

## Sister Mary Joseph's nodule:

an unusual but important physical finding characteristic of widespread internal malignancy

### INTRODUCTION

Sister Mary Joseph's nodule is an eponymous term referring to a malignant metastatic umbilical nodule. It is a rare but important physical finding as it is a sign of advanced stage of malignancy. This case report describes an 82-year-old female diagnosed with metastatic pancreatic adenocarcinoma presenting with a Sister Mary Joseph's nodule. This is an unusual presentation of pancreatic malignancy.

### CASE REPORT

An 82-year-old woman presented to the GP with a 4-week history of an umbilical nodule (Figure 1). This was suspected to be a neoplastic lesion and she was referred to the dermatology department for further assessment. She was otherwise asymptomatic. She had a past medical history of type 2 diabetes mellitus, essential hypertension, and anaemia due to chronic kidney disease. On examination, she looked well with no abdominal discomfort. There was a 3 cm umbilical nodule and bilateral inguinal lymphadenopathy. The working diagnosis was a Sister Mary Joseph's nodule with underlying internal malignancy.

Urgent outpatient investigations were arranged. Blood tests demonstrated normocytic anaemia, raised tumour markers, renal impairment, and a normal liver function test. Imaging with a staging computerised tomography scan of her chest, abdomen, and pelvis, and biopsy of the umbilical nodule confirmed the diagnosis of metastatic pancreatic adenocarcinoma. She was referred to the hepatobiliary multidisciplinary team; the malignancy was judged to be inoperable due to the extent of the disease, and she was referred to the Macmillan Team for continuing palliative care in the community. Six weeks after initial presentation to secondary care, she developed gastric outlet obstruction symptoms and died 5 weeks later.

This case raised the question of how to diagnose Sister Mary Joseph's nodule and how to distinguish it from other causes of umbilical nodules?

### BACKGROUND

Metastatic umbilical nodule was first described in the literature in 1864.<sup>1</sup> This clinical sign later became known as Sister Mary Joseph's nodule, named after Sister Mary Joseph Dempsey (1856–1939), surgical assistant of Dr William James Mayo, who first noticed the association between abdomino-pelvic malignancies and metastatic umbilical nodules.<sup>2</sup>

### EPIDEMIOLOGY

Sister Mary Joseph's nodule is uncommon, with an estimated 1–3% cases of abdomino-pelvic malignancy metastasising to the umbilicus.<sup>3</sup> Sister Mary Joseph's nodule is usually associated with primary neoplasm of the gastrointestinal (35–65%) and genitourinary tract (12–35%).<sup>4</sup> Other reported sites included lung, pancreas, liver, gallbladder, lymphoma, breast, kidney, penis, prostate, and testicles. The source of the primary neoplasm may not be found in up to 30% of patients.<sup>4</sup>

### AETIOLOGY

The aetiology of Sister Mary Joseph's nodule remains unclear and the proposed

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Figure 1. A Sister Mary Joseph's nodule.



**Table 1. The differential diagnosis of an umbilical nodule**

Differential diagnosis	Associated features	Management
Primary umbilical neoplasm	<ul style="list-style-type: none"><li>• May be indistinguishable from a Sister Mary Joseph's nodule</li></ul>	Referral to secondary care
Sister Mary Joseph's nodule	<ul style="list-style-type: none"><li>• Symptoms and physical signs suggestive of malignancy may be present</li><li>• History of a previously treated primary neoplasm in some cases</li></ul>	Referral to secondary care
Umbilical and paraumbilical hernia	<ul style="list-style-type: none"><li>• The hernia may be congenital or acquired after abdominal surgery</li><li>• Uncomplicated hernias are reducible and associated with a cough impulse</li></ul>	Referral to secondary care
Umbilical endometriosis	<ul style="list-style-type: none"><li>• Occurs in females of reproductive age</li><li>• Presents with a bleeding umbilical nodule especially during menstruation</li></ul>	Referral to secondary care
Keloid	<ul style="list-style-type: none"><li>• History of skin trauma or navel piercing</li></ul>	Options include no treatment, silicon gel, intralesional steroids, surgery, and laser
Omphalith	<ul style="list-style-type: none"><li>• Poor personal hygiene leading to hardening of exfoliated skin and sebum accumulated within the umbilicus</li></ul>	Personal hygiene advice
Pyoderma gangrenosum	<ul style="list-style-type: none"><li>• Typically associated with an underlying disorder such as inflammatory bowel disease, rheumatoid arthritis, and myeloproliferative disorder</li></ul>	Referral to secondary care
Foreign body	<ul style="list-style-type: none"><li>• Identification of a foreign body</li></ul>	Removal of foreign body

hypothesis includes direct extension of tumour to the umbilicus, lymphatic, or haematogenous spread.<sup>5</sup>

### PRESENTATION

Sister Mary Joseph's nodule typically presents as an umbilical or paraumbilical nodule with a firm consistency, varying in size between 0.5–15 cm.<sup>5</sup> The nodule may be painful and discharge fluid. It is important to bear in mind that Sister Mary Joseph's nodule may be the only presenting complaint in an otherwise well patient; other patients can present in a poor clinical state with additional physical signs such as ascites and pleural effusion.<sup>6</sup> A focused history and examination of the chest, abdomen, and regional lymph nodes may reveal the source of the primary neoplasm. Patients who presented with Sister Mary Joseph's nodule following treatment of a known neoplasm raise the possibility of its recurrence.

### DIFFERENTIAL DIAGNOSIS

Consideration of the differential diagnosis of a patient presenting with an umbilical nodule should include primary umbilical neoplasm, Sister Mary Joseph's nodule (metastatic umbilical nodule), umbilical hernia, umbilical endometriosis, omphalith,

keloid, pyoderma gangrenosum, and foreign body. Distinguishing between these differential diagnosis can be difficult and Table 1 provides a guide to facilitate GPs with their diagnosis and management of these conditions.

### MANAGEMENT

The finding of a Sister Mary Joseph's nodule (or a suspicious looking umbilical nodule) in primary care should prompt urgent referral to secondary care for further assessment. Patients who are debilitated by their illness may require hospital admission for investigation and management. Biopsy of the umbilical nodule provides a convenient way of obtaining tissue sample for histological diagnosis of the disease. Imaging with CT and/or MRI scan will establish the extent of the malignancy.

The prognosis of patients presenting with Sister Mary Joseph's nodule is generally poor as it is a sign of advanced malignancy. Management of the disease should take into account patient preference, the clinical state of the patient, and the aetiology of the primary malignancy. Palliative management may be the only option for some patients, whereas in carefully selected cases, patients may benefit from more aggressive treatment such as surgery, chemotherapy, and radiotherapy.<sup>4</sup>

### CONCLUSION

Sister Mary Joseph's nodule is an uncommon but important physical finding. Therefore, this is an important differential diagnosis for GPs to consider in patients presenting with umbilical nodules.

### Patient consent

The patient provided written consent for the photograph to be published. The patient's next of kin provided written consent for this article to be published.

### Provenance

Freely submitted; externally peer reviewed.

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