Case for diagnosis / Caso para diagnóstico

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DISEASE HISTORY

Sixty-five year-old female, brown skin, married, housewife, natural of Paraíba and coming from Praia Grande, SP, sought a Dermatology outpatient clinic with the complaint of a "nodule on the tip of the nose" for six months, which presented with rapid and progressive growth.

Personal and familar history: Virchowian Leprosy in the 15th month of treatment with Multibacillary Multidrug Therapy. Denied neoplasms or other relevant diseases in family history.

At the dermatological examination, the patient displayed a bright, infiltrated, skin-colored nodule, of hard consistency and granulous bleeding central ulceration, measuring 1.3 cm of diameter, located at the nasal dorsum. The patient had no cervical lymphadenomegalies (Figure 1).

A biopsy was performed for histopathological examination (Figure 2), which revealed tumoral cells with nuclear pleomorphism, prominent nucleoli and multilobuled cytoplasm. Immunohistochemistry (Figure 3) revealed strong expression of Epithelial Membrane Antigen (EMA).

Twenty days after the first visit, the pacient underwent surgery and a complete investigation in order to exclude systemic diseases.

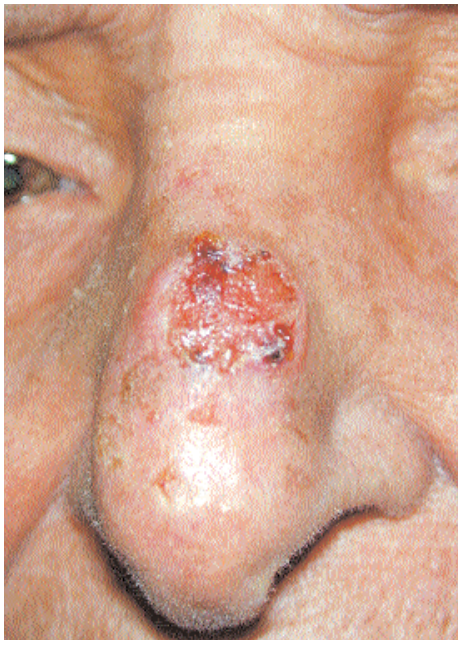

Figure 1: Skin-colored nodule, with a bleeding central ulceration on nasal dorsum

DIAGNOSIS AND COMMENTS

Extraocular sebaceous carcinoma

Sebaceous carcinoma is a malignant tumor derived from sebaceous glands anexial epithelium. Its most frequent locations are head and neck, especially in sebaceous glands of the ocular anexa.[1]

It can be classified as either ocular or extraocular, being roughly 25% extraocularly located.[2] The tumor incides more in females (2:1) who are in ave-

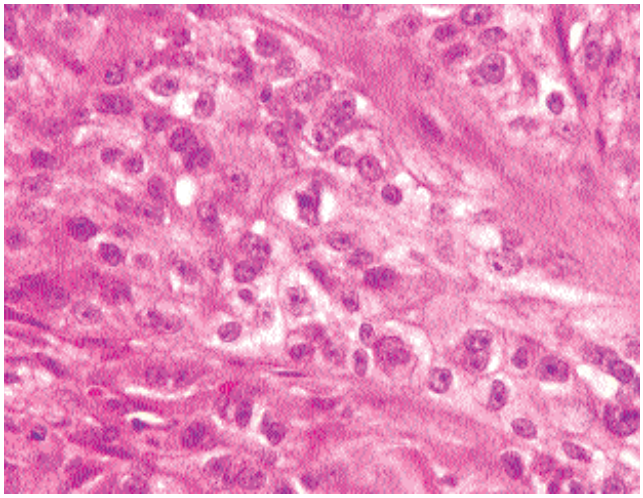

Figure 2: Tumoral cells with nuclear pleomorphism, prominent nucleoli and multilobuled cytoplasm (HE 400x)

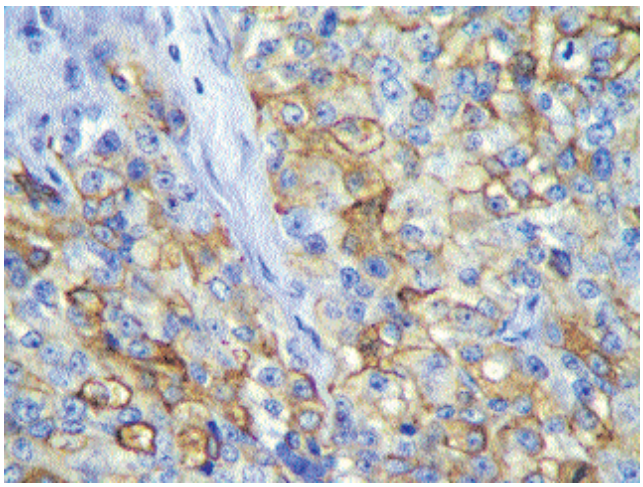

Figure 3: Tumoral cells presenting strong expression of Epithelial Membrane Antigen (EMA) infiltrating a neural bundle (400x).

rage 65 years old.[3] The clinical presentation of the disease is not specific; usually, the extraocular tumor is described as nodular, pinkish to yellowish red, and measuring 6-20 mm.[1]

The treatment of choice is surgery, always performed with wide safety margins, or Mohs' micrographical surgery. Radiation therapy, along with chemotherapy, can be used as palliation in case there are metastases.[4]

The prognostic is reserved, since the sebaceous adenocarcinoma is an aggressive tumor that can be multicentric and it has a great tendency to relapse. Metastases occur in a percentage ranging from 14 to 25% of instances, and can be hematogenic, lymphatic or through the lacrimal system in ocular cases. The most affected organs are the liver, lungs, brain and bones. One of the most significant prognostic factors is the time elapsed between the onset of lesion and treatment. Periods from 1 to 6 months imply a mortality rate of 14%, a figure that increases to 38% in cases with a time course over 6 months.[1] Extraocular sebaceous carcinomas have a better prognosis, with a lower risk of relapsing and metastizing.

Most sebaceous carcinomas have no defined ethiology; however, in some instances they may be associated with Muir-Torre's Syndrome, dominant autossomic genodermatosis characterized by sebaceous tumors (either benign or malignant) or ceratoacantomas associated with visceral malignancies,[5] gastrointestinal and genitourinary tumors being the most frequent ones.[1,5] Thus, patients with sebaceous carcinoma should be investigated both clinically and laboratorially to rule out systemic neoplasms.

The need for a quick and accurate diagnosis must be highlighted, since the prognosis of the disease is intimately related to its time of evolution. In addition to that, the need for an investigation to exclude systemic neoplasms, as well as Muir-Torre's Syndrome should be remembered, not to mention the importance of an oncological follow-up of these subjects, as this is an aggressive and relapsing tumor. ❑

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