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Sister Mary Joseph Nodules: A Case Report About A Rare Location Of Skin Metastasis

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Abstract: *Sister Mary Joseph's nodule is an exceptional metastatic site of cancer, often pelvic, with a poor prognosis due to its delayed diagnosis. We report the case of a patient who presented to the Hassan II University Hospital in Fez with a bulging umbilical mass: Sister Mary Joseph's nodule. A biopsy of this nodule was performed with histopathological and immunohistochemical results in favor of a secondary cutaneous localization of a high-grade serous adenocarcinoma of very probable gynecologic origin. Sister Mary Joseph's nodule remains a rare tumor of metastatic origin, most often from a digestive cancer. The prognosis is still very poor, requiring early and systematic screening. This involves a biopsy of any umbilical nodule or mass to determine the nature of the pathological lesion. The aim of our work is to specify the diagnostic difficulties that practitioners face at the clinical, radiological, and pathological stages, particularly in determining the primary origin of this metastasis*

Keywords: sister Mary Joseph Nodule, MRI, Histology, Adenocarcinoma

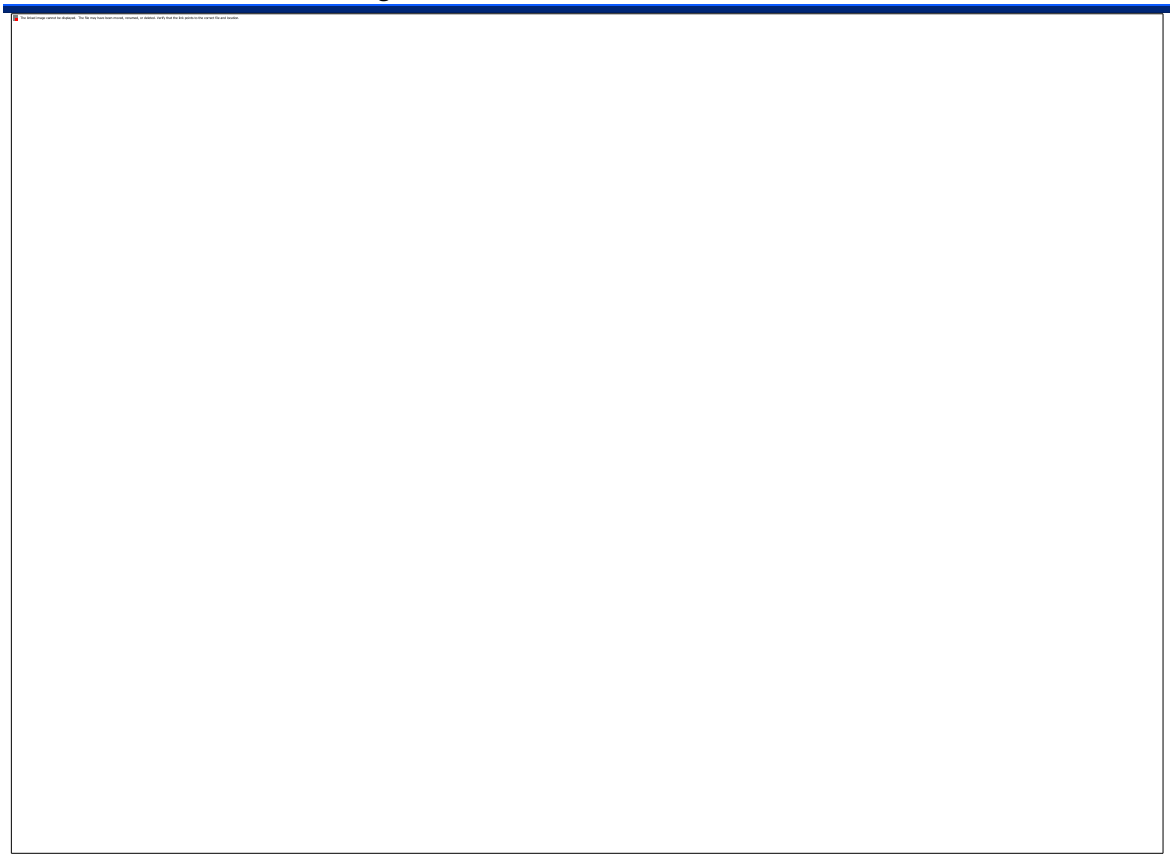
Introduction:

Sister Mary Joseph's nodule is an exceptional metastatic site of cancer, originating from intra-abdominal or pelvic tumours, with a poor prognosis due to its delayed diagnosis. The importance of a thorough examination of the umbilicus should, therefore, not be overlooked. The aim of our work is to highlight the variety of primary sites for this metastasis which could pose difficulties for practitioners in determining the primary origin.

Observation:

We are reporting the case of 60 years old woman who was admitted to our department for the onset of an umbilical nodule associated with abdominal distension and chronic pelvic pain, all evolving in a context of preserved general condition

clinical examination**:** the patient was conscious and stable in terms of hemodynamics and respiration, with the presence of an umbilical swelling: Sister Mary Joseph's nodule, and on gynecological examination, perception of a mass filling the cul-de-sac of douglas.



Sister Mary Joseph Nodule of the umbilicus

Radiological examination**:** The patient underwent a pelvic MRI revealing:

Multiple solid-cystic peritoneal masses. The largest mass involves the cul-de-sac of Douglas, measuring 84 x 75 x 50 mm, and presents the following relationships and extensions:

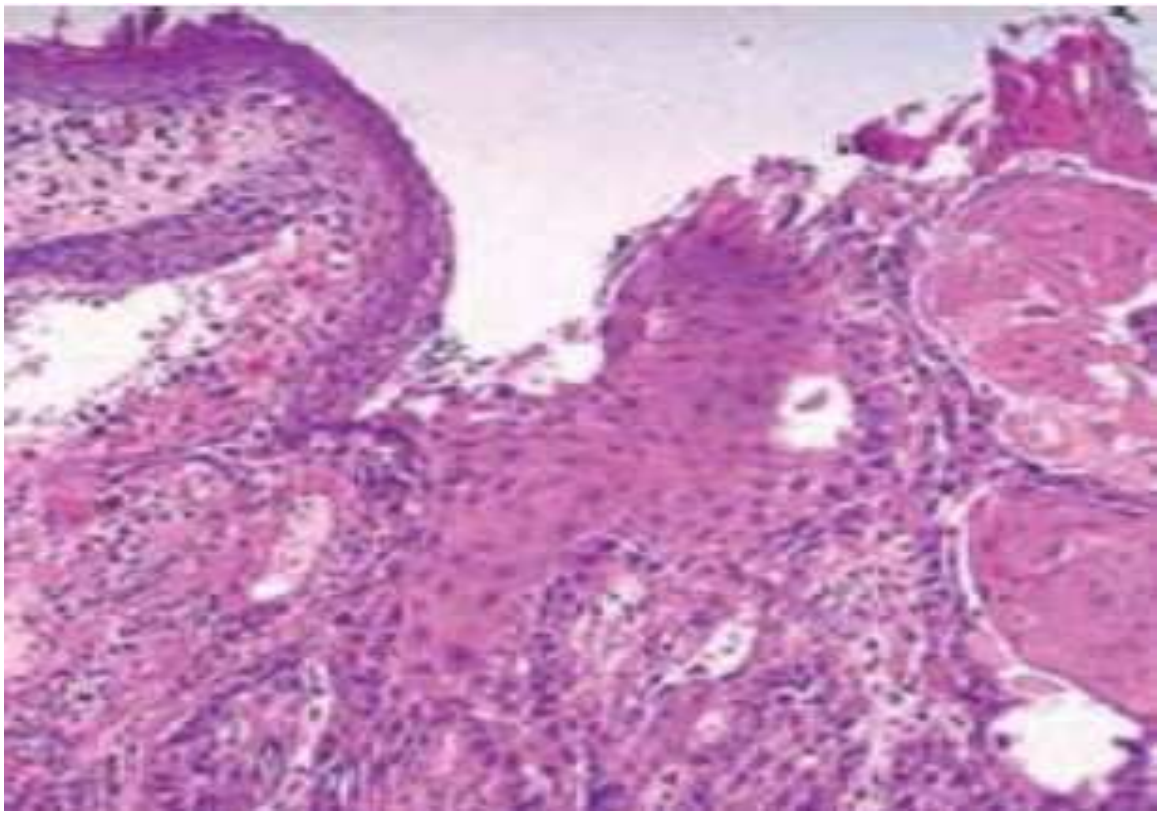
- Anteriorly, it invades the cervico-isthmic region.
- Below, it appears to invade the posterior vaginal cul-de-sac.
- Posteriorly, it is in intimate contact with the rectum without a clear fatty separation line, with uncertainty about the presence of invasion.
- It invades both parametria without ureteral invasion.
- A mass is adhered to the anterior wall of the uterus, measuring 21 mm, with uncertainty about uterine invasion.
- A left lateral uterine mass, measuring 42 mm in the major axis, of difficult-to-precise ovarian or peritoneal origin, comes into intimate contact with the sigmoid without a clear fatty edge and without obvious signs of invasion.
- Associated with this is a moderately abundant intraperitoneal effusion, with nodular peritoneal thickening in some areas.
- Secondary peritoneal, intra- and retroperitoneal lymph node locations.
- Parietal umbilical mass involving subcutaneous fat, with the same semiotic characteristics as the aforementioned peritoneal masses, measuring 25 mm, appearing secondary.
- Intra- and retroperitoneal adenopathies and lymph nodes."

the thoraco abdominal Pelvic CT Scan revealing:

- Thickening of the antral gastric digestive wall and another probable focal rectal thickening, requiring correlation with endoscopic data.
- Peritoneal carcinosis appearance above and below the mesocolon, with multiple masses and nodules of pelvic carcinosis, associated with suspicious-looking external and internal iliac lymph nodes.
- Mediastinal and bilateral pulmonary hilar lymphadenopathy that may be part of the same condition.
- Lesion on the hepatic dome requiring correlation with targeted ultrasound data.
- Macronodular thyroid requiring correlation with targeted ultrasound data."

Anatomopathology: A biopsy of the Sister Mary Joseph's nodule was performed with histopathological and immunohistochemical results in favor of a secondary cutaneous localization of a high-grade serous adenocarcinoma of very probable gynecologic origin. The patient subsequently underwent an upper endoscopy and a colonoscopy with gastric and rectal mucosal biopsies, which did not reveal any suspicious tumor cells.

The ovarian origin has been established.



Histopatholgy of adenocarcinoma

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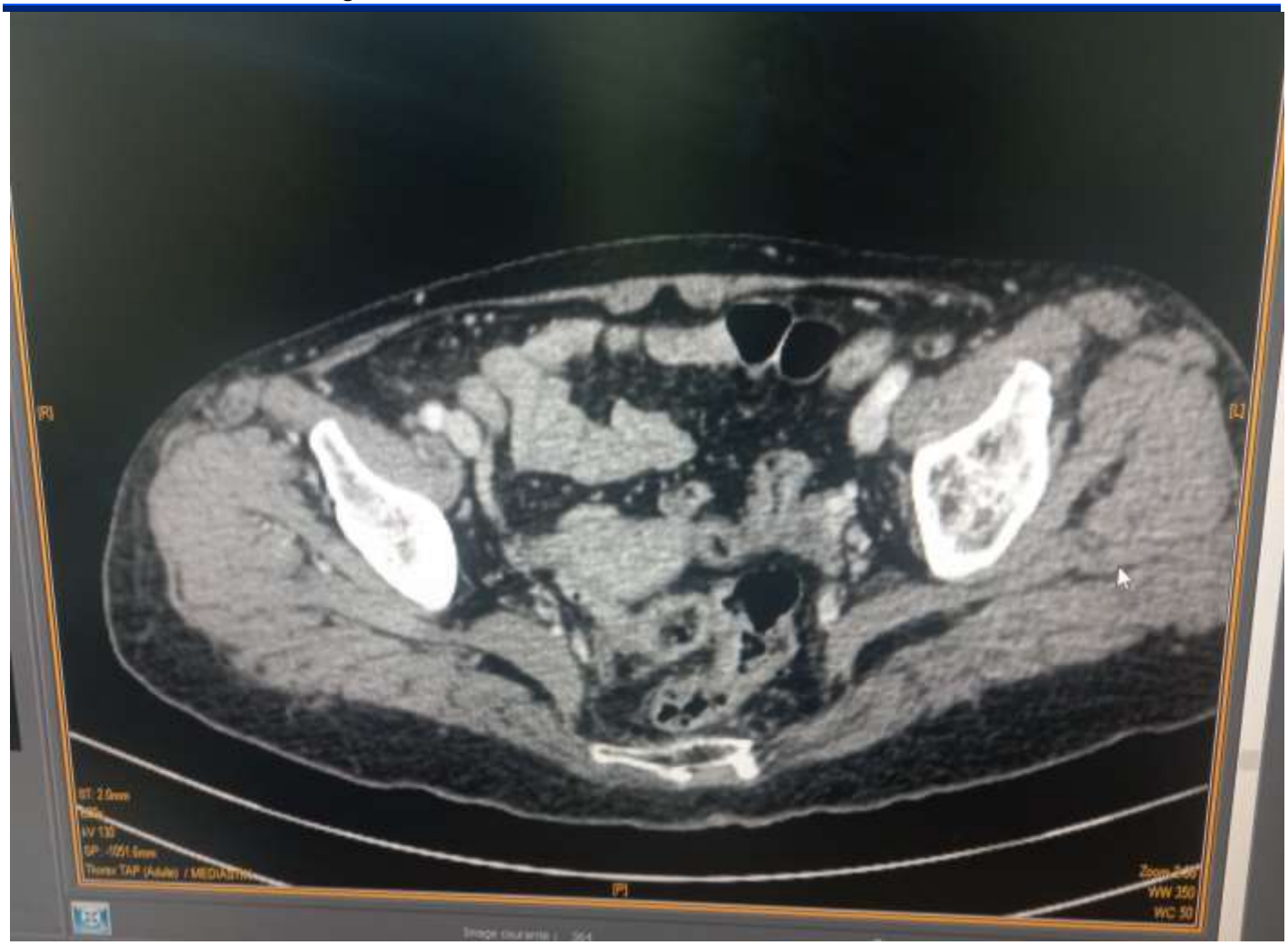
Neoadjuvant treatment: The patient reported the complete disappearance of the umbilical nodule after just 2 sessions of chemotherapy, and control CT scan showing regression of ovarian masses.



CT scan showing the regression of the ovarian masses after chemotherapy

Discussion:

The sign of Sister Mary Joseph nodule has been extensively described in literature

The most common origins of Sister Mary Joseph nodule are gastrointestinal (52%), gynecologic (28%), stomach (23%) and ovarian (16%) carcinomas

In 30% of patients the source of the primary neoplasm may not be found.2 This is more frequently encountered in females. The commonest histological type is adenocarcinoma (about 75% of cases), and is more rarely epidermoid, undifferentiated, or carcinoid. [1]

A review by Majmudar in 1991 of 25 cases of metastatic carcinoma of the umbilicus in women showed that there were no cases of primary umbilical malignancies identified. Eighteen of 25 patients had a gynecologic primary tumor, with 11 ovarian, 3 endometrial, 2 cervical, and one uterine mixed Mullerian and vulvar cancer. Microscopic examination of the tumor showed adenocarcinoma in 19 cases (76%), with 4 of them showing mucin production by mucicarmine stain and 4 had papillary features . A patient with a pancreatic primary demonstrated adenosquamous carcinoma, while another showed undifferentiated carcinoma. There were 3 cases of squamous cell carcinoma (12%), 2 originating from the cervix and one from the vulva. There was one case of mixed Mullerian tumor arising from the uterine corpus and invading the umbilicus [2].

Although umbilical malignancies are rare, primary umbilical tumors are much more so compared to metastatic tumors. Historically, the first large review by Cullen in 1916 on umbilical carcinoma had 22 cases of primary umbilical malignancies while 72 were due to metastatic disease [3]. A review of 667 cases of umbilical lesions by Barrow [4] showed 29.7% to be metastatic with an 8.4% incidence of primary malignant tumors. The remaining cases were benign primary tumors or endometriosis. In Steck's review of malignant umbilical tumors, 8 were primary lesions while 40 were metastatic [5].

Conclusion:

Sister Mary Joseph's nodule remains a rare tumor of metastatic origin, most often from a digestive cancer. The prognosis is still very poor, requiring early and systematic screening. we recomended that in all patients a careful examination of the umbilicus be performed, especially in those with known malignancies. All suspicious nodules should be biopsied to determine whether the nodule represents metastatic tumor and the primary site determined.

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