Sister Mary Joseph Nodes: About three cases

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Key words: Ombilical metastasis, Pancreatic adenocarcinoma, Gall blader cancer.

ISSN: 2070-254X

Abstract

Sister Joseph nodule is a metastatic umbilical lesion secondary to a primary malignancy of any viscera. The majority of cases originate from gastrointestinal or ovarian cancer. It is an uncommon clinical sign, which reveals the poor prognosis of these malignancies. Here, we present 3 such cases, with Sister Joseph nodule metastasing from bilo-pancreatic tract. The clinicopathologic features of all 3 patients are discussed, and the related literature is briefly reviewed.

Introduction

The Sister Mary Joseph’s nodule (SMJN) is a periumbilical metastatic tumor originating from advanced metastatic intra-abdominal and intrapelvic malignancies. The term SMJN was first used by Sir Hamilton Bailey in honor of Sister Mary Joseph, a surgical assistant who found that patients with abdominal and pelvic malignant neoplasm occasionally have an umbilical metastasis [1]. Here, we report 3 cases of SMJN encountered at our institute.

Observations

Observation 1
A 70 years old female presented with 6-months history of painful red periumbilical swelling. Examination of the patient revealed tender erythematous firm umbilical nodule about 2 cm in diameter (figure 1). CT abdomen revealed pancreatic mass with peritoneal carcinomatosis. Fine-needle aspiration cytology of the umbilical nodule revealed metastasis of a well-differentiated adenocarcinoma suggestive of pancreatic origin. The patient is currently undergoing gemcitabine based chemotherapy.

Observation 2
A 71 years old female presented with 3 months history of umbilical nodule associated with diffuse abdominal pain. CT abdomen revealed a pancreatic mass with liver metastases. Biopsy from the liver and umbilical nodule revealed adenocarcinoma. The patient received Gemcitabine based chemotherapy with progression of the disease. Patient then received capcitabine associated with Oxaliplatin as second line chemotherapy then she was lost of view.

Observation 3
A 60 years old woman presented with history of umbilical swelling associated with anorexia nausea and weight loss. General examination of the patient revealed poor general condition, cachexia with performance status of 3. Abdominal examination revealed painless firm indurated subcutaneous umbilical nodule measuring 3cm. MRI and CT abdomen revealed gall bladder mass with liver metastases, peritoneal carcinomatosis and ureteral invasion. Umbilical nodule biopsy revealed a carcinoma of biliopancreatic tract. The evolution was fatal before starting chemotherapy.

Discussion

In 1928, Sister Mary Joseph, a head surgical nurse of William James Mayo, first pointed out the umbilical metastatic nodule [2]. However, the Sister Joseph’s nodule was first described only in 1949 when Bailey mentioned the term in his book [3]. This lesion may be the first clinical sign of an underlying advanced intra-abdominal or intrapelvic malignancy. It’s diagnosed in only 1%–3% of all intra-abdominal or pelvic malignancies [4,5]. The mechanism of umbilical seeding from primary tumours is not clearly understood; however, authors have proposed several hypotheses [6].

- Direct invasion, through continuous extension, of the anterior peritoneum or along the embryonic ligaments. As in two of our cases CT scan revealed peritoneal carcinomatosis indicating invasion of the peritoneum.
- Lymphatic spread via the axilliary, inguinal, para-aortic, internal mammary and external iliac lymph nodes;
- Hematogenous spread through arterial and venous seeding;

Sister mary joseph nodule usually presents with an irregular and painful umbilical nodule or even just induration of the subcutaneous tissue. It may develop ulceration and bloody or serous discharge. The diagnosis of SMJN is based on histopathology because of the large number of possible differential diagnoses. When a patient presents with an umbilical nodule, fine-needle aspiration biopsy is adequate to establish an easy and early diagnosis. In most cases the umbilical neoplasm, is a metastasis of an adenocarcinoma followed by squamous cell carcinoma, undifferentiated malignant tumors and carcinoids [1,6,7]. The most common origins are the stomach and colon in males and the ovaries in females. In our case series the bilo-pancreatic tract was the primary site in all the cases [1,6,8].
The umbilical metastatic nodule is a sign of advanced malignancy and poor prognosis. Mean overall survival is 2–11 months without treatment [8,9, 10]. Recent reports have suggested that prognosis could be improved to 17.6–21 months with aggressive management (surgery and adjuvant chemotherapy) [9]. However, surgery should be used only in patients with a solitary umbilical metastasis. In our cases, only palliative chemotherapy could be indicated and the evolution was fatal in two of the three patients because of widespread disease.

Conclusion

Sister Mary Joseph nodule is an uncommon manifestation of visceral malignancies. It is important to identify these lesions during routine imaging, as they may be the unique metastatic site.

Figures

Fig. 1: Umbilical erythematous nodule with necrosis measuring 2 cm

References