Undifferentiated Sarcoma of Spermatic Cord: A Case Report

Santosh Koppal, Vishwash Pai, Gajanan Bhat
Department of surgery, St Ignatius hospital, Honavar, India.
Corresponding Author: Santosh Koppal, consultant surgeon, st Ignatius hospital, prabhatnagar, honavar, Karnataka state, india.

Abstract: Sarcomas are rare variety of malignant tumours arising from mesodermal cells. The paratesticular sarcomas constitute major proportion of primary tumours of paratesticular area. These tumours often pose diagnostic difficulty and it is wise to consider histological examination before going for surgery. We report a case of undifferentiated sarcoma in an 84 year old male patient presented as slowly growing mass in the left inguinal region.

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I. Introduction

Although soft tissue sarcoma (STS) of the spermatic cord comprises almost 30% of the neoplasm derived from cord structures, it is still an uncommon disease, which presents differently in children, adolescents and in adults. In children and adolescents rhabdomyosarcomas and embryonal sarcomas are common whereas in adults commonest type of sarcomas are liposarcoma followed by leiomyosarcomas, undifferentiated sarcomas. In adults, wide local excision with radical orchidectomy with high ligation is considered the procedure of choice for spermatic cord STS, retroperitoneal lymph node dissection, as well as adjuvant radiation therapy and chemotherapy remain controversial treatments. The lack of standard treatment protocol makes treatment decisions more complicated.

II. Case Report

84yrs male known diabetic and hypertensive, presented with painless gradually increasing swelling in the left groin since last 4months. There was previous history of left orchidectomy 8years back. Examination revealed a diffuse firm mass of size about 10x8cms in the left inguinal region extending into left hemiscrotum. Right testis was normal. Absent cough impulse and no secondary skin changes. Complete blood count, blood urea, serum creatinine, RBS and urine routine examination were normal. Ultrasonography of abdomen revealed an irregular mixed echoic growth of size about 9.7x7.6cms in the left inguinal region with infiltration to surrounding area and extending upto root of scrotum, with highly irregular margins and moderate vascularity. Irregular thickening of left spermatic cord noted. Right testis and paratesticular area is normal, more likely tuberculosis of spermatic cord, secondaries from prostate/testis. CT scan of abdomen showed multicystic lesion with multi variate density with infiltration to surrounding tissues except peritoneum and intra abdominal organs, correlating with ultrasound features of chronic inflammatory process.

Fig1. Ct scan
FNAC of the swelling done which revealed moderate cellularity comprising malignant cells with pleomorphic spindle shaped nucleus with coarse chromatin with multinucleate tumour giant cells suggestive of pleomorphic sarcoma. After informed consent patient underwent wide local excision with high ligation of cord with reconstruction using prolene mesh.
Histopathology of specimen (Left spermatic cord, mass, wide excision) showed: Pleomorphic sarcoma, infiltrating adjoining fat. Spermatic cord resection margin is negative for tumour. All nine lymph nodes are negative for tumor metastasis (0/9). Report: Sections show a tumor arranged in sheets, interlacing bundles and as singly scattered cells. Tumor cells show moderate to marked pleomorphism having irregular nuclear membrane, hyperchromatic nuclei and moderate amount of cytoplasm. Many bizarre forms along with multinucleated giant cells seen. Few atypical mitotic figures noted. Focal area shows necrosis.
Immunohistochemistry revealed the final diagnosis as UNDIFFERENTIATED SARCOMA. AS Immunopositivity for SMA, Vimentin, CD68, patchy positive for Desmin and CK supports the rendered diagnosis. Negative staining for S-100 rules out liposarcoma and neural origin of the tumor. Negative staining for SATB2 rules out osteosarcoma. Negative staining for CD99 and TLE-1 rules out synovial sarcoma. The tumor cells are positive for SMA, Vimentin, CD68, patchy positive for Desmin and CK with a low Ki67 proliferation index (5%).

III. Discussion

The majority of malignant tumors in the nonepididymal extratesticular soft tissues are sarcomas that arise from the spermatic cord. The most common scrotal sarcomas are rhabdomyosarcoma, liposarcoma, leiomyosarcoma, and MFH, all of which are rare clinical entities. Rhabdomyosarcoma most commonly affects children where as liposarcomas, leiomyosarcomas and undifferentiated sarcomas occur in adults. Myxoid liposarcoma is characterized by slow growth. The tumor is usually highly differentiated, tends to recur locally, rarely metastasizes, and has a good prognosis following complete removal. The scrotum and the spermatic cord are the third most common sites of Undifferentiated liposarcoma, which can occur in late adult life. Most of these tumors present as de novo lesions, whereas the remainder develop as a late complication of a pre-existing well-differentiated liposarcoma after an average interval of 7.7 years. MFH is also a rare subtype of STS of the spermatic cord. Long-term survival is possible and local lymph node involvement is rare. Definitive treatment is radical orchiectomy 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. Retroperitoneal lymph node dissection is advocated in patients with MFH of the spermatic cord. Liposarcoma of the spermatic cord can be characterized as a painless slow-growing scrotal swelling that can present with sudden enlargement of the mass. Inguinal orchiectomy is an adequate surgical approach. Retroperitoneal lymphadenectomy is not indicated owing to the low malignancy potential of these sarcomas. Postoperative radiation therapy is an optional adjuvant treatment. Recurrences are frequent, owing to incomplete surgical removal of the tumor. The role of chemotherapy in recurrent disease remains to be elucidated. The course of embryonic sarcoma and rhabdomyosarcoma is rapid; regional nodal involvement and early hematogenous spread are common. Surgery alone seems to be inadequate for disease control, and adjuvant chemotherapy and radiation therapy may be indicated. Rare histologic subtypes of STS arising in the spermatic cord include leiomyosarcoma and primary extraosseous osteosarcoma. The etiology for spermatic cord sarcoma is unknown.

Soft tissue sarcoma of the spermatic cord usually presents as a firm, non-tender mass, located in the scrotum. The preoperative diagnosis is often incorrect and can include inguinal hernia, hydrocele, and epididymal cyst. Solid extratesticular masses can be evaluated by ultrasonography. No sonographic features of masses were useful for distinguishing benign from malignant lesions. STS, especially dedifferentiated liposarcoma, of the spermatic cord can also be assessed by fluorodeoxyglucose positron emission tomography (FDG-PET) imaging. It was suggested that FDG-PET seemed to be useful in differentiating recurrent tumor from radiation necrosis in patients with liposarcoma after therapy. The treatment of spermatic cord sarcomas is based primarily on surgery. The role of adjuvant radiation therapy has not been determined. Radical
orchidectomy is considered as the procedure of choice, followed postoperatively by regional ipsilateral iliac and inguinal node irradiation with or without scrotal irradiation.

IV. Conclusion

Purpose of presenting this case is to highlight the rarity of this malignancy and stress the importance of complete treatment of cancer in geriatric population.

V. References