Proximal-type epithelioid sarcoma - Case report*

Sarcoma epitelióide tipo proximal - Relato de caso

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Abstract: Epithelioid sarcoma, first described by Enzinger in 1970, is a rare soft-tissue sarcoma typically presenting as a subcutaneous or deep dermal mass in distal portions of the extremities of adolescents and young adults. In 1997, Guillou et al. described a different type of epithelioid sarcoma, called proximal-type epithelioid sarcoma, which is found mostly in the pelvic and perineal regions and genital tracts of young to middle-aged adults. It is characterized by a proliferation of epithelioid-like cells with rhabdoid features and the absence of a granuloma-like pattern. In this paper we present a case of proximal-type epithelioid sarcoma with an aggressive clinical course, including distant metastasis and death nine months after diagnosis.
Keywords: Diagnosis; Diagnosis, differential; Sarcoma; Therapeutics

Resumo: O sarcoma epitelióide, primeiramente descrito por Enzinger, em 1970, é uma neoplasia de partes moles que ocorre principalmente nas extremidades distais de adolescentes e adultos jovens. Em 1997, Guillou e cols. descreveram um tipo diferente de sarcoma epitelióide, que afetava frequentemente a região pélvica, períneo e áreas genitais de pacientes de média idade, com exame histológico caracterizado pela proliferação de células com aspecto epitelióide. Neste trabalho, descreve-se caso de paciente que apresentava há três meses duas lesões na região glútea, cujo exame histológico confirmou diagnóstico de sarcoma epitelioide do tipo proximal, já com presença de metástases pulmonares e cerebrais e que foi a óbito nove meses após o diagnóstico.
Palavras-chave: Diagnóstico; Diagnóstico diferencial; Sarcoma; Terapêutica

INTRODUCTION

Epithelioid sarcoma, first described by Enzinger in 1970, is a rare soft-tissue sarcoma typically presenting as a subcutaneous or deep dermal mass in distal portions of the extremities of adolescents and young adults. It is a slowly growing neoplasm with a strong propensity for local recurrence and, ultimately, metastasis.[1] Microscopically, most tumors are characterized by a granuloma-like pattern: nodules of spindled and epithelioid cells circumscribe areas of central hyalinization and necrosis. Fibrous histiocytoma-like and angiomatoid subtypes have also been reported as less common histologic variants.[2] In 1997, Guillou et al. described a different type of epithelioid sarcoma, called proximal-type epithelioid sarcoma, which is found mostly in the pelvic and perineal regions and genital tracts of young to middle-aged adults and is characterized by a proliferation of epithelioid-like cells with rhabdoid features and the absence of a granuloma-like pattern.[3]

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CASE REPORT

In 2009, a 25-year-old man was referred to the dermatology department with a 3-month history of a rapidly growing painful mass on his right buttock which subsequently ulcerated. Skin examination revealed an erythematous infiltrated plaque and an ulcerated lesion with fibrinous exudate in the gluteal region (Figure 1). There were no lymphadenopathies or other abnormalities on physical examination. A skin biopsy of the ulcerated lesion revealed proliferation of pleomorphic epithelioid cells with eosinophilic cytoplasm and vesicular nuclei with prominent nucleoli. Scattered rhabdoid cells characterized by abundant glassy cytoplasm, eccentric nuclei and prominent nucleoli were observed throughout the lesion (Figure 2). Immunohistochemistry showed positivity for vimentin, cytokeratin, EMA, CAM 5.2, MIB and CD34 for the latter marker positivity was focal confirming the diagnosis of proximal-type epithelioid sarcoma (Figure 3). Blood count and serum biochemistry were unremarkable, and culture for mycobacteria, fungi

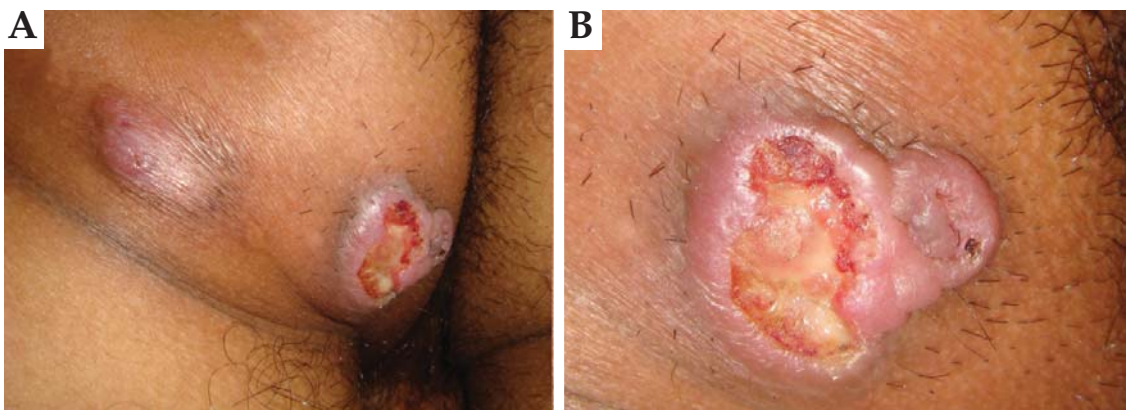

Figure 1: Two lesions located on the gluteus: the first, which is 6 cm in diameter, is ulcerated and has elevated, infiltrated edges; the second, a plaque, is reddish-purple with a fibrous consistency and 4 cm in diameter

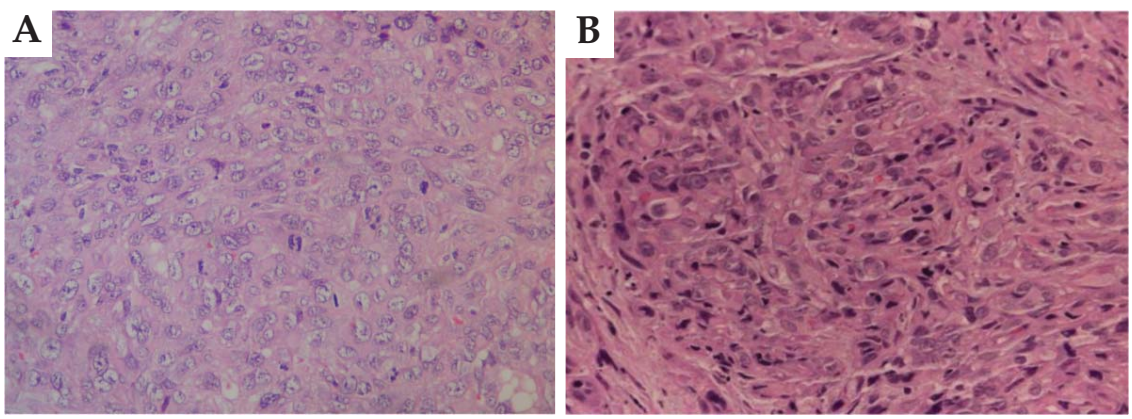

Figure 2: Epithelioid cells with moderate pleomorphism, eosinophilic cytoplasm and prominent vesicular nuclei and scattered rhabdoid cells with hyaline intracytoplasmic inclusions displacing the nucleus eccentrically

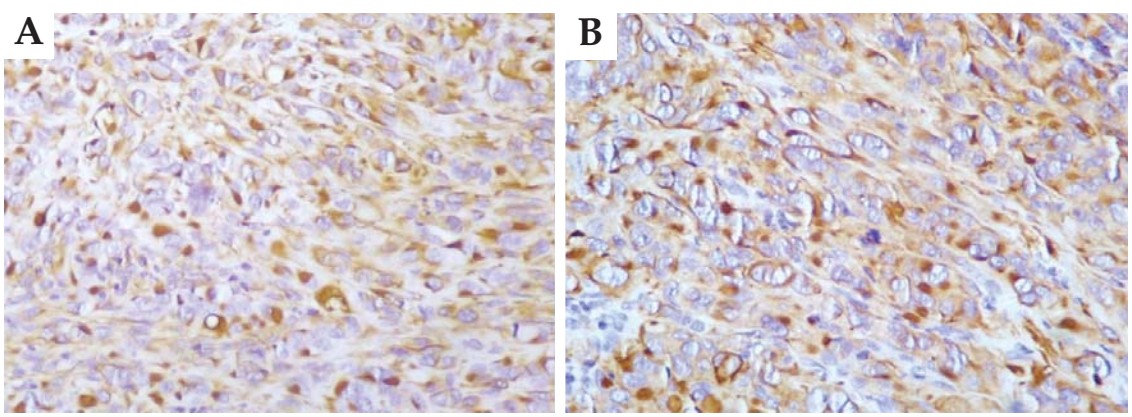

Figure 3: Epithelioid and rhabdoid cells exhibiting positivity for vimentin and cytokeratin

and aerobic bacteria were negative. At the time of diagnosis, the tests to define clinical staging, which included a computed tomography scan, revealed numerous metastatic lesions in the brain and lungs (Figure 4). The patient received chemotherapy at a reference oncology institute but died in the ninth month of follow-up.

DISCUSSION

The clinical, morphological and immunohistochemical features of the case described here are in accordance with those of proximal-type epithelioid sarcoma. Clinically, the proximal-type variant differs from the classic form of epithelioid sarcoma in that it frequently occurs in older patients in a proximal/axial often deep-seated location, preferentially involving the pelvic, perineal and genital regions, it is apparently more aggressive and metastasizes earlier than the conventional distal-type epithelioid sarcoma[4] Histologically, it tends to be characterized by predominantly epithelioid cells, marked cytologic atypia, the frequent occurrence of rhabdoid features and the absence of a granuloma-like pattern.[5] All variants of epithelioid sarcoma co-express vimentin and cytokeratin. The histogenetic origin of epithelioid sarcoma is unknown, but a hypothesis that it originates from

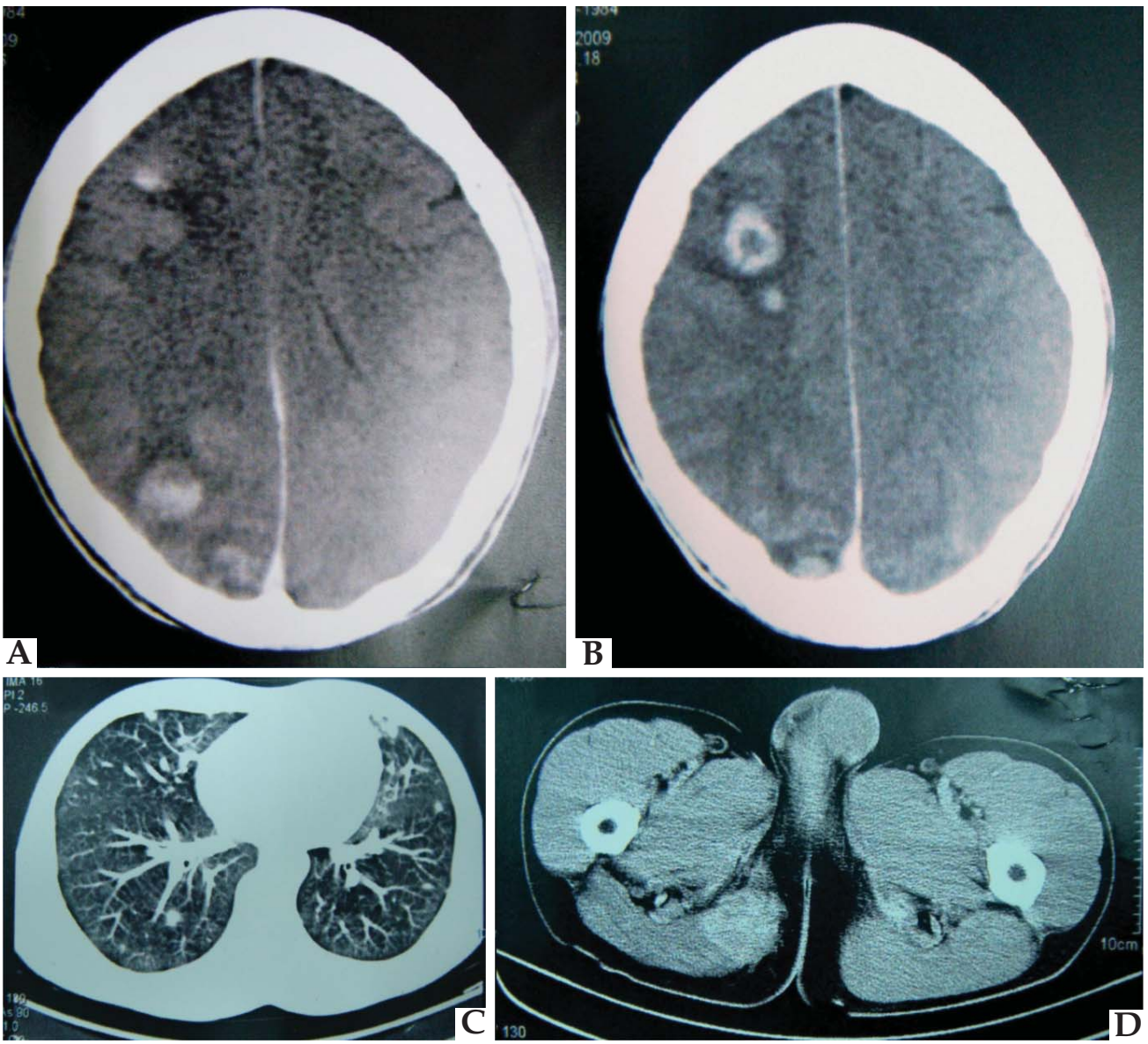

Figure 4: Tomography scans showing lung and brain metastases

mesenchymal cells undergoing epithelial differentiation has been favored.[6,7] Differential diagnosis of proximal-type epithelioid sarcoma includes epithelioid malignant peripheral nerve-sheath tumor, clear cell sarcoma, epithelioid hemangioendothelioma, angiosarcoma, epithelioid leiomyosarcoma, synovial sarcoma, rhabdomyosarcoma and metastatic carcinoma. In an endemic area for leishmaniasis, it should also be differentiated from an ulcerated lesion with infiltrated edges caused by leishmaniasis.[8] In this paper we present a case of proximal-type epithelioid sarcoma with an aggressive clinical course, including distant metastasis and death nine months after diagnosis. ❑

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