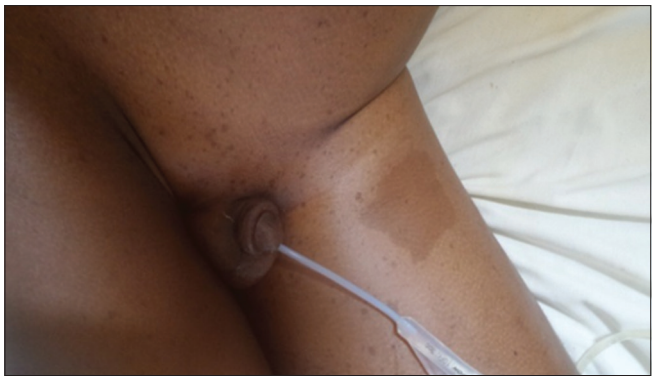

Figure 1: Café au lait spots on the skin

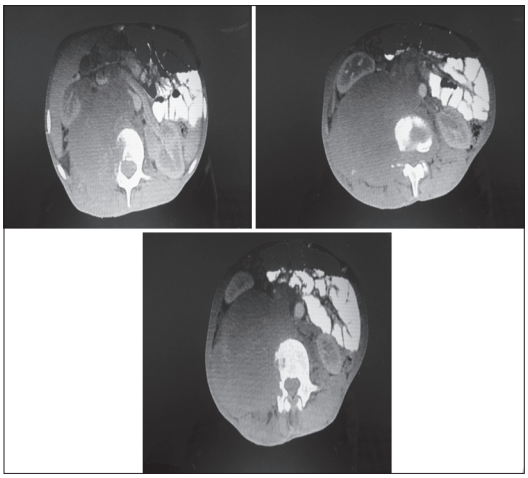

Figure 2: Abdominal computerized tomographic scan

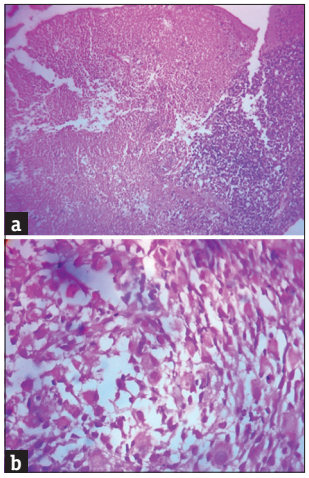

Figure 3: (a) Photomicrograph of H and E. Tumor composed of alternating hypercellular and hypocellular myxoid area (b) malignant cells having slightly pleomorphic hyperchromatic nuclei with scanty cytoplasm. There were areas showing rhabdomyoblastic differentiation with foci of coagulative necrosis

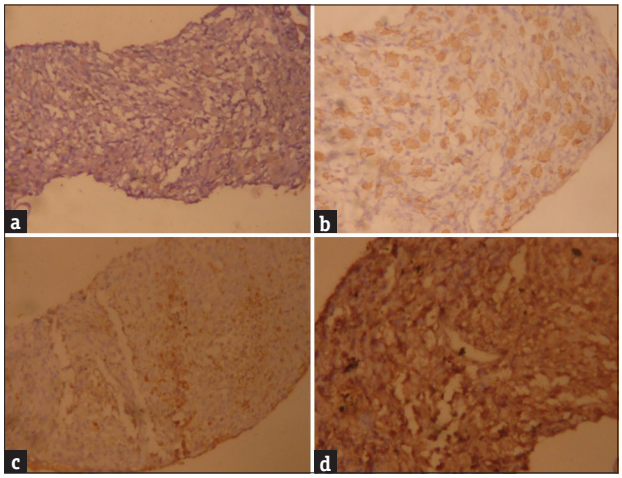

Figure 4: Immunohistochemical stains. (a) Epithelial membrane antigen EMA (negative) suggesting that it is not epithelial in origin. (b) Desmin (positive) suggesting rhabdomyosarcoma and other tumours with myoid differentiation. (c) Myogenin (positive), usually expressed in rhabdomysarcomas. (d) S-100 protein (positive) suggests neural origin

that was poorly delineated, tender and hard. Power on both limbs was Grade 0 and there was a right lumbar scoliosis. The full blood counts and serum electrolytes with blood urea and creatinine assays were all within normal ranges.

Abdominal computerized tomography with contrast was performed and demonstrated a huge right lumbar and paraspinal mass extending from the level of L1 vertebrae down to the pelvis measuring 17.0 cm by 11.4 cm and 10.6 cm in widest cranio-caudal, antero-posterior and transverse dimensions, respectively, compressing the inferior vena cava and displacing the abdominal aorta to the left with stretching of the left renal pedicle. There was intraspinal involvement with widening of the right neural foramina of L1/L2, L2/L3, and L3/L4 with lytic destruction of T11-L3 vertebra [Figure 2]. There were nodular lesions of varying sizes in both lungs.

Considering the inoperable nature of the patient due to his poor clinical status, a conservative care was adopted while an ultrasound guided core needle biopsy was carried out in the radiology suite. He was placed on analgesics, subcutaneous heparin, and prophylactic antibiotics. The histological report read malignant peripheral nerve tumor composed of alternating hypercellular and hypocellular myxoid area. These malignant cells have slightly pleomorphic hyperchromatic nuclei with scanty cytoplasm. There were areas showing rhabdomyoblastic differentiation with foci of coagulative necrosis, on immunohistochemical studies, the slides stained positive for desmin, myogenin, S-100 and negative for epithelial membrane antigen (EMA) [Figures 3 and 4]. He developed breathlessness and died 31 days after admission.

Discussion

MTTs are extremely rare tumors arising from the peripheral nerve sheaths and they constitute a variant of MPNST with rhabdomyoblastic differentiation.[4] There are <170 cases reported worldwide and most are case reports and case series with 9 cases as the only single largest report in literature.[6,9,10] Among the scanty reports of peripheral nerve sheath tumors from Sub-Saharan Africa, none was specified as MTTs.[11-13] Retroperitoneal triton tumors are even rarer and <10 have been reported in literature so far out of which only one was in a child.[6,7]

Woodruff *et al*.[14] first used the word triton tumor to describe these variants of MPNSTs and developed a criteria for its diagnosis which includes: (1) Tumor arising from a peripheral nerve in a patient with NF-1 or location typical of MPNST, or represents a metastases from such a tumor, (2) tumors demonstrating growth characteristics of Schwann cells, (3) tumors contain rhabdomyoblasts that appear to arise from within the body of the peripheral nerve tumor and not extension from extrinsic rhabdomyosarcoma. Daimaru *et al*.[15] further included tumors in patients without NF-1 that are microscopically compatible with malignant schwannoma and contain focal rhabdomyoblas-ts or patients with tumors consisting of rhabdomyoblastic differentiation with focal Schwann cell elements within a nerve or in patients with NF-1. This captures both the sporadic form and those associated with NF-1.

The index patient was a 12-year-old male who presented with a short history of bilateral thigh pains, progressive weakness of both lower limbs and a large abdominal mass. This appears to be the usual progression of retroperitoneal triton tumors as in previous reports.[11,16] Even though, the parents and the referring doctors did not observe hyperpigmented patches or nodules on the skin, these were found on examinations as shown in Figure 1. MTT occurring in patients with NF-1 is more common, usually more than 50% of the cases; however, there are some sporadic cases not associated with NF-1.[1,6-8,10,15,17] Some have been suggested to arise following irradiations.[6] Those associated with NF-1 have been found to occur more in younger male patients and are more aggressive as seen in the index case.[6]

Ducatman and Scheithauer[18] however reported a case of retroperitoneal MTT with NF-1 in a 12-year-old female, that again was the only report of MTT occurring in retroperitoneal space in a child as previously reported.[7,18] Retroperitoneal MTT is a locally invasive tumor but also metastasizes to the lungs.[7,11,16,19] The index case had metastasis to the lungs as picked up by the computerized tomographic scan. Imaging with computerized tomographic scan and magnetic resonance imaging could be elucidatory in terms of tumor location and extension but diagnosis is confirmed by tissue histology and immunohistochemical staining for S-100 protein, desmin, actin.[7,17] Our patient had an abdominal computerized tomographic scan as shown in Figure 2. The findings of a huge retroperitoneal mass with extension into the spine, adherence to the inferior vena cava and aorta with stretching of the left renal pedicle coupled with the unstable clinical status precluded the option of operative intervention at the time. We then carried out a core needle biopsy which was strongly suggestive of triton tumor on routine hematoxylin and eosin stains and confirmed with positive immunohistochemical stains for S-100, myogenin and desmin and negative to EMA [Figures 3 and 4]. Negativity to EMA suggests that it is not epithelial in origin.[19] Core needle biopsy has been strongly suggested to give a good tissue yield for MTT.[20]

Even though there is no standardized, guideline for treatment of MTT, Surgical extirpation has been found to be the mainstay of treatment.[1,8,11,17] Adjuvant chemotherapy and radiotherapy have been used with doubtful effects.[17] Despite treatments, MTT has grave outcomes with high mortalities worse with the retroperitoneal lesions, most cases die within months of diagnosis as a result of late detection, local invasion, and metastasis.[1,6,11,20] This has prompted some authors to suggested that MTT be described as a different entity from MPNST because of its more aggressive tumor characteristic and attendant dismal outcome.[8]

Conclusion

MTT of the retroperitoneal space is an extremely rare tumor of the nerve sheaths and none has been reported in Nigeria. It is often associated with NF-1. Perhaps, a more routine and close surveillance on these patients could aid early detection and treatment.

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Conflicts of interest

There are no conflicts of interest.

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