International Journal of Biomedical Research
ISSN: 0976-9633 (Online); 2455-0566 (Print)
Journal DOI: 10.7439/ijbr
CODEN: IJBRFA

Case Report

Monophasic Synovial sarcoma mimicking inguinal hernia in a young female

Manveen Kaur[1], Varsha Dalal[1], Amar Bhatnagar[2] and Anju Bansal[1*]

[1]*National Institute of Pathology (ICMR), Safdarjang Hospital Campus, New Delhi -110029.*
[2]*Department of Cancer Surgery, Safdarjang Hospital, New Delhi – 110029.*

***Correspondence Info:**
Dr. Anju Bansal
Scientist D,
National Institute of Pathology (ICMR),
Safdarjang Hospital Campus, New Delhi -110029.
E-mail: dranjubansal@yahoo.com

Abstract

Synovial sarcoma is an aggressive soft tissue tumor, accounting for 5-10 % of all sarcomas, and occurs mainly in the para-articular region of extremities with a predilection of lower limbs. Monophasic inguinal synovial sarcomas are extremely rare. We present here a case of synovial sarcoma in a 17 year old female presenting as an inguinal mass.

Keywords: Sarcoma, Synovial Sarcoma, Monophasic Synovial Sarcoma, Inguinal mass.

1.Introduction

Synovial sarcoma (SS) is an aggressive soft tissue tumour, usually seen in adolescents and young adults between the age of 15 and 35 years. It accounts for 5-10% of all soft tissue sarcomas, with the most common sites being deep soft tissue of extremities. Synovial sarcomas have also been reported in unusual locations including the head and neck, mediastinum, lung, abdominal wall, abdomen, kidney and retroperitoneum. Isolated rare cases have been mentioned in the vulva, skin, blood vessels and nerves.[1,2]

Synovial sarcoma presenting as inguinal mass is very rare. Here, we present a case of synovial sarcoma in a 17 year old female presenting as an inguinal mass.

2. Case report

A 17 year old female was admitted with complaint of progressively increasing mass in the inguinal region since two months without any obvious cause. On physical examination, a swelling was found in the left inguinal region measuring 8x6x6 cm. The swelling was hard, fixed and did not disappear under pressure. A clinical diagnosis of femoral hernia/ femoral abscess with inguinal lymphadenopathy was made and MRI and CT angiography (CTA) scans were conducted. MRI revealed a well- defined enhancing heterogenous mass lesion with haemorrhage in the subcutaneous planes, anterior to the pubic symphysis and medial to femoral vessels (Figure 1a). CT angiography showed a heterogeneously enhancing mass lesion measuring 9.5x7.5x6.6cm in left inguinal region, abutting medial end of adductor longus muscle. The mass was seen to receive blood supply from both the femoral and internal iliac artery. An impression of mesenchymal tumour was rendered. Histopathological correlation was advised.

Surgical excision of the mass was performed. On gross examination, the specimen was 10x8x4cm in size. Cut surface was firm and grey white to tan (Figure1b). Histopathological examination showed a tumour arranged as short fascicles, bundles and sheets (Figure 1c). The tumour cells were moderately pleomorphic with spindle shaped nuclei (Figure 1d). Brisk mitotic activity was noted with more than 10 mitoses/10 hpfs (Figure 1d inset). Areas of haemorrhage and hyalinised stroma were seen. On extensive sampling, no epithelial component was identified.

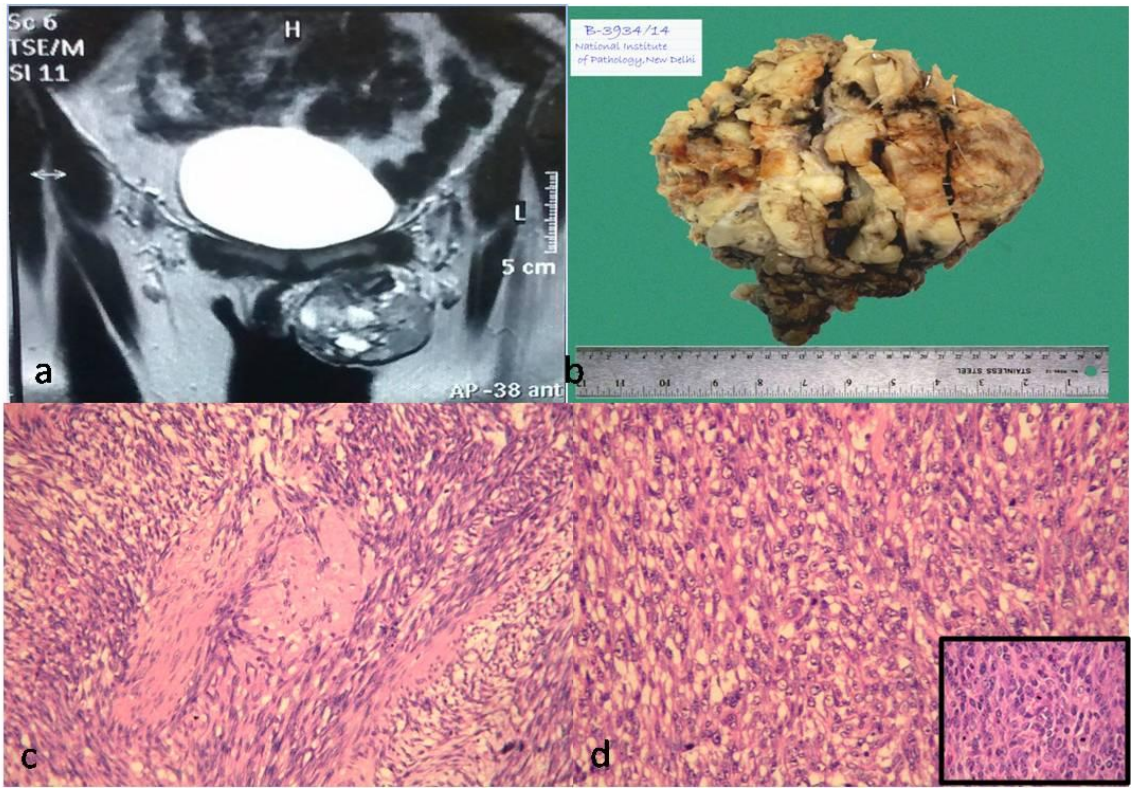

Figure 1a - Preoperative MRI study showing well defined heterogenous lesion anterolateral to pubic symphysis and medial to femoral vessels
Figure 1b - Soft tissue mass measured 10x8x4cm. On serial sectioning, grey white tumor with areas of hemorrhage was seen.
Figure 1c - Tumor composed of spindle cells arranged in interlacing fascicles and sheets with areas of hyalinization (H&E 100x)
Figure 1d - Tumor cells showing moderate nuclear atypia (H&E 200x). Frequent mitotic activity noted (inset 400x)

Immunohistochemically, the tumour cells were positive for vimentin (Figure 2a), epithelial membrane antigen (EMA) (Figure 2b), cytokeratin (CK), Bcl-2 (Figure 1c) and CD99 (Figure 1d) but negative for smooth muscle actin (SMA), CD68 and S100.

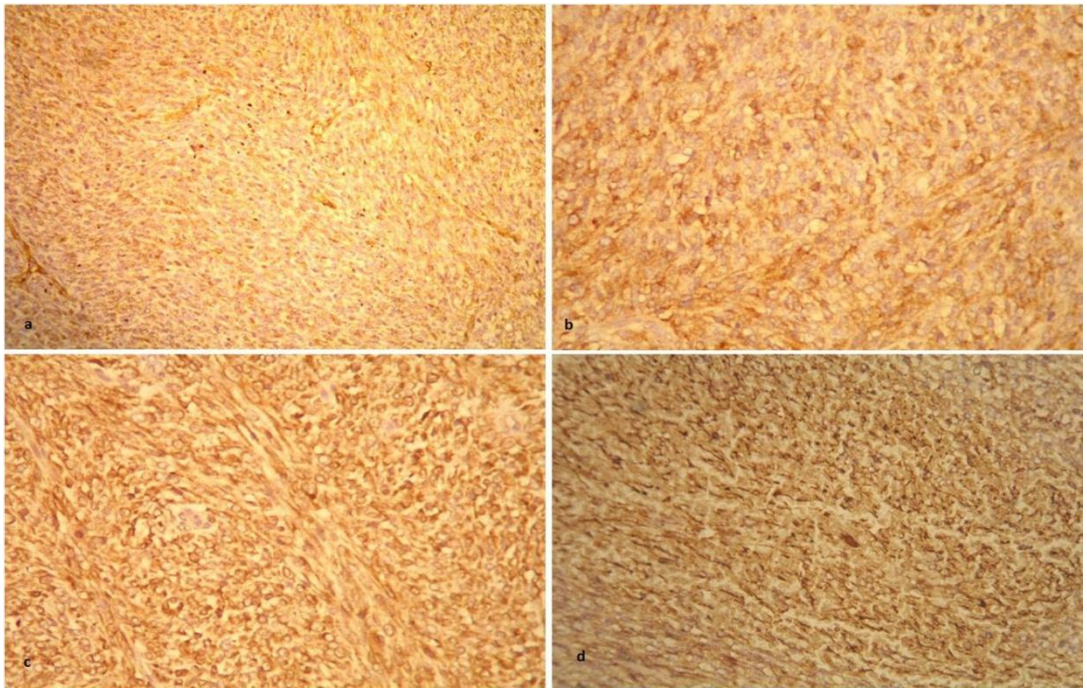

Figure 2a - Tumor cells showing positivity for Vimentin
Figure 2b - Tumor cells showing membranous positivity for EMA
Figure 2c - Tumor cell showing strong cytoplasmic positivity for Bcl-2
Figure 2d - Tumor cells showing positivity for CD99

Based on these findings, a diagnosis of monophasic synovial sarcoma was made. Further work up included a CECT chest which revealed two well defined nodules with ground glass attenuation in the anterior segment of right upper lobe and right middle lobe of lung, suggestive of metastasis.

Patient was started on chemotherapy with doxorubicin and ifosphamide. Following three cycles of chemotherapy, the patient has been doing well.

3. Discussion

Synovial sarcomas are uncommon soft tissue tumours accounting for 5-10% of the soft tissue sarcomas. They occur both in the paediatric and adult age groups and represent the most common non-rhabdomyosarcoma soft tissue sarcomas in children. Approximately 95% of SS occur in the para-articular regions of the extremities, usually in close association with tendon sheaths, bursae and joint capsules.[1,2,3]

The common presenting features of synovial sarcoma include a palpable mass or swelling, associated with pain in almost half of the patients. The sites of involvement are the lower extremities (60%), with a predilection of area around the knee, the upper extremities (23%), the head and neck region(9%), and the abdominal wall or retroperitoneum (8.1%).[2,4] SSoccuring in the inguinal region are exceedingly rare. Exhaustive search revealed a handful of cases.[5,6,7]

In our case, the patient presented with an inguinal mass which was clinically suspected to be hernia or abscess. The differential diagnoses of such a mass in a young female include both benign and malignant entities. Benign causes include inguinal hernia, haematoma, abscess, neurofibroma, leiomyoma and desmoid tumour. The most common clinical differential is inguinal hernia, as in our case. Malignant lesions include liposarcoma, fibrosarcoma, leiomyosarcoma, synovial sarcoma, epithelioid sarcoma and rhabdomyosarcoma.[5]

Two major histologic subtypes of SS exist: monophasic and biphasic. The classical SS has a biphasic appearance with varying proportions of epithelial and spindle cells components. The epithelial cells may form glands or papillary structures, solid cords, nests or rounded clusters.[1] The monophasic SS consists solely of sarcomatous components and its diagnosis is often challenging. It may also present as a poorly differentiated round cell sarcoma often arranged in a pericytomatous pattern, representing a form of tumour progression rather that a distinct subtype.[1] Immunohistochemistry is a useful diagnostic tool. Positive immunohistochemical staining for cytokeratin or epithelial membrane antigen and vimentin helps in the diagnosis of monophasic synovial sarcoma. Other markers, including CD99, CD56, and Bcl-2, may display positivity. Synovial sarcomas are genetically characterized by the presence of a specific chromosomal translocation, t(X;18)(p11;q11), resulting in formation of either of the two fusion genes: *SYT-SSX1* or *SYT-SSX2*.

Primary treatment of synovial sarcoma is surgical resection. Adjuvant radiation therapy and chemotherapy are increasingly being used, especially for patients with tumours larger than 5 cm.[4] Prognosis is poor with local recurrence in 28% to 49% of patients and metastases in almost 50% of the patients. Five-year survival rates range from 36% to 51%. Young age, female gender and tumour size less than 5 cm are features associated with favourable prognosis. There is no difference in survival between the monophasic and biphasic subtypes. The presence of lung metastases implies an unfavourable prognosis.[1,4]

4. Conclusion

Monophasic synovial sarcoma in inguinal region is an extremely rare entity. The present case was clinically diagnosed as inguinal hernia. The knowledge of occurrence of synovial sarcoma in unusual locations is necessary to avoid its misdiagnosis as a benign lesion which may lead to delay in treatment and tumour metastasis.

References

[1] Weiss SW, Goldblum JR. Synovial sarcoma. In: Weiss SW, Goldblum JR, editors. Enzinger and Weiss's soft tissue tumours. 5th ed. London: Mosby; 2008, p. 1161-82.

[2] Xu J, Wang J, Cui L, Wu X. Malignant inguinal monophasic synovial sarcoma: report of a case and review of the literature. *World J Surgical Oncol* 2010; 8:102.

[3] Sultan I, Rodriguez-Galindo C, Saab R, Yasir S, Casanova M, Ferrari A. Comparing children and adults with synovial sarcoma in the Surveillance, Epidemiology and End Results Program, 1983 to 2005: an analysis of 1268 patients. *Cancer* 2009; 115: 3537–47.

[4] White BE, Kaplan A, Lopez-Terrada DH, Ro JY, Benjamin RS, Ayala AG. Monophasic synovial sarcoma arising in the vulva: a case report and review of the literature. *Arch Pathol Lab Med* 2008; 132(4): 698-702.

[5] Bhosale PR. The Inguinal Canal: Anatomy and Imaging Features of Common and Uncommon Masses. *Radio Graphics* 2008; 28:819–835.

[6] Lee PJ, Chu CH, Shieh SJ. Groin synovial sarcoma with intraluminal femoral sheath involvement. *Form J Surg* 2014; 47(1): 36-41.

[7] Karplus G, Krasin MJ, Rodriguez-Galindo C, McCarville B, Jenkins J, Rao B *et al*. Retrospective study of the surgical management and outcome of nonrhabdomyo sarcoma soft tissue sarcomas of the groin and axilla in children. *J Pediatr Surg* 2009; 44:1972-1976.