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Caso aislado

"Proximal-type" epithelioid sarcoma: A case report with immunohistochemical study

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RESUMEN

Introducción: Se presenta un caso de tumor de partes blandas de patrón epitelioide, con rasgos clínicos, histológicos e inmunohistoquímicos acordes con la recientemente descrita "variante proximal" de sarcoma epitelioide. Material y métodos: Un varón de 28 años debuta con un tumor profundo localizado en la región perineal. Resultados: Microscópicamente, mostró hábito epitelioide, con marcada atipia citológica y frecuentes rasgos rabdoides, siendo el patrón de inmunorreactividad superponible al del sarcoma epitelioide clásico. Conclusiones: Ante un tumor con rasgos intermedios entre un sarcoma epitelioide clásico, un tumor rabdoide y un carcinoma indiferenciado, hay que pensar en la variante proximal del sarcoma epitelioide. **Rev Esp Patol 2000; 33(3): 245-249.**

Palabras clave: Sarcoma epitelioide - Tumor rabdoide - Inmunohistoquímica

SUMMARY

We report a case of an epithelioid-appearing malignant soft-tissue tumor with clinical, histological and immunohistochemical features consistent with the recently described entity "proximal-type" epithelioid sarcoma. A 28-year-old man presented with a deep-seated tumor in the perineal region. The tumor had an epithelioid appearance, marked cytonuclear atypia and frequent rhabdoid features. The immunoreactivity observed was in keeping with the pattern of reactivity of classic epithelioid sarcoma, including reactivity for carcinoembryonic antigen. The tumor displayed features resembling those of an epithelioid sarcoma and those of a rhabdoid tumor or undifferentiated carcinoma. We concluded that the described features best fit with a proximal type of epithelioid sarcoma. **Rev Esp Patol 2000; 33(3): 245-249.**

Key words: Epithelioid sarcoma - Rhabdoid tumor - Immunohistochemistry

INTRODUCTION

"Proximal-type" epithelioid sarcoma is a newly described variant of conventional epithelioid sarcoma (1), the morphology of which ranges from that of atypical epithelioid sarcoma to that of a rhabdoid tumor or undifferentiated carcinoma.

We report a case of an aggressive malignant soft tissue neoplasm, which had been misdiagnosed as a rhabdomyosarcoma, in the perineum of a young adult. The

clinical, morphological and immunohistochemical features of the tumor were consistent with the proximal variant of epithelioid sarcoma described by Guillou *et al.* (1). The characteristic immunophenotype of epithelioid sarcoma is emphasized (1-8).

CASE REPORT

A 28-year-old man with no previous history of neoplasm presented with a tumor in the perineal region. A local excision was performed at another hospital, at which time the diagnosis of rhabdomyosarcoma was made. The report described a deep-seated, patternless proliferation of cells with rhabdoid morphotype. The tumor invaded the deep soft tissues and had no contact with the skin. Immunohistochemistry was not performed. No histological sections of the tumor were available. The patient underwent chemotherapy.

Three years after the initial presentation, a local recurrence was biopsied at our hospital and considered to be epithelioid sarcoma. He was treated with surgical resection. Eight months after the second excision, he showed extensive local recurrence.

MATERIALS AND METHODS

The material used corresponded to the first recurrence. Formalin-fixed, paraffin-embedded tissue was subjected to immunohistochemical analysis using monoclonal antibodies to the following antigens: vimentin (Dako, clone V9, 1:500); cytokeratin AE1/AE3 (Dako, 1:400); epithelial membrane antigen (Dako, 1:100); carcinoembryonic antigen (Dako, prediluted); CD34 (Biogenex, prediluted); muscle-specific actin (Dako, 1:400); myoglobin (Dako, 1:200); desmin (Biomeda, prediluted); S-100 protein (Dako, 1:300); and factor VIII-related antigen (Dako, 1:20). Tissue sections for immunohistochemistry were developed using the avidin-biotin-peroxidase complex method.

RESULTS

Gross findings

The tumor developed within the deep soft tissues of the perineum and ischiorectal region and appeared as multiple confluent white nodules scattered throughout the fibrofatty and muscular tissue.

Light microscopic findings

On microscopic examination, the tumor showed an infiltrating pattern and invaded the deep soft tissues. Uncohesive aggregates of epithelioid cells with rhabdoid features were present with areas of compartmentation. Pseudoalveolar, adenoid and myxoid areas and isolated irregularly shaped nodules with central necrosis (Fig. 1) were also present. These cells were large, poorly cohesive and had abundant, eosinophilic cytoplasm and a vesicular nucleus with prominent nucleoli (Fig. 2). In other areas, solid layers of cohesive cells displayed an undifferentiated carcinoma-like appearance. The nuclei were pleomorphic and hyperchromatic (Fig. 3). Mitotic count ranged from 10 to 35 mitoses per 10 high-power fields (HPF). Areas of geographic necrosis and perineural invasion were observed. There was no associated spindle-cell component.

Immunohistochemical findings

All the tumor cells were diffusely positive for keratin, vimentin, CD34 and epithelial membrane antigen (Fig. 4). Areas of undifferentiated carcinoma-like appearance showed patchy membranous staining for carcinoembryonic antigen. Positively staining rhabdoid cells were observed focally. None demonstrated positivity for desmin, smooth-muscle actin, myoglobin, S100 protein and factor VIII-related antigen.

DISCUSSION

The differential diagnosis was of epithelioid-appearing malignant soft-tissue neoplasms (9), including the following: epithelioid malignant peripheral nerve sheath tumor, melanoma, epithelioid rhabdomyosarcoma, epi-

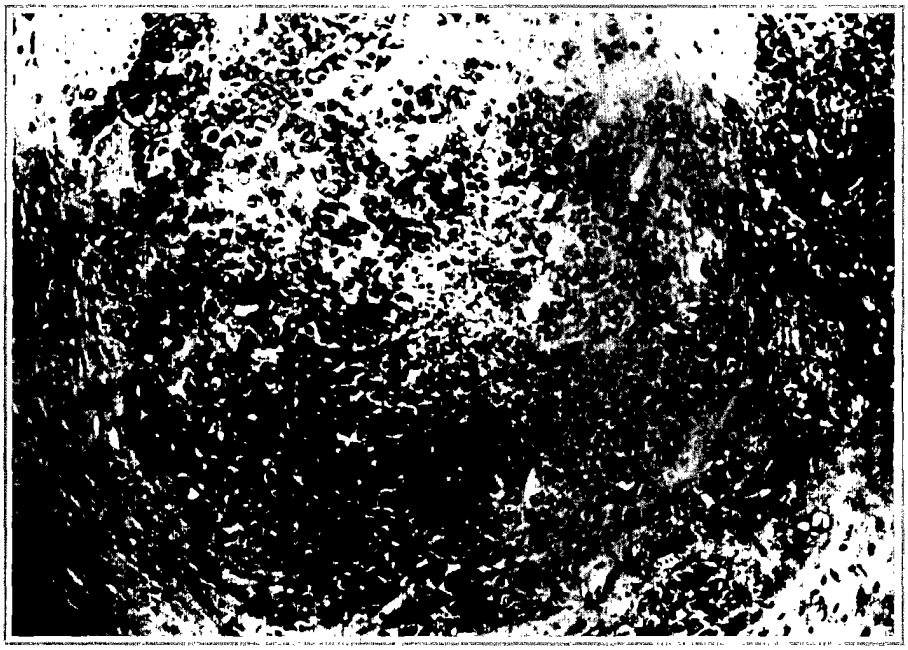

Figure 1. Irregularly shaped nodules with central necrosis were observed focally (original, HE ×100).

thelioid monophasic synovial sarcoma, undifferentiated carcinoma, extrarenal rhabdoid tumor and conventional epithelioid sarcoma. Most of these were discarded on the basis of immunohistochemical results, with the exception of extrarenal rhabdoid tumor, undifferentiated carcinoma and conventional epithelioid sarcoma.

The so-called extrarenal rhabdoid tumor seemed to be a phenotype rather than an entity (10-13). This diagnosis was based on the cytologic resemblance of the constituent cells to those of classic malignant rhabdoid tumor of the kidney, characterized by filamentous cytoplasmic inclusions, macronucleoli and abundant cyto-

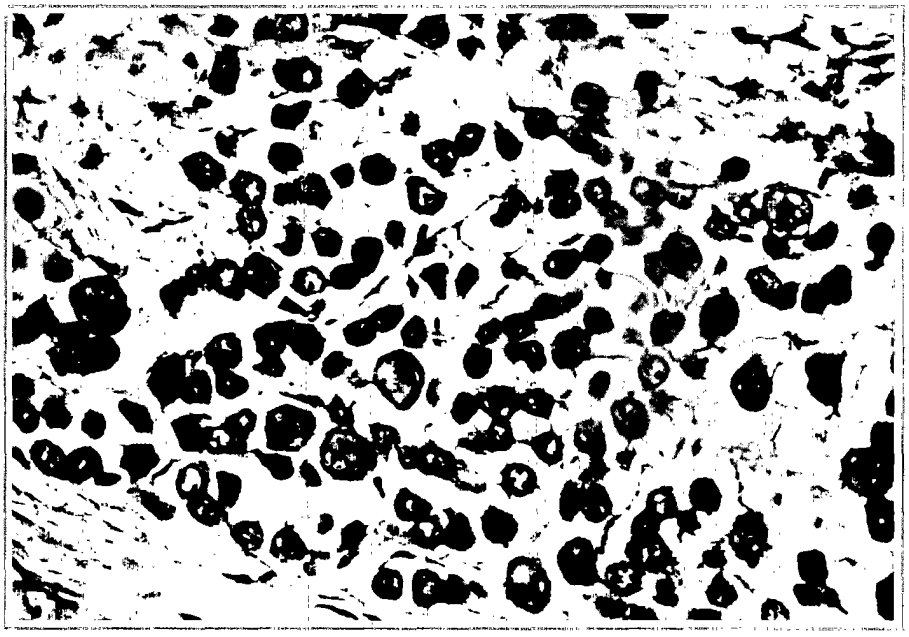

Figure 2. Most tumor cells show rhabdoid features, *i.e.*, the presence of abundant, eosinophilic cytoplasm displacing the nucleus, and vesicular nucleus with prominent nucleoli (original, HE ×200).

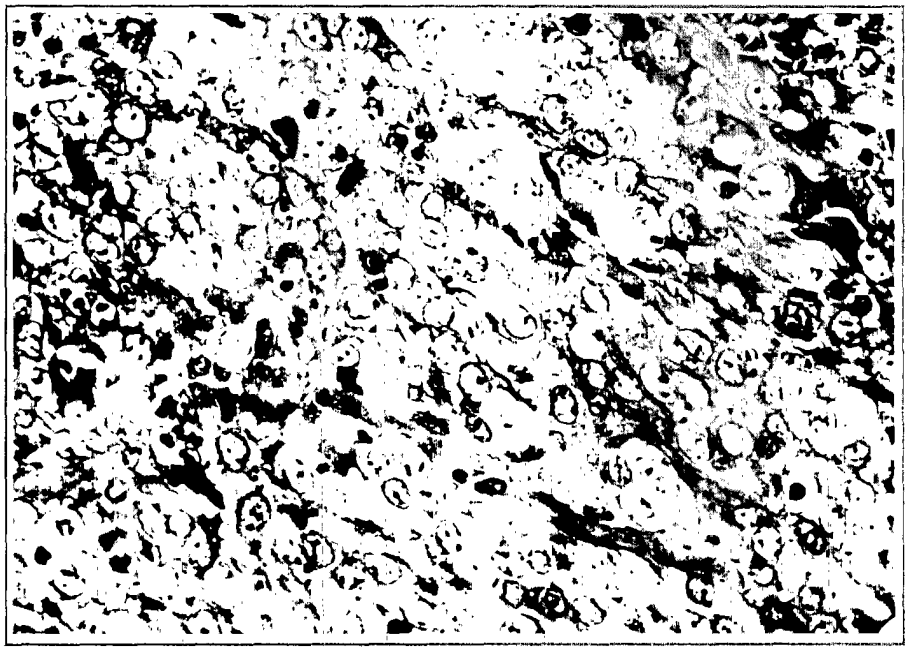

Figure 3. Areas of an undifferentiated carcinoma-like appearance, with large cell size, marked cytologic atypia and pleomorphic nuclei (original, HE ×200).

plasms. This phenotype can be made up of a variety of unrelated neoplasms that reveal specific differentiation. It seems appropriate to add the term rhabdoid as a phenotypic descriptor.

From a morphological point of view, this tumor could be considered an undifferentiated carcinoma (14). Immunohistochemistry and electron microscopy were unhelpful in differentiating it from epithelioid sarcoma. Reactivity for carcinoembryonic antigen was of little help because it has been observed in both tumor types. The deep-seated location of the tumor, the lack of a previous history of primary or metastatic carcinoma, the

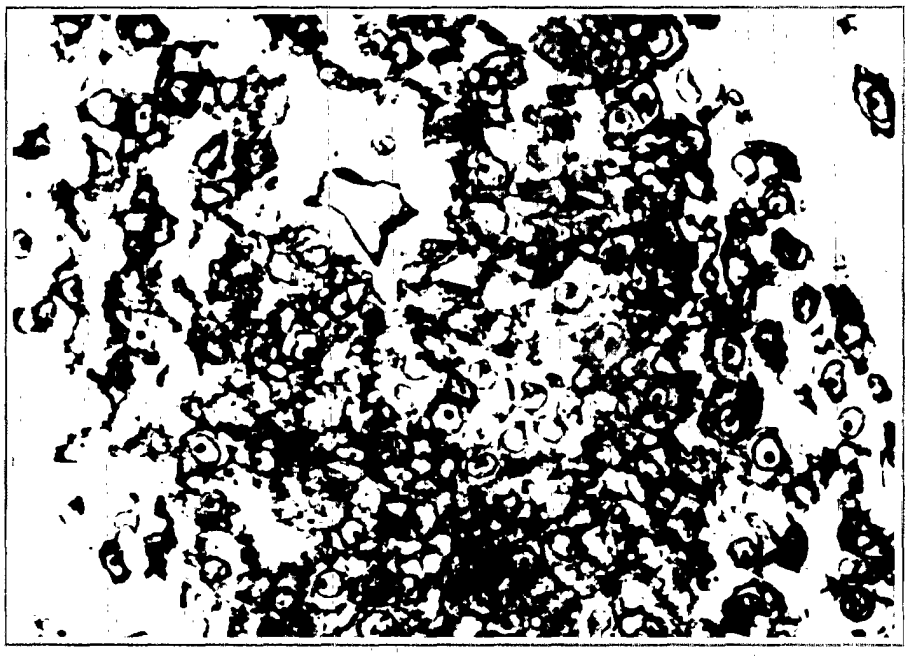

Figure 4. Tumor cells show strong membranous immunopositivity for CD34 (original, immunoperoxidase ×200).

presence of CD34 reactivity and the same strong and diffuse immunoreactivity for vimentin and cytokeratin support a diagnosis of epithelioid sarcoma.

The classic epithelioid sarcoma (15-17) occurs in the distal extremities of young adults as a slow-growing soft-tissue neoplasm. Histologically, a multinodular pattern is characteristic, with occasional necrosis in the center of the nodules. There is a combination of eosinophilic epithelioid and spindle-shaped cells with minimal atypia, vesicular nuclei and single, central nucleoli. The clinical course is usually protracted.

We have described the case of a tumor with clinical, morphological and immunohistochemical features that support a diagnosis of epithelioid sarcoma. The described features best fit with the recently described entity "proximal-type" epithelioid sarcoma. Unlike conventional epithelioid sarcoma, the proximal variant is deep-seated and usually metastasizes earlier. Histologically, the marked cytonuclear atypia, the prominence of the epithelioid component, the frequent occurrence of rhabdoid features and the usual absence of a granuloma-like pattern are distinctive features.

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