

Umbilical metastasis of unknown primary presenting as umbilical hernia: a case report

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Abstract: Umbilical metastasis is well known to be a late stage of malignancy and is associated with a poor prognosis. The majority of such cases are associated with primary gastrointestinal and gynecologic malignancies, and fewer cases are noted to derive from the thoracic cavity and the urinary tract. In this report, we describe a rare case of metastasis that presented as incarcerated umbilical hernia with no primary sites found. Extensive work-up, including tumor markers, imaging studies, endoscopies, and immunohistochemical analysis, failed to identify the primary source of this malignancy; this was a rare case of Sister Mary Joseph's nodule with unknown primary. Therefore, it is very important for a surgeon to consider metastasis among the differential diagnosis of umbilical hernia.

Keywords: umbilical metastasis, Sister Mary Joseph's Nodule, hernia

Case report

A 77-year-old female with history of hypothyroidism, hypertension, and hypercholesterolemia, was seen initially in the office with complaints of umbilical swelling associated with mild discomfort for 1 month, but without other constitutive gastrointestinal symptoms such as pain, nausea, vomiting, early satiety, diarrhea, constipation decreased appetite, or weight loss. Diagnosed with incarcerated umbilical hernia, she presented to ambulatory surgery for elective umbilical hernia repair. The patient had no family history of malignancy. Physical exam was significant for a soft, 4 cm nontender umbilical bulge with irreducible contents. No other abdominal masses were found and the liver, spleen, thyroid, cervical, and supraclavicular lymph nodes were nonpalpable. Rectal exam did not reveal any palpable masses or firmness. Gynecology consultation was performed and revealed no ovarian mass. Previous supracervical hysterectomy was for uterine fibroids. Cervix was unremarkable. Basic laboratory work-up including liver function tests did not show any abnormalities.

During the operation for the proposed umbilical hernia repair, a 3 cm soft tissue mass attached to the omentum and herniating through an umbilical defect was found and completely resected. A few lymph nodes measuring about 1 to 2 cm were found attached to the mass. A frozen section of this specimen was reported as adenocarcinoma. The patient developed atrial fibrillation while in the operating room and hence the procedure was terminated with repair of the umbilical defect without further exploration. Postoperatively the patient was admitted for treatment of the atrial fibrillation and work-up for malignancy.

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Image studies

Computed tomography (CT) of the abdomen and pelvis revealed 2 small soft tissue nodules (3.5 cm and 2.2 cm, respectively) in the left iliac region, a 4.3×3.6 cm right hepatic cyst, a 2.4×1.7 cm left hepatic cyst, and multiple bilateral renal cysts. There was no abdominal lymph node enlargement and no gastric or intestinal masses. Bilateral adrenal nodules measuring up to 2.7 cm were identified and due to the patient's medical condition, biopsy could not be performed. Endoscopy revealed hiatal hernia and mild chronic gastritis. Colonoscopy was performed and revealed unremarkable but moderate to severe diverticulosis and benign polyp. A whole body positron emission tomography/CT scan revealed a 4.4×3.6 cm malignant mass within the umbilical region extending into the peritoneal space, and a second malignant soft tissue opacity measuring 2.9×2.2 cm adjacent to the right lobe of the liver. However, due to the patient's medical condition, biopsy could not be performed. A third hypermetabolic malignant-appearing nodule adjacent to the urinary bladder measured 2.8×2.4 cm. Multiple hypermetabolic left external iliac lymph node compatible with malignancy were seen. Breast exam and mammogram were unremarkable.

Pathology findings

Grossly the specimen measured $4.2 \times 3.5 \times 3$ cm in greatest dimension. Cut section revealed a fleshy dense tan surface. Microscopic examination revealed sheets of hypercellular malignant cells characterized by large pleomorphic nuclei with coarse chromatin or hyperchromasia, irregular nucleoli, and abundant mitotic figures. The tumor formed a vague acinar or trabecular pattern (Figure 1). Immunohistochemical study revealed that tumor cells were positive for CK7 and negative for CK20, TTF-1, CDX2, CA125, and CA19.9 (Figure 2). Serum CEA level was normal. The final pathology was reported as metastatic adenocarcinoma without a specific primary site.

Discussion

Since its initial use by Hamilton Bailey in 1949, many articles and reviews have been published about the eponym Sister Mary Joseph's Nodule, a rare clinical finding.¹⁻³ Possible routes of metastasis to the umbilicus may include arterial, venous, and lymphatic channels; however, the direct extension of tumor through the peritoneum appears to be the most favorable pathway.^{2,4-6} Clinical history, histological studies combined with immunohistochemical stains, and tumor markers analysis usually help in identifying the primary site.^{2,7,8}

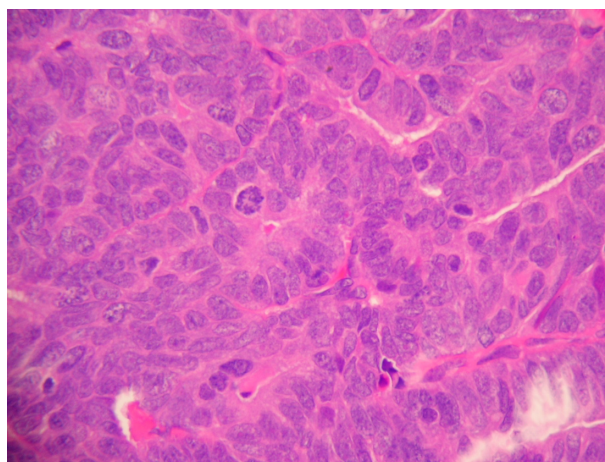


Figure 1 Microscopic examination (H&E stain, 40 \times) of the umbilical nodule reveals sheet of highly malignant cells showing hyperchromatic pleomorphic irregular nuclei with glandular formation. Mitotic activity is easily identified.

Most tumors are noted to be adenocarcinomas followed by squamous cell carcinomas and undifferentiated tumors,^{2,5,9} the majority of cases deriving their primaries from gastrointestinal tract (about 50%), gynecologic organs (about 28%), and genitourinary and respiratory tracts.⁹ It was reported that the commonest origins are stomach, ovary, colon, and pancreas.⁸ About 15% to 30% of cases are reported to have occult primaries.^{1,4,9} As in our case, this may also be the only clinical manifestation of malignancy, or the recurrence or relapse of a known malignancy,^{2,4,5,7,10} and may present as a frank umbilical hernia.¹¹

Based on the origin of most primaries, consideration for extensive work-up including CT scan of the abdomen¹² and chest, and upper and lower gastrointestinal endoscopy with biopsy of suspicious lesions, and serum assay for tumor markers should be ordered in the evaluation of any mass lesions of

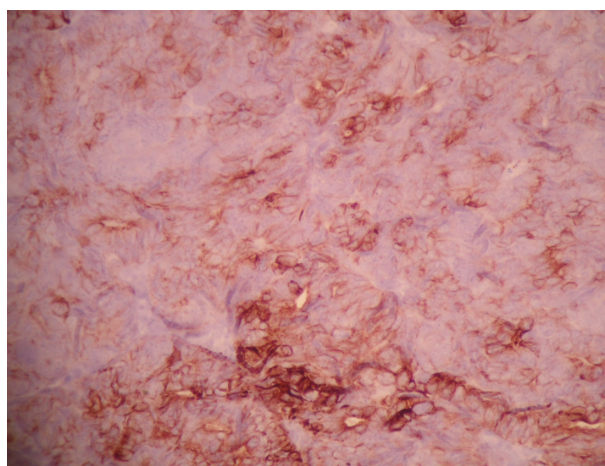


Figure 2 Immunohistochemical stain (20 \times) showing tumor cells strongly positive for CK7.

the umbilicus. Metastatic malignancy should be among the differential diagnosis of umbilical hernia. It is important to remember that fine needle aspiration biopsy, which has a high diagnostic yield,^{4,13} should be performed in the evaluation of any suspicious umbilical mass prior to operative intervention. With appropriate surgery, adjuvant therapy, and timely follow-up, average survival rates will be improved.^{1,4,9}

Conclusion

Umbilical metastasis (Sister Mary Joseph's Nodule) is well known to be a late stage of malignancy and is associated with a poor prognosis. Umbilical metastasis should be among the differential diagnosis of umbilical hernia.

Conflicts of interests

The authors declared no conflicts of interests

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