CASO CLÍNICO

Sister Mary Joseph's nodule: a sign of internal malignancy*

Nódulo da Irmã Maria José: um sinal de neoplasia maligna interna

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Abstract: The umbilicus can be affected by various skin diseases including metastasis of internal malignancies, at which point it is known as Sister Mary Joseph's Nodule. The authors report a case of Sister Mary Joseph's Nodule as the first sign of internal malignancy and highlight the importance for the dermatologist to recognize the condition.
Keywords: Neoplasm metastasis; Ovarian neoplasms; Umbilicus

Resumo: A cicatriz umbilical pode ser acometida por diversas dermatoses inclusive metástase de neoplasias internas quando então recebe a denominação de nódulo da Irmã Maria José. Os autores relatam um caso de nódulo da Irmã Maria José como primeiro sinal de neoplasia interna destacando desta forma a importância do dermatologista no reconhecimento desta entidade.
Palavras-chave: Metástase Neoplásica; Neoplasias Ovarianas; Umbigo

INTRODUCTION

Sister Mary Joseph, Superintendent Nurse at St. Mary´s Hospital in Rochester, found that patients with abdominal and pelvic malignant neoplasm occasionally have an umbilical nodule indicating umbilical metastasis. [1, 2]

Hamilton Bailey, an English surgeon, first described this as Sister Mary Joseph's Nodule in 1949 in his book *Demonstrations of Physical Signs in Clinical Surgery*. [2]

The first survey of cancer relating to the umbilical area was conducted in 1846 by W H Walshe, who studied 9118 cancer and mortuary registers. This survey identified only two cases. [3]

CASE REPORT

81-year-old female patient with a history of having started bleeding in the umbilical region one month previously (associated with the appearance of a local tumor). Received emergency care and was prescribed cephalexin and acetaminophen for seven days. Failed to improve and was sent to the Dermatology Department. Examination revealed a painless *ulcerated erythematous-violaceous* umbilical nodule measuring 2x1cm. No bleeding was observed during the examination (Figure 1). A biopsy was performed with anatomopathology, which revealed an infiltrating carcinoma. An immunohistochemistry examination was next performed which showed positive for AE1, AE3, Cytokeratin 7, WT-1, estrogen receptor and CA 125, suggesting a tumor of the genital tract (uterus or ovaries) as the primary site (Figure 2). A transvaginal ultrasound showed the left-side adnexal region with a *heterogeneous hypoechoic* lesion with *cystic and solid* components (Figure 3). A *contrast* CT scan of the abdomen and pelvis showed a complex *solid* and *cystic* mass with *heterogeneous enhancement* located in the region of the uterus and ovary, with 103x96 mm displacement of the posterior bladder wall and thickening of the skin in the abdominal region (Figures 4 and 5). The patient was referred

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Figure 1: Umbilical violaceous erythematous nodule with ulceration

reagentes

marcador (anticorpo)	clone	expressão
WT-1	6F-H2	positiva (figura 4)
TTF-1	SPT24	negativa
Receptor de estrogênio	SP1	positiva (figura 3)
CK7 (citoqueratina 7)	OV-TL 12/30	positiva (figura 2)
CK20 (citoqueratina 20)	Ks20.8	negativa
CDX-2	EPR2764Y	negativa
Ca 125	OC125	positiva
BRST - 2, GCDFP-15	23A3	negativa
AE1 + AE3	AE1/AE3	positiva

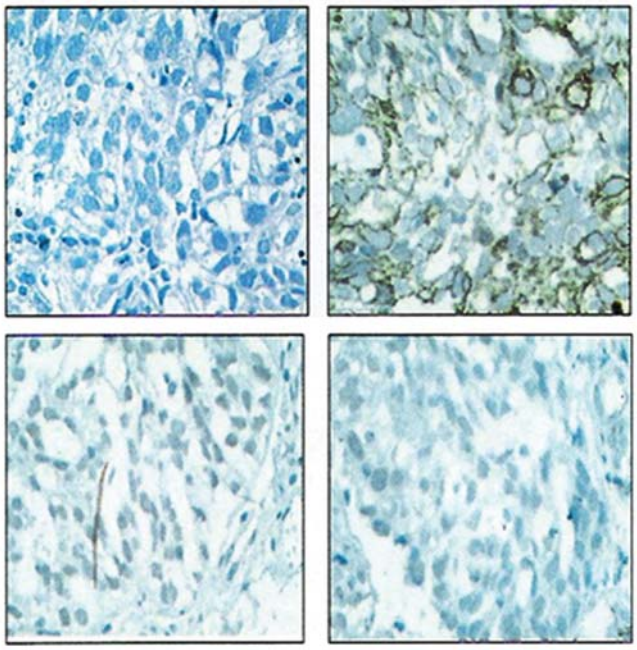

Figure 2: Immunohistochemistry

to the gynecology-oncology service where she is still being followed up.

DISCUSSION

Sister Mary Joseph´s Nodule is a form of metastasis to the umbilical region where the majority of cases are metastatic adenocarcinoma malignancies. [4] These account for 83% of all malignant tumors in the umbilical region[5]. Cutaneous metastases occur in between 1% and 9% of cases of malignancies, with around 10% affecting the umbilical region.[6] Sister Mary Joseph´s Nodule suggests widespread internal neoplasia, usually of the abdominal cavity.The most common primary sites are the gastrointestinal tract (52%) or are gynecological (28%)[7]. In around 15-29% of cases the primary site is unknown. [8]

Clinically characterized by an irregular nodule, painful or not, the nodule may present with ulceration and bloody discharge, purulent or serous. It may also present as an induration of the subcutaneous tissue. [9]

Three hypotheses for the metastases in the umbilical region exist. The most common form results from contiguity of the internal neoplasia as well as from *lymphatic* or *hematogenous dissemination.*

Discovery of this nodule on physical examination may be the first sign of internal malignant neoplasm. Symptoms such as nausea, epigastric pain, weight loss and abdominal distension are often present. [4]

Differential diagnoses include umbilical hernia, cutaneous endometriosis, pyogenic granuloma, melancocytic nevus, keloid, melanoma, squamous cell carcinoma and basal cell carcinoma. [2,10,11]

In cases where an internal malignancy resection cannot generally be performed the average survival

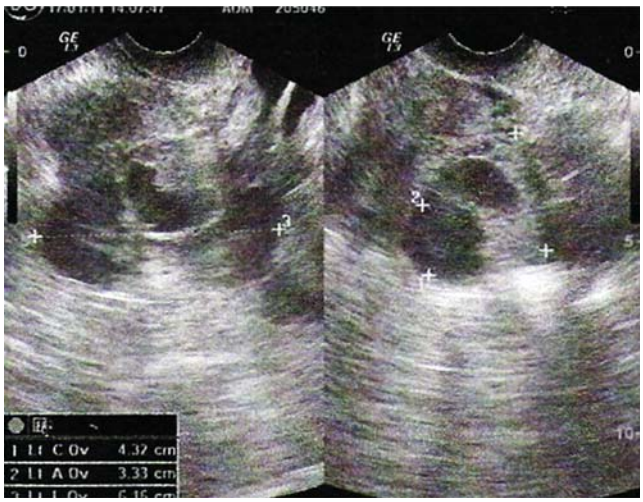

Figure 3: Transvaginal ultrasound showed left adnexal region with heterogeneous hypoechoic lesion with cystic and solid components

Figure 4: Presence of the CT with umbilical nodule (arrow)

Figure 5: TC with complex solid and cystic mass located in the region of the uterus and ovary with displacement of the posterior bladder wall

rate is 11 months. Fewer than 15% patients survive after two years. [12] Treatment usually involves palliative care, but the possibility does exist of surgical resection and radiotherapy/chemotherapy. [4]

Benign umbilical nodules are called 'pseudo-Sister Mary Joseph´s nodules'. [2,13]

In this case, the Sister Mary Joseph´s Nodule was considered to be the first sign of internal neoplasia, highlighting the need for dermatologists to be vigilant.

The umbilical metastatic cutaneous nodule and anti-CA-125 positivity revealed in the immunohistochemical test, together with the imaging methods employed (ultrasound and CT scan procedures), enabled us to diagnose metastatic ovarian adenocarcinoma. ❑

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