

Sister Mary Joseph's Nodule: An important Physical Finding.

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Research Article

Keywords: umbilical lesions, Sister Mary Joseph's nodule, intraabdominal malignancy, Prognosis

Posted Date: June 25th, 2021

DOI: <https://doi.org/10.21203/rs.3.rs-629180/v1>

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Abstract

Background: Sister Mary Joseph's nodule is a rare but important physical finding as it is a sign of an advanced stage of malignancy.

Case presentation: This is a case of an apparently clinically well but immunodeficient 62-year-old African woman whose main complaint was an enlarging violaceous discharging umbilical nodule. A focused history and physical examination complemented by an abdominal computerized tomography (CT) scan was suggestive of an umbilical cutaneous manifestation (Sister Mary Joseph's nodule) of an underlying disseminated ovarian malignancy. This was confirmed histologically by an excision biopsy of the umbilical nodule.

Conclusions: Sister Mary Joseph's nodule is an important differential diagnosis for an umbilical nodular lesion as it is a sign of advanced malignancy with a generally poor prognosis.

Background

Sister Mary Joseph's (SMJ) nodule is an eponymous term referring to a malignant umbilical nodule. It was first described in the literature in 1864 [1] and, was named after Sister Mary Joseph Dempsey (1851–1939) a surgical assistant to Dr William James Mayo who first noticed the association between abdomino-pelvic malignancies and metastatic umbilical nodules [2]. It is a rare but important physical finding as it is a sign of an advanced stage intra-abdominal malignancy. A clinical presentation of Sister Mary Joseph's nodule as an umbilical cutaneous manifestation of a disseminated ovarian carcinoma in a highly active antiretroviral treatment (HAART)-compliant human immunodeficiency virus (HIV) infected patient is reported herein.

Case Presentation

A 62-year-old African woman presented at the outpatient clinic with a 4-week history of a rapidly progressing umbilical nodular lesion that was associated with some discomfort and a clear fluid discharge. She felt generally well except for gradual weight loss and anorexia in the past months. She was an HIV-infected patient but compliant with highly active antiretroviral treatment (HAART). On examination she was thin and listless with abdominal bloatedness and discomfort. The abdomen was tense and with a shifting dullness on percussion suggestive of ascites and a tender 5 cm hepatomegaly. She had a violaceous non-tender 4 cm umbilical nodule which was firm in consistency and irreducible. The working diagnosis of an SMJ nodule with an underlying intrabdominal malignancy was made (Fig. 1). The differential diagnosis included (1) a complicated umbilical or paraumbilical hernia being not reducible and had no associated cough impulse, (2) a primary umbilical neoplasm which may be indistinguishable from a SMJ nodule, (3) umbilical endometriosis but occurs in females of reproductive age and presents with a bleeding umbilical nodule especially during menstruation, (4) a pyoderma gangrenosum but typically associated with inflammatory bowel disease,

rheumatoid arthritis and myeloproliferative disorders. (5) a foreign body but usually associated with an identifiable foreign body, (6) an omphalith and (7) a keloid. Blood tests revealed a microcytic anaemia with a haemoglobin level of 8g/dl. A computed tomography (CT) contrast scan of the abdomen and pelvis demonstrated a large ovarian tumour, an umbilical nodule measuring 4cm in diameter, some ascites and, a segment IV liver metastases (Fig. 2). She underwent an excision biopsy of the umbilical nodule under local anaesthesia. The tumour tapered intraperitoneally, thus allowed a gush of ascitic fluid following its excision. While awaiting a definitive histopathological report the macroscopic appearance was suggestive of a metastatic ovarian adenocarcinoma (Fig. 3).

Discussion

This case report demonstrated that SMJ nodule although uncommon is a sign of advanced malignancy [3]. Similar umbilical cutaneous metastases from ovarian carcinoma have been reported by Touraud et al in 2000 [4], Calongos et al in 2016 [5] and Carrey et al in 2018 [6]. SMJ nodule is uncommon with an estimated 1–3% cases of abdomino-pelvic malignancy. Of these 35–65% involves primary caecinas of the gastrointestinal tract and 12–30% involves genitourinary tract carcinomas [7]. Other reported sites include lung, pancreas [3, 8, 9], oesophagus [10], stomach [11], liver, gallbladder, caecum [12], breast, uterus [13], kidney, penis, testicles and prostate[14]. However, the source of the primary neoplasm may not be found in up to 30% of patients [7, 15]. The mechanism of metastatic spread to the umbilicus remains unclear but include contiguous, lymphatic or haematogenous spread. It typically presents as an umbilical or paraumbilical nodule with a firm consistency varying in size between 0.5-15cm [3, 12]. The nodule may be painful and discharging fluid and, may be the only complaint by the patient as in this case. Other patients can present in a poor clinical state with additional physical signs such as ascites and pleural effusion [16]. A focused history and examination of the chest, abdomen and regional lymph nodes may suggest and reveal the source of the primary neoplasm. Patients who present with SMJ nodule following treatment of a known neoplasm raises the possibility of tumour recurrence, thus acting as part of a paraneoplastic syndrome [12, 15, 17, 18]. Imaging with CT and/or magnetic resonance imaging (MRI) scan will establish the extent of the malignancy (i.e. tumour staging) [3, 19]. Biopsy of the umbilical nodule will provide tissue sample for histological diagnosis of the type of malignancy prior to treatment [3]. The management of the disease should take into account patient preference, the clinical state of the patient and the aetiology of the primary malignancy [3, 7]. Palliative treatment may be the only treatment for some patients, whereas carefully selected patients may benefit from more aggressive treatment with surgery, chemotherapy and radiotherapy [7].

Conclusion

Sister Mary Joseph's nodule is an important physical finding being a sign of advanced intra-abdominal malignancy with usually a poor prognosis. It is a differential diagnosis to consider in a patient with an umbilical nodule.

List Of Abbreviations

SMJ. Sister Mary Joseph. HIV, Human immunodeficiency virus. HAART, Highly active retroviral treatment

Declarations

Ethics approval and consent to participate: Not required as being a case report

Consent for publication: Written informed consent was obtained from the patient for publication of this case report. A copy of this written consent is available for review by the editor-in-chief of this journal.

Availability of data and materials: Not applicable

Competing interests: The authors declare that they have no competing interests

Funding: None

Authors' contributions: EPW is the main author and surgeon, MMT contributed to literature search, JCM referred the patient and contributed to literature search.

Acknowledgement: I acknowledge the nursing staff of the surgical unit of the Regional Hospital Limbe for the concern and care given to the patient.

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Figures



Figure 1

Sister Mary Joseph's nodule

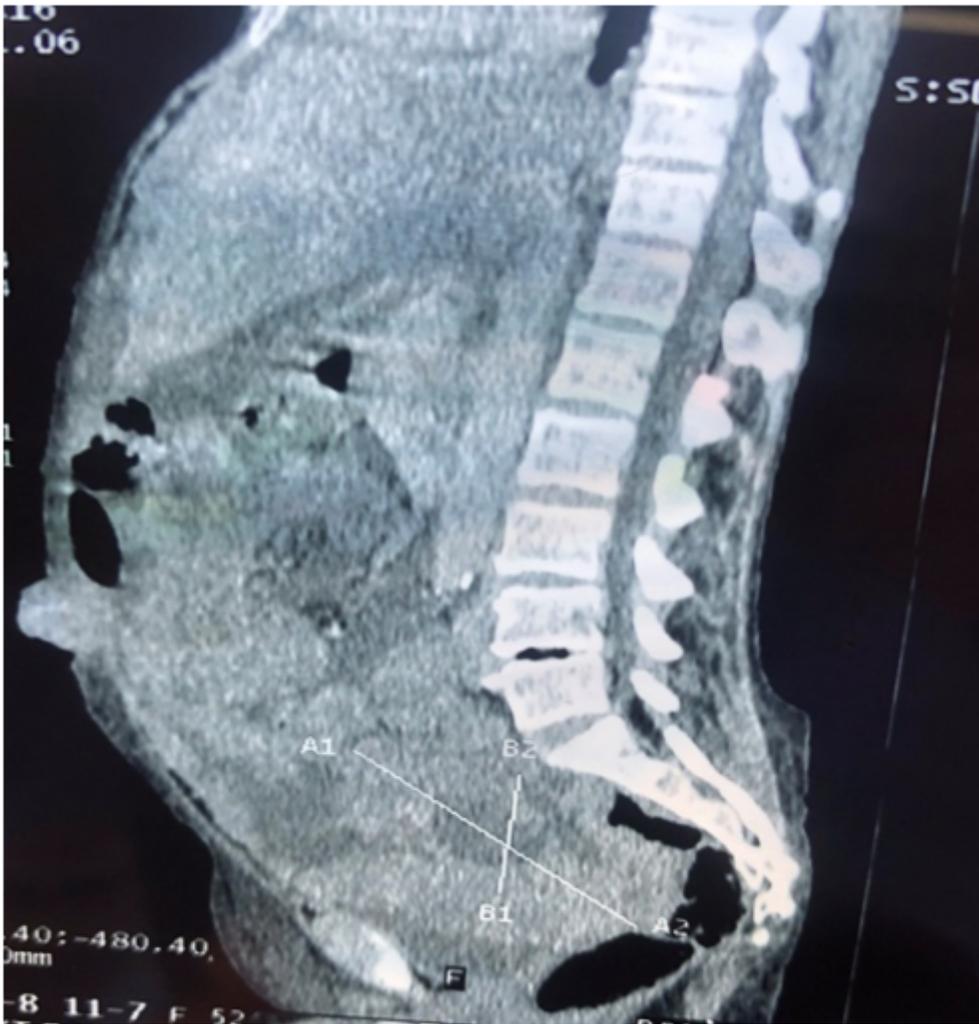


Figure 2

A CT scan of Abdomen demonstrating pelvic tumour and an umbilical nodule

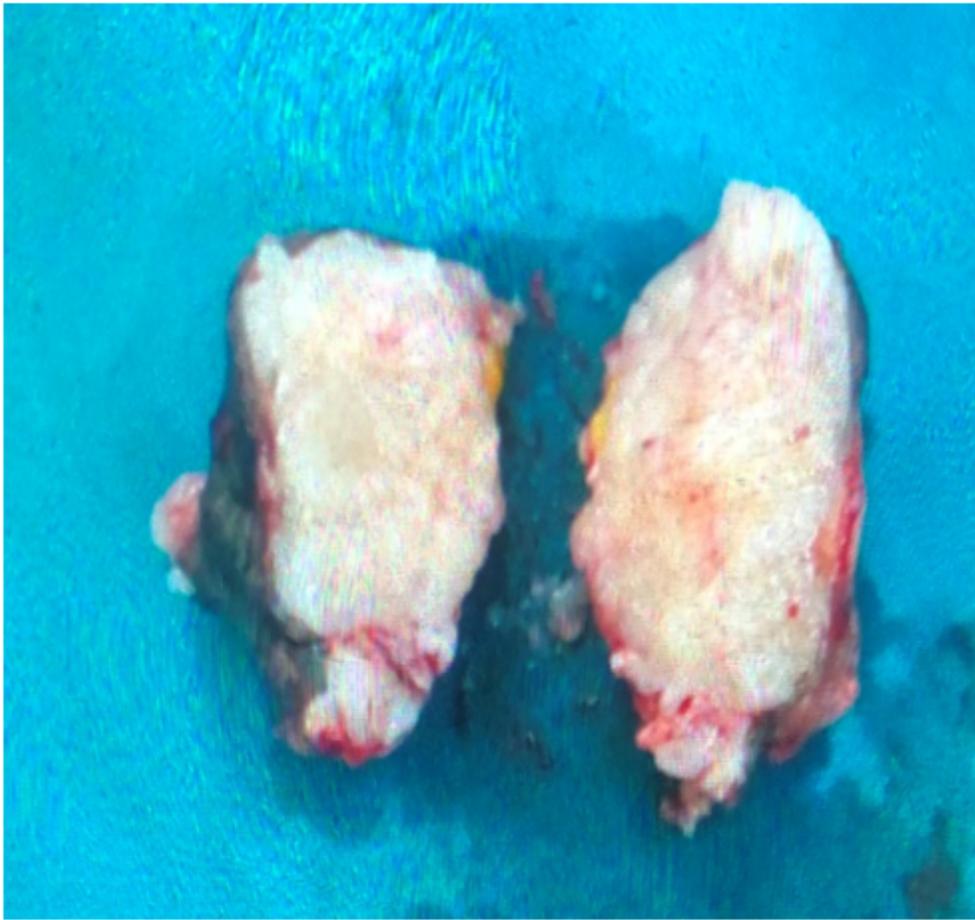


Figure 3

Macroscopic appearance of excised nodule with the consistency of a carcinoma