Case for diagnosis

Caso para diagnóstico

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CASE REPORT

An 82-year old Caucasian woman presented with a lesion on her face. Approximately 1 year earlier she had noticed a small, circular tumor on the left side of her lower jaw. She reported localized itching, as well as bleeding following mild trauma. Examination revealed a single, small pink tumor of 2 cm in diameter on the left side of her lower jaw. The lesion was well defined, mobile, with an irregular surface, ulceration and a central crust (Figure 1). A biopsy was performed of the lesion, revealing an ulcerated exophytic tumor encroaching on the reticular dermis, albeit without affecting the hypodermis. The lesion consisted of bizarre spindle cells, a diffuse proliferation of epithelioid cells and atypical mitoses with severe pleomorphism and hyperchromatic nuclei (Figures 2 and 3). At immunohistochemistry, the tumor tested positive for vimentin and negative for S-100, CD-99 and pan-cytokeratin. The Ki-67 proliferative index was high, over 60%. Based on these findings, the tumor was then excised, leaving a wide margin of 5 mm.

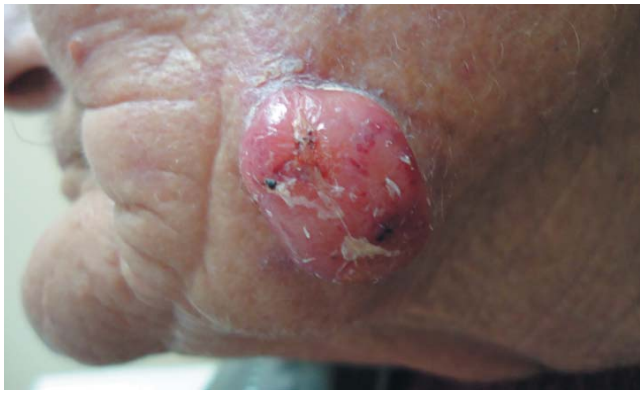

FIGURE 1: A mobile, well-defined tumor of approximately 2 cm in diameter with an irregular surface, areas of ulceration and the formation of crusts on the surface

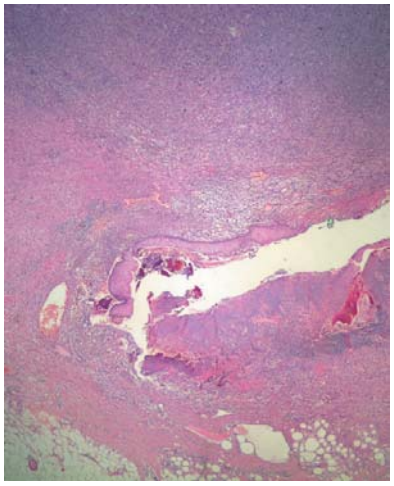

FIGURE 2: Exophytic neoplasm with invasion up to the reticular dermis (hematoxylin-eosin, magnification 2x)

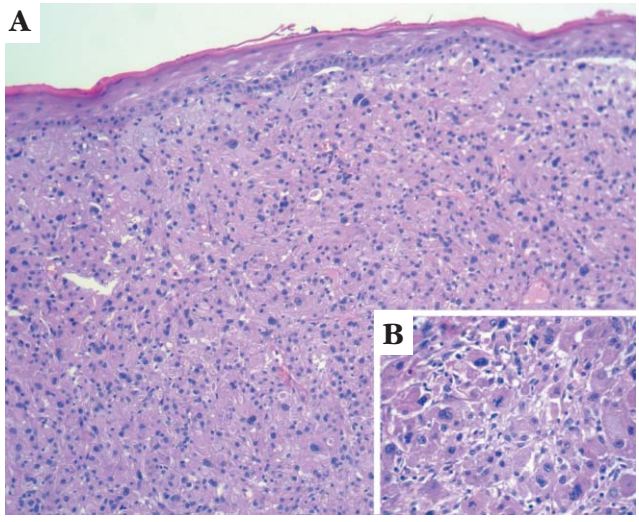

FIGURE 3: Neoplasia consisting predominantly of epithelioid cells of largely eosinophilic cytoplasm, pleomorphic nuclei and a high degree of atypia. A: hematoxylin-eosin, magnification 100x (2.0x optic zoom). B: Magnification 400x (2.0x optic zoom)

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DISCUSSION

Atypical fibroxanthoma (AFX) is a rare malignant skin neoplasm. These tumors are most common in the elderly population and usually develop on sun-exposed areas of the skin. They are found almost exclusively in Caucasians and are twice as common in men as in women. The tumor usually presents as a solitary papule or nodule. Ulceration and bleeding may develop. Most lesions measure less than 2 cm in diameter, with a mean diameter of 1.1 to 1.5 cm. AFX is most commonly confined to two distinct anatomic sites: on the head and neck (sun-exposed areas) of elderly patients and, less commonly, on the trunk and limbs (non-sun-exposed areas) of younger individuals. [1,2,3] Clinically, AFX are nonspecific tumors and indistinguishable from other malignancies such as squamous cell carcinoma (SCC), basal cell carcinoma (BCC) and keratoacanthoma; therefore, biopsy is essential in order to reach a diagnosis. [1,4,5] Histologically, AFX is considered a malignant fibrous histiocytoma, with dermal cell proliferation of bizarre spindle cells, epithelioid cells or multinucleated giant cells. Atypical mitoses with severe pleomorphism and hyperchromatic nuclei are often seen. [6,7] AFX has been considered a superficial variant of undifferentiated pleomorphic sarcoma (UPS), previously known as the pleomorphic variant of malignant fibrous histiocytoma (MFH). [4,5,7] Some authors believe that if a tumor is larger than 2 cm in diameter, if it involves the deeper subcutis, if it penetrates fascia or muscle or if there is necrosis or vascular invasion, it should be diagnosed as UPS rather than AFX, since these conditions are indistinguishable at histopathology. None of these factors was found in the present case. In spite of these histological characteristics of malignancy, AFX is considered a low-grade malignancy and there have been few reported cases of metastases. [1,4,5,6,7] Immunohistochemistry is essential in order to confirm diagnosis. In this case, immunohistochemistry was negative for the S-100 protein and pan-cytokeratin (AE1AE3) and positive for vimentin and Ki-67. AFX is usually negative for cytokeratin, unlike epithelial neoplasms such as fusiform SCC in which it is usually positive. Unlike spindle cell melanoma, AFX typically tests negative for S-100 protein. [5,7,8,9] CD-99 has been used as an adjuvant for differentiating between MFH and AFX, since positivity for CD-99 is more common in the latter. [8,9] Ki-67 is a marker of cell proliferation. The positivity for Ki-67 found in the present case indicates a high mitotic index in neoplasia. Vimentin is a marker of mesenchymal cells and is positive in neoplastic cells in AFX. [1] It is now generally accepted that the malignant potential of AFX is intermediate, with metastasis being rare. [1,7] Over the years, AFX has been treated in various manners, including wide local excision (WLE), Mohs micrographic surgery (MMS), radiation therapy, cryotherapy and electrocauterization. However, local recurrence rates of up to 20% have been found with WLE. In addition, clinicians have used a modified MMS technique (slow Mohs) as an alternative treatment for AFX, with rates of local recurrence that range from 0 to 6%. [2,4,5,7] The patient in the present report was submitted to surgical removal of the lesion, leaving wide margins of 5 mm. She has now been followed-up for two years and there are no signs of local recurrence. ❑

Abstract: Atypical fibroxanthoma is a rare cutaneous tumor found mainly in elderly people on sun-exposed areas of the body. Histologically, atypical fibroxanthoma is considered a malignant fibrous histiocytoma with bizarre neoplastic cells, marked pleomorphism, hyperchromatic nuclei and abundant mitoses. It must be differentiated from other skin tumors, usually by immunohistochemistry, since its diagnosis is made by exclusion.
Keywords: Malignant histiocytic disorders; Histology; Skin neoplasms

Resumo: O fibroxantoma atípico é um tipo de neoplasia cutânea maligna rara, encontrado principalmente em idosos, em áreas fotoexpostas. Na histologia, o fibroxantoma atípico é uma neoplasia fibro-histiocítica dérmica, de células fusiformes e epitelioides, algumas vezes bizarras, com acentuado pleomorfismo, apresentando núcleos hipercromáticos e mitoses abundantes. Deve ser diferenciado de outros tumores de pele, principalmente através da imunoistoquímica, já que seu diagnóstico é de exclusão.
Palavras-chave: Histologia; Neoplasias cutâneas; Transtornos histiocíticos malignos

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