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Abstract: Muir-Torre syndrome is a rare genodermatosis characterized by the occurrence of at least one sebaceous tumor associated with visceral neoplasia, but with no predisposing factors. The sebaceous neoplasm may appear before, during or after the diagnosis of colorectal cancer. As it is regarded as a subtype of nonpolyposis hereditary colorectal cancer, it is important to evaluate the patient's first-degree relatives. The clinical course of the neoplasm is usually more indolent and the syndrome has a good prognosis. We report the case of a patient who, after a ten-year diagnosis of colorectal cancer, presented with multiple sebaceous neoplasms.
Keywords: Sebaceous gland neoplasms; Colorectal neoplasms, hereditary nonpolyposis; Skin Neoplasms; Syndrome; Muir-Torre Syndrome

CASE REPORT

A 56-year-old male patient had a 4-year history of skin lesions on the face and trunk. He reported having had a partial colectomy and chemotherapy in 2001 due to descending colon adenocarcinoma. His mother, maternal uncle and son had colonic neoplasia.

Dermatological examination revealed multiple erythematous yellowish papules with central depression, pearly edges and telangiectasias, distributed on the trunk and face (Figure 1).

Histopathological examination of the lesions revealed a proliferation of sebaceous glands surrounding a dilated follicular infundibulum. At the periphery of the glands, there is a layer of germinative basaloid cells and, in their center, there are mature sebocytes, a finding compatible with sebaceous adenoma(Figures 2 and 3).

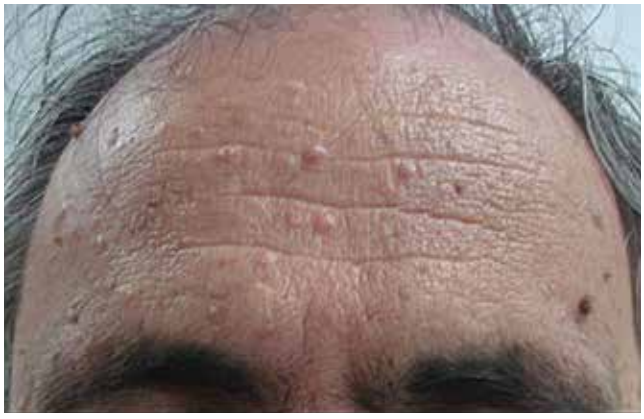

Figure 1: Yellowish erythematous papules with central depression on the face

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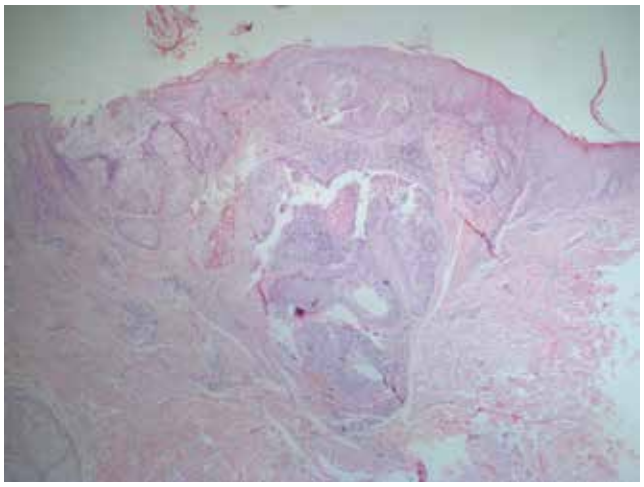

Figure 2: Histopathology: proliferation of sebaceous glands surrounding a dilated follicular infundibulum. At the periphery of the glands, there is a layer of germinative basaloid cells and, in their center, there are mature sebocytes, a finding compatible with sebaceous adenoma

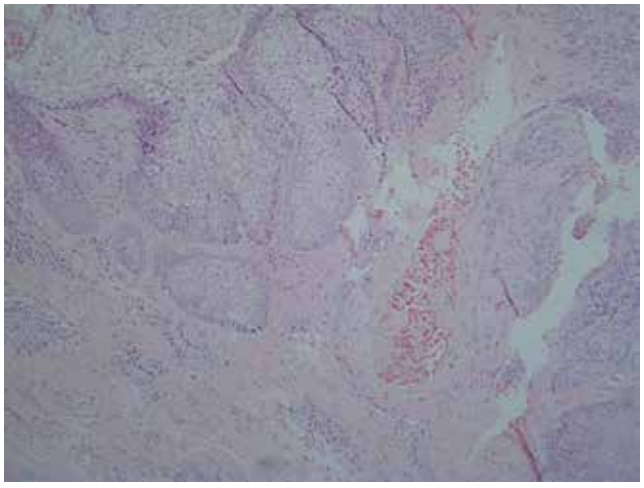

Figure 3: Tumor with mature sebaceous cells and basaloid cells. Sebaceous adenoma

The patient also had a one-month history of ill-defined erythematous maculae on the upper limb and trunk. Histopathological examination revealed an epidermis with mild hyperplasia. In the upper dermis there is an inflammatory infiltrate composed of lymphocytes that blur the junction boundary, forming intraepithelial groups compatible with mycosis fungoides.

DISCUSSION

Muir-Torre syndrome (MTS) is a rare genodermatosis characterized by the occurrence of at least one sebaceous tumor (adenoma, epithelioma or carcinoma) associated with visceral neoplasia, but with no predisposing factors. The syndrome may occur spontaneously. However, most cases found are a variant of the autosomal-dominant disorder hereditary non-polyposis colorectal cancer (HNPCC).[1] The underlying genetic mutations responsible for the syndrome are associated with changes in in the DNA mismatch–repair genes MSH2 and MLH1, which are also found in HNPCC. [2]

Sebaceous tumors are rare, and their identification alerts to a possible association with MTS. Sebaceous adenomas are the most common tumors. Skin lesions may appear before, during or after the diagnosis of internal malignancy.[2] Among visceral tumors, gastrointestinal cancers occur in 50% of cases and genitourinary cancers occur in 25% of cases.[2] Colorectal cancer associated with the syndrome usually occurs proximal to the splenic flexure, unlike most sporadic colorectal cancers. These cancers tend to have a more indolent course, when compared with sporadic tumors.[1]

There are reports of associations with other malignancies such as breast, hematologic, head, neck and lung.[1] Our patient was also diagnosed as having mycosis fungoides. However, to date, there are no reports of an association between mycosis fungoides.and MTS.

The syndrome has a good prognosis (over 50% survival in 10 years). Nevertheless, recurrence is frequent and approximately 60% of cases develop metastasis. [3]

Surgical treatment is preferred in patients with skin and visceral tumors. Use of oral isotretinoin may help prevent the appearance of new sebaceous lesions.[3] In patients with MTS or non-polyposis colorectal cancer, it is recommended to perform a colonoscopy every 1 to 2 years beginning at age 20 to 25 years of age or 10 years earlier than the youngest age of colon cancer diagnosis in the family. In women, transvaginal ultrasound or endometrial biopsy should be performed. [2] It is suggested the evaluation of first-degree relatives, due to the possibility of family involvement.[4]

Our patient was referred to the medical clinic for gastrointestinal revaluation The revaluation evidenced a malignant tumor of the biliary tract of which he died.❑

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