



WESTERN NIGERIA JOURNAL OF MEDICAL SCIENCES (WNJMS)

**GIANT SYNOVIAL SARCOMA OF THE POSTERIOR THIGH IN AN ADULT AFRICAN
MALE: A CASE REPORT**

GIANT SYNOVIAL SARCOMA OF THE POSTERIOR THIGH IN AN ADULT AFRICAN MALE: A CASE REPORT

Babalola OF^{1,2}, Salawu AI^{1,2}, Okunlola AI^{1,2}, Erinomo OO^{3,4}, Omoseebi O^{3,4}

1. Department of Surgery, Federal Teaching Hospital, Ido – Ekiti, Nigeria
2. Department of Surgery, Afe Babalola University, Ado – Ekiti, Nigeria
3. Department of Morbid Anatomy, Federal Teaching Hospital, Ido – Ekiti, Nigeria
4. Department of Morbid Anatomy, Afe Babalola University, Ado – Ekiti, Nigeria

Correspondence to: Babalola OF, Department of Surgery, Afe Babalola University Ado – Ekiti, Nigeria.
Telephone: +234-8033858309; **E-mail:** olakunleabs@yahoo.com

ABSTRACT

Introduction: Synovial sarcoma represents 5-10% of adult soft tissue sarcomas and is noted to have an aggressive biological behavior. In sub-Saharan Africa, the diagnosis is rarely made largely due to scarcity, high cost and inaccessibility of immunohistochemical facilities.

Case Report: A 65-year-old man presented with 5-month history of a painless progressive posterior right thigh swelling with no impairment of ambulation. Examination revealed a huge mass of the posteromedial thigh measuring 32cm x 24cm x 16cm in the widest dimensions. A provisional diagnosis of soft tissue sarcoma was made and was supported by the magnetic resonance imaging findings of heterogenous consistency of the mass. He had a wide local excision done with a gross tumor free margin and split thickness skin grafting of the residual defect at a second surgery 3 weeks after. The histopathological diagnosis was initially categorized under the broader classification of undifferentiated pleomorphic sarcoma with histologically free tumor margin noted. Immunohistochemical staining further characterized the lesion as poorly differentiated synovial sarcoma. The post-operative recovery was initially smooth with a well-healed defect. However, the patient declined further treatment and died 4 months post-tumor excision from an early aggressive recurrence.

Conclusion: The occurrence of an aggressive, rarely diagnosed, synovial sarcoma in an adult African man is highlighted. A fatal outcome from an aggressive early recurrence occurred despite histologically clear resection margins.

Keywords: synovial sarcoma; soft tissue sarcoma; posterior thigh mass; adult; African

INTRODUCTION

Synovial sarcoma accounts for 5-10% of adult soft tissue sarcomas.^{1,2,3} Proportionately, soft tissue sarcoma represents less than 1% of all malignancies. Hence, synovial sarcoma constitutes approximately 0.05 – 0.1% of malignancies. Synovial sarcoma is believed to arise from primitive mesenchymal tissues of tendon sheaths, bursae, fascia, membranes, etc. and rarely in association with synovium of joints.² It derived its name from the synovial like appearance of the cells on microscopy and is commonly found in the extremities of young adults (15-40 years) with no gender or racial predilection.⁴

It is a rare and aggressive malignancy that requires skilled histopathological, immunohistochemical and cytogenetic facilities for its unequivocal diagnosis and subsequent management. In Sub-Saharan Africa, where these facilities are sparsely distributed and rarely of the state of the art, the diagnosis of synovial sarcoma presents enormous challenges.⁵ Immunohistochemical

analysis of soft tissue sarcoma received at Lagos University Teaching Hospital, Nigeria over a period of five years showed that Kaposi sarcoma was the commonest soft tissue sarcoma and no single case of synovial sarcoma was found.⁶ Similarly, Ukekwe et al. noted that synovial sarcoma is hardly reported in the subcontinent of West Africa prior to their case report of a mediastinal primary tumor occurrence.¹ While it is known that synovial sarcoma can develop at any anatomical site, about 80% of this tumor occur primarily in the extremities with only 20% favoring non-extremity sites.² In this case report, we present the rare occurrence of synovial sarcoma of the posterior thigh in a sexagenarian male Nigerian highlighting the peculiarity of late presentation, unfavorable pathology and poor prognosis.

CASE PRESENTATION

A 65-year-old man presented to the plastic surgery out-patient clinic with a painless,

progressively increasing right thigh swelling of 5 months duration. He had no associated anorexia or weight loss. There was no preceding trauma or other swellings. He had no chronic cough, breathlessness, bone pains or jaundice. He had no history of tobacco smoking or alcohol intake and there was no co-morbid illness. He was not on any routine medication and there was no known drug allergy.

On physical examination, there was a 32cm x 24cm x 16cm mass on the posteromedial aspect of the right thigh involving the proximal three-fourth of the thigh and encroaching the right gluteal fold. The mass had a smooth surface with hyperpigmentation of the central area of the overlying skin. It was non-tender, had well defined edges and was hard in consistency. The mass was attached to the overlying skin and underlying hamstring muscles (Figure 1). There were no peripheral lymph nodes enlargement, and the neurovascular status of the limb was intact.



Figure 1: Clinical images of the posterior thigh synovial sarcoma. a. pre-operative clinical photograph showing the huge mass in the posterior thigh. b. the specimen with adherent overlying skin following wide local excision. c. three weeks post excision defect ready for coverage. d. three weeks after split thickness skin grafting of post excision defect

Ultrasonography showed a heterogenous hypoechoic mass in the posterior compartment of the right thigh. Magnetic resonance imaging revealed an encapsulated mass in the posterior compartment of the right thigh with some cystic and solid components (Figure 2). Plain radiograph of the right thigh did not reveal any bony involvement while plain chest radiograph showed normal findings. Abdominal ultrasound scan did not reveal liver or intraperitoneal metastasis.

He had a wide local excision of the mass with 5cm gross tumor free margin under General Anaesthesia with Endotracheal Intubation. The intra-operative finding was an encapsulated soft tissue tumor attached to the skin and encasing the bellies of some of the posterior thigh muscles. The sciatic nerve, adhered to the anterior surface of the mass by loose areolar tissue was easily freed and covered with the salvaged, uninvolved segments of the biceps femoris. The excised tumor mass weighed 3.8kg and measured 37cm x 27cm x14cm.

Cut sections on pathologic examination showed greyish-brown variegated and haemorrhagic surfaces as well as focal cystic degeneration containing gelatinous materials. Microscopically, histopathologic sections showed tumor mass comprising an admixture of spindle shaped fibroblast cells arranged in a storiform pattern within myxoid stroma. The tumor cells show varying degree of pleomorphism, hyperchromatism and mitotic activity. Multinucleated bizarre tumor giant cells were present. Diffuse infiltrates of mixed inflammatory cells comprising predominantly polymorphs were also seen. The superior, inferior, lateral, medial and deep resection margins were free of tumor cells. Histopathologic diagnosis was reported as undifferentiated pleomorphic sarcoma.

Immunohistochemistry showed tumor cells positive for pancytokeratin AE 1/3 and vimentin. Tumor cells were negative for desmin, S100 and synaptophysin. Overall features were those of poorly differentiated synovial sarcoma (Figure 3).

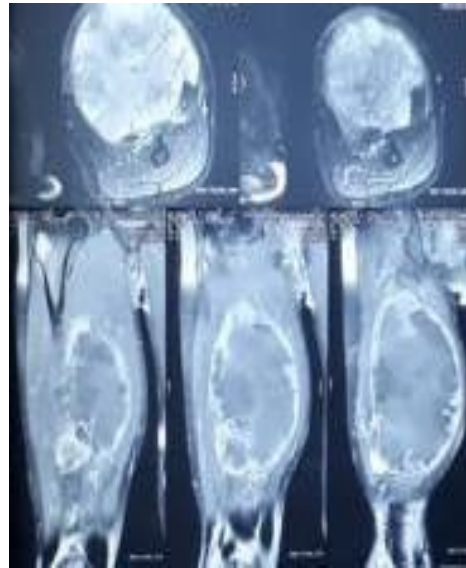


Figure 2: Magnetic resonance imaging (MRI) showing an encapsulated mass in the posterior compartment of the right thigh with some cystic and solid component

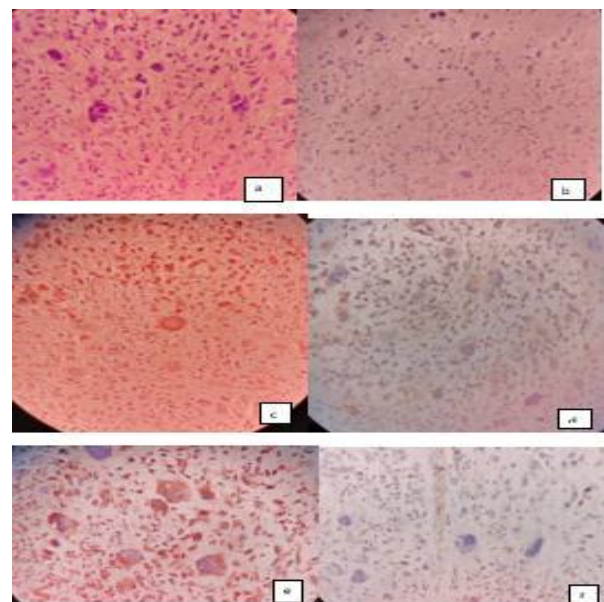


Figure 3: Photomicrograph (x40 magnification) of the histology and immunohistochemical studies a. photomicrograph of the histology showing malignant tumor cells b. photomicrograph showing negative immunohistochemical staining for desmin. c. photomicrograph showing positive immunohistochemical staining for pancytokeratin d. photomicrograph showing negative immunohistochemical staining for synaptophysin e. photomicrograph showing positive immunohistochemical staining for vimentin f. photomicrograph showing negative immunohistochemical staining for S10

Three weeks post-tumor excision, he had coverage of the defect with split thickness skin graft harvested from the contralateral thigh. Graft take was satisfactory and there was no evidence of recurrence at 6th week post-excision when he was discharged to the surgical out-patient clinic for follow up and adjuvant chemotherapy. The patient declined further treatment and did not honor the follow-up appointments. The wife was however contacted on his mobile phone number 6 months after hospital discharge; she disclosed that he had tumor recurrence 4months post-operation with associated bleeding from the tumor bed and died 2weeks post-emergence of the recurrence.

DISCUSSION

Synovial sarcoma, first reported in 1893, represents the fourth most common histological type of soft tissue sarcoma.^{4,7} Typically, three different histological subtypes are identifiable; the monophasic (50-60%) composed of mainly spindle cells, the biphasic ones (10 – 30%) has an admixture of spindle and epithelial cellular components, and the poorly differentiated subtype (15-25%) characterized by epithelioid morphology, high mitotic activity and areas of geographic necrosis.⁷ Expectedly, this latter subtype, discovered in our patient is also associated with poor prognosis.

Synovial sarcoma can occur in any anatomical region of the body including glandular organs with 80 -85% found in the lower extremities particularly the knee region.^{2,4,8} They are often proximally located, subfascial and deep seated within the extremities.⁹ Our index patient, a 65year old man, had a proximal, deep seated giant tumor involving the hamstring muscles and resting on the sciatic nerve with no gross motor or sensory deficit at presentation. Even though the clinico-radiologic and pathologic features of synovial sarcoma have been well characterized, they are not pathognomonic. These features can be mimicked by other soft tissue sarcomas such as Ewing Sarcoma, Peripheral Nerve Sheath

Tumour, Undifferentiated Pleomorphic Sarcoma (UPS) and other Spindle cell sarcomas.⁷

Immunohistochemical and cytogenetic studies are invariably needed to establish the diagnosis. In this index patient, the appearance of heterogenous tumor with cystic degeneration on magnetic resonance imaging (MRI) and the characteristic histological findings suggested an initial diagnosis of UPS. Malignant Fibrous histiocytoma, now grouped as UPS were identified by Adigun et al. and Ayandipo et al. as the 2nd and 3rd most common soft tissue sarcoma respectively in two different Sub-Saharan retrospective studies.^{5,10} The presence of tumor cells positive expression of keratin in conjunction with spindle cell histological appearance is diagnostic of synovial sarcoma.⁷ Further immunohistochemical studies in our patient revealed positivity of tumor cells for pancytokeratin AE and vimentin which confirmed the diagnosis of synovial sarcoma. It is therefore instructive that with more widely accessible and affordable immunohistochemical facilities in Sub-Saharan Africa, more reports of synovial sarcoma diagnosis may emerge from this region. Additionally, unequivocal confirmation of synovial sarcoma requires cytogenetic molecular studies. The occurrence of t(X:18) chromosomal translocation and fusion of SYT-SSX genes have been reported as a consistent hallmark of synovial sarcoma.^{7,11} Our patient was financially constrained and molecular diagnostic facilities are still sparsely distributed in our sub-region, hence, the inability to investigate this important genetic make-up of the tumor cells.

The current mainstay of treatment of extremity synovial sarcoma is wide local excision with a minimum of 3cm gross tumor free margin as a planned curative resection to reduce the incidence of loco-regional recurrence.^{8,12} The index patient's huge tumor, possibly the largest size ever recorded, occurred in the lax and accessible tissues of the posterior thigh permitting a curative resection of 5cm gross tumor free margin except on the anterior surface loosely adhered

to the sciatic nerve. Split thickness skin grafting of the residual defect was uneventfully done three weeks post resection when the histologically clear margin was ascertained.

Notably, tumor size $\geq 5\text{cm}$ has been linked with adverse prognosis and mortality in a malignancy traditionally known for aggressive biological behavior.^{8,13} The recognition of this ominous outlook emphasizes the need for adjuvant treatment despite a histologically clear margin of resection. While the role of adjuvant radiotherapy in reducing locoregional recurrence and improving overall survival is established in extremity soft tissue sarcoma, the impact of adjuvant chemotherapy remains uncertain.¹² However, some studies have advocated the use of ifosfamide-based regimen in improving the disease free survival for patients with synovial sarcoma and even regarded it as the recommended treatment for metastatic disease.⁸ Our patient however declined further treatment despite adequate counselling. The financial and psychological exhaustion following an apparently successful resection might have been partly responsible coupled with the overall grim prognosis of the pathology. It is however unknown if adjuvant radiotherapy and chemotherapy would have prolonged the disease-free survival of this patient considering the early recurrence. The need for a collaborative and multimodal approach to the care of patients with soft tissue sarcoma as advocated by several authors cannot be overemphasized.^{8,12}

CONCLUSION

This report has highlighted the occurrence of synovial sarcoma in the elderly age group in Sub-Saharan Africa with unfavorable pathology and poor prognosis particularly following a unimodal form of treatment. There is need to improve the diagnostic facilities, build experience and support in the management of this disease.

Patient's Informed Consent

Informed verbal consent obtained from patient for academic publication of this case report prior to discharge from hospital.

Conflict of Interest: Nil

Source of funding: Nil

REFERENCES

1. Ukekwe FI, Ezemba N, Olusina DB, Igbokwe U, Ngene C. Giant primary synovial sarcoma of the anterior mediastinum: A case report and review of literature. *Niger J Clin Pract.* 2016; 19(2): 293-297.
2. Eilber FC, Dry SM. Diagnosis and Management of Synovial Sarcoma. *J Surg Oncol.* 2008; 97(4):314-320.
3. Mahesh KT, Ponnuswamy IA, David MP, Shivhare P, Puttaranganayak MI, Sinha P. Synovial sarcoma of the buccal mucosa: a rare case report. *Case Rep Dent.* 2013; 2013:938291. doi.org/10.1155/2013/938291.
4. Vlok SSC, Wagener GWW, Zaharie D. Synovial sarcoma. *S Afr J Radiol.* 2014; 18(2):a704. doi:10.4102/sajr.v18i2.704.
5. Adigun IA, Rahman GA, Buhari MO, Ogundipe KO, Omotayo JA. Soft-Tissue Sarcoma in Black Africans: Pattern, Distribution and Management Dilemma. *J Natl Med Assoc.* 2007; 99:88-93
6. Ikeri NZ, Akinjo AO, Ajayi OO, Banjo F, Adekunbiola A. Histological Types of Soft-Tissue Sarcomas at the Lagos University Teaching Hospital. *Nig Postgraduate Med J.* 2017; 24(4):205-209. DOI: 10.4103/npmj.npmj_146_17
7. Murphey MD, Gibson MS, Jennings BT, Crespo-Rodriguez AM, Fanburg-Smith J, Gajewski DA. Imaging of Synovial Sarcoma with Radiologic-Pathologic Correlation. *Radiographics.* 2006; 26(5):1543-65.
8. Heckman J, Witkiewicz A, Berger AC. Cystic Synovial Sarcoma of the Lower Extremity: A Case Report and

- Review of the Literature. Surg Sci. 2011; 2:62-5
9. Jones BC, Sundaram M, Kmansdorf MJ. Synovial Sarcoma: MR Imaging Findings in 34 Patients. AJR 1993; 161:827-830.
 10. Ayandipo OO, Afuwape OO, Soneye OY, Orunmuyi AT, Obajimi GO. A sub-Saharan African experience in the surgical management of soft tissue sarcomas in an oncology unit in: a retrospective cohort study. Pan Afr Med J. 2019; 33:207-17.
 11. Rekhi B, Jambhekar NA, Desai SB, Basak R, Puri A, Agrawal M. A t(X; 18) SYT-SSX2 positive synovial sarcoma in the pelvis of a young adult male: a rare case report with review of literature. Indian J Cancer. 2008; 45(2):67-71.
 12. Gamboa AC, Gronchi A, Cardona K. Soft-tissue sarcoma in adults: An update on the current state of histiotype-specific management in an era of personalized medicine. CA Cancer J Clin. 2020; 70(3):200-229.
 13. Vilanova JC. WHO Classification of Soft Tissue Tumors. In: Vanhoenacker FM, Parizel PM, Jan L. Gielen JL Eds. Imaging of Soft Tissue Tumors. 4th edition. New York: Springer. 2017; pp187-196.