Case for diagnosis / *Caso para diagnóstico*

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DISEASE HISTORY

65 year-old female patient, homemaker. Six years ago, she noticed the appearance of a lesion in the left malar region, assymptomatic, with progressive growth.

At dermatological examination, there was an infiltrated papule in the left malar region, with erythemathous violet borders, and a diameter of 1.5 cm, with superficial teleangiectasias and depressed center (Figure 1).

The anatomopathological study of skin biopsy revealed the presence of dermic infiltrative neoplasic proliferation, composed by blocks of cells and ducts of various shapes and forms, covered by polygonal cells with a discrete shape and nuclear size variation, the latter displaying pleomorphism and evident nucleoli (Figure 2). Perineurial infiltration by the neoplasic cells was also present in the sample (Figure 3). Such findings lead to the establishment of a syringomatous carcinoma as diagnosis.

The patient was submitted to surgical exeresis of the tumor, with wide margins. Anatomopathological exam confirmed the previous diagnosis of syringomatous carcinoma. Surgical margins were free, and a neural compromise was noticed, with the presence of neoplasic cells in the perineuria.

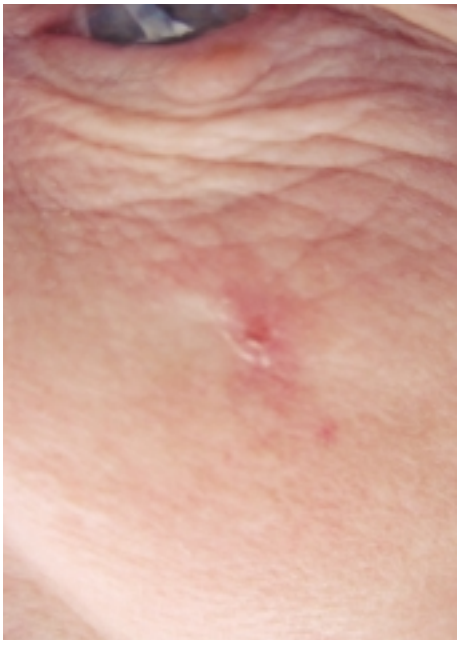

FIGURE 1: Infiltrated papule in the left malar region

COMMENTS

Syringomatous carcinoma is a cutaneous malignant neoplasia with probable origin in eccrine glands. It characteristically presents slow and destructive growth, with local, perineurial and muscular invasion.[1]

It affects men and women alike, and generally occurs between the fourth and seventh decades of life.

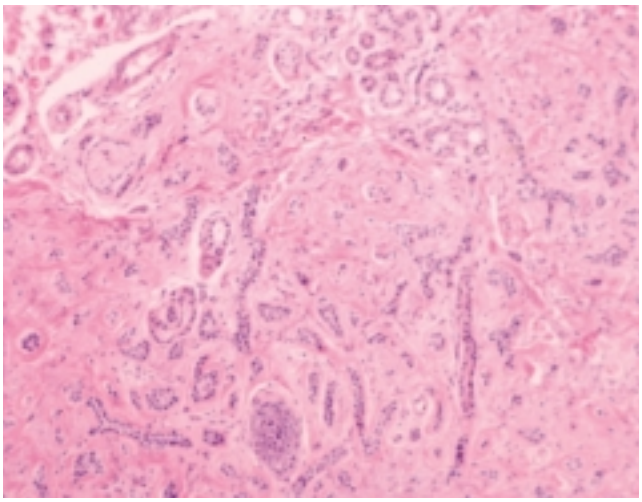

FIGURE 2: Infiltrative neoplasic proliferation in dermis, composed by blocks of cells and ducts of various shapes and sizes, covered by polygonal cells with discrete variation in the size and shape of the nuclei, displaying pleomorphism and evident nucleoli (HE 40x)

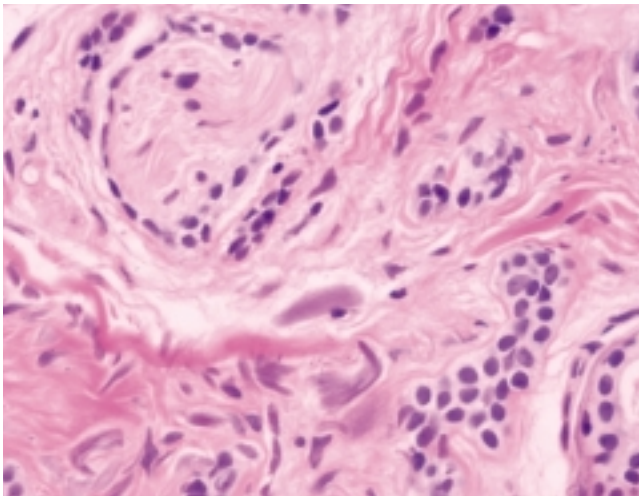

FIGURE 3: Perineurial infiltration by the neoplasic cells (HE 400x)

Clinically, it appears as a yellowish plaque or nodule, solitary, of slow growth, superficial teleangiectasias and poorly defined borders. The occurrence of ulceration is rare and found only in the older lesions. The most affected places are face, palpebrae and scalp, sometimes forming an alopecia plaque in the latter.[2]

The main histological features include formation of ducts and small cysts containing horny cells in the lumen. It displays an infiltrative growth pattern with extension of the neoplasia to the subcutaneous, invasion of perineurials, presence of demoplasia and rare or absent mitoses.[1] Diagnosis is generally made with the hematoxilin-eosin stain, although it can be complemented by an immunohistochemical analysis.

This immunohistochemical test reveals positivity to the carcinoembrionary and membrane epithelial antigens.

Histological differential diagnoses of the syringomatous carcinoma include: sclerodermiform basocellular carcinoma, microcystic anexial carcinoma, desmoplasic tricoepithelioma, and skin metastatic adenocarcinoma, the latter generally having a mammary origin.

In sufficiently superficial or small biopsies, obtained by either *shaving or punching*, there is occasional difficulty in the establishment of an accurate diagnosis, and syringoma should be considered in the differential diagnosis. In such instances, an ample biopsy is indicated for the establishment of the correct diagnosis based on an architectural pattern indicating malignancy.[3,4]

The treatment of choice is a wide and deep surgical exeresis, with rigorous histological analysis of the borders. If not totally resected, the neoplasia persists in the site and spreads surreptitiously. Radiotherapy is seldom used. Forty to sixty percent of pacients submitted to surgery will have a relapse of the lesion. Regional lymph nodular and pulmonary metastases may occur.[3,5]

This clinical story calls attention to the occurrence of an uncommon malignant neoplasia and to the necessity of an anatomopathological study of malignant-looking skin lesions in order to establish a definite diagnosis and thus provide the patient with better therapeutical options. ❑

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Dear colleague,

The "What is your Diagnosis?" section aims to present clinical cases in which the final diagnosis is questionable. If you have an article that fits this section, please contribute to the Anais Brasileiros de Dermatologia by sending it to us, our address is:

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