Malignant Fibrous Histiocytoma of Left Spermatic Cord: A Postmortem Case Report

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Abstract

Malignant fibrous histiocytoma or undifferentiated high grade pleomorphic sarcoma is a rare tumour of the spermatic cord. It presents as a painless swelling in the inguinal canal which is usually difficult to diagnose.The prognosis is poor if not detected early and treated promptly, therefore physicians need to be on the lookout for this tumour in any inguinal swelling in adults in the fifty years and above age group. We hereby present a case of a 53 year old man that was never diagnosed to have this tumour until autopsy.

Keywords: malignant fibrous histiocytoma, spermatic cord, inguinal swelling

Introduction

In 2002, the World Health Organization (WHO) modified the nomenclature for soft tissue neoplasms, with the most significant changes being in the nomenclature for fibrous and lipomatous malignancies. The so-called fibrohistiocytic tumours were classified by the WHO into benign, intermediate, and malignant types [1]. The percentage of malignant form is small and it is referred to as malignant fibrous histiocytoma (MFH) in this article. MFH was first described by Ozzelo *et al* in 1963 and then O'Brien and Stout in 1964 [2, 3].MFH is the most common soft tissue sarcoma in late adult life [4] with male to female ratio of approximately 1.2:1[1]. It commonly occurs in the lower extremities and the retroperitoneum and rarely involves the spermatic cord [5-9]. We present a postmortem case of this tumour occurring at a rare location, the left inguinal region with metastasis to the pelvis, parietal peritoneum and the mesentery.

Case Report

A 55 year old businessman presented initially at the Nigerian Airforce Medical Centre Ikeja, Lagos State with a left inguinal swelling of 10 months duration. Fine needle aspiration cytology was performed in June, 2008 which was reported as 'suspicious of malignancy'. However, he was lost to follow-up. The swelling progressively enlarged so he re-presented at the same medical centre from where was referred to the Lagos State University Teaching Hospital (LASUTH), Ikeja.

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He was admitted in LASUTH on 17th September, 2008 on an account of abdominal pain, abdominal distension and constipation of 4 days duration as well as cough and dyspnoea of 2 days duration.

On physical examination, he was conscious but acutely ill-looking and was in respiratory distress with mild pallor, was dehydrated and afebrile with a left pitting oedema up to the knee. There was a left inguinal swelling that measured 20.0×16.0 cm. The swelling was firm, non-reducible, and tender. No superficial lymph nodes in the inguinal region were palpable. The abdomen was distended but soft with mild tenderness and rectal examination was unremarkable. The cardiorespiratory systems examination revealed tachycardia, tachypnoea and fine crepitation in the left hemithorax.

He was managed with intranasal oxygen, nasogastric tube, intravenous fluids, antibiotics and analgesics. He died on the third day of admission. It is important to note that he was managed as a case of fibrosarcoma with no histological diagnosis available until his death. Postmortem examination shows a middle-aged negroid male with left lower limb pitting oedema up to the hip. There was a left firm inguinal swelling that measured 12.0cm ×8.0 cm which was attached to the underlying structures. The abdominal cavity reveals haemorrhagic ascites (750ml) and presence of multiple, discrete, greyish white soft to firm nodules on the mesentery, the parietal peritoneum, and the pelvis. The nodules have an average diameter of 10cm. There was also pulmonary congestion and oedema.

Histological sections show cellular tissue with plump malignant fibroblasts exhibiting a storiform pattern with moderate pleomorphism. The fibroblasts also have a high nuclear-cytoplasmic ratio with three mitotic figures per high power field. Admixed with the fibroblasts are histiocyte-like cells and a few multinucleated giant cells. The intervening fibrocollagenous stroma is scanty and there are foci of necrosis. Many congested thin-walled blood vessels and lymphatic channels are present in the sections. Immunohistochemical study showed negative response to CD 34, desmin, CD 117, epithelial membrane antigen, cytokeratin (CK AE1/AE3) antibodies but mild positivity to S100. This result does not support a putative origin from smooth or skeletal muscle, adipose tissue, melanocytes, or epithelium.

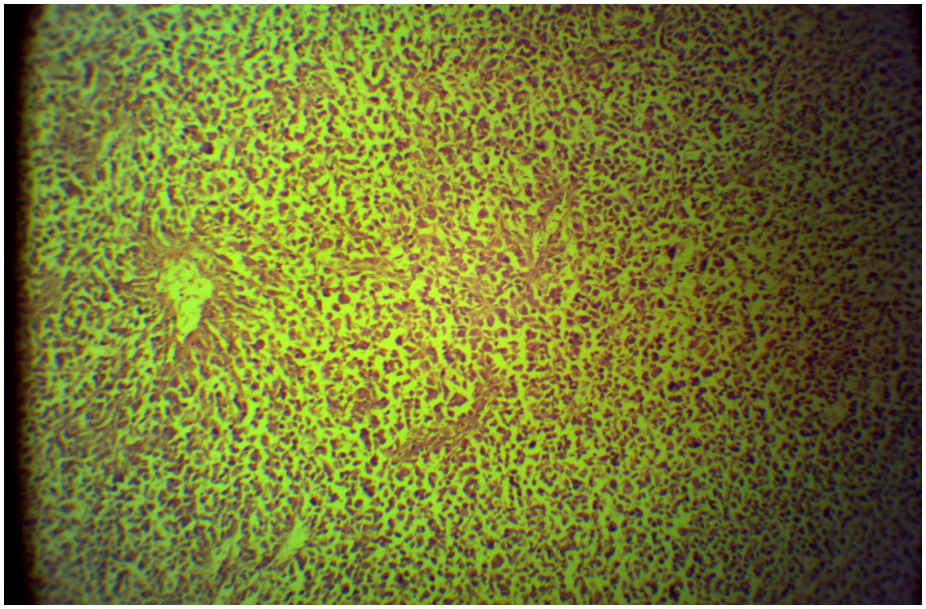

Figure 1 (H & E x100) shows loose sheets of malignant fibroblastic cells with variable sized deeply basophilic nuclei forming a storiform pattern around a central hub on the right.

Discussion

MFH is the commonest malignant soft tissue tumour in adults mostly involving the lower extremities (70%) and the retroperitoneum (16%) and MFH of the spermatic cord is rare [4-9]. Prior to 2002, there were only 36 reported cases presented in English and Japanese literatures [5,10]. To our knowledge this is the first reported case in this sub-region.

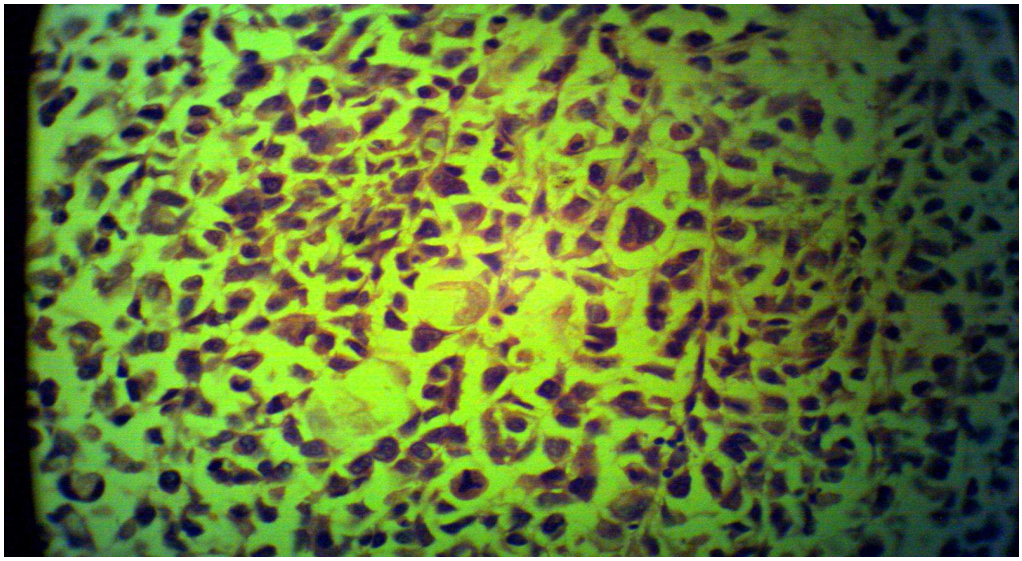

Figure 2 (H&E x400) shows loose malignant fibroblastic cells with highly pleomorphic hyperchromatic nuclei with many bizarre mitotic figures seen.

The incidence of MFH peaks in the 5th and 6th decades of life [11] and most cases presented at the 6th decade. Orio *et al* in their case report presented a patient aged 61 years and the two cases presented by Lin et al were aged 65 and 70 years [5,10]. However, Lin et al in their review of literature found out the age range of presentation was between 32 years and 84 years with most patients presenting after 50

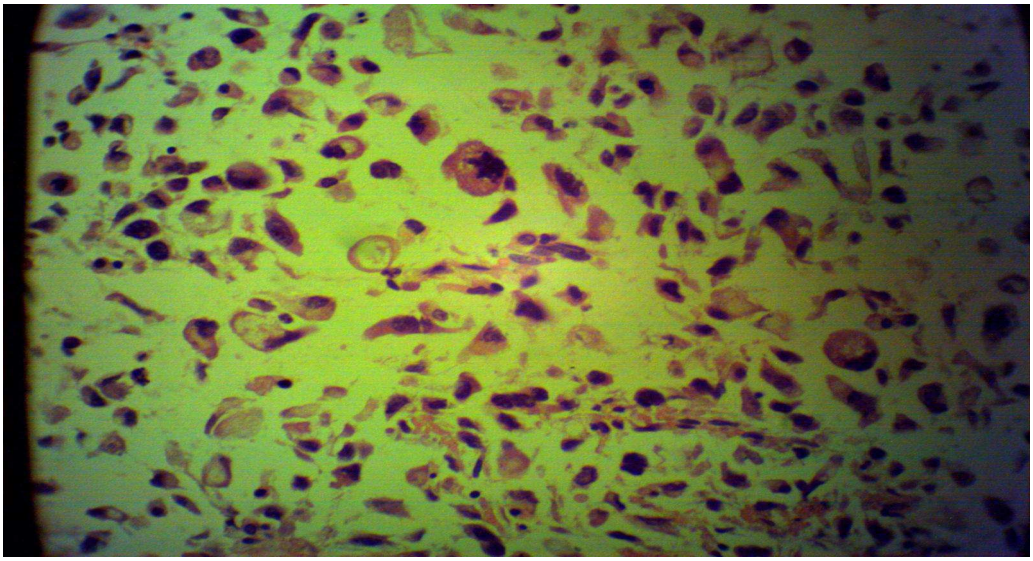

Figure 3 (H&E x400) shows loose fibroblastic cells with many bizarre mitotic figures and few chronic inflammatory cells infiltration

years of age and that MFH was not common in the 40 years and below age group[5]. Our case also conforms to the 6th decade of presentation they found out that there was no significant difference on the side of presentation[5].

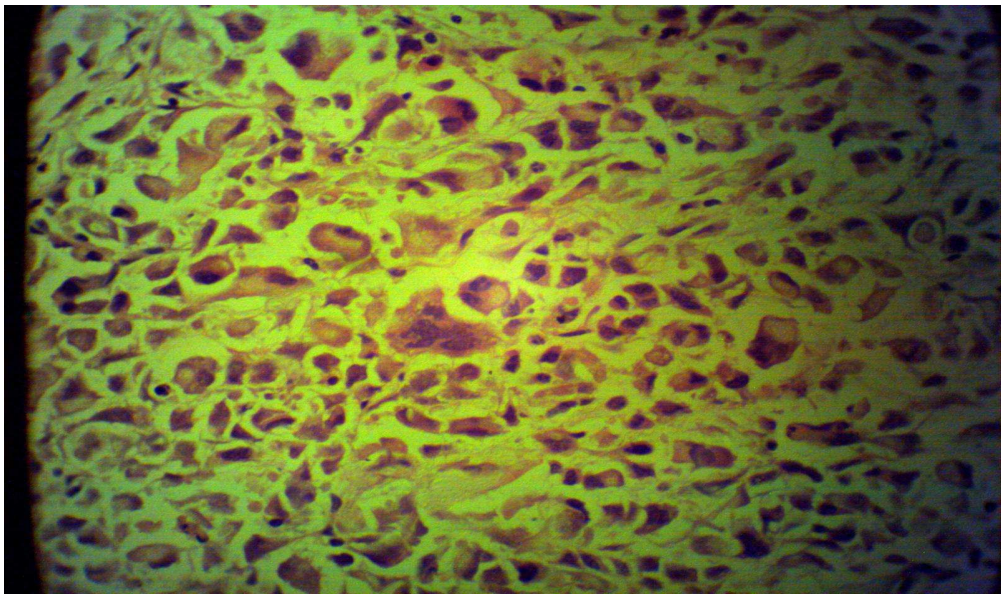

Figure 4 (H & E x 400) shows a tumour giant cell in the centre of the field as well as histiocyte-like cells

(53 years) and was located on the left inguinal canal like the case presented by Orio *et al*[10]; however in the analysis of 38 cases by Lin *et al*, The pre-operative clinical diagnosis of a spermatic cord tumour is difficult without ancillary investigations [6,9]. The lesion can be

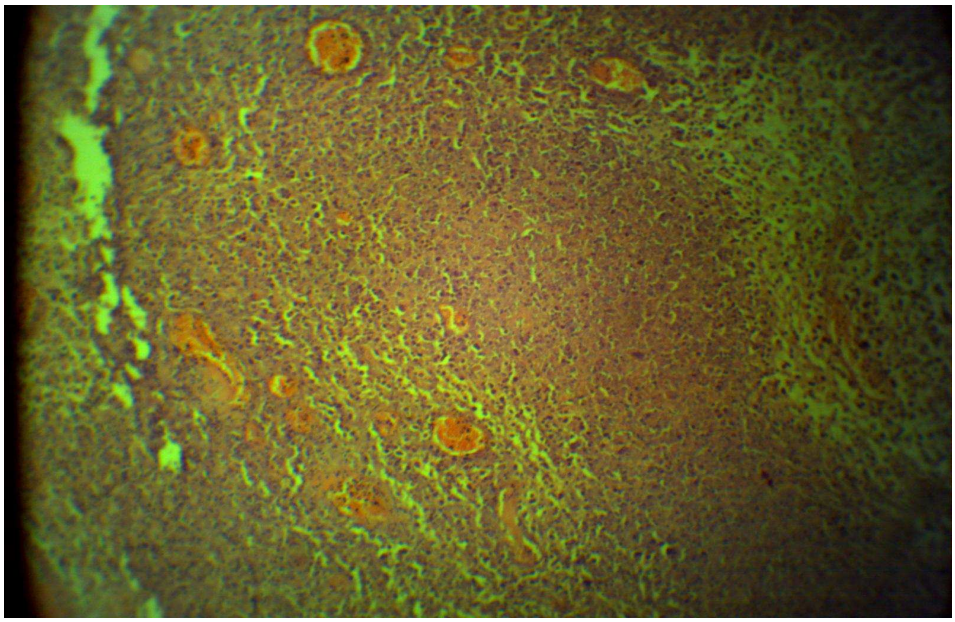

Figure 5 (H&E x100) shows focal areas of necrosis and many congested blood vessels within the tumour cells

confused with an inguinal hernia, spermatocele, epididymo-orchitis and testicular tumour [9], which may be because the commonest mode of presentation is a painless slow growing

various numbers of tumour giant cells were seen in some cases with only few of them being classified as Giant cell type. Other variants include the Myxoid type and those with focal areas of myxoid change. The presence of

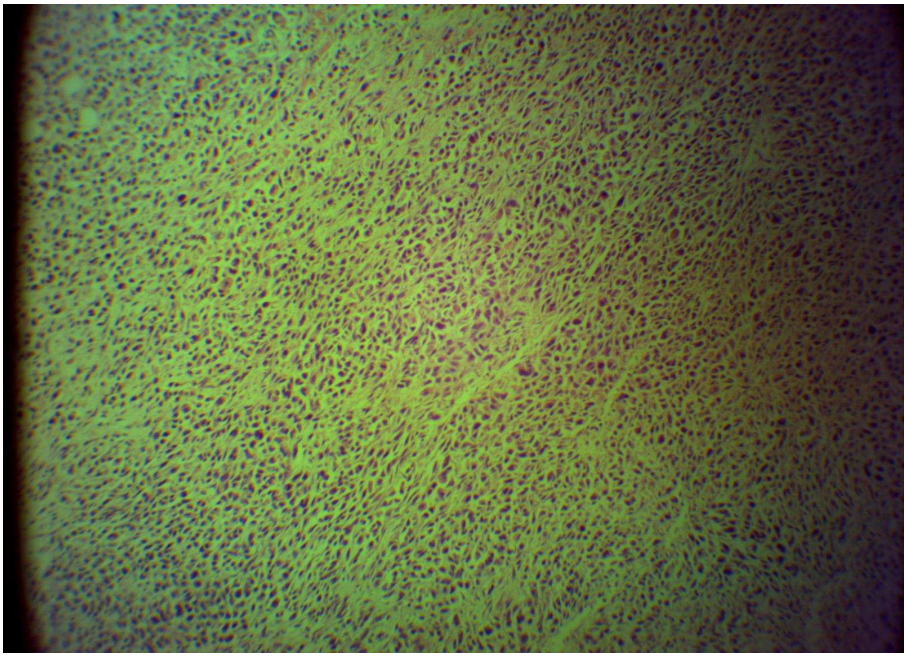

Figure 6 (H&E x100) shows cart wheel appearance around central hub.

inguinal mass. The size of the tumour ranges between 1cm and 20 cm with majority falling between 2cm and 10 cm[5]. The mean maximum diameters in cases from Japan and in other parts of the world were 6.1cm and 7.9cm respectively [5]. In our case, the maximum diameter was 12cm with multiple satellite tumour nodules in the pelvis and abdominal cavity.

The histological type seen in this case was the storiform-pleomorphic type which was recently classified as pleomorphic sarcoma; this is interspersed by some tumour giant cells which are not however enough to categorize it as the Giant cell type. The pleomorphic type is the commonest type of MFH as confirmed in the case series presented by Lin et al which showed most tumours were of the storiform-pleomorphic type[5]. They also found that moderate acute or chronic inflammation was also noted in some cases by Lin et al and two cases were actually classified as Inflammatory type [5]. The index case also showed areas of acute inflammatory cell infiltration.

Originally, MFH was defined as a neoplasm showing both fibroblastic and histiocytic differentiation; however, more recent evaluation has shown no evidence of true histiocytic differentiation. Furthermore, the morphological pattern seen with pleomorphic MFH is shared by a variety of poorly differentiated malignant neoplasms [1] including the current WHO classification. Undifferentiated high grade pleomorphic sarcoma is the most common type while the Giant cell type and the Inflammatory type are much less common[1,6]. Lin et al found in immunohistological investigation on their two cases that the tumor cells were positive for

vimentin, CD68, and lysozyme and were negative for cytokeratin, S100, desmin, smooth muscle actin, muscle-specific actin, CD34, epithelial membrane antigen, and CD117. These results are consistent with fibroblastic or histiocytic lineage[5]. This index case was also negative for ctyokeratin AE1/AE3, desmin, CD34, epithelial membrane antigen (EMA), CD 117 but weakly positive for S100. The result in our case shows no support for origin from smooth or skeletal muscle, adipose tissue, melanocytes, or epithelium.

Weiss and Enzinger in their analysis of 200 cases of MFH reported a 12% rate of lymph node metastasis [12]. It has been shown that evidence of distant metastasis is not common at the time of diagnosis in most cases of MFH of the spermatic cord [9], most likely the result of early diagnosis of the reported cases unlike what was seen in our case in which the patient presented at an advanced stage with large left inguinal swelling and multiple intra-abdominal metastasis as well as features of intestinal obstruction.

Orio *et al* presented 5 prognostic factors as reviewed by the WHO that are independently associated with decreased disease specific survival rate to include, Union for International Cancer Control/American Joint Committee on Cancer (UICC/AJCC) cancer stages III and IVA; residual microscopic disease after primary local therapy; deep tumour location; non myxoid histology and age over 50years[10]. Our patient showed positivity in four out of the five bad prognostic factors and this may be responsible for the fatal outcome of the illness, in addition to the late histological diagnosis and lack of appropriate therapy.

Conclusion

MFH of the spermatic cord is rare and usually presents as a painless growing tumour which may be overlooked by the patient. It is imperative that the attending physician be on the lookout for this condition especially when assessing cases of inguinal swelling in male patients in the fifth and sixth decades of life. This is very important because of the poor prognosis of the tumour if not detected early and the requisite therapy given. Therefore, MFH of the spermatic cord should be a differential diagnosis of an inguinal swelling in adults particularly in our sub region where reported cases of MFH of the spermatic cord are almost non-existent.

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