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Sister Marie Joseph's Nodule Revealing an Adenocarcinoma of Excreto-Biliary Origin: Apropos of a Case.

F. Zahra Cheikhna * [1], Cherif Mini [2], M Yeslem Madi [3], Ahmed Ballati [4]

1,2,3,4. Gastroenterology Department Visceral Surgery Department, Imaging Department, Military Hospital, Nouakchott Mauritania.

Corresponding Author: F. Zahra Cheikhna, Gastroenterology Department Visceral Surgery Department, Imaging Department, Military Hospital, Nouakchott Mauritania.

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Abstract

The Sister Marie-Joseph nodule (SMJ) is an umbilical cutaneous metastasis of cancer, most often digestive. The primary tumor is usually an adenocarcinoma, rarely a squamous cell carcinoma, a melanoma or a sarcoma. It is a rare metastatic umbilical lesion, which occurs in 1 to 3% of abdominopelvic adenocarcinomas. Most often it is the stomach, the ovary, the colon and the pancreas. Bile duct and small intestine involvement are very rare locations. Its discovery should lead to the performance of a skin biopsy and an abdomino-pelvic CT scan. Therapeutic possibilities include surgery and/or chemotherapy, but most often the treatment is palliative. The prognosis remains poor because the discovery is most often late.

Key words; *Umbilical nodule, metastasis, adenocarcinoma, Sister Marie-Joseph, Excretory-biliary tumor.*

Introduction

The nodule of Sister Mary Joseph is a metastasis umbilical from intra-abdominal cancer **[**1**]** . It represents 30% of umbilical tumors and in 88% of cases, it it is an adenocarcinoma **[**2**]**. The site of the tumor primitive is most often the stomach, the ovary or the colon. **[**3**]** Very few observations of hepatic tumors with Umbilical metastasis have been described in the literature.

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We report an exceptional case of this lesion superficial revealing hepatocellular carcinoma. The Sister Marie-Joseph nodule is a cutaneous metastasis located at the level of the umbilicus, present in 1 to 3% of abdominopelvic adenocarcinomas (1). It was described by Sister Marie-Joseph Dempsey, an operating nurse at William J. Mayo in Rochester, Minnesota (1) . Late onset in the course of the disease, it is associated with a poor prognosis. In a third of cases, it represents the first manifestation of the cancerous disease (2, 3).

We report a case of an umbilical skin metastasis revealing an excreto-biliary adenocarcinoma in a patient admitted to the gastroenterology department of the Nouakchott military hospital in Mauritania.

Observation

A 68-year-old patient who consulted in the gastrological emergency department for a painful umbilical skin lesion that appeared one month before her consultation. At the interview, she has no particular pathological history, she reports a notion of epigastralgia associated with nausea and anorexia evolving for 2 months without any notion of weight loss. On clinical examination, a conscious patient is observed, hemodynamically and respiratory stable PS at 2, jaundiced, abdominal examination reveals an umbilical swelling 2 cm in diameter, firm, painful, erythematous and slightly oozing.

Faced with this table, a blood test was done showing hepatic cytolysis, total bilirubin at 50mg/l predominantly conjugated, CA19-9 and high ACE an abdominal CT scan was performed showing a secondary hepatic mass of metastatic appearance with nodules of peritoneal carcinomatosis . She benefited from a FOGD which objectified an erythematous pangastritis. A biopsy of the umbilical nodule was carried out and the Histological and immunohistochemical aspect was in favor of an adenocarcinomatous process of excreto-biliary origin. The patient was proposed for palliative chemotherapy.

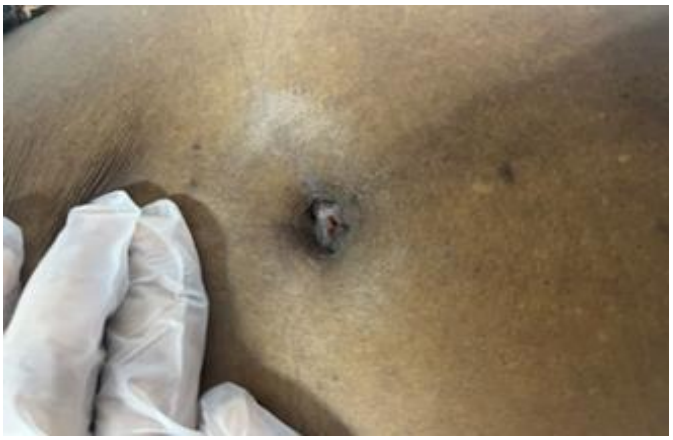

Figure 1: image of the umbilical nodule of our patient

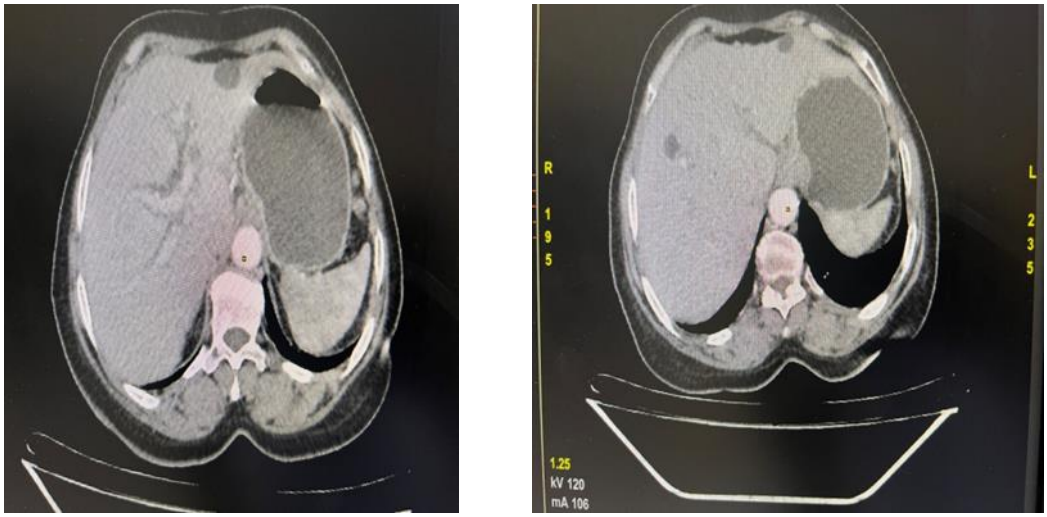

Figure 2: Abdominal scan of the patient

Discussion

The nodule of SMJ is a nodule of the umbilicus most often secondary to a metastasis of an abdomino-pelvic cancer (4), a squamous cell carcinoma, a melanoma or a sarcoma are found more rarely (10%). It was sister and nurse Marie-Joseph who, in 1928, at the Mayo Clinic in Minnesota, discovered the association between this nodule and stomach carcinoma and gave it her name. It was first described by Sir Hamilton Bailey in 1949 in his book "Physical signs in clinical surgery" (3,4).

Epidemiologically, umbilical metastases are rare. They represent 3 to 4% of all secondary tumor locations and 10% of all secondary skin lesions (5,6). Primary lesions should be sought primarily in the stomach (25%), ovary (12%), colon (10%) or pancreas (7%). The endometrium, cervix, bile ducts and small intestine are rare locations (3). In a third of cases, the origin of the primary cancer is not identified. The pathophysiology of Sister Marie-Joseph's nodule is still incompletely elucidated. Indeed, there are four possible ways of dissemination of neoplastic cells towards the umbilicus which are: hematogenous dissemination, extension from the round hepatic ligament to the middle umbilical ligament of the urachus, retrograde lymphatic reflux and dissemination by contiguity from the anterior surface of the peritoneum via the vessels of the umbilicus, which explains the appearance of the Sister-Marie-Joseph nodule in the advanced stages of deep-seated cancers often accompanied by peritoneal carcinomatosis. A few iatrogenic cases following laparoscopy have been reported in patients who probably had occult intra-abdominal tumors (1,8,9). The average age of onset of these metastases is 60 years with no difference in distribution by sex(7). The age of our patient matches the data of the literature with an age of 68 years.

This nodule appears as a rounded, irregular, indurated swelling, often painful and oozing, sometimes itchy. It can take different colors: white, purple, red, brown. It usually measures between 5 and 20 mm in diameter, but can reach up to 10 cm. Sometimes ulcerated, fissured or necrotic, its evolution is sometimes characterized by a discharge of blood, pus or serous fluid (2,3). Imaging is insufficient to distinguish these benign or malignant lesions from each other. Biopsy, often easy to perform without or with ultrasound guidance, is then the investigation of choice to support the diagnosis.

The differential diagnosis of an umbilical nodule includes benign conditions such as botryomycoma, omphalitis, hernias and umbilical locations of Crohn's disease or endometriosis (1,2).

With an average life expectancy of ten to eleven months, the prognosis after the diagnosis of NSMJ is unfavorable (10). A radical surgical procedure with adjuvant chemotherapy is associated with a better prognosis with a longer life expectancy compared to surgery or chemotherapy alone (11)

Conclusion

The SMJ nodule is a metastasis of a cancer most often of digestive origin. It is often the only sign of an underlying oncological disease. The prognosis is poor requiring systematic screening.

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