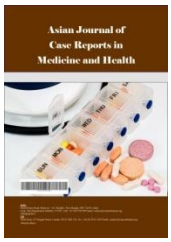

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Epithelioid Sarcoma of Hand Masquerading as Radial Styloid Osteomyelitis: A Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Epithelioid Sarcoma of the hand is a rare, high grade soft tissue sarcoma. It accounts for less than 1% of soft tissue sarcoma. It is often misdiagnosed and is a great mimicker of multiple diseases due to its deceptively harmless appearances during the initial stages of the disease.

We are presenting a case of a 24-year-old gentleman presented to our centre with history of nodular swelling and pus discharge over radial side of left wrist for 3 months. He was diagnosed with osteomyelitis of left radial styloid and treated with 6 weeks of antibiotic after wound debridement. He presented to us 2 months later with worsening wound over surgical site for which he underwent wound debridement and another 6 weeks course of antibiotics. Patient presented to us 2 weeks later with complaint of swelling and bleeding from operative site. On examination we noted a wound measuring 3x3 cm with friable granulation tissues over radial aspect of left wrist and multiple nodular lesions over left palm with haemoserous discharges. X ray of left wrist showed juxta-articular osteopenia with focal bone lysis. Patient underwent wound debridement and intraoperative histopathological sample sent came back as Grade 3 epithelioid sarcoma. Patient underwent above elbow amputation with postoperative radiotherapy after 3 months being treated for osteomyelitis of radial styloid.

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This case should alert treating surgeons to include soft tissue sarcoma as differential diagnosis of harmless looking, occasional painful firm nodules localised on the hand and wrist region of young adults especially when there are osteolytic changes in x-rays.

Keywords: Epithelioid sarcoma; soft tissue sarcoma.

1. INTRODUCTION

Epithelioid sarcoma of the hand is often misdiagnosed as other benign or malignant conditions. This disease typically presents on the distal end of upper limbs, in male who are in second and third decades of their lives [1]. Overall 5- and 10-years survival rates are 70% and 42% respectively [2]. We are presenting a case of a 24 year old gentleman who came to us for a total of 3 times with initial impression of left radial styloid osteomyelitis in which incision and drainage and wound debridement were done before eventually during third visit came with bleeding, ulceration and multiple firm nodular satellite lesions which hinted us the possibility of malignancy and histopathological result which showed epithelioid sarcoma and we proceeded with above elbow amputation and planned for radiotherapy postoperatively.

2. PRESENTATION OF CASE

A 24 years old gentleman otherwise well and healthy presented to our centre initially with history of nodular swelling and pus discharge over radial side of left wrist for 3 months. He was diagnosed with osteomyelitis of left radial styloid. Inflammatory markers were all elevated. X ray of left wrist showed juxta-articular osteopenia and focal bone lysis as shown in Fig. 1a and 1b. Patient underwent wound debridement and completed 6 weeks course of antibiotics. Patient then presented to us 2 months post operatively with worsening wound over previous surgical site for which he underwent wound debridement and another 6 weeks course of antibiotics. He presented to us 2 weeks later for the third time with complaint of swelling and bleeding of the previous operative site. On examination, we noted a wound measuring 3x3 cm with friable granulation tissues over radial aspect of left wrist and multiple small nodular satellite lesions with rolled edges over left palm with haemoserous discharge as shown in Fig. 2a and 2b. Inflammatory markers were all elevated. X ray of left wrist showed juxta-articular osteopenia, focal bone lysis and periosteal reactions. MRI of left wrist and forearm showed soft tissue mass involving the hand, wrist joint and forearm with no significant bony infiltration. Soft tissue malignancy needs to be considered. Patient underwent wound debridement and intraoperative histopathological sample sent came back as Grade 3 epithelioid sarcoma (high grade sarcoma). The histopathological specimen showed tumour tissue covered with stratified squamous epithelium. It is composed of sheets of malignant cells with extensive tumour necrosis and suppuration (>50%; score: 2). The malignant

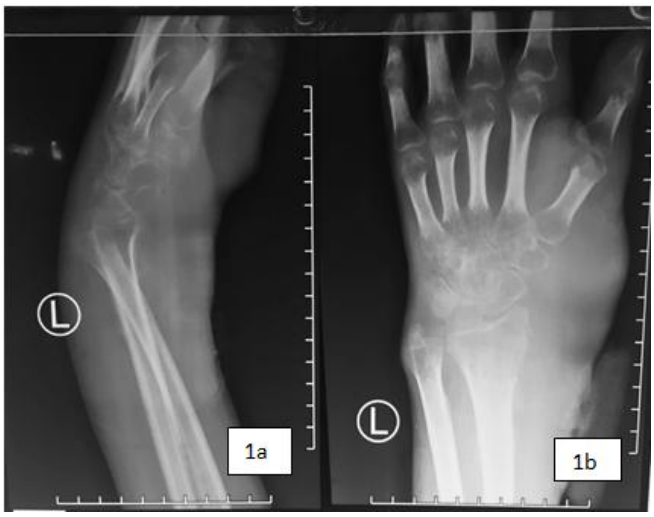

Fig. 1a and 1b showing X rays of left wrist taken during first presentation showing juxta-articular osteopenia, focal bone lysis and periosteal reactions

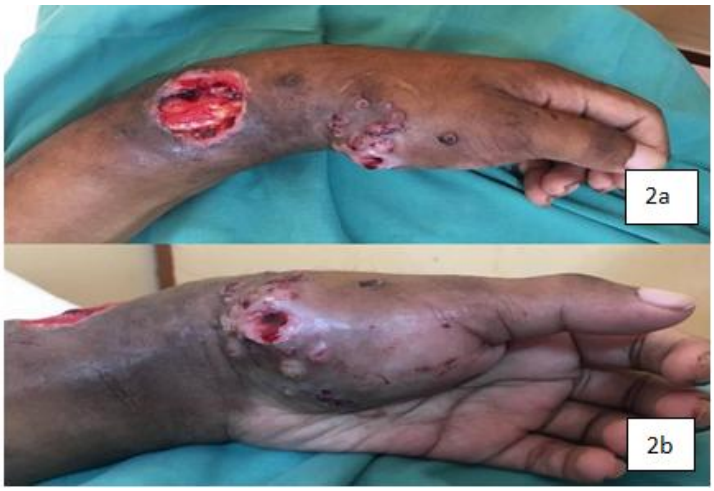

Fig. 2a and 2b showing photographic images of left wrist showing previous operative site and multiple firm nodular satellite lesions with rolled edges over radial side of left wrist

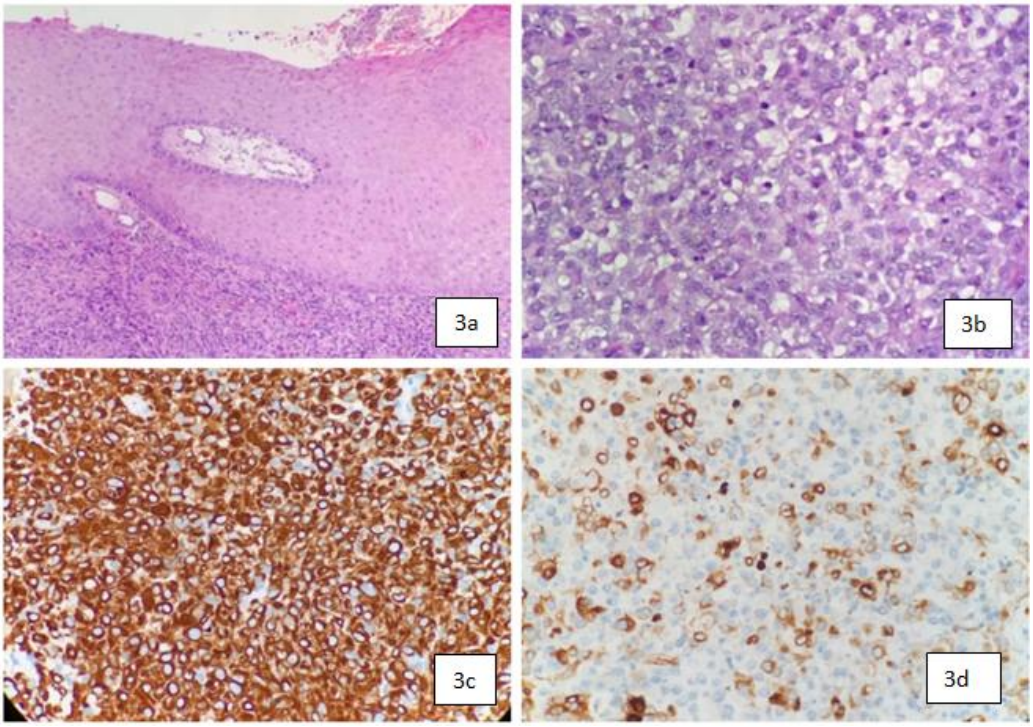

Fig. 3a and 3b showing histopathological images showing tumour tissue covered with stratified squamous epithelium. The malignant cells exhibit large cells with abundant clear cytoplasm, marked nuclear pleomorphism, vesicular nuclei and prominent nucleoli
Fig. 3c and 3d showing immunohistochemical staining positive for vimentin and epithelial membrane antigen respectively

cells exhibit large cells with abundant clear cytoplasm, marked nuclear pleomorphism, vesicular nuclei and prominent nucleoli as shown in Fig. 3a and 3b. Mitotic figures are frequently seen (15/10 HPF; score: 2). No lymphovascular invasion. Immunostains show positive for epithelial membrane antigen, Pan-CK, CD10, vimentin, CK8/18, CD34, cylin D1 and focal positivity for synaptophysin as shown in Fig. 3c and 3d; negative for S100, Desmin, actin, caldesmon, CD99, CD56, CD30, CD45, CD31, chromogranin, CD117, CK5/6, CK20, CK7 and myogenin. Ki67 shows 70% proliferative index. FNCLCC grading has a total score of 7/8 which

corresponds to Grade 3 (high grade sarcoma). Patient underwent above elbow amputation with postoperative radiotherapy after 3 months being treated for osteomyelitis of radial styloid.

3. DISCUSSION

Epithelioid sarcoma is a rare, malignant condition, first described by Enzinger in 1970. He found out that the tumour tends to grow slowly and frequently misinterpreted on histopathology and misdiagnosed as granulomatous disease, synovial carcinoma, wart, ulcerating squamous cell carcinoma, amelanotic melanoma, clear cell sarcoma, epithelioid hemangioendothelioma or metastasis from occult epithelial tumours [3]. In epithelioid sarcoma, the epithelioid cells in the lesions are larger and more sharply defined than in granulomas. The epithelioid cells are also more eosinophilic in staining and less mature in nature. It can be differentiated from squamous cell carcinoma by the absence of keratin pearls and dyskeratosis in the surrounding epithelium.

Jawad, et al. identified 441 cases, and the male-to-female ratio was 1.309:1 (250:191) with the major peak incidence in the 17- to 60-years age range [4]. Epithelioid sarcoma can be histologically categorized into classical, spindle and mixed forms. Furthermore, the tumour can be classified as two recognised subtypes, the distal type and the less frequent proximal type based on their location. Both entities are predominant in young male adults. Distal-type epithelioid sarcoma has a high tendency to occur in the extremities, especially in the upper limb. Conversely, proximal-type epithelioid sarcomas most commonly affect trunk or deep tissue sites. Tumour grading of epithelioid sarcoma is based on the Federation Nationale des Centres de Lutte Contre le Cancer system (FNCLCC) and is considered as a relevant prognostic factor [5,6,7]. Based on the reported literature apparently the proximal epithelioid sarcomas have a more aggressive clinical course and worse outcomes [8]. Adverse prognostic factors include male sex, older age, and proximal/axial location [9].

Epithelioid sarcoma has a high local recurrence rate (87%) and frequent late metastasis (30%). Baratti et al reported that up to 50% of cases develop metastatic lesions, especially in the lung and regional lymph nodes [10]. The tumour often presents as superficial nodules or ulcers. However, deep seated lesions can occur rarely. Like other soft tissue sarcomas, tissue biopsy is the diagnostic modality of choice which should be supported with ancillary use of immunohistochemical antibodies. It exhibits immunohistochemical reactivity for epithelial markers such as keratins and epithelial membrane antigen, and for mesenchymal markers such as vimentin and CD34 [11]. Tissue biopsy of epithelioid sarcoma shows typical histological features of distinct nodular aggregates of epithelioid and spindle cells with zonal necrosis. Approximately, 50% of cases of epithelioid sarcoma show CD34 and epithelial membrane antigen positivity. Desmin and S100 negativity eliminates rhabdomyosarcoma and melanoma as possible differential diagnoses, while CD31, Factor VIII, and FL- 1 are useful in vascular tumours [12]. Immunohistochemical markers like Ki67 can also be used for determine the proliferation rate [13]. There is no documented specific radiological diagnostic finding, though tumour extent and metastatic foci can be demonstrated.

Several hypotheses emerge regarding the origin and tumorigenesis of epithelioid sarcoma. It has been proposed that epithelioid sarcoma is a mesenchymal tumour derived from primitive mesenchymal or myofibroblastic cells [14]. Another hypothesis argues that epithelioid sarcoma originates from naive synovial cells and represents a variant of synovial sarcoma [15]. Modena et al. showed that, analogous to malignant rhabdoid tumours, proximal epithelioid sarcoma exhibits an allele deletion of the tumour suppressor gene SMARCB1/INI1, suggesting the possibility that epithelioid sarcoma belongs to the rhabdoid tumour family [16]. Zhang, et al. found that epithelioid cells arrange in a fascicular and sheet-like pattern, similar to the patterns of mesenchymal cell-derived tumours. Meanwhile, the positivity for the epithelial markers cytokeratin and epithelial membrane antigen reflects their epithelial phenotype. They also propose that epithelioid sarcoma derives from mesenchymal stem or progenitor cells, followed by the malig-nant transformation through the mesenchymal-epithelial transition [17]. The transitioning tumour cells are capable of co-expressing epithelial and mesenchymal markers [18].

Prognosis of the disease is dependent on the depth of tumour in relation to the deep fascia, local recurrence and regional lymph node involvement. The size of the primary lesion is not a reliable indicator of prognosis. However, smaller tumours are associated with significantly better distant metastasis free interval. Distal

limbs also have been reported to have better prognosis than proximal limb and axial tumours.

Adequate treatment requires wide or radical local excision, which may be combined with adjuvant radiotherapy or chemotherapy [19]. Elective regional lymph node dissection has been suggested because lymph node metastasis is a fairly common recurrence. Chemotherapy are recommended for metastatic disease with standard regimen comprises of ifosfamide or doxorubicin, sometimes in combination. Amputation can be considered if there are multiple recurrences or there is no significant loss of function [20]. Our patient underwent above elbow amputation followed by radiotherapy.

4. CONCLUSION

Epithelioid sarcoma is a great mimicker of various diseases. Early recognition and treatment are essential for better outcome for patients. This case should alert treating surgeons to include soft tissue sarcoma as a differential diagnosis of harmless looking, occasional painful firm nodules localised on the hand and wrist region of young adults especially when there are osteolytic changes in x-rays.

CONSENT AND ETHICAL APPROVAL

Informed consent and ethical approval has been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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