

Epithelioid Cell Histiocytoma: a Case Report and Brief Review of the Literature, with an Emphasis on Differential Diagnosis

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Citation

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Abstract

Epithelioid cell histiocytoma (ECH) is a benign tumor of the dermis which can closely resemble other benign and malignant tumors showing epithelioid features. Correct diagnosis is needed to prevent unnecessary treatment. A case of ECH and a brief review of the literature are presented, with emphasis on differentiating ECH from other benign dermal neoplasms with epithelioid features.

CASE REPORT

A 44-year-old woman presented with a 5-mm, smooth, flesh-colored papule of the cheek. The lesion had been present for an unknown duration.

Microscopically (Figures 1 and 2), the epidermis was unremarkable. The upper dermal tumor was composed of large epithelioid cells containing bland nuclei with occasional large nucleoli. There were no mitotic figures or necrosis. There were a few multinucleated giant cells but no foam cell. The tumor cells were strongly positive for Vimentin (Figure 3) and focally positive for CD68 (Figure 4) and were negative for CK AE1/3, S-100, MITF, SMA, Myogenin, and CD34.

Figure 1

Figure 1. Low magnification, H&E stain showing normal epidermis with an upper dermal epithelioid tumor.

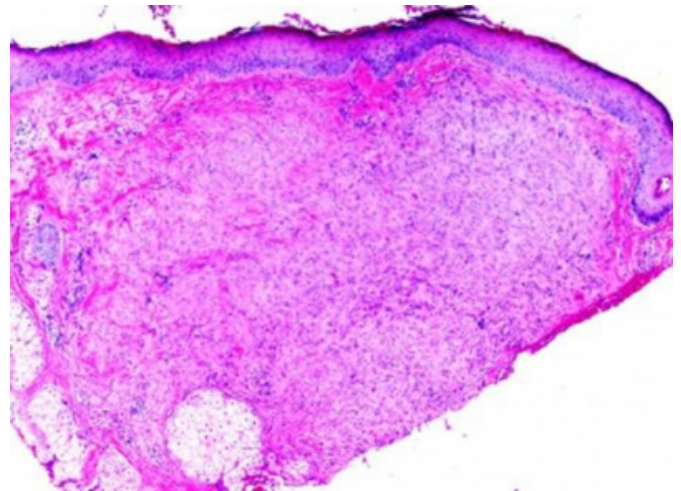


Figure 2

Figure 2. High magnification, H&E stain showing large epithelioid tumor cells with slightly pleomorphic nuclei and occasional large nucleoli. No increased mitosis or necrosis is present.

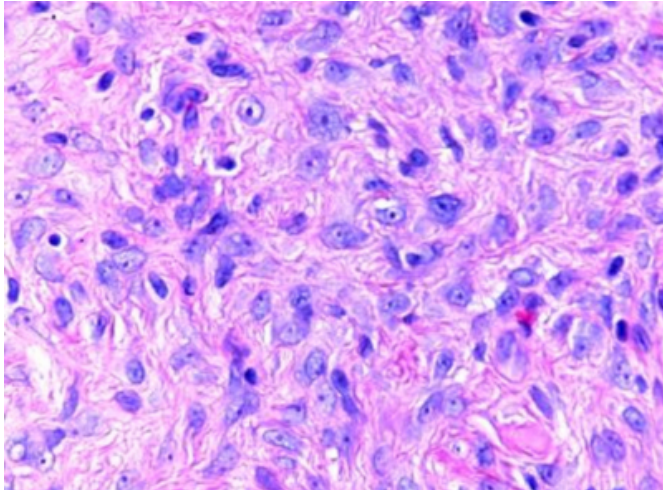


Figure 3

Figure 3. Tumor cells are strongly positive for Vimentin.

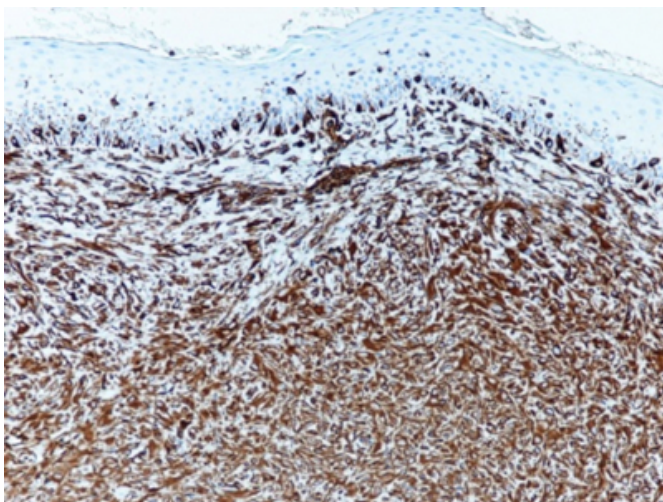
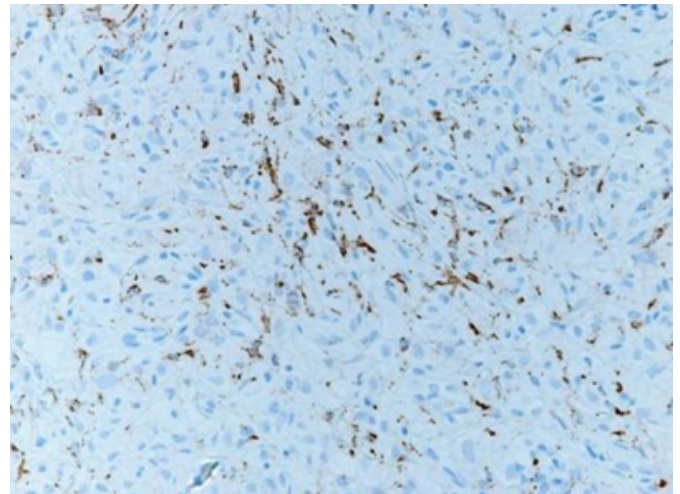


Figure 4

Figure 4. Tumor cells are focally positive for CD68.



Diagnosis: Epithelioid cell histiocytoma (ECH)

COMMENT

Epithelioid cell histiocytoma (ECH) is a rare variant of dermatofibroma, the histogenesis of which has not been determined with certainty. Because ECH may histologically resemble other benign and malignant dermal lesions with epithelioid features, the pathologist must be familiar with this entity in order to render a correct diagnosis.

Clinically, ECH occurs in all age groups, with a median age of 35-40 years [1, 2]. There is a slight male preponderance. Lesions most commonly present on the trunk and upper and lower extremities, followed by the head and neck and other sites [1, 2]. Tumors appear as circumscribed, solitary papules or pedunculated lesions measuring 0.1 to 2.0 cm in greatest dimension [1, 2]. A case with multiple ECH has recently been reported [3]. Excision is usually curative, with rare recurrences [2].

Histologically, ECH differs from usual benign fibrous histiocytoma in several ways: the predominance of epithelioid cells, relative lack of secondary elements (giant cells, foam cells, hemosiderin-laden macrophages), relatively sharp circumscription, prominent vascularity, and location within the papillary dermis in most cases [1, 4]. Lesions are not typically ulcerated [1]. More polypoid tumors show an epidermal collarette [2]. The epithelioid cells are polygonal or round and contain abundant, eosinophilic cytoplasm with variable numbers of lymphocytes and neutrophils [1, 2]. Multinucleated forms may be present. Vesicular nuclei with small, eosinophilic

nucleoli are characteristic [2]. Mild to no nuclear atypia and a low mitotic rate is the rule [1]. Rarely, ECH may show prominent cellular whorls or increased cellularity [4, 5].

Immunohistochemically, positivity for CD163, CD68, and vimentin in the epithelioid cells has been reported [1, 3, 6]. Focal nuclear immunoreactivity for microphthalmia transcription factor and focal reactivity for Factor XIIIa and S-100 protein can occur [1, 3, 6, 7]. Membrane positivity for CD3, CD20, CD30, HMB45 is variable [1, 8]. Epithelioid cells are negative for CD3, CD 10, CD20, CD30, S-100 and keratins [1, 8, 9].

ECH may be confused with other benign lesions, such as desmoplastic Spitz nevus, epithelioid and/or cellular blue nevus, cellular neurothekeoma, and even pyogenic granuloma. In contrast to ECH, desmoplastic Spitz nevus generally lacks an overlying hyperplastic epidermis and the epithelioid cells usually lie singly in a dense, eosinophilic, collagenous stroma [10]. In addition, desmoplastic Spitz nevus shows diffuse immunoreactivity with S-100 [11]. Cellular blue nevi may attain a much larger size and extend into subcutaneous adipose tissue when compared to ECH [10]. Blue nevi are also differentiated from ECH by S-100 positivity and the presence of melanin pigment-containing macrophages. The lesions of cellular neurothekeoma are papules or nodules primarily involving the face of adolescents and young adults [12, 13, 14]. Unlike ECH, cellular neurothekeoma involves the reticular dermis and tends to be poorly circumscribed; low-grade cytologic atypia and mitotic activity are common [14, 15].

Immunohistochemical analysis may not be helpful in the distinction between ECH and cellular neurothekeoma, and immunohistochemical and histologic similarities have led some authors to suggest that cellular neurothekeoma may be a variant of dermatofibroma [12, 16].

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