Dear Editor,

Sister Mary Joseph’s nodule refers to a malignant metastatic umbilical or paraumbilical nodule and is a sign of advanced stage of malignancy. It is usually associated with primary neoplasm of the gastrointestinal and genitourinary tract. Here, we report an unusual case of cutaneous metastases secondary to an unknown primary malignancy, localizing to the umbilicus.

A 45-year-old woman was admitted to the surgery department for abdominal pain of 2 months duration and associated history of weight loss. Patient had undergone cholecystectomy and cesarean section in the past. A referral was sent for lesions over the umbilicus. There was no history of breathlessness, hemoptysis, or chest pain indicative of pulmonary malignancy. No history of nausea, vomiting, abdominal distension, constipation, diarrhea, bloating, or per rectal bleeding (melena or hematochezia) was present suggesting gastrointestinal involvement. Also symptoms of urinary, genital, and ovarian malignancy were absent, except for the vague abdominal pain, reduced appetite, and weight loss which was of recent onset. Cutaneous examination revealed multiple lichenoid and few erythematous papules and nodules with rough surface and hard consistency, in and around the umbilicus and extending to cesarean section scar over the infra-umbilical region [Figure 1]. Considering a diagnosis of warts but to rule out malignancy, a punch biopsy of the papule was done which revealed features of metastatic mucinous adenocarcinoma [Figure 2a and b]. The immunohistochemistry showed positivity for CK7 and CDX2 [Figure 3a and b] and was negative for CK20, TTF-1, and PAX8. Contrast enhanced computed tomography abdomen showed extensive liver and lymph node metastases with ascites, omental caking, and peritoneal deposits with vascular invasion. Ultrasound-guided fine needle aspiration from liver mass suggested metastatic poorly differentiated epithelial malignancy, possibly adenocarcinoma. Level of CA-125 were 2624 IU which was significantly raised. Eventually, a diagnosis of cutaneous metastases secondary to an occult primary, possibly intra-abdominal malignancy was made.

Around 1–3% cases of abdominal and pelvic malignancies metastasize to the umbilicus, out of which gastrointestinal (52%) and gynecologic (28%); especially stomach (23%), and ovarian (16%) carcinomas are the common sites of origin. However, about 15–29% of umbilical metastasis has an unknown origin. Though the mechanism of spread of cancer to the umbilicus is unknown, multiple hypothesis have been proposed such as direct transperitoneal spread via lymphatic running along the obliterated umbilical vein, hematogenous spread, or via remnant structures like median umbilical ligament, falciform ligament, or a remnant of the umbilical duct.

Differential diagnoses considered for an umbilical nodule include primary umbilical neoplasm, Sister Mary Joseph’s nodule, umbilical endometriosis, pyoderma gangrenosum, keloid, omphalith, etc. Primary umbilical neoplasms are usually painless long standing nodules. Umbilical endometriosis presents as cyclical pain over the umbilicus during menstruation. The umbilical nodule can present as a hard and irregular nodule or a soft nodule which may ulcerate.

In our case, metastasis to the umbilicus were lichenoid in appearance with transepidermal elimination of tumor cells
on histopathology. As far as ascertained on an extensive search, no such case has been described in English literature till date and this makes our case unique.[4,5]

In light of the immunohistochemistry profile (CK7+, CDX2+ and CK20-), high CA-125 levels and presence of peritoneal carcinomatosis, primary origin of metastasis to umbilicus in our case could possibly be the ovary.[6] Although positivity to CDX2 is a marker of intestinal malignancy, high expression of CDX2 has been reported in mucinous carcinoma of ovarian origin. There was transepidermal elimination of tumor cells. But it was not visible clinically. Possibly, there were microerosions.

Our case epitomizes the value of a careful physical examination and thorough radiological and pathological investigations keeping a high degree of suspicion for diagnosing an internal malignancy.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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