
Sister Mary Joseph's Nodule: An Indicator Of Intraabdominal Malignancy

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Abstract

Umbilical metastasis is one of many characteristic signs of extensive neoplastic disease. It suggests advanced distant metastasis and is associated with poor prognosis; mean survival is approximately 10–12 months, although long-term survival has been reported, but only in the presence of a solitary metastatic umbilicus nodule. Sister Mary Joseph's nodule has traditionally been considered a sign of ominous prognosis (survival of 10 months on average) and suggests therapeutic abstinence. We present such a case with a detailed review of the literature.

INTRODUCTION

Cutaneous metastases from carcinoma are relatively uncommon in clinical practice, but they are very important to recognize. They may herald the diagnosis of an internal malignancy. Early recognition can lead to accurate and prompt diagnosis and timely treatment, but a high index of suspicion is required because the clinical findings may be subtle. The recognition of cutaneous metastases often dramatically alters therapeutic plans, especially when metastases signify persistence of cancer originally thought to be cured. Some tumors metastasize with predilection to specific areas. Recognition of these patterns can be useful in directing the search for an underlying tumor (1).

Most cutaneous metastases occur in a body region near the primary tumor. The most common presentation of cutaneous metastases is nodules. The nodules are often non painful, round or oval, firm, mobile, and rubbery in texture. The nodules are usually flesh colored, although they may also be other colors (e.g., from flesh colored to brown or blue-black) (2). They vary in size from barely perceptible lesions to large tumors. In this article, we have summarized the topic "Sister Mary Joseph's Nodule as an indicator of intra-abdominal malignancy".

CASE REPORT

A 53-year-old male presented with history of an umbilical swelling of four months duration. It was insidious in onset and was not associated with pain but was progressively increasing in size and associated with ulceration with serous

discharge. He gave no history of loss of weight or appetite. There was no other positive contributory history.

General and systemic examination was normal. Abdominal examination revealed a single, hard, ulcerated, non tender swelling, measuring 3x2 cms at the umbilicus with partial destruction of the umbilicus. A seropurulent foul smelling discharge was also seen. There were no masses, free fluid or intra-abdominal lymphadenopathy. The supraclavicular lymph nodes were not enlarged. A per rectal examination was normal. Hence, a tentative clinical diagnosis of umbilical metastasis/umbilical granuloma was made.

Routine investigations (hemogram, CXR, LFT, stool) were normal. A FNAC of the umbilical swelling was performed and was reported to contain adenocarcinoma cells. Hence, diagnosis of metastatic adenocarcinoma with suspicion of a primary tumor in the GIT was considered. An UGI scopy done was normal and so a colonoscopy was performed. It showed an irregular mucosa in the ascending colon, which was biopsied. The final histopathology report was consistent with adenocarcinoma. The patient was also subjected to an abdominal CT scan which revealed an enhancing soft tissue density lesion in the right lateral wall of caecum, suggestive of malignancy.

As the patient was asymptomatic and had an umbilical deposit he was advised to receive chemotherapy and was given four doses of chemotherapy (Adriamycin and Mitomycin C). He tolerated this well, has been under constant follow-up and is doing fine at the moment with a

good quality of life.

DISCUSSION

Sister Mary Joseph (head nurse and eventually skilled surgical assistant to William Mayo, MD), was the first to note that the presence of a periumbilical nodule heralded an advanced intra-abdominal malignancy (3). The cutaneous finding is a firm, non tender nodule of red or purple hue that represents a metastasis from the primary tumor. The nodule most often results from contiguous extension of the tumor from the peritoneum, although lymphatic and venous pathways likely contribute in some cases (4).

As recognized by Sister Mary Joseph, the presence of the umbilical nodule is associated with advanced intra-abdominal malignancies. Approximately 90% of these malignancies are adenocarcinomas; with gastric and ovarian being the most commonly discovered primary malignancies (5). Other cited primary sites include the pancreas and bowel. Because the nodule is so easy to biopsy and because most associated cancers are inoperable at the time of diagnosis, the ability to identify this lesion may save a patient an unnecessary diagnostic surgery (6).

The spread of metastatic cancer to the umbilical region has been hypothesized to occur in several ways. Contiguous spread of peritoneal cancer is thought to be the most simplistic and important method (7, 8). One report described an ovarian carcinoma that presented with a Sister Joseph's nodule; by histologic sectioning, the authors were able to prove that the spread of carcinoma occurred by direct extension, a finding that had not been shown before in the literature. Hematogenous spread through the arterial and venous systems is another postulated method by which metastasis occurs, with the persistence of the paraumbilical veins being a possible conduit for metastatic spread.

Lymphatic spread to the umbilical region is also a probable method of metastasis, considering that 4 sets of lymphatics pass from the umbilical region. In particular, metastases from pancreatic cancer are believed to cause Sister Joseph's nodule

in this manner, because pancreatic cancer rapidly invades the lymphatic system. Lastly, direct extension along the ligaments of embryonic origin is a presumed mode of metastatic spread, including the round ligament of the liver, the urachus, the vitellointestinal duct remnant, and the obliterated vitelline artery. This method may be important in metastatic tumors of the small intestine (6).

The diagnosis of metastatic carcinoma hinges on histopathologic evaluation of involved skin. Tumors may show characteristics of the underlying tumor, or they may have a more anaplastic appearance. In the situation of an anaplastic tumor, immunohistochemical marker studies and ultrastructural examination may help delineate the tissue of origin (8).

Effective treatment depends on treatment of the underlying tumor. Palliative care is given if lesions are asymptomatic and the primary cancer is untreatable. In many cases, cutaneous metastasis can cause disfigurement or social embarrassment, or it can diminish the quality of the patient's life. Excision and removal of metastasis may be warranted to enhance the patient's quality of life, but they do little to increase survival. Simple excision is usually the treatment of choice (8). Patients with Sister Joseph's nodule who were treated aggressively with both surgery and adjunctive therapy lived an average of 17.6 months, which was more than with surgery alone (7.4 months), adjunctive therapy alone (10.3 months), or no treatment (2.3 months). In any event, the overall prognosis for patients presenting with Sister Joseph's nodule is generally poor, and the treatment plan for each patient must be individualized (7,8,9).

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