Giant Sister Mary Joseph nodule in gastric carcinoma

Ergin Erginöz 1, Ahmet N. Şanlı 1, Deniz E. Şanlı 2, Server S. Uludağ 1
1Department of General Surgery, Istanbul University, Cerrahpasa School of Medicine, Istanbul
2Department of Radiology, Acibadem Kozyatagi Hospital, Istanbul, Turkey

Abstract

Nodules in the periumbilical region are a rare finding as the differential diagnosis varies widely from umbilical hernia to malignant metastatic cancer. Therefore, any suspicious looking periumbilical nodules warrant further investigation of an underlying gastrointestinal or a genitourinary disease, as these usually indicate an advanced stage of malignancy. A Sister Mary Joseph (SMJ) nodule is one of these types of nodules that, once diagnosed, it must be promptly referred to the associated specialty to begin standard therapy for the patient. It is still unclear how these intra-abdominal malignancies spread towards the periumbilical region. Swift actions must be taken to make a diagnosis. In this case report, we present a 37-year-old male patient with a 15-cm Sister Mary Joseph nodule with an underlying gastric signet ring cell carcinoma.

Keywords

Sister Mary Joseph; Gastric carcinoma; Periumbilical nodule
Introduction
Sister Mary Joseph nodule was named after Sister Mary Joseph Dempsey, an assistant of Dr. Mayo at the Mayo Clinic, who observed that patients presenting to the clinic with a palpable nodule in the umbilical region usually presented with intra-abdominal malignancy [4]. The SMJ nodule is a palpable, cutaneous metastatic nodule that is malignant in nature originating from a tumor in the gastrointestinal tract [7]. These tumors often include adenocarcinomas of the gastrointestinal system (usually gastric, colon, and pancreas) and genitourinary tumors (ovarian and endometrial), carcinoid tumors, or undifferentiated tumors [1,2]. These nodules usually indicate advanced stage of the disease and are often associated with poor prognosis [6,7]. The pathophysiology remains unclear, but includes different routes towards the umbilical region. Some of the proposed hypotheses include hematogenous spread via branches of the external iliac artery and internal mammary artery, venous drainage including branches of the internal mammary vein and femoral vein, lymphatic drainage around the umbilical region, or via remnant structures such as the falciform ligament and the median umbilical ligament [3,5]. In the clinic, an SMJ nodule usually presents with an irregular lump in the periumbilical region varying from pink to brown in appearance. The nodule may appear necrotic, vascular in nature, and hard in consistency. The size of the nodule is usually less than 2 cm, but it can reach to sizes up to 10 cm, as seen in this case report [3]. The differential diagnosis includes an umbilical hemia, SMJ nodule, neoplasm of the umbilicus, endometriosis of the umbilical region, melanoma, or a keloid [1,2]. Therefore, any nodule in the periumbilical region requires prompt referral to the corresponding secondary care centers.

Case Report
A 37-year-old male patient presented to our clinic with symptoms of fatigue, weight loss, and a periumbilical nodule. In physical examination, the patient was fully cooperated and appeared pale. The nodule in the periumbilical region was nearly 15 cm in size with irregular borders which was elevated above the umbilical region (Figure 1). On palpation, the nodule was firm. Percussion and auscultation of the abdomen did not reveal any abnormality. The laboratory evaluation revealed anemia. Kidney and liver function tests were normal and electrolyte levels were within normal limits. Viral serology results were negative. The tumor markers included CA 19-9 849.80 U/ml (normally <39 U/ml), CEA 4.63 ng/ml (normal <6.5 ng/ml), AFP 16.03 ng/ml (normal <15 ng/ml), PSA 0.60 ng/ml (normal <4 ng/ml). MRI of the abdomen revealed an umbilical protrusion (Figure 2).

CT of the abdomen also showed similar results with MRI. Whole body PET scan showed increased 18F-fluoro-2-deoxy-d-glucose (FDG) uptake along the greater curvature of the stomach and a linear FDG uptake along the distal esophagus near the gastro-esophageal junction (corresponding with esophagitis). Gastroscopy was performed and the biopsy of the greater curvature of the stomach was consistent with signet ring cell carcinoma of the stomach. Incisional biopsy of the periumbilical nodule was also consistent with a signet ring cell carcinoma infiltration.

Discussion
SMJ nodule usually presents with a periumbilical mass with varying morphological features such as pink to brown in color, and may include ulceration or vascularization. The pathophysiology of the SMJ nodule may involve the hematogenous, venous, lymphatic, or the remnant pathways, however, it is still unclear how intra-abdominal malignancy spreads through the periumbilical region [3,5]. To reach a definite conclusion, history of the patient, thorough physical examination, and proper imaging modalities are necessary. Once an SMJ nodule is diagnosed, the patient must be thoroughly examined to search for any underlying gastrointestinal or a genitourinary malignancy, since a SMJ nodule often indicates an advanced stage of malignancy. Therefore, any suspicious lesions should be biopsied and promptly referred to the associated specialty since, as presented in this case report, advanced stage of malignancy may be present in the patient.

Scientific Responsibility Statement
The authors declare that they are responsible for the article’s scientific content including study design, data collection, analysis and interpretation, writing, some of the main line, or all of the preparation and scientific review of the contents and approval of the final version of the article.
Animal and human rights statement
All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. No animal or human studies were carried out by the authors for this article.

Conflict of interest
None of the authors received any type of financial support that could be considered potential conflict of interest regarding the manuscript or its submission.

References

How to cite this article: