Case Report

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Epithelioid Cell Histiocytoma: A Report of Two Cases

B.		Dezfoulia ^a
A.F.		Nikkels ^a
C.		Piérard-Franchimont ^{a,b}
G.E.		Piérard ^a

^aDepartment of Dermatopathology, University of Liège, and ^bSection of Dermatology, Centre Hospitalier Hutois, Huy, Belgique

Key Words

Epithelioid cell histiocytoma
Factor XIIIa
Dendrocytes

Abstract

Epithelioid cell histiocytoma is a rarely reported tumor derived from factor-XIIIa-positive dermal dendrocytes. Two additional cases are presented including their clinical, histologic and immunohistochemical features.

Dr. G. Piérard, Service de Dermatopathologie, CHU du Sart Tilman, B-4000 Liège (Belgium)

Epithelioid cell histiocytoma is an uncommon tumor. Less than 50 cases have been reported in the literature so far [1 -4]. The lesion is characterized by its occurrence as a solitary elevated nodule often with a vascular appearance. The histologic aspect resembles the intradermal form of Spitz nevus. Most of the characteristic angulated epithelioid cells that form the tumor are factor XIIIa positive and therefore categorize the lesion as a variant of fibrous histiocytoma [1].

Case Report

Case 1

A 38-year-old man, without particular medical history, noticed an erythematous papulonodular lesion on his thigh growing relatively rapidly and reaching 8 mm in diameter within 3 months. A biopsy specimen was obtained, and the histological examination revealed an infiltration of the reticular dermis by angular epithelioid cells with large and pale nuclei (fig. 1). The vasculature was prominent. Some mitotic figures were scattered throughout the tumor. Inflammatory cells of much smaller size were rare, focally infiltrating the lesion. This histologic aspect suggested an intradermal Spitz nevus or a malignant melanoma, perhaps metastatic. An immuno-pathological study revealed, however, that the epithelioid cells were negative for S-100 protein, and HMB45 and NKI-C3 antibodies. Among the small-sized cells looking like inflammatory cells, many were positive for S-100 protein, suggesting a langerhansian origin. The positive immunoreactivity of about 50% of the epithelioid cells with the anti-factor-XIIIa antibody was the key for the diagnosis of epithelioid cell histiocytoma.

Fig. 1. Epithelioid cell histiocytoma. Accumulation of large angulated epithelioid cells admixed with a few small lymphoid cells.

Case 2

A 46-year-old man presented with an exophytic papule on the neck. The histologic examination revealed a symmetrical ovoid nodule that projected above the level of the surrounding skin.

Large angular and polygonal epithelioid cells were clustered around blood vessels. Their nuclei were pale with vesicular chromatin. Mitoses were rare. Most of these cells were factor XIIIa positive, confirming the diagnosis of epithelioid cell histiocytoma.

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Discussion

Histiocytomas contain histiocytes, fibrocytes, dendro-cytes and extracellular matrix in various proportions. They may be single, multiple or eruptive. Among different histo-logic types of histiocytomas, there is a distinct one described for the first time by Wilson Jones et al. [1] and named 'epithelioid cell histiocytoma'. The tumor occurs in adults with an average age of 40 years; women are slightly more affected than men in a ratio of 1.4:1. The lesions are usually solitary. Presented as a firm, sessile or polypoid nodule or papule, their diameter ranges between 0.5 and 1.5 cm. Generally, the lesions are circular or ovoid, dome-shaped with a smooth surface, reddish or pigmented, with a vascular appearance. Their most frequent localizations are the limbs, but they may occasionally be seen on the trunk and the head. The duration of the lesions is greatly variable ranging from a few months to more than 3 years.

Epithelioid cell histiocytoma is an intradermal tumor made of large and angulated epithelioid cells with abundant and eosinophilic cytoplasm [1-4]. Multinucleated giant cells are not prominent. Cellular atypia and mitotic figures are few. The vascularization is rich. The epidermis above the lesion is usually normal in thickness; it may sometimes be acanthotic laterally and hyperpigmented. Characteristically, an ingrowing collar of epidermis at the outer and lower margins encloses the tumor at its base. Skin appendages are sometimes bowed or displaced laterally at the margins of the nodules.

S-100 protein immunoreactivity is positive in only 5-10% of the cases, showing Langerhans cells or related cells dispersed within the tumor which is itself negative. Common leukocyte antigen is positive in about 25% of the cases. Vimentin and α -antitrypsin are present in the histiocytoid cells. Factor XIIIa positivity in the epithelioid cells reveals a type 1 dermal dendrocyte lineage [1, 5]. Immuno-reactivities to von Willebrand factor, PAL-E, Ulex euro-paeus agglutinin 1 and EN4 are all positive in the blood vessels but absent in the tumoral cells. Cytokeratin markers PKK1, LP34, CAM 5.2 and markers of activated or neoplastic melanocytes such as HMB45 and NKI-C3 are all negative. Mac 387, a marker of macrophages, is also negative. Epithelioid cell histiocytoma may clinically resemble many lesions including bacillary angiomatosis, histiocytoma, dermatofibroma, pyogenic granuloma, blue nevus, nodular malignant melanoma, metastatic melanoma and Spitz nevus. Histologically, this form of histiocytoma could be mistaken for Spitz nevus, solitary reticulohistiocytoma or atypical fibroxanthoma.

At the difference with Spitz nevus, epithelioid cell histiocytoma has no junctional melanocytic hyperplasia and nesting. The stainings for S-100 protein and HMB45 are negative while the positive staining for vimentin, α -anti-trypsin and factor XIIIa confirms their dermal dendrocytic nature. Solitary reticulohistiocytoma contains multinucleated giant cells. The presence of inflammatory cells, including eosinophils, and a greater variability in the shape of histiocytes and multinucleated cells in solitary reticulohistiocytoma make the distinction possible. Atypical fibroxanthoma shares a polypoid configuration with epithelioid cell histiocytoma. Tumoral cells are however different, having bizarre nuclei and numerous mitotic figures.

In summary, epithelioid cell histiocytoma is a recently recognized entity that enters the spectrum of lesions related to the factor-XIIIa-positive dendrocytes [5]. The histologic recognition of the lesion is of importance because the differential diagnosis includes atypical and malignant neoplasms.

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Dezfoulan/Nikkels/Piérard-Franchimont/ Piérard
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