CASE REPORT

Synovial Sarcoma Vulva: A Case Report

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ABSTRACT
Introduction: Contrary to its name, synovial sarcoma does not arise from the synovial membrane but from multi-potent stem cells and can present in any part of the body. Very few cases of vulval synovial sarcoma have been reported in the literature; we report on such a presentation. These tumors can present as painless lumps, which must be completely excised to give the best prognosis. Therefore the diagnosis of synovial sarcoma should always be kept in mind in the management of vulval masses, especially in young patients.

Keywords: Multi-potent stem cells, Synovial sarcoma, vulva

INTRODUCTION

Synovial sarcoma is the fourth most commonly occurring sarcoma, accounting for 7-8% of all sarcomas. It most frequently occurs in young adults, with up to 30% being manifested during the first two decades of life, and a median age of 13 years at presentation.[1] There is a slight male predominance. The term synovial sarcoma was coined in reference to tumors arising near tendon sheaths and joint capsules.

CASE PRESENTATION

A 35 yr old asaian woman, *ramzana bibi* was referred to us, presenting with one month history of a right vulval swelling, which was becoming increasingly uncomfortable. (Fig-1) Patient gave history of swelling in right vulvar region which was not associated with pain, itching or discharge. She was treated in a private hospital and her excision biopsy was taken and sent for histopathology. Her histopathology report showed biphasic synovial sarcoma of vulva. Her CT-Scan & MRI confirmed synovial sarcoma with femoral lymph nodes involvement. She received 8 cycles of radiotherapy from institute of nuclear medicine and oncology Lahore in 2010.In year 2012 she was referred with recurrence of vulvar swelling, associated with pricking pain and itching. She presented to Jinnah hospital gynae unit 2 on 9.02.2012.

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Clinical examination revealed a healthy woman with a well-defined deep firm mass 10x10cm on right labium majus extending towards mons pubis with excoriation of skin in right iliac fossa.

Histopathology report of 20 -3-2010 done in Shaukat khanum hospital Lahore demonstrated a biphasic tumor (measuring 3x2.5x1.8cm) consisting of epithelial cells and fibroblast-like spindle cells, with the presence of glandular structures and mitoses. The epithelial areas consist of trabeculae, nests and tubules lined by large cells with pale cytoplasm and vesicular nucleus. Bcl2 & Mic2immunohistochemical stains were recommended which confirms positivity in spindle cell component. Findings were consistent with biphasic synovial sarcoma.[2] (Fig-2)

The immune-histochemical stains LCA, CD68, NSE, CD34, CAM 5.2, SMA, desmin, and S100 were recommended with limited availability of all markers available.

A decision to re-excise the vulval mass was taken, as there was doubt about the completeness of the first excision.(See image show Fig-3a-3p). A pre-operative computed tomography (CT) scan of the chest, abdomen with i.v contrast was done on 7-2-2012 showing minimal bronchiectatic changes noted in right middle lobe of lungs and no mediastinal, axilliary lymphadenopathy noted. No pleural effusion seen. Cardiac chambers pericardium aortic arch and great vessels were unremarkable. Thyroid appeared normal. Liver normal in size and contours.A 35mm cyst noted in left adenexa. No obvious solid component. Mild free fluid in cul.de.sac. A 37mm heterogeneous mass noted in rightt upper vulva.

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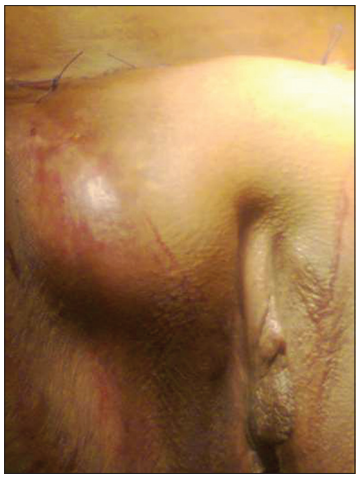

Figure 1: Most common site of tumor

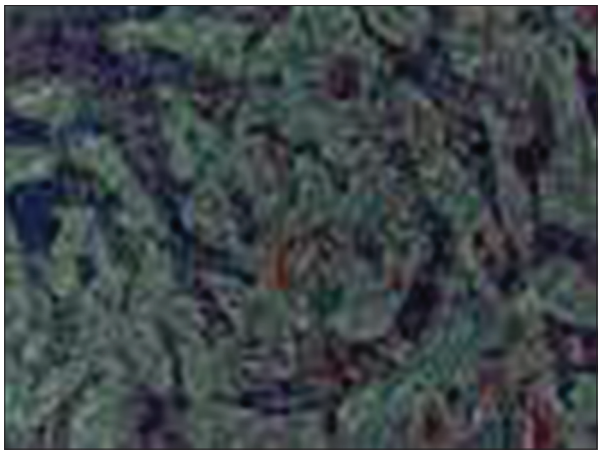

Figure 2: Synovial sarcoma showing epitheloid component with glandular and/or tubular formation, surrounded by malignant spindle cell component

Visible bones show no suspicious focus. After discussion at our oncology, it was felt that radical wide excision of mass with remaining negative margins should be done and follow up should be done in the radiotherapy and chemotherapy unit of Jinnah hospital. The radical vulvectomy with femoral & inguinal lymph nodes clearance was done, followed by flap reconstruction by the plastic surgery department of Jinnah hospital Lahore. She was then referred to Oncology unit for opinion regarding further chemotherapy and radiotherapy. Her histopathology after vulvectomy came out as margins free from cancer cells. She was operated by Plastic surgery department for flap reconstruction in Jinnah hospital.

DISCUSSION

Synovial sarcoma is a soft tissue sarcoma and is divided into three subtypes:[3-5] biphasic, monophasic and poorly differentiated. The biphasic type contains epithelial and spindle cell elements, which occur in varying proportions, whilst the monophasic type contains only spindle cells. The poorly differentiated type shares features of both the monophasic or biphasic types, and has varying proportions of poorly differentiated areas characterised by high cellularity, pleomorphism, numerous mitoses and often necrosis. All morphological subtypes are characterised by a specific t(X; 18) (p11.2; q11.2) chromosomal translocation.[6]

It is difficult to recognize synovial sarcoma on the basis of only its histological appearance. In most cases, it can only be unambiguously identified by immunohistochemical analysis, ultrastructural findings and the demonstration of the specific chromosomal translocation mentioned above. However we have to consider limitations of research and laboratory diagnostic facility available.

It most commonly features as a slowly growing, painless mass, with the most common sites of presentation being at the extremities. The lower extremities are more frequently affected than upper extremities, often in the region of the thigh and knee. The peri-articular regions are especially affected, usually in close association with the tendon sheath, bursae and joint capsules, but they rarely involve the articular surface. However, there is a broad spectrum of locations where they have presented, including head, neck, trunk, lungs, esophagus, intestine, mediastinum and retroperitoneum. The major sites of metastatic spread are the lungs and, less often, regional lymph nodes, bone and bone marrow.[6-8]

A definitive diagnosis can only be established by an adequate tissue biopsy, but radiological investigations may be useful to characterise the tumor. On a CT scan, the tumor usually shows as a heterogeneous septate mass with a mixed solid and cystic appearance; calcification can be seen in one third of them.[9] Magnetic resonance imaging (MRI) provides a greater contrast between tumor and normal tissue and can show neurovascular or regional lymphatic involvement Radiological investigations are important in the detection of metastatic disease, as the presence of Secondary spread influences both prognosis and management. The presence of metastases, a tumor size greater than 5 cm, invasiveness, high histological grade, positive surgical margins and poor histological differentiation are all associated with adverse prognostic significance.[10,11] The complete surgical excision of the tumor is the treatment of choice. The presence of positive microscopic margins gives a greater likelihood of local recurrence and is associated with an increased risk of metastatic spread and decreased length of disease-free survival. Therefore, the surgeon should aim at obtaining negative margins, although there is no clear evidence on the extent of negative margin required. Primary re-excision

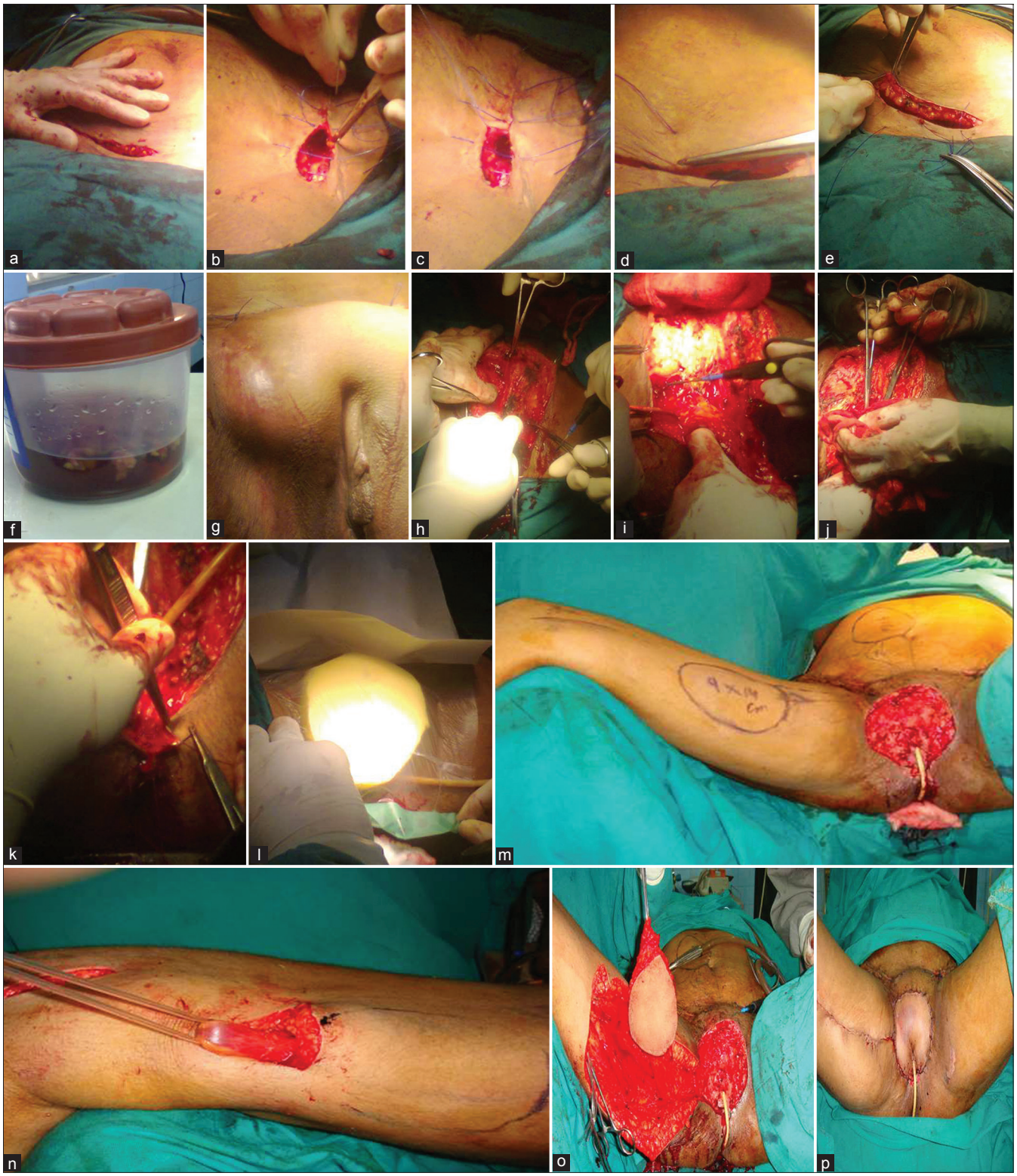

Figure 3: (a,b) Inguinal lymph nodes dissection (c,d) Bilateral inguinal lymphnodectomy (e,f)Inguinofemoral lymph nodes biopsies (g,h) Primary site of synovial sarcoma vulva (i,j) Radical vulvectomy and urethral preservation (k,l) Primary closure and suction application (m,n) Secondry closure and flap removal for reconstruction of vulva (o,p) Flap reconstruction of vulva

should be carefully considered in all patients with a gross residual tumor, as re-excision decreases the risk of local recurrence[10-12]

Radiotherapy appears to have a role in the treatment of patients newly diagnosed with synovial sarcoma, especially children with minimal primary tumor or following surgery.

Radiotherapy should improve local control of the disease but does not affect overall survival. The role of adjuvant chemotherapy in the management of synovial sarcoma remains controversial.[10,12,13]

CONCLUSION

This is a rare case of synovial sarcoma of the vulva, the appearance of which can be easily mistaken for a lipoma. The overall prognosis for our patient should be good given the complete excision of the tumor. Other favorable factors affecting the prognosis include a tumor size of less than 5 cm and no evidence of metastatic spread.

CONSENT

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

COMPETING INTERESTS

The authors declare that they have no competing interests.

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