

Sister Marie-Joseph's Nodule Revealing A Pancreatic Adenocarcinoma: About A Case with Review of the Literature

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Abstract: The Sister Marie-Joseph nodule (SMJ) is an umbilical cutaneous metastasis of a cancer of the gastrointestinal tract or of the gynecological sphere [1]. Stomach and colon are the common gastrointestinal cancers associated with the SMJ nodule. The pancreas is a rare primary site of umbilical metastasis [1]. In pancreatic cancer, the Sister Mary Joseph nodule is a sign of advanced disease associated with a poor prognosis [2].

Keywords: Cholestatic jaundice, umbilical nodule, tumoral process of Winslow's small pancreas.

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INTRODUCTION

Sister Marie-Joseph's nodule is an umbilical cutaneous metastasis of a cancer of the gastrointestinal tract or of the gynecological sphere. It was described by Sister Marie-Joseph Dempsey, an operating nurse at William J. Mayo in Rochester, Minnesota [3]. Pancreatic cancer accounts for 7-9% of reported cases of Sister Mary Joseph's nodule, with nearly 90% of tumors occurring in the body and tail of the pancreas [4]. We report the case of a patient with SMJ nodule as the initial clinical presentation of pancreatic cancer.

PATIENT AND OBSERVATION

Patient information

This is a 61-year-old patient with no particular pathological history who consults the emergency department for a periumbilical umbilical

collection with outcome of serous fluids initially treated with antibiotics in a local hospital without improvement. The evolution is marked three weeks later by the appearance of progressive cholestatic jaundice evolving in a context of apyrexia and weight loss of 5 kg for which he was hospitalized in our gastroenterology department at the University Hospital of Marrakech.

Clinical Result

On admission, the patient was conscious, normocardic at 81 beats/minute and eupneic at 17 beats/minute, afebrile at 37°C. Her body mass index was 20kg/m². The jaundice was frank. Abdominal examination found indurated, ulcerated umbilical nodule with issue of serosities measuring approximately 3 cm, painful on palpation (Image 1). There were no peripheral adenopathies.

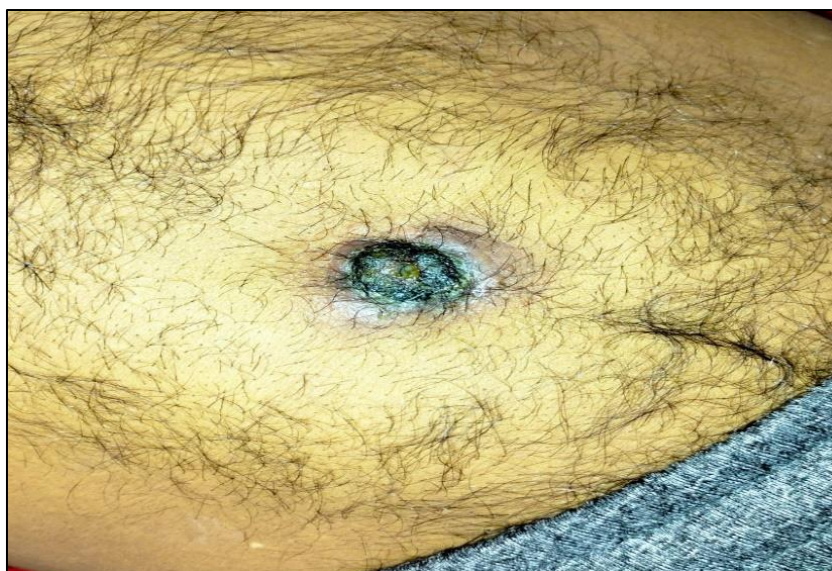


Image 1: Clinical appearance of the SMJ nodule

Diagnostic Approach

The patient benefited from an assessment biological objectifying a cytotoxicity with AST at 64 IU/L (1.2N) ALT: 63 UI/L (1.5N) and biological cholestasis with GGT levels at 133 IU/L (1.3N), alkaline phosphatase was at 272 IU/L (2.1N) and the total bilirubin level were at BT 184.2mg/l predominantly conjugated with BC: 168mg/l.

An abdominopelvic scan revealed the presence at the level of the soft parts of a collection measuring 19.4*19.4mm, a localized dilation of the intrahepatic bile ducts with localized peri-portal infiltration of segment II of the liver and a lesion of the head of the pancreas with a cystic appearance. The decision was to complete with a bili-MRI objectifying an evolving aspect of the locally infiltrating small Winslow pancreas tumor process with vascular sheathing and infiltration of the retroportal lamina, responsible for significant dilation of the upstream intra and extra hepatic bile ducts. Associated with secondary-looking retroperitoneal lymphadenopathy with an infectious-looking periumbilical collection (Image 2). Chest CT was normal. A biopsy of the umbilical nodule was performed, showing a dermal location of a moderately differentiated and infiltrating adenocarcinoma with, in addition, an immunohistochemical appearance in favor of a location of a moderately differentiated and infiltrating adenocarcinoma of gastro-bilio-pancreatic origin gastroscopy did not show any abnormality.

Therapeutic intervention and follow-up:

A cholangiography performed showed an impassable stenosis of the hepatic convergence with

significant dilation of the intrahepatic bile ducts (IBVH) upstream.

External radiological biliary drainage of the left VBIH was performed using an 8F coiled pigtail drain upstream of the stenosis with fixation of the drain to the skin. Subsequently, the patient was referred to oncology for palliative care.

DISCUSSION

The nodule of SMJ is a palpable nodule of the umbilicus secondary most often to a metastasis of an abdominopelvic cancer. It is a discreet and rare clinical sign. Its incidence is low, since it is estimated that 1 to 3% of patients with abdominopelvic neoplasia may present with an SMJ nodule [5]. The most reported digestive origins in descending order are: stomach (25%), colon or rectum (10%) and pancreas (7%) [6]. 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 [7]. The discovery of such a nodule must imperatively lead to the realization of an abdominopelvic scanner and a biopsy of the lesion in order to obtain an anatomopathologic diagnosis. The prognosis is more often dark with the presence most often of an advanced oncological situation as in the observed case of our patient. Treatment is often palliative. The late diagnosis in our study can be partly attributed to the discrete nature of the SMJ nodule, often trivialized by patients, and to a lack of knowledge of the disease by practitioners.

This lesion makes it possible to discuss the differential diagnosis with umbilical endometriosis, pyogenic or foreign body granulomas, hemangioma, umbilical localization of Crohn's disease or melanoma [8]. Therefore, only the anatomopathological study can confirm the diagnosis. The preponderant mode of extension is by contiguity from the anterior surface of the peritoneum, otherwise the extension can be venous, lymphatic or around embryonic residues (round ligament, urachus, vestige of the vitelline artery) [9].

This rare but characteristic umbilical metastasis should be known to the practitioner. It is easily accessible for clinical examination. Its recognition as a secondary lesion of primary abdomino-pelvic cancer can allow early diagnosis and treatment.

CONCLUSION

Sister Marie Joseph's nodule is rare. It may be the only warning sign towards a primary gastrointestinal tumour, thus posing the diagnostic problem. The presence of this nodule is a pejorative element of the prognosis. This rare but characteristic umbilical metastasis must be known to the practitioner to allow early diagnosis and treatment.

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