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GLOBAL JOURNAL OF
Medical and Clinical Case Reports

ISSN: 2455-5282 DOI: https://dx.doi.org/10.17352/gjmccr

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**Received:** 12 September, 2019
**Accepted:** 04 November, 2019
**Published:** 05 November, 2019

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Case Report

# An unusual presentation revealing pancreatic carcinoma: Sister Mary Joseph Nodule

## Abstract

Sister Mary Joseph Nodule (SMJN) is a cutaneous nodule resulting from metastasis of malignant tumors affecting the umbilicus; it had been reported in 0.7e10.4% of patients.

This case report describes an 82-year-old male diagnosed with metastatic pancreatic adenocarcinoma and confirmed by the biopsy of Sister Mary Joseph's nodule.

The aim of this presentation is too aware of such a specific manifestation, enables the physicians to investigate the underlying malignancy and to show the role of biopsy of the nodule for the diagnosis of the primitive cancer.

## Introduction

Peri-umbilical metastases are uncommon symptoms of advanced cancers mostly abdominal locations (typically stomach, large bowel, and ovary) [1]. The term is named after Sister Mary Joseph, she was a nurse at Saint Mary's hospital; she recognized that patients with an umbilical lump often died from cancer [2].

This case report describes an 82-year-old male, diagnosed with metastatic pancreatic adenocarcinoma and confirmed by the biopsy of Sister Mary Joseph's nodule.

## Case Report

A.M, is a 82 years old patient with no particular pathological history. Who presented abdominal pain with an umbilical induration evolving for 8 months. Evolution was marked by the appearance of an abdominal distension of average abundance. Everything evolves in a context of weight loss and anorexia. Clinical examination found an umbilical nodule of 3 cm diameter, hard consistency with ascites of moderate abundance (Figure 1).

During his hospitalization, the patient had a general assessment. An iron deficiency anemia has been found associated with hypocholesterolemia and hypoalbuminemia. Cholestasis parameters (γ-glutamyl transpeptidase, 108U/L; alkaline phosphatase, 357U/L; bilirubin, 4.2mg/dl) were markedly elevated. CA 19-9 was increased (2139U/ml). Ascetic fluid analysis showed a chylous ascites.

CT scan (Computed Tomography) was done. A mass of 5cm diameter in the pancreatic tail that invades the vessels was detected (Figure 2).

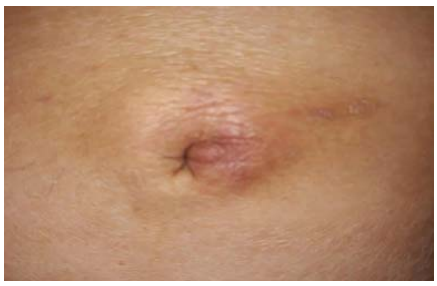

**Figure 1:** Umbilical nodule.

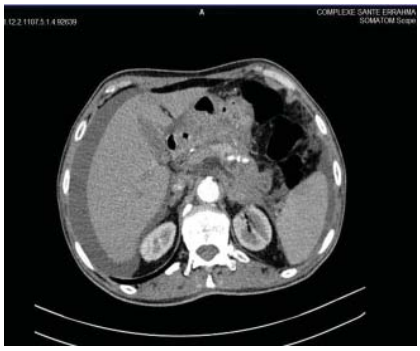

**Figure 2:** Tumor at the pancreatic tail with ascites.



**Citation:** Sghir H, Jellal S, Errami AA, Oubaha S, Samlani Z (2019) An unusual presentation revealing pancreatic carcinoma: Sister Mary Joseph Nodule. Glob J Medical Clin Case Rep 6(2): 032-034. DOI: https://dx.doi.org/10.17352/2455-5282.000076

Diagnosis of pancreatic adenocarcinoma was confirmed by biopsy of the umbilical mass, and immunohistochemical staining was positive for cytokeratin 7, CK20, CK19 and pancytokeratin, and negative for CDX2.

The patient refused any further treatment. He died 40 days after the diagnosis.

## Discussion

Sister Mary Joseph Nodule (SMJN) is a term describing metastatic umbilical nodule. Observed firstly by a nurse at Saint Mary's Hospital but first description was done by Hamilton Bailey in the 11th edition of 'Physical Signs in Clinical Surgery [3].

Differential diagnoses should include umbilical hernia, cutaneous endometriosis, benign tumors such as foreign body granuloma, melanocytic nevi, papilloma, fibroma and primary umbilical carcinoma which are exceedingly rare, accounting for 17% of the cases and including melanoma, squamous and basal cell carcinoma. Since many benign conditions can mimic this umbilical metastasis, a histological or cytological study of the umbilicus is not only mandatory, but it also guides the clinician to search for potential primary site [4,5].

This nodule became an alarm and poor prognosis sign of many malignant tumors. The main primary sites described in the literature are the stomach, ovary, colon and pancreas. Other rare sites are reported. In 15% the primary tumor is unknown [1,6].

There are many possible mechanisms of the spread of metastasis to the umbilicus. The most known are; direct extension, haematogenous spread and lymphatic spread. The most common primary sites are intra-abdominal. Other causes has also been reported like arterial embolization, and Tumor implantation at the umbilicus after laparoscopic cholecystectomy for unsuspected gallbladder carcinoma [7].

Clinically, this type of nodule is most often painful, fibrotic consistency to hard and irregular margins. Its surface can be ulcerated and necrotic with serous or purulent discharge [8,9].

Immunohistochemical markers are very important to define the origin of unknown primary cancer. Monoclonal antibodies to cytokeratin 7 and cytokeratin 20 are the most used because of their distinct expression by different organs as pancreatic adenocarcinoma [10]. Other serum markers can be used as CA19.9; there elevation can be considered as a strong evidence of pancreatic adenocarcinoma [11].

Imaging is very helpful to establish diagnosis. Ultrasonography, CT scan, MRI, and PET scan. A biopsy: fine needle aspiration cytology or excision is necessary to pose diagnosis and to find the primary cancer once Sister Mary Joseph's nodule is diagnosed [12].

The presence of Sister Mary Joseph's nodules usually signifies an advanced, metastasizing cancer. And almost certainly establishes the inoperability of the patient [13].

The treatment is commonly palliative. Several authors have advocated wide excision [14], radiotherapy [15] and surgery with adjuvant therapy [16].

Majmudar and al have estahblished the superiority of surgery with adjuvant therapy: an average of 17.6 months survival, vs 7.4 months for surgery alone, 10.3 months for adjuvant therapy alone and 2,3 months when no treatment [16]. Gabriele and al found longer survivals with surgery and chemotherapy (17-21 months) [17].

The main indication of surgical excision is a solitary metastasis or in case of complications, and have no indication in case of unresectable disseminated cancer.[1] The prognosis is related with the primary tumor, the SMJN caused by ovarian cancer have a better survival rate [18,19].

In our case, based on clinical presentation, the results of imaging studies and cytokeratin staining, it is clinically rational to conclude that the primary tumor site was the pancreas, without performing biopsy of the pancreas mass.

The presence of SMJN often means that the Survival rate is very poor. The survival time without treatment ranged from 2 months to 11 months, in our case it was 40 days [6].

## Conclusion

In conclusion, SMJN is unusual, Awareness of such a specific manifestation enables the physicians to investigate the underlying malignancy promptly, and its superficial location makes it amenable to biopsy, which can assist in making a specific tissue diagnosis.

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