



Clinical Case

Sister Mary Joseph Nodule: Three Case Reports of Unusual Skin Metastasis

Nodule de Soeur Mary Joseph: A propos de trois cas de métastases cutanées inhabituelles

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ABSTRACT

Sister Mary Joseph's nodule is a metastatic umbilical nodule usually associated with the primary neoplasm of the gastrointestinal and genitourinary tract. Umbilical metastasis is rare in clinical practice. It can be the first sign of an undiagnosed neoplasm or can appear as a recurrence or progression of an already known tumor, and usually indicate poor prognosis. We report three cases of Sister Mary Joseph's nodule which emphasize the importance of correctly diagnosing an umbilical nodule.

RÉSUMÉ

Le nodule de Soeur Marie-Joseph est une métastase cutanée ombilicale d'un cancer du tractus gastrointestinal ou de la sphère gynécologique. C'est une lésion rare qui peut être inaugurale ou apparaître dans le cadre d'un cancer connu et signe une récurrence ou une progression néoplasique. Sa présence est souvent associée à un mauvais pronostic. Nous rapportons 03 cas de nodule de soeur Mary Joseph qui soulignent l'importance de diagnostiquer correctement un nodule ombilical.

INTRODUCTION

Sister Mary Joseph's nodule refers to a palpable nodule bulging into the umbilicus as a result of cancer in the abdomen or pelvis [1]. Sister Mary Joseph's nodules are seen in 1-3% of all intraabdominal or pelvic malignancies [2,3]. It is an uncommon clinical sign which reveals the poor prognosis of these malignancies [3, 4].

We report three cases of Sister Mary Joseph's nodule seen in our department, as the first sign of malignant disease.

CASE 1

A 20 years-old patient was referred to surgery emergency because of a five-month history of abdominal pain and umbilical mass noticed two weeks ago. The patient also complained of loss of appetite, weight loss, and progressive abdominal distension.

The ultrasound showed an umbilical mass measuring 23mmx17 mm thought to be an irreducible umbilical hernia. Laparotomy was performed. Surgical abdominal exploration revealed multiple nodules disseminated on the peritoneum and the handles, ascites, and a tumoral mass involving the umbilic. The umbilical mass has been resected

and the peritoneal nodule was biopsied. The pathologic examination of the surgical specimen showed secondary localization of adenocarcinoma infiltrating the epidermis (figure 1).

In the postoperative period, the patient was sent to the oncology unit where further investigations were done. Computed Tomography of chest, abdomen, and pelvis showed peritoneal carcinosis and ascites but any primary tumor site was found. Both upper and lower gastrointestinal endoscopy showed no abnormalities. Tumor markers including the Carbohydrate Antigen 19-9 and carcinoembryonic antigen were in high- levels.

Immunohistochemistry was not performed as it was not available in our context. Finally, this case was diagnosed as an umbilical metastasis of unknown digestive primary tumors.

Palliative chemotherapy with FOLFOX regimen (5-Fluorouracil 1200 mg/ m² continuously for 46h, levofolinate 200 mg/m², oxaliplatin 85 mg/m²) was proposed and permitted to obtain the decrease of ascites and peritoneal nodules after six cycles. Three months after the treatment, the patient presented recurrence and died approximately twelve months after the diagnosis.

CASE 2

A 25 years old patient presented with 6 months history of the umbilical nodule with diffuse abdominal pain. He had no medical history.

Clinical examination found an ulcerated umbilical tumor painful and bleeding on contact measuring 50x 45mm. (figure 2).

The assessment with an abdominal Computed Tomography scan revealed an umbilical heterogenous mass associated with mesenteric nodule measuring 15.4x 8.5 mm and ascites. The percutaneous biopsy of the umbilical nodule revealed metastasis of a moderately differentiated adenocarcinoma. Fibroscopy detected a gastric tumor and the pathologic examination of this lesion showed gastric adenocarcinoma.

The diagnosis of gastric adenocarcinoma revealed by umbilical metastasis was retained. The patient underwent two cycles of palliative chemotherapy (Capecitabine 625 mg/m² twice daily during 21 days, oxaliplatin 130 mg/m²). The evolution was fatal four months after the diagnosis.

CASE 3

A 63-year-old man presented with a growing periumbilical mass during the last two months treated unsuccessfully as an abscess and progressive cholestatic jaundice appeared two weeks ago. His medical history includes hypertension. He reported abnormally dark urine and light-colored stool. He had pruritis, loss of appetite, and constipation. On clinical examination he was icteric. The abdomen was distant and the umbilical lesion fixed to the underlying tissues. Liver function tests were abnormally elevated.

A Computed Tomography scan showed an umbilical mass measuring 29x28 mm involving the percutaneous tissue (figure 3), associated with heterogeneous hepatomegaly, dilatation of the bile duct, and an endoluminal tumoral lesion extending to the right and left hepatic bile ducts. There was an irregular focal parietal thickening of the gallbladder measuring 22x 88 mm and multiple Celio mesenteric lymph nodes.

Laparotomy was performed. The surgery consisted of resection of the umbilical mass and palliative bypass bile duct. The pathological examination identified the umbilical lesion as a secondary localization of an adenocarcinoma. Based on the results of imaging and the surgical exploration of this patient, the gallbladder was considered as the primary tumor site without performing a further biopsy.

Palliative chemotherapy (Gemcitabine 1250 mg/m², Cisplatin 25 mg/m²) was proposed and the treatment is ongoing.

DISCUSSION

Cutaneous metastases can appear in any region of the skin. The Sister Mary Joseph Nodule refers to an umbilical cutaneous metastasis of internal malignancies [5]. Although it is a rare clinical finding seen in only 3% of metastatic cancers, Sister Mary Joseph nodules have been described in

several case reports [6]. However, to our knowledge, these cases are the first reported in Togolese patients.

The term Sister Mary Joseph nodule was first used by sir Hamilton Bailey in honor of Sister Mary Joseph [7]. Julia Dempsey (1856-1939) also known as Sister Mary Joseph Dempsey was an American Catholic nun, surgical nurse, and assistant to the famous surgeon William James Mayo who found that patients with abdominal and pelvic malignant neoplasm occasionally have an umbilical metastasis [1].

Classically the nodule is usually firm, irregular, and ulcerated and can be associated with induration, bleeding, discharge, and secondary infection [5,8]. It is typically less than 5 cm in diameter but occasionally enlarges enough to form a protruding tumor [9]. Some cases up to 10 cm have been reported [5].

Sister Mary Joseph nodules are usually detected during or years after the diagnosis of the primary neoplasm [8], however in up to 30% of cases, the nodule is the first and only sign of neoplasm [10].

As in the cases shown here, the most common histology is adenocarcinoma but it can also be squamous cell carcinoma or undifferentiated cancer [3].

Searching for the sites of primary malignant tumors is essential when the diagnosis of Sister Mary Joseph's nodule is made. In men, the commonest primary site is the gastrointestinal tract of which the stomach is the most common entity while gynecological malignancies, particularly epithelial ovarian tumors are the most common primary sites in women [3, 6].

The origin of the primary tumor is unknown in 15-30% of patients [11]. The possible mechanism of tumor spread to sister Mary Joseph could be through lymph ducts, blood vessels, contiguous extension, or embryologic remnants in the anterior abdominal wall [4].

Imaging techniques (ultrasound, CT scan, magnetic resonance imaging) are essential to determine the origin of the primary neoplasm. Fine Needle Aspiration Cytology or core biopsy of the tumor is invaluable in the diagnosis and helps to exclude a benign umbilical lesion [9].

Differential diagnosis should include benign causes such as endometriosis, fibroma, omphalitis keloid, foreign body or umbilical hernia as well as primary malignant umbilical skin tumors [12].

Management of the disease should take into account the clinical state of the patient and the etiology of the primary malignancy. Sister Mary Joseph Nodule suggests advanced distant metastasis and is associated with poor prognosis [3, 4]. Mean survival following diagnosis varies between 2 and 11 months [2,10]. Fewer than 15% of patients survive more than 2 years after diagnosis [4] An aggressive treatment combining surgical excision, radiotherapy, and chemotherapy can be proposed [13]. However, as the disease is usually advanced or metastatic, only palliative treatment is offered like it was done in our cases.

CONCLUSION

Sister Mary Joseph nodules are uncommon but crucial physical examination findings of intraabdominal and pelvis malignant neoplasm. All physicians need to be aware of such a rare entity. A careful examination of all umbilical lesions is recommended to avoid delayed management of the underlying neoplasm.

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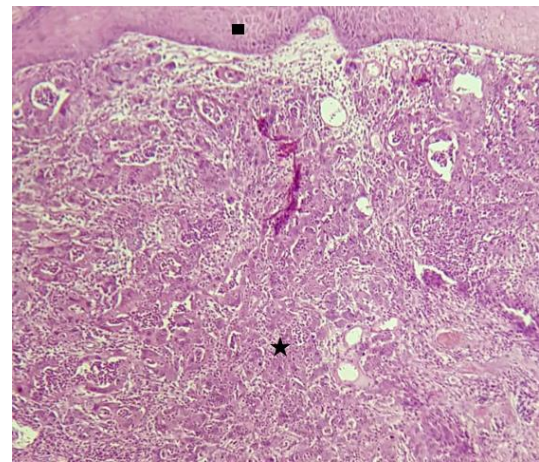


Fig1 : Pathological specimen from umbilical nodule showing an adenocarcinomatous proliferation (star) involving a regular epidermis (Haematoxylin and Eosin staining ; x25)



Fig 2 : Ulcerated Sister Mary Joseph's nodule (white Arrow)

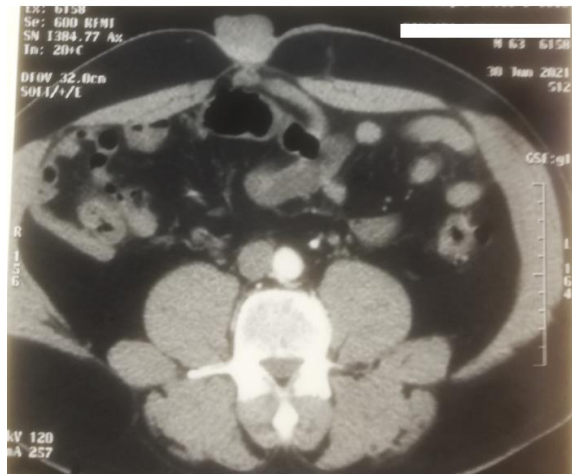


Figure 3: CT scan of the abdomen showing sister Mary Joseph nodule measuring 29x28 mm (white arrow)