Case Report

Management of Metastatic Paratesticular Tumour in a Resource-Poor Setting

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Abstract

Paratesticular tumours are tumours that arise from the testicular tunics, spermatic cord, epididymis, or vestigial remnants. The tumours are rare and account for approximately 5% of intrascrotal neoplasms. About 75% of these tumours arise from the spermatic cord. Paratesticular tumours most commonly manifest as painless scrotal masses. Alternatively, the tumour may be incidentally noticed when a scrotal ultrasound scan is done for another intrascrotal pathology such as hydrocele, inguinoscrotal hernia, epididymo-orchitis, or suspected testicular tumour. We present a case of metastatic paratesticular tumour in a 21-year-old Nigerian male, who presented at the University of Nigeria Teaching Hospital, Ituku-Ozalla, Enugu, in September 2018 at the age of 19 years with a painless right hemiscrotal mass. The patient was clinically evaluated with scrotal ultrasonography, testicular tumour markers, and liver function test. Biopsy specimen obtained was ignorantly discarded by the patient who was subsequently lost to follow-up. Histologic diagnosis of mesenchymal tumour (myxoid liposarcoma) was made two years after his initial presentation when he developed both inguinal and retroperitoneal lymph node metastasis at the age of 21 years. He was evaluated as clinical stage IV disease and then commenced on chemotherapy after baseline investigations. Our objective of presenting this report is to highlight the effect of delayed diagnosis in the management outcome, challenges in the provision of resources in low- and middle-income countries, and to emphasise the rarity of the tumour in our subregion.

Keywords: Delayed diagnosis, lymphadenopathy, metastasis, Nigeria, paratesticular tumour

Introduction

Paratesticular tumours are tumours which arise from the epididymis, spermatic cord and its coverings, and the testicular tunics. They are rare and account for approximately 5% of intrascrotal neoplasms and roughly 75% of which arise from the spermatic cord.[1] These tumours affect patients of all ages and there is no racial discrimination. Paratesticular tumours are histologically classified into benign and malignant tumours. Most of the tumours are benign and the most common is adenomatoid type.[2] Adenomatoid tumour is of mesothelial origin and 75% of it arises from the epididymis. The tumour occurs in men between 30 and 40 years with a mean age of 36 years.[2] Other common benign variants include lipoma and leiomyoma, while fibroma, hemangioma, papillary mesothelioma, and paraganglioma are less common. The most common malignant paratesticular tumour is liposarcoma and other common variants are leiomyosarcoma and rhabdomyosarcoma. The less common malignant types include malignant fibrous histiocytoma, malignant lymphoma, and metastatic carcinoma. The most common malignant tumour in infants and children is rhabdomyosarcoma.[3] Paratesticular rhabdomyosarcoma arises from testicular tunics, epididymis, or spermatic cord. This accounts for about 40% of paratesticular tumours and represents 7%–10% of genitourinary rhabdomyosarcoma. It has a bimodal peak age distribution at one to five years and around 16 years, respectively. Malignant mesenchymal tumours are soft-tissue tumours which are derived from

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mesenchyme. Mesenchymal stem cells can differentiate into blood and lymph vessels, muscles, and bones. The histological types of paratesticular tumours have varied characteristics and subtypes. Immunohistochemical staining of adenomatoid tumour cells shows mesothelial markers such as calretinin and Wilms tumour 1 (WT1). Calretinin is a calcium-binding protein originally found in neurons. It is overexpressed in most types of malignant mesothelioma. Pathologists use calretinin as a selective marker to diagnose mesothelioma,[4] while researchers are testing the protein as a target for cancer therapy. WT1 protein has been noted to be predominantly expressed in the cytoplasm of malignant adenomatoid tumours.[4] Alfa-fetoprotein is the relevant tumour marker in prepubertal testis tumours and elevated alfa-fetoprotein (AFP) is associated with yolk sac tumours. Other markers for testicular tumours such as beta-human chorionic gonadotrophins (bHCG) and lactate dehydrogenase (LDH) have not been found to be very useful in the diagnosis or monitoring of the recurrence of paratesticular tumours. However, increased levels of LDH reflect tumour burden, growth, and cellular proliferation.

Staging of paratesticular tumours is similar to tumour–node–metastasis staging of soft-tissue sarcomas.

The mode of spread may be locally to the scrotal wall or through lymphatics to the inguinal, pelvic, or retroperitoneal lymph nodes. Lymphatic spread is the most common mode of metastasis. Haematogenous spread to the lungs and liver can occur but less common mode of metastasis.

The history of painless swelling or mass distinct from the testis may be the only complaint if the patient presents early. Other symptoms could be related to the stage of the disease. Groin or inguinoscrotal swelling associated with cough, chest pain, or haemoptysis suggest lung metastasis. Findings on physical examination depend on the stage of the disease. Firm, cystic, or hard mass or nodule distinct from the testis may be the only significant finding in early disease. Lymphadenopathy, hepatomegaly, and abnormal chest findings may point to metastatic disease.

The paratesticular tumours must be differentiated from testicular tumours, inguinoscrotal hernias, epididymo-orchitis, testicular torsion, varicocele, spermatocele, etc.

Ultrasound scan of the scrotum and its contents can differentiate between testicular and paratesticular mass(es) with a sensitivity of over 95%.[5] Ultrasound scan is a standard and very useful imaging modality in resource-poor countries.

In situations where the diagnosis is equivocal, Magnetic Resonance Imaging (MRI) can be used to characterise the lesion further, especially in defining the location of the tumour and its relationship with paratesticular structures.[6,7] On the other hand, computed tomography (CT) of the abdomen and pelvis with or without the thorax is typically the imaging modality of choice for staging the tumour.[8]

The treatment of paratesticular tumours is surgical and this involves radical inguinal orchiectomy with high ligation of the spermatic cord at the internal inguinal ring. Patients younger than 10 years old, who have evidence of positive nodes on CT scan should undergo staging ipsilateral retroperitoneal lymph node dissection (RPLND); if all tumour is removed, the patients are classified as stage II.[9] A nerve-sparing ipsilateral technique is recommended by Children's Oncology Group protocols. Patients who have grossly positive lymph nodes on staging CT scan can be managed by confirmatory biopsy and are considered stage III. Children older than 10 years should always undergo staging ipsilateral RPLND regardless of negative CT findings.[9] Adjuvant chemotherapy and radiation therapy are employed in patient management.

Approval was obtained from the Research and Ethics Committee of University of Nigeria Teaching Hospital and written consent was obtained from the patient to use his clinical details for this report which we deem necessary due to its rarity in our subregion.

Case Report

Our patient was a male teenager who presented at the University of Nigeria Teaching Hospital, Ituku Ozalla, Enugu, at the age of 19 years with a two-month history of right hemiscrotal swelling. The swelling was insidious in onset, painless, and progressively increased in size. There was no history of ulcer on the scrotal skin of the ipsilateral side. There was no history of trauma to the ipsilateral hemiscrotum and no history of urethral discharge or swelling of the contralateral hemiscrotum. There was no prior history of testicular swelling or undescended testis.

The patient did not have fever, cough, chest pain, weight loss, or jaundice. There was no prior history of testicular surgery or irradiation [Table 1].

Clinical examination on presentation revealed a teenager in no obvious form of distress. He was afebrile, not pale, anicteric, no peripheral lymphadenopathy, and no pedal edema. His temperature was 36.9°C, pulse rate was 78 beats/min full volume and regular, respiratory rate was 18 cycles/min, while his blood pressure was 140/90 mmHg. He had a weight of 66 kg and a height of 1.7 m with a body mass index of 22.83 kg/m^2.

On examination of the urogenital system, a 3 cm by 3 cm soft fluctuant mass was found, located medially and inferior to the right testis. The mass was mobile and was not attached to the underlying structures or overlying skin.

The left hemiscrotum was normal. There was no palpable significant inguinal and other peripheral lymphadenopathy. Digital rectal examination did not reveal any abnormality.

Abdominal, chest, and other systemic examinations were done and no abnormal findings were noted.

A clinical diagnosis of the right epididymal cyst was made. However, a scrotal ultrasound scan done before the presentation showed a well-circumscribed hypoechoic mass

at the inferior end of the right testis. Differential diagnosis was right epididymal cyst and seminoma.

Some of the requested investigations (abdominopelvic CT scan, liver function test, and tumour marker assays) were not done due to financial constraints. The patient and his father were counseled for scrotal exploration and informed consent was obtained.

In view of the finding on clinical examination, the patient had scrotal exploration. Intraoperative finding was an extratesticular soft-to-firm oval-shaped mass that measured 3 cm by 2.5 cm. The mass appeared to be arising from the distal half of the epididymis. This was excised and the specimen was given to the patient to send for histologic examination. However, there was no histologic diagnosis because the patient discarded the specimen due to financial constraints and he was lost to follow-up.

The patient presented again two years later with a recurrent right scrotal mass and right inguinal swelling. He noticed both swellings about six months before representation but started taking herbal concoction but there was no improvement in his condition.

Clinical examination showed a hard, tender mass in the right hemiscrotum. This was superior to the ipsilateral testis. There were also palpably enlarged matted nontender ipsilateral inguinal lymph nodes that measured 4 cm by 3 cm [Figure 1].

Abdominopelvic ultrasound scan showed markedly enlarged para-aortic and retroperitoneal lymph nodes, the largest of which measured 8.6 cm in its greatest dimension.

Full blood count, liver function test, and kidney function tests done at this point were within normal limits. AFP was 19.17 ng/mL (normal ≤8.5 ng/mL).

bHCG was 0.5 IU/mL (normal ≤5.0 IU/mL).

The patient had trucut biopsy of the right hemiscrotal mass three weeks after he represented, and at this time, the masses had increased to more than twice the previous sizes [Figure 2].

The clinical diagnosis at this time was testicular tumour. However, the mass was huge and not mobile thus delivering it through a groin incision would be extremely difficult and hence the reason for a trucut biopsy. Histopathological review showed features of a malignant mesenchymal tumour (myxoid liposarcoma) [Figure 3a-c]. The effort to do S100 immunohistochemistry was not successful due to financial constraints.

The patient was subsequently started on chemotherapy using Adriamycin (30 mg/m^2), dacarbazine (200 mg/m^2), cisplatin (35 mg/m^2), and etoposide (100 mg/m^2).

The patient tolerated the chemotherapy and the paratesticular and inguinal masses started regressing after two courses. However, the patient failed to turn up for subsequent courses of chemotherapy due to financial constraints and he died a short time after as confirmed by his relatives.

Discussion

Extratesticular masses mostly arise from the paratesticular tissues. These paratesticular tissues include the spermatic cord, testicular tunics, epididymis, and vestigial remnants, which include the appendices epididymis and appendices testis.[10] Neoplasms arising from these regions, therefore, form a heterogenous group of tumours with different behavioral patterns.

Paratesticular tumours need to be differentiated from inguinoscrotal hernia, hydrocele, epididymo-orchitis, undescended testis, and torsion of the testis. In most cases, this can be accomplished on clinical grounds alone. In some other cases, scrotal ultrasound scan is required to differentiate paratesticular tumours and testicular tumours from these other conditions.[11]

Malignant paratesticular tumour could cause a diagnostic dilemma as in our patient. Examination of the mass gave an impression of an epididymal cyst. Hence, a high index of suspicion is required for diagnosis. Ultrasound of the

Figure 1: Clinical photograph of patient on his second presentation

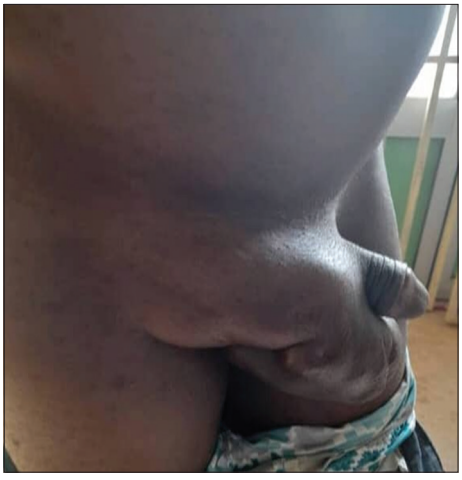

Figure 2: Clinical picture of patient three weeks after second presentation

Table 1: Relevant laboratory results of master OD on presentation at University of Nigeria Teaching Hospital

Patient's parameters	Results	Normal ranges
FBC	Normal	
Kidney function test	Normal	
Liver function test	Normal	
AFP (ng/mL)	19.17	<8.5
bHCG (IU/mL)	0.5	<5.0
Transabdominal ultrasound scan	Enlarged para-aortic retroperitoneal lymph node. The largest measured 8.6 cm in its greatest dimension	
Histology	Malignant mesenchymal tumour (myxoid liposarcoma)	

bHCG: Beta-human chorionic gonadotrophin, AFP: Alfa-fetoprotein, FBC: Full blood count

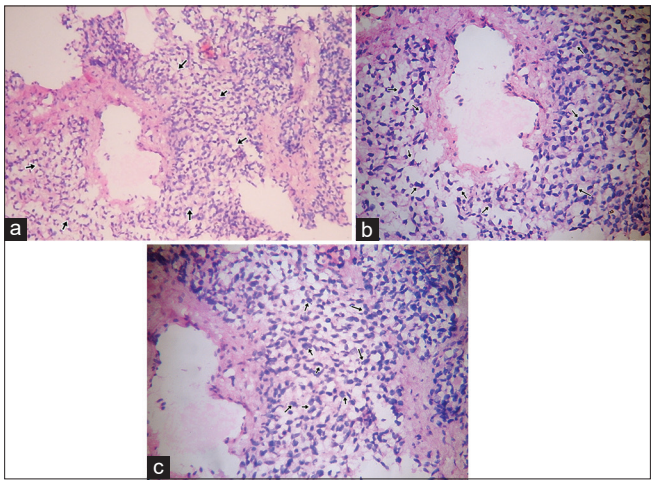

Figure 3: (a) Photomicrograph showing a malignant mesenchymal proliferation consisting of sheets of lipoblasts with large hyperchromatic nuclei, indented by single or multiple vesicles of fat in a mucoid background (short arrows) (H and E, ×10 objective). (b) Photomicrograph showing a malignant mesenchymal proliferation consisting of sheets of lipoblasts with large hyperchromatic nuclei, indented by single or multiple vesicles of fat in a mucoid background (short arrows) (H and E, ×40 objective). (c) Photomicrograph showing a malignant mesenchymal proliferation consisting of sheets of lipoblasts with large hyperchromatic nuclei, indented by single or multiple vesicles of fat in a mucoid background (short arrows) (H and E, ×40 objective)

testis, however, gave two differential diagnoses of the right epididymal cyst and right testicular seminoma. MRI could have clarified this mix-up before surgery but this could not be obtained due to financial constraints. Furthermore, follow-up assays for tumour markers could not be done still because of paucity of funds. This buttresses the challenges of medical care in resource-poor settings.

Lymph node metastasis develops in approximately one-third of cases with paratesticular tumours.[12] This was evident in the index patient who had obvious palpable inguinal lymphadenopathy and ultrasound confirmed retroperitoneal lymph node enlargement which was compressing the abdominal aorta.

Haematogenous metastasis to distant sites, such as the lungs, liver, bone, and bone marrow, is present in 20% of patients at initial diagnosis.[13] However, the index patient was not fully evaluated for metastasis due to scarcity of funds.

Paratesticular rhabdomyosarcomas are treated with combined protocols which include inguinal radical orchiectomy, local radiotherapy to eliminate microscopic and macroscopic residual tumours, and multiple chemotherapy protocols.[14] However, these protocols were not observed in the index patient due to the already-mentioned challenges.

Illiteracy, ignorance, and financial constraint contributed immensely to the patient discarding the specimen and absconding from follow-up visits, resulting in a late diagnosis. These affected the management outcome and the unexpected early demise of such a promising young adult.

Conclusion/Recommendation

This case illustrates the impact of late diagnosis and financial constraints in the management of paratesticular tumours. Hence, the importance of early diagnosis and proper treatment before lymph node or distant metastasis occurs. This case also highlights the importance of an integrated approach which includes clinical findings, imaging studies, and histopathological examination with immunohistochemistry in the definitive diagnosis of paratesticular tumours. In cases where the suspicion of malignancy cannot be ruled out, radical orchidectomy should be done. In very rare cases of bilateral paratesticular tumours in young patients with the early curable disease, sperm banking should be advocated before bilateral radical orchidectomy.

We recommend that nongovernmental organisations, International Organisations, wealthy individuals, and philanthropists should assist low- and middle-income countries in providing funds and facilities (food, drugs, and medical equipment including human capital) as aids to these countries to assist in the early diagnosis, proper and adequate management of the indigent population.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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