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Case Study of the Month

Proximal Type Epithelioid Sarcoma of the Scrotum: A Source of Diagnostic Confusion That Needs Immediate Attention

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Abstract

Epithelioid sarcoma is a rare soft-tissue sarcoma which classically presents as a subcutaneous or deep dermal mass in distal extremities of adolescents and young adults. A more aggressive ''proximal'' or ''large-cell'' type has been described in rare cases to occur as a deep soft-tissue mass at proximal body sites which tends to recur and metastasize earlier than the conventional epithelioid sarcoma. The correct diagnosis of this type is essential since these lesions can easily be misdiagnosed as other epithelioid lesions. Its prognostic factors also have not yet been fully investigated. We herein report a case of metastatic proximal-type epithelioid sarcoma arising from the scrotum of a 50-year-old man managed by radical surgery and adjuvant chemotherapy.

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1. Case report

A 50-year-old man presented with an ulcerating mass on the left hemiscrotum that arose as a small ulcerating lesion 2 weeks prior to admission. The patient had received local treatments in different clinics and was admitted based on the fast progression of the ulcerating process. He had no history of primary or metastatic carcinoma. On physical examination, a 10-cm ulcerating mass arising from the left hemiscrotum was noted (Fig. 1). The testes were normal on palpation. General physical examination and blood biochemistry results were also unrevealing. Results of a computed tomography (CT) scan of the abdomen and the chest were reported as normal. Debridement of the necrotic tissues with incisional biopsy of the scrotal lesion was performed. Cultures for aerobic, anaerobic, and mycotic pathogens as well as investigations for mycobacteria were negative. The patient was then offered

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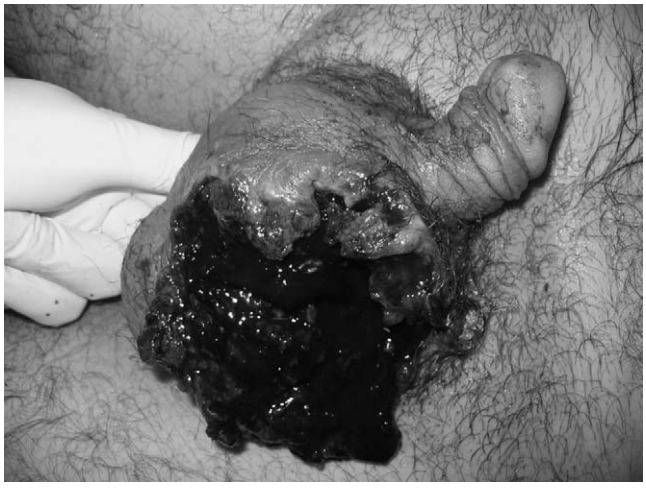

Fig. 1 – Ulcerating mass arising from the left hemiscrotum during initial presentation.

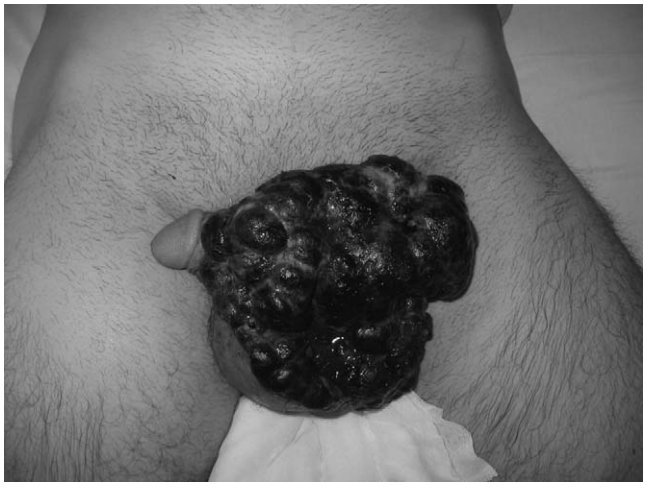

Fig. 2 – The lesion is shown to grow to a huge size extremely quickly.

extensive surgery, but he refused immediate surgery and was discharged when the wound healed. He returned 2 weeks later with a 15-cm vegetating and fragile mass originating from the previous site and extending to the right hemiscrotum and base of the penis (Fig. 2). A 3 × 2-cm firm, enlarged lymph node was palpated at the left groin. Repeat CT of the abdomen did not show any pathology although two peripheral nodules of 1.5 cm each were demonstrated at the base of the left lung on chest CT. The patient then underwent extensive surgery with complete resection of the scrotum bearing the huge mass and scrotal contents with a safety margin of 1–1.5 cm as well as dissection of the left 3 × 2-cm

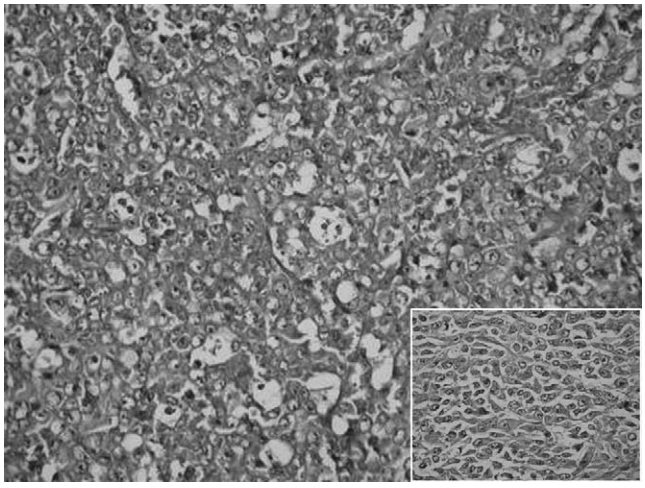

Fig. 3 – Histopathologic examination of the primary lesion demonstrates malignant tumor cells with epithelioid areas and rhabdoid phenotype (*inset*). Hematoxylin & eosin staining, ×400.

superficial inguinal lymph node. The mass was in close proximity to the corpus spongiosum at the penile base but no macroscopic invasion was evident; therefore, the penis was preserved. The perineal defect was covered with right superficial epigastric artery-based inguinal flap and the penis was covered with split-thickness skin graft harvested from the left thigh. Results of the histopathologic examination are shown in Fig. 3. Immunohistochemical studies revealed diffuse positivity with vimentin, patchy positivity with cytokeratin, CD99, and CD34. Immunoreactivity was negative with S100, HMB45, melan-A, CD31, factor VIIIa, desmin, and smooth muscle actin (SMA).

2. EU-ACME Question

Please visit www.eu-acme.org to answer the below EU-ACME question on-line (the EU-ACME credits will be attributed automatically). The answer will be printed after next month's *Case Study of the Month*.

Question:

What is the most likely diagnosis in this patient?

A. Blastomyces infection of the scrotum
B. Paratesticular rhabdomyosarcoma
C. Proximal-type epithelioid sarcoma
D. Malignant melanoma