Malignant fibrous Histiocytoma of Spermatic Cord: A rare case of Paratesticular Tumor

Manish Swarnkar[1]

[1] Associate Professor,Department Of Surgery,Jawaharlal Nehru Medical College,Datta Meghe Institute Of Medical Sciences,Sawangi(Meghe), Wardha,Maharashtra,442001

ABSTRACT

Malignant fibrous histiocytoma (MFH) is a morphologically ill-defined tumour of the soft tissues and may involve nearly every organ of the body but Malignant fibrous histiocytoma (MFH) of the spermatic cord represents an uncommon location for the most common soft-tissue tumor in adults , MFH of the spermatic cord is such a rare entity that only 33 case reports have been described reported in English-language journals to date and only one case from Indian literature. Therefore, this case report is presented in anticipation of adding numerical power to future analysis for correct treatment planning, mortality rates, and prognostic indications of a somewhat unknown subset of a common disease.

Key words: malignant fibrous histiocytoma, pleomorphic, spermatic cord, orchidectomymalignant

Corresponding author address: Manish swarnkar, associate professor,department of surgery,jawaharlal Nehru medical college,datta Meghe institute of medical sciences,sawangi(Meghe), Wardha,maharashtra,442001 M: 9763703920,

E-Mail: mswarnkar1971@gmail.com

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INTRODUCTION

Soft tissue sarcomas are rare malignant tumours that represent < 1% of malignancies.(1)Malignant fibrous Histiocytoma is the most commonly diagnosed subtype of soft tissue sarcoma, and was first reported in 1964 by O'Brien and Stout.(2) Some pathologists consider malignant fibrous Histiocytoma to be a discrete entity that may be sub classified five or six histological subtypes (pleomorphic-storiform, giant- cell, inflammatory,

fibrous, myxoid and angiomatoid), which partially differ both clinically and prognostic ally (3, 4). The Pleomorphic subtype of MFH is regarded as the most common soft tissue sarcoma of adulthood. However, there are currently no definable criteria for its diagnosis. In fact, several recent studies have expressed considerable doubts regarding MFH, or at least its Pleomorphic type, as a distinct entity and have suggested that it represents a common morphology of a group of poorly differentiated sarcomas(5, 6) and, more rarely, other neoplasms. Malignant fibrous Histiocytoma arises mainly in the deep soft tissues of the extremities (70%) and retro peritoneum (16%) and, occasionally, in the inguinal region;4 it rarely involves the spermatic cord, and was first described at this site by Cole et al. in 1972.(7) A literature search revealed that 33 cases of spermatic cord malignant fibrous Histiocytoma have been reported in English-language journals.(8)

The rarity of spermatic cord malignant fibrous Histiocytoma has made its diagnosis, staging and treatment difficult. This report describes the clinical and histological features of spermatic cord malignant fibrous Histiocytoma, and discusses the recommended management of, and prognosis for, this tumor.

CASE REPORT

A 50-year-old male was admitted to the Department of surgery associated to Acharya Vinoba Bhave Rural Hospital Sawangi, Wardha with complaint of swelling in right inguinal region since 10 month and vague pain during walking Physical examination revealed a painless mass in the right inguinal region, of 8×4.5cm size which was fixed to the right spermatic cord. The surrounding skin was of normal appearance and no superficial inguinal lymph nodes were palpated. Physical examination was otherwise unremarkable. Blood chemistry revealed no abnormalities. Levels of tumor markers (such as carcinoembryonic antigen, α-fetoprotein and prostate specific antigen) were within normal ranges. Ultrasonography revealed a hypo echoic mass (9.5x5x4.8 cm), with a clear border, arising from the right spermatic cord. The testes and epididymides were normal.

The patient underwent right inguinal exploratory surgery. A tumor involving the spermatic cord was identified, and a frozen section was examined and considered to be malignant. Right radical Orchidectomy was subsequently performed with wide dissection of the mass. Removal of any spermatic cord remnant was performed as high as possible up to the deep inguinal ring, and the proximal extent of the cord was labeled for pathological margin evaluation. The testicle was found to be of normal size and was not involved by the tumor. The tumor was 10 × 6 × 4.8 cm with an intact external surface. The cut surface of the tumor was grey-yellow and elastic . Microscopically the tumor consisted of highly Pleomorphic spindle tumor cells arranged in a storiform pattern with numerous multinucleated giant cells and frequent mitoses (mitotic count of approx 8- 10/10 hpf) along with areas of necrosis. Immunohistochemical analysis showed cd 68 positivity. Post-operative abdominal computed tomography demonstrated no retroperitoneal and mediastinal lymph-node enlargement. Surgical margins were noted to be tumor negative, and the postoperative recovery was

uneventful. He was followed up regularly for 16 months, with no evidence of recurrence or metastasis.

Figure 1: Represents the cut section of the orchidectomy specimen which shows the fleshy tumor arising from spermatic chord and normal testis

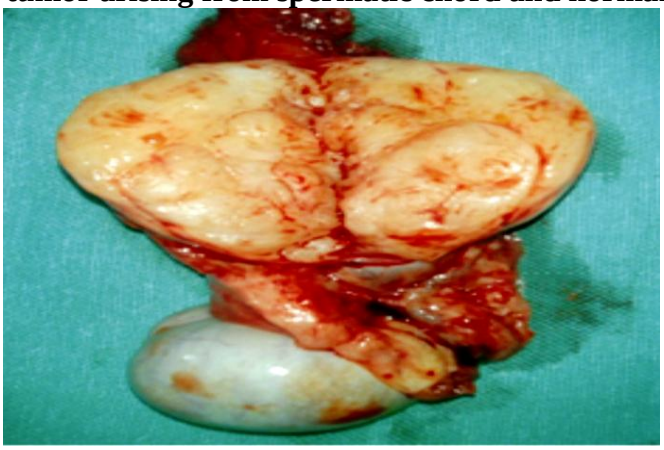


Figure 2: Low-power photomicrograph showing tumor cells with unusual morphology and severe nuclear Pleomorphism

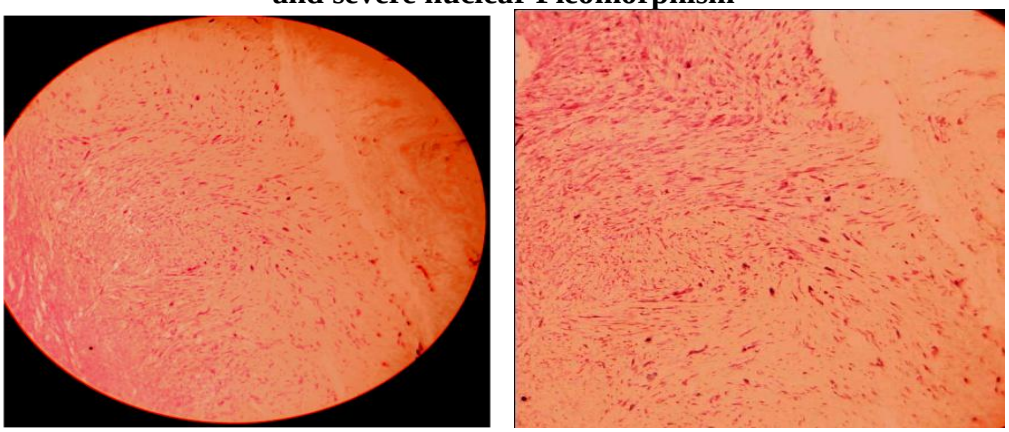


Figure 3: Represents microscopic picture of malignant fibrous Histiocytoma with the marked Pleomorphism from spindle cells to giant cells (arrow)

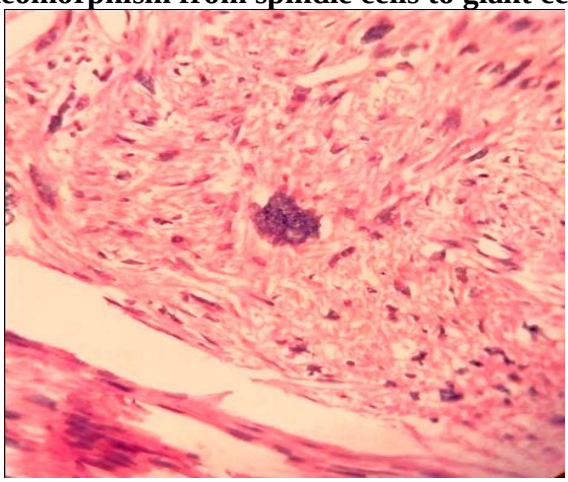
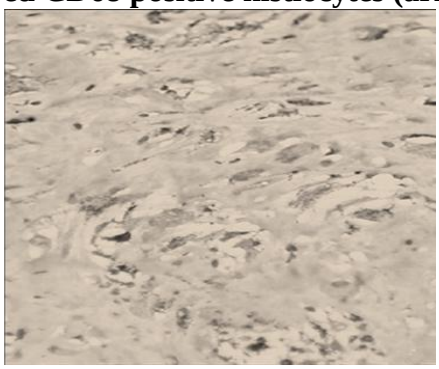


Figure 4: Anti-CD68 immunohistochemical staining of tumour tissue.Tumour cells showed CD68 positive histiocytes (arrow).



DISCUSSION

Since malignant fibrous Histiocytoma was first reported, its status as a tumor entity has been the subject of dispute and re-evaluation.(9) Malignant fibrous histiocytoma, particularly in its storiform Pleomorphic and myxoid forms, has been regarded as the most common type of soft tissue sarcoma for the last 30 years.(10) Malignant fibrous Histiocytoma arising from the spermatic cord is rare and has been reported to represent only 11% of adult spermatic cord sarcomas.(11) A literature search of the PubMed database found that 33 cases of spermatic cord malignant fibrous Histiocytoma have been reported in English literature till date (8) and only one case published in Indian literature by Sethi s et al in 2003.(12)

The pathogenesis of malignant fibrous Histiocytoma is still unknown. Tumor cells have features suggestive of fibroblastic, myofibroblastic and/or histiocytic origin, and may have developed from the fibrous tissue of the spermatic cord.(13) Histopathologically, spermatic cord malignant fibrous Histiocytoma can be divided into four morphological subtypes, depending on the predominant cellular components (in decreasing order of prevalence): storiform–Pleomorphic type (83%), giant-cell type (9%), inflammatory type (6%) and, very rarely, myxoid type.(14) MFH may mimic the histopathologic appearance of other sarcomas, e.g., de- differentiated and Pleomorphic liposarcoma, pleomorphic leiomyosarcoma.(14) MFH has no specific markers, but can show focal positivity for CD68, S100 and vimentin and is a diagnosis of exclusion.(14) The rarity of malignant fibrous Histiocytoma of the spermatic cord has led to difficulties in its diagnosis and management

DIAGNOSIS

Preoperative diagnosis of spermatic cord malignant fibrous histiocytoma is difficult, as clinical presentation and physical examination alone usually cannot determine the origin and nature of an inguinal mass. Currently, excisional biopsy and microscopic examination of tissue specimens are considered to be the gold-standard method for the diagnosis of MFH.

Grossly, the specimen is frequently a well-circumscribed, yellowish solid tumor. Spermatic cord MFH may infiltrate the vas deferens, pampiniform plexus and adjacent adipose tissues (15). The epididymis and testis are characteristically not infiltrated, with the exception of neglected cases and the giant-cell subtype of MFH (15-18). Recently, a combination of fine-needle aspiration (FNA) for cytological examination and immunohistochemistry was applied to examine a patient with spermatic cord mass and surprisingly demonstrated a Pleomorphic subtype of MFH. (19) A Pleomorphic appearance and marked anaplasia as well as vimentin reactivity were found on cytological examination of these fragments, confirming the diagnosis of spermatic cord MFH of the storiform subtype. This finding, later confirmed by surgical excision and histological examination, confirmed the results obtained from FNA. The authors suggested that a FNA combined with immunohistochemical studies may replace the gold-standard open biopsy in the diagnosis of a spermatic cord MFH (19). Ultrasonography is helpful in defining neoplasms and can reveal satellite nodules. It is accurate in distinguishing testicular from non-testicular masses and eliminating other diagnostic possibilities.(20, 21) Although the imaging findings are rather nonspecific, CT and magnetic resonance imaging (MRI) studies are essential for preoperative staging and surgical planning in patients with spermatic cord malignant fibrous Histiocytoma.(22, 23) CT is necessary for staging of the pelvis and abdomen, especially when the tumor is not well differentiated.(21) MRI is an excellent modality for evaluating the extent of inguinal masses;(24) such images often show heterogeneous signal intensity on all pulse sequences, reflecting the complex histological components of the tumor.(25)

PROGNOSIS AND TREATMENT

MFH tends to recur locally thereby the treatment of choice is radical Orchidectomy, with complete removal of all seminal cord structures. Ligation of the spermatic cord should be as high as possible. Despite complete resection, local recurrence has been reported to be 51%.(14) Prognostic factors of spermatic cord malignant fibrous Histiocytoma are difficult to assess because of the relative paucity of cases and the short follow-up. Coleman et al.(11) did not identify any specific prognostic factors associated with recurrence or disease-free survival

in adult spermatic cord sarcomas. Based on a careful review of previous reports and the present case study, we suggest that tumor biology, adequacy of surgical resection and adjuvant treatment could be the major factors that affect local control rates. Histopathological grade and size are known to be significant prognostic factors for malignant fibrous Histiocytoma,(26, 27) and the presence of satellite nodules at initial surgery increases the overall recurrence rate.

Adjuvant loco regional (ipsilateral pelvic and groin nodes and scrotum) radiation reduces the risk of local recurrence.(28) Fagundes et al., reported no local recurrence in patients receiving adjuvant radiation, compared with 37% local failure in those treated with Orchidectomy alone.(29) Modest activity with doxorubicin and dacarbazine based chemotherapy has been reported with MFH.(30, 31)

CONCLUSION

Spermatic cord MFH, a rare tumor of this location, should be considered in the differential diagnosis of scrotal, inguinal and/or spermatic cord masses. Malignant fibrous Histiocytoma presents earlier in the spermatic cord than at other sites and seems to have a more favorable prognosis. Definitive treatment is radical Orchidectomy but localized radiotherapy may decrease local recurrence rates. Satellite lesions at surgery indicate a poorer prognosis. Metastases may develop late in the lungs or mesentery, and so long-term follow-up is necessary.

CONFLICTS OF INTEREST

The authors had no conflicts of interest to declare in relation to this article.

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