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CASE REPORT

Alveolar Soft Part Sarcoma in a 44-year-Old Female: A Case from Uganda

Charles Malisaba Posite[1,2], Biruk Legesse Tadesse[1], Mirna Batista Santos[1], Raymond Atwine[1,3]

[1]Department of Pathology, Kampala International University, Ishaka, Uganda; [2]Department of Pathology, Université Catholique du Graben, Butembo, Democratic Republic of the Congo; [3]Department of Pathology, Mbarara University of Science and Technology, Mbarara, Uganda

Correspondence: Charles Malisaba Posite, Email charles.malisaba@studwc.kiu.ac.ug

Background: Alveolar Soft Part Sarcoma (ASPS) is a rare, highly vascular soft tissue sarcoma characterized by the specific ASPSCR1-TFE3 fusion, typically affecting adolescents and young adults. Despite its often-indolent growth, it has a high propensity for late hematogenous metastasis. Management for localized disease is centered on wide surgical excision.

Case Presentation: We report a case of ASPS in a 44-year-old female from the Ankole tribe, presenting with a slow-growing 8 cm mass in her right upper arm over two years. This age of presentation is outside the typical peak incidence. The diagnosis was established solely on classic histopathological features-a pseudoalveolar pattern, large polygonal cells with abundant eosinophilic cytoplasm, and PAS-positive, diastase-resistant intracytoplasmic granules-due to resource limitations precluding molecular confirmation (TFE3 IHC/FISH). She underwent wide local excision with negative margins and remains free of recurrence at 12 months post-surgery.

Conclusion: This case highlights the occurrence of ASPS in an older-than-typical patient and underscores the enduring reliability of classic histopathology for definitive diagnosis in resource-limited settings like Uganda. Although surgically cured locally, the patient's large tumor size and age confer a high risk for future metastasis, mandating rigorous, long-term surveillance.

Keywords: Alveolar Soft Part Sarcoma, soft tissue sarcoma, case report, histopathology, Uganda, upper extremity

Introduction

Alveolar Soft Part Sarcoma (ASPS) is a rare and enigmatic soft tissue sarcoma (STS) of indeterminate histogenesis, accounting for fewer than 1% of all STS diagnoses.[1,2] Since its initial characterization by Christopherson et al in 1952, it has been recognized for its distinct clinicopathological and molecular features.[3] Epidemiological studies, including analyses of large databases, show that ASPS predominantly affects adolescents and young adults, with a median age at diagnosis in the third decade of life and a clear predilection for females.[4–6] Clinically, it typically manifests as a slow-growing, painless, and highly vascular mass in the deep soft tissues. The most common primary site is the lower extremities, particularly the thigh, head and neck, trunk, and upper extremities.[2,4,6] Despite its often indolent growth, ASPS has a profound propensity for early hematogenous metastasis, with up to 50% of patients presenting with or developing metastatic disease, most frequently in the lungs, brain, and bone.[7,8]

The diagnostic pathway for ASPS integrates advanced imaging and definitive pathological analysis. ASPS classically appears as a soft tissue mass with high signal intensity on both T1- and T2-weighted images of magnetic resonance imaging (MRI), and prominent signal voids due to its rich vascular network.[2,9] Definitive diagnosis relies on histopathology, which reveals a pathognomonic pseudoalveolar or organoid architecture. The tumor is composed of large, polygonal cells with abundant, granular, eosinophilic cytoplasm and round-to-oval nuclei. These cells contain characteristic intracytoplasmic, PAS-positive, diastase-resistant rhomboid or rod-shaped crystals.[2,3,10] The molecular hallmark of ASPS is a specific, non-reciprocal translocation, t(X;17) (p11.2;q25), which fuses the Alveolar Soft Part Sarcoma Chromosome Region Candidate 1 (ASPSCR1) gene to the Transcription Factor E3 (TFE3) gene.[4,6] The resultant

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ASPSCR1-TFE3 fusion protein acts as an aberrant transcription factor, driving tumorigenesis and serving as a highly specific diagnostic marker detectable by TFE3 immunohistochemistry or fluorescence in situ hybridization (FISH).[2,9]

Management of localized ASPS is centered on wide surgical excision with negative margins, which remains the only curative modality.[1] The tumor is notoriously resistant to conventional cytotoxic chemotherapy and radiation therapy, with response rates being exceedingly low.[2,6] For advanced, unresectable, or metastatic disease, the therapeutic landscape has evolved significantly. Given the tumor's hypervascular nature, systemic treatment now focuses on anti-angiogenic agents. Multi-targeted tyrosine kinase inhibitors (TKIs) that inhibit the vascular endothelial growth factor receptor (VEGFR) have demonstrated significant clinical activity. Cediranib, in a Phase 2 trial, showed a 35% objective response rate and a median progression-free survival of 10.7 months.[1,6] Similar efficacy has been reported in retrospective and prospective studies for sunitinib and pazopanib.[4,6] More recently, immune checkpoint inhibitors have shown promise, with a phase 2 trial of atezolizumab demonstrating an objective response rate of 42% in treatment-naïve patients.[2,10] Despite these advances, the overall prognosis for ASPS remains guarded, especially in the metastatic setting. Key negative prognostic factors include age over 25, male sex, tumor size greater than 5 cm, and the presence of metastases at diagnosis.[1,9] Long-term survival rates are poor, underscoring the aggressive biology of the disease despite its indolent presentation.[3,6] We present a case of ASPS in a 44-year-old female belonging to Ankole tribe, notable for its late-age presentation and its definitive diagnosis based exclusively on classic histopathology in a low-resource environment.

Case Presentation

A 44-year-old female of the Ankole tribe presented to the Surgical Department at Kampala International University Teaching Hospital with a primary complaint of a mass in the anterior aspect of her right upper arm, which had been present for two years. The lesion was initially small and asymptomatic but had progressively enlarged, recently causing compressive symptoms in the distal forearm. The patient denied any constitutional symptoms such as fever, weight loss, or night sweats. Her personal and family medical histories were non-contributory for malignancy.

On physical examination, the patient's vital signs were stable and within normal physiological limits. A firm, non-tender mass measuring approximately 8×6 cm with irregular margins was palpated in the anterior compartment of the right upper arm. There was no associated erythema or warmth of the overlying skin. Neurological and vascular examinations of the distal limb were intact. An ultrasound scan was performed, which delineated a soft tissue mass with high vascularity and irregular, infiltrative margins relative to surrounding tissue planes.

A decision was made for surgical intervention. A wide local excision of the mass was performed under general anesthesia. Intraoperatively, the mass was well circumscribed. No gross invasion of adjacent neurovascular bundles or bone was observed. Hemostasis was secured, and the surgical bed was irrigated. No palpable regional lymphadenopathy was noted during the procedure. The resulting defect was closed primarily. The blood loss was minimal. The specimen was submitted for histopathological analysis.

The Histopathology Department received a single, unoriented, ovoid tissue specimen measuring 8×6×4 cm. Gross examination revealed a firm, white-gray tissue with interspersed brown and yellow areas. The cut surface was notably solid and white, featuring distinct nodular brown foci. Microscopic examination with Hematoxylin and Eosin (H&E) staining revealed a neoplastic proliferation of polygonal to round cells arranged in a distinctive pseudoalveolar and pseudoglandular pattern, separated by thick fibrous septa containing sinusoidal vascular channels. The tumor cells were characterized by abundant granular, eosinophilic cytoplasm and round to oval nuclei, some of which contained prominent nucleoli. Periodic Acid-Schiff (PAS) staining was positive and demonstrated diastase-resistant intracytoplasmic granules. The closest surgical margin measured 0.3 cm from the tumor. All other margins were clear with distances varying from 0.5 to 1 cm. Therefore, all margins were free from the tumor (Figure 1). These histomorphological features were consistent with a definitive diagnosis of Alveolar Soft Part Sarcoma.

The patient had an uneventful postoperative course. She was enrolled in a review program of two years: three-month interval for the first year, then, six-month interval for the second year. At the latest follow-up, 12 months post-surgery, the patient was free of local recurrence. She had no complaint.

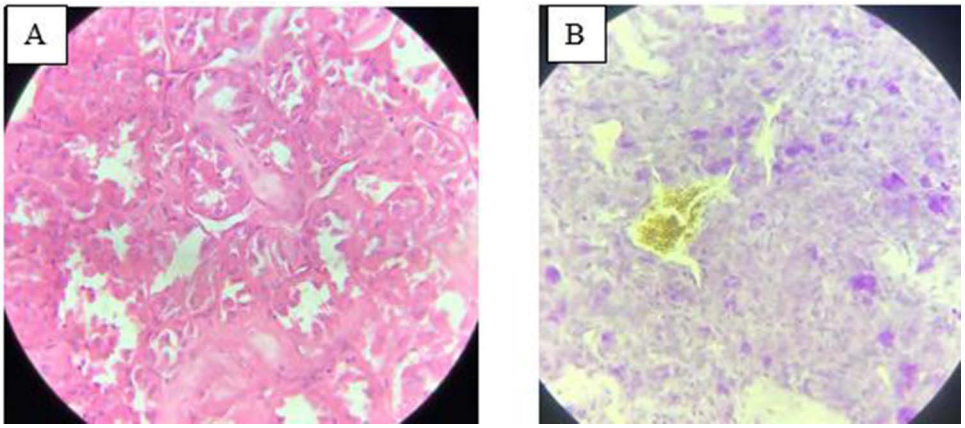

Figure 1 H&E-stained sections showing neoplastic proliferation of polygonal to round cells arranged in a distinctive pseudoalveolar and pseudoglandular pattern. The tumor cells are characterized by abundant granular, eosinophilic cytoplasm and round to oval nuclei, some of which contain prominent nucleoli (A; x400). PAS staining is positive and demonstrates diastase-resistant intracytoplasmic granules (B; x400).

Discussion

We present a case of ASPS in a 44-year-old female, which is noteworthy for its presentation in an age group outside the typical adolescent and young adult peak. This report underscores the classic histopathological features that enable diagnosis, even in settings where advanced molecular testing is unavailable, and highlights the clinical management considerations dictated by the tumor's well-established natural history.

The demographic and clinical features of this case present both typical and atypical aspects when compared to established literature. The patient's sex is consistent with the known female predominance of ASPS.[5–7] However, at 44 years of age, she is considerably older than the median age of onset, which is typically between 15 and 35 years.[6,8,11] While less common, the diagnosis of ASPS in middle-aged and older adults is well-documented, and some studies suggest that older age at diagnosis may be an independent adverse prognostic factor.[3,9,11] The tumor's location in the upper extremity is also less common than in the lower limbs but represents a significant site of origin, accounting for approximately 20–30% of cases in some series.[2,4,6] The clinical history of a slow-growing, initially asymptomatic mass spanning two years is a classic feature of ASPS, often leading to a delayed diagnosis and presentation with a large tumor burden, as seen in this patient with an 8 cm mass.[3]

The modern diagnostic pathway relies heavily on advanced imaging, with MRI being the gold standard for evaluating soft tissue masses, including ASPS.[9,12] The mass in ASPS typically exhibits high signal intensity on T1- and T2-weighted images and prominent signal voids due to its high vascularity.[2,9] Furthermore, the lesion frequently displays a characteristic ovoid configuration identifiable on MRI, which is consistent with the specimen's gross morphology in our case. While the ultrasound in our setting indicated hypervascularity, confirming the need for surgical excision, this feature can also be seen in other vascular lesions.[6,9,12]

The diagnostic process in this case relied on foundational clinical, radiological, and pathological methods. The ultrasound finding of a hypervascular mass is a key characteristic of ASPS, reflecting its rich sinusoidal vascular network.[9] This feature is critical for the differential diagnosis but can also be misleading, as ASPS may mimic vascular malformations or other hypervascular tumors.[6,9] The definitive diagnosis was secured by histopathology. The observed features (a pseudoalveolar architecture, large polygonal cells with abundant eosinophilic granular cytoplasm, and the presence of PAS-positive, diastase-resistant granules) are pathognomonic for ASPS.[9] These intracytoplasmic granules correspond to the crystalline structures that are a hallmark of this sarcoma.[5,9] In a resource-equipped setting, this diagnosis would typically be confirmed by demonstrating the underlying molecular alteration, the ASPSCR1-TFE3 gene fusion, most commonly via immunohistochemistry for TFE3 nuclear expression or by FISH analysis.[9] However, due to our limited resources, the immunohistochemistry could not be done. The strong correlation between the classic histology and the specific translocation means that in settings like Uganda, a confident diagnosis can be made based on morphology alone, highlighting the enduring importance of expert histopathological interpretation.[6,9]

The management and prognosis of this patient must be considered in light of established prognostic factors. The primary treatment, wide local excision, is the undisputed standard of care for localized ASPS and offers the only potential for cure.[3,5] The success of surgery is heavily dependent on achieving negative histological margins, as positive margins are a strong predictor of local recurrence.[5,6] For this patient, two significant adverse prognostic indicators are present: a tumor size greater than 5 cm and an age older than 25 years.[3,4] These factors place her at a high risk for developing distant metastases despite successful local control. The indolent but relentless metastatic potential of ASPS necessitates rigorous, long-term surveillance, primarily involving serial chest imaging to detect pulmonary metastases, which are the most common site of distant spread.[3,9]

Currently, there is no established role for adjuvant systemic therapy for high-risk, localized ASPS following complete resection.[1] However, given the high risk of relapse, this remains an area of active investigation. Should this patient develop metastatic disease, the treatment paradigm has shifted from ineffective conventional chemotherapy to targeted therapies.[6,9] The profound vascularity of ASPS provides a strong rationale for using anti-angiogenic agents, and multiple studies have confirmed the clinical activity of TKIs such as sunitinib, pazopanib, and cediranib in the metastatic setting.[4,6,7,9] Furthermore, emerging data on the efficacy of immune checkpoint inhibitors, which may leverage the unique tumor microenvironment of ASPS, offer a promising new therapeutic avenue for patients with advanced disease.[1] The management of sarcoma in low- and middle-income countries (LMICs) presents unique challenges, including limited access to advanced imaging, molecular diagnostics, and novel systemic therapies, making complete surgical resection even more critical and heightening the need for adaptable surveillance protocols.[2,3]

Conclusion

This case is notable for the diagnosis of ASPS in a 44-year-old female, an age group outside the usual peak incidence. Most critically, the definitive diagnosis relied exclusively on classic histopathology (pseudoalveolar architecture and PAS-positive crystals), highlighting the enduring and fundamental role of expert morphological diagnosis in LMICs where molecular confirmation is unavailable. Despite achieving complete surgical clearance, the presence of significant adverse prognostic factors (age > 25 years and tumor size > 5 cm) underscores the necessity for rigorous and adaptable long-term surveillance protocols, which are paramount given the unique challenges of follow-up and advanced therapy access in such resource-constrained environments.

Ethics Approval

Institutional ethical approval was not required to publish the case details.

Consent for Publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Author Contributions

All authors made a significant contribution to the work reported, took part in drafting and reviewing the article, gave final approval of the version to be published, have agreed on the journal to which the article has been submitted, and agree to be accountable for all aspects of the work.

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Disclosure

The authors declare that they have no conflicts of interest.

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