Mycosis fungoides mimicking a nodule of Sister Mary Joseph

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INTRODUCTION

The umbilicus constitutes a complex anatomic structure, closely related to the intraabdominal organs. It can be the elective site of inflammatory, infectious, tumoral, or malformative dermatoses. We report an exceptional case of mycosis fungoides taking the form of an umbilical tumor.

CASE REPORT

A 45-year-old woman with a history of breast carcinoma in complete remission for 2 years and mycosis fungoides, undergoing UV-B phototherapy for 1 year, presented at her follow-up visit for a painless umbilical lesion.

Clinical examination revealed erythematous, scaly, nonpruritic papules involving the whole body but sparing the face and a blackish, sessile umbilical tumor that was hard at palpation (Fig 1, *A*). Dermatoscopic examination revealed a nonmelanocytic lesion with comedo-like openings suggestive of seborrheic keratosis (Fig 1, *B*). Findings of the remaining clinical examination were normal, including that of the examination of the lymph nodes.

A histologic study of an excised biopsy of the umbilical lesion after application of a keratolytic agent revealed an epidermotropic lymphoid infiltrate composed of medium-sized lymphocytes with cribriform notched nuclei arranged in a discohesive or single-file pattern (Fig 2). Immunohistochemical analysis revealed $CD2^{+}$, $CD3^{+}$, $CD5^{+}$, and $CD7^{-}$ staining (Fig 3).

A workup was performed, including a complete blood cell count and flow cytometry, a thoracic-abdominal-pelvic computed tomography scan, and a bone-medullary biopsy, with no abnormalities found. Therefore, we retained the diagnosis of plaque mycosis fungoides.

DISCUSSION

The etiologies of the umbilical nodule are diverse. They depend on the terrain and the clinical presentation.[1] In the case of children, a pyogenic granuloma should be considered first; it is often observed in newborns after cord separation. The condition may also be caused by the delayed and irregular separation of the cord stump. Umbilical granulomas generally develop within the first few weeks of life and can present as a solid, soft, velvety red mass with serosanguineous discharge, approximately 1 mm to 10 mm in diameter. They are usually treated with 75% silver nitrate or can be surgically excised. Failure to respond to silver nitrate application differentiates these granulomas from congenital malformations such as urachal or vitelline duct anomalies.[2]

In adults, the etiologies of umbilical tumors are dominated by umbilical cutaneous metastasis. The term "Sister Mary Joseph nodule" was first described by Sister Mary Joseph in 1928.[3] The Sister Mary Joseph nodule is usually irregular, firm, and blue-purple or brownish-red in color. It may be fissured or ulcerated and associated with a bloody, mucinous, serous, or purulent discharge with a diameter of <5 cm; however, it may enlarge and form a protruding tumor.[2] Therefore, a histopathologic evaluation is necessary to confirm the diagnosis, which usually reveals a metastatic adenocarcinoma. Most often, metastasis occurs

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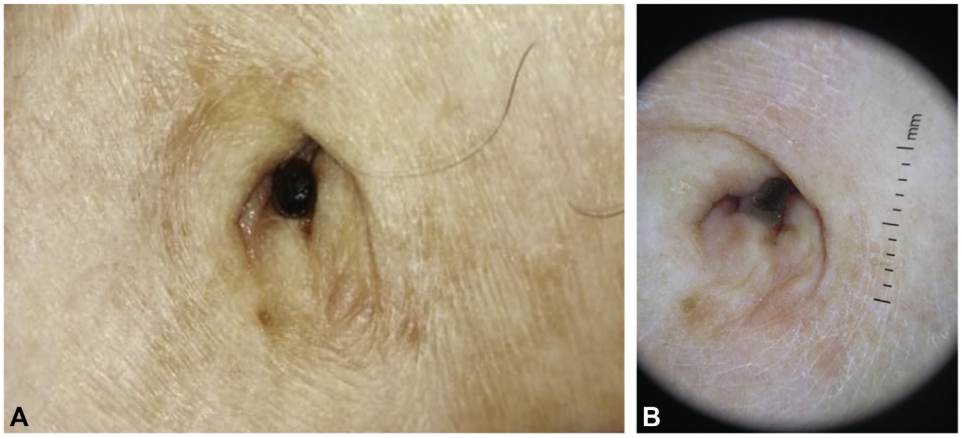

Fig 1. A, Blackish tumor of the umbilicus. **B,** Dermatoscopy: nonmelanocytic lesion with comedo-like openings.

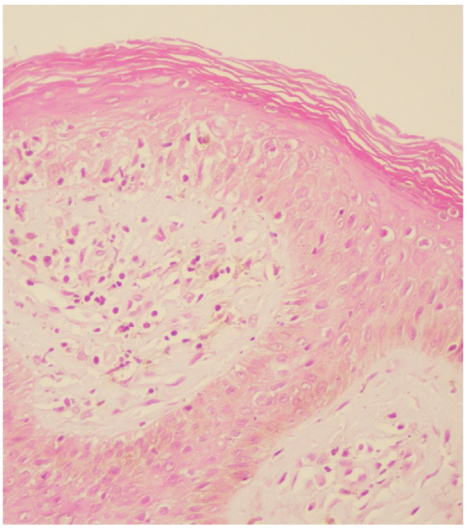

Fig 2. Epidermotropic lymphoid infiltrate composed of medium-sized lymphocytes with cribriform notched nuclei and eosinophilic cytoplasm. These cells are arranged in rows and theca. The dermis is fibrous, with the same cells described above arranged in a subepidermal band.

from gastric adenocarcinoma in men and ovarian cancer in women; however, metastases from sarcoma, melanoma, and mesothelioma have also been reported. It signifies advanced metastasizing malignancy associated with a poor prognosis and, therefore, requires urgent attention.

Benign tumors, such as omphaloliths,[4] seborrheic keratoses, dermatofibromas, epidermal cysts, dermal nevi, hypertrophic scars, keloid, and verrucae vulgares, should also be considered in adults. Lymphomas historically seemed to reject the umbilicus as a site of cutaneous spread. The few rare cases found in the literature reported umbilical metastasis of systemic lymphomas; in total, 12 cases have been reported (Table I).[5-16] Although it is not possible to draw firm conclusions regarding the outcome of these patients with umbilical lymphomatous deposits, their prognosis is definitely much better than that of their carcinomatous counterparts.

To our knowledge, this is the first reported case of primary cutaneous lymphoma presenting as an umbilical nodule. It should be noted that dermatoscopy was not helpful in this case and did not show any specific pattern.

In conclusion, our experience with this patient illustrates an important message: not all umbilical tumors are attributable to malignancies with a reserved prognosis. Although mycosis fungoides is exceptionally the cause of umbilical nodules, it should be suspected and a biopsy of any umbilical tumor should be performed.

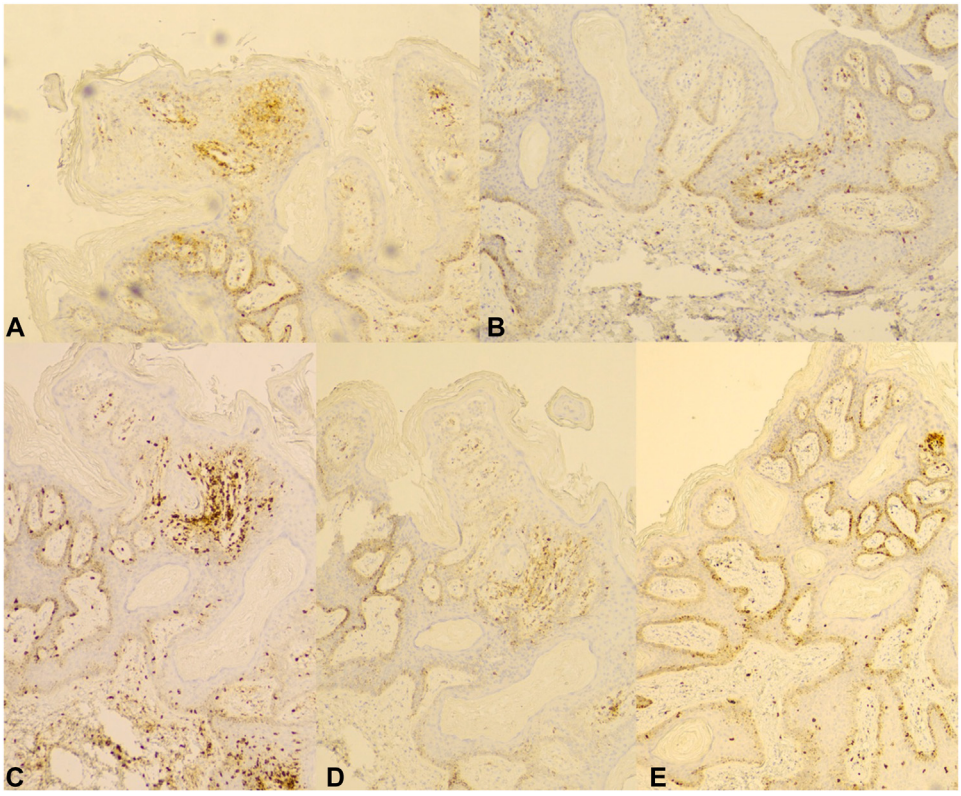

Fig 3. Immunohistochemical study findings with positive labeling of (**A**) anti-CD4, (**B**) anti-CD8, (**C**) anti-CD3, and (**D**) anti-CD5 antibodies and negative labeling of (**E**) anti-CD7 antibody.

Table I. Literature review of reports on patients with umbilical metastasis of systemic lymphomas

Patient	Age (y)/sex	Type	Treatment	Outcome	Reference
1	44/M	B cell	CHOP	Remission achieved after therapy	5
2	63/M	Large cell	NA	Remission achieved after therapy	6
3	63/M	Centrocytic	CHOP	Alive in remission for 4 y	7
4	79/M	Small cell	NA	NA	8
5	61/F	DLBCL	R-CHOP	Alive in remission for 1 y	9
6	78/F	High-grade B cell	NA	Alive in remission for 1 y	10
7	NA	NA	NA	NA	11
8	73/F	DLBCL	CEOP followed by ICE and IF XRT	Died 12 mo later	12
9	40/M	DLBCL	CHOP	Alive in remission for 4 y	13
10	30/M	Intermediate grade	NA	NA	14
11	72/M	DLBCL	R-CHOP	Died 6 mo later	15
12	72/M	MCL	R-CHOP/R/bortezomib/ gemcitabine-carboplatin-dexamethasone-R	Died 11 mo later	16

CEOP, Cyclophosphamide, epirubicin, vincristine, prednisone; *CHOP*, cyclophosphamide, doxorubicin, vincristine, prednisone; *DLBCL*, diffuse large B-cell lymphoma; *F*, female; *ICE*, ifosfamide, carboplatin, etoposide; *IF XRT*, involved-field radiotherapy; *M*, male; *MCL*, mantle cell lymphoma; *NA*, not available; *R*, rituximab.

Conflicts of interest

None disclosed.

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