Case Report

Umbilical cutaneous nodule: A diagnostic dilemma

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ABSTRACT

Umbilical cutaneous nodule can be due to infection, skin ailments and tumors. It can be primary or secondary in origin. When it is secondary due to metastasis from visceral malignancy then it is named as Sister Mary Joseph nodule. It can be a presenting symptom and sign of hidden malignancy and thus poses a diagnostic dilemma for physician. Our case presented to us with ulcerated umbilical nodule and subsequent ultrasound revealed accompanied urinary bladder malignancy. Mostly, these nodules are adenocarcinoma but our was an unusual case of transitional cell carcinoma.

Key words: Sister Mary Joseph nodule, transitional cell carcinoma, urinary bladder, visceral malignancy

INTRODUCTION

Periumbilical cutaneous metastasis from hidden visceral malignancies has been named as "Sister Mary Joseph's (SMJ) nodule." It was named in respect of SMJ in 1928. SMJ was first observer to establish a correlation between umbilical nodules and carcinoma. She was first assistant of Dr. William J. Mayo a surgeon at St. Mary's hospital at Rochester, Minnesota (presently the Mayo Clinic).[1] Primary malignancy in the gastrointestinal tract and ovary can spread to umbilicus. Rarely, malignancy of endometrium, prostate, lung, urinary bladder and from unknown primaries can also spread to umbilicus.[2,3] It is more common in females. In a report of 34 cases, male outnumbered females by the ratio of 1.4:1.[4] In one study, in 29% of umbilical metastasis, the origin of the primary lesion was unknown,[5] thus umbilical nodule poses a diagnostic dilemma for clinician.

CASE REPORT

A 65-year-old woman presented with an ulcerated umbilical nodule with yellowish discharge for the last 2 months. On examination, there was a 2 cm × 1 cm hard ulcerated, brownish nodule occupying lower half of the umbilicus [Figure 1]. Urine examination showed field full of RBC/high power field. Blood investigations revealed hemoglobin level as 7 g, blood urea 45 mg/dl and serum creatinine 2.2 mg/dl. Fine-needle aspiration (FNA) cytology of the umbilical nodule revealed transitional cell carcinoma [Figure 2].

Subsequent ultrasound of the abdomen showed growth occupying whole of the urinary bladder with thickening at dome [Figure 3]. There were metastases in the liver. Due to poverty, patient refused for computerized tomography. Cystoscopy and biopsy of bladder growth was carried out under local anesthesia. Whole bladder was affected with growth including dome. Histopathology of bladder growth revealed transitional cell carcinoma. Adjuvant chemotherapy was planned due to disseminated disease. A combination of cisplatin, methotrexate and vinblastine was planned. Patient refused treatment due to the high cost of drugs and was lost to follow-up.

DISCUSSION

The umbilical nodule can occur due to various diseases. About 38% umbilical nodules are primary tumors of the umbilicus; 32% are due to endometriosis and 30% are due to metastatic disease from a distant location.[6]

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Sometimes, the SMJ nodule is the sole indication of an occult malignancy. An SMJ nodule was discovered prior to the diagnosis of the primary tumor in 152 of 368 cases in one study.[7] In another study, 45 of 85 patients developed an SMJ nodule 1-12 months prior the diagnosis of the primary malignancy.[8] Sometime, umbilical metastases are the first sign of a systemic malignancy as in our case. Rarely discharge from umbilical nodule can be the first sign of recurrence in carcinoma ovary.[9] For these reasons, the presence of a peri-umbilical nodule necessitates assessment for an occult intra-abdominal malignancy.

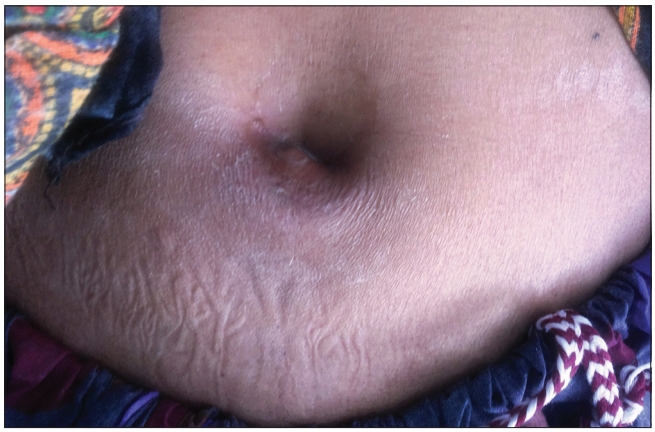

Figure 1: Umbilical ulcerated nodule in patient

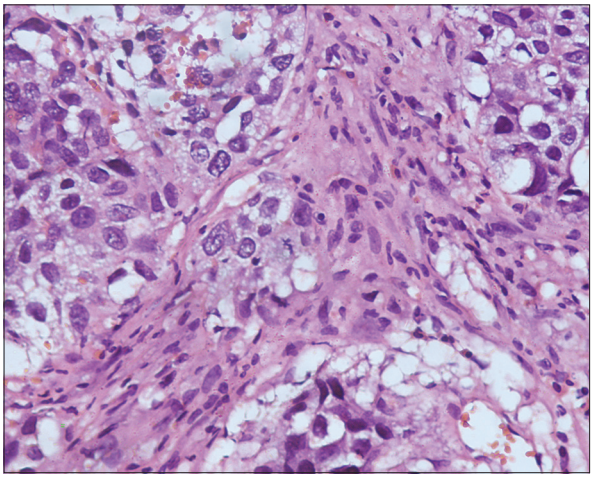

Figure 2: Fine-needle aspiration cytology of umbilical nodule showing transitional carcinoma of urinary bladder

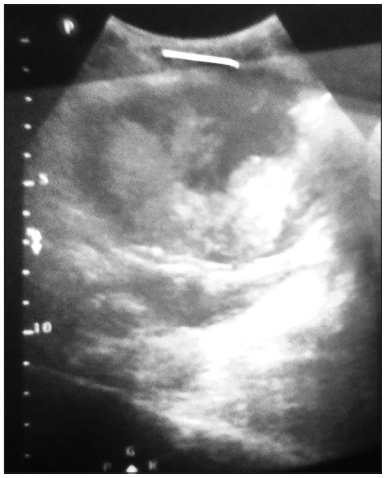

Figure 3: Ultrasound showing urinary bladder growth

The incidence of SMJ nodule is only 1-3% of all intra-abdominal or pelvic malignancies. Studies have shown that most of the umbilical lesions are adenocarcinoma, rarely a squamous carcinoma, melanoma or sarcoma,[10] but uncommonly it can be transitional cell carcinoma as in our case.[11]

Among all reported cases of SMJ nodule, 35-65% metastasize from gastrointestinal malignancies; 12-35%, from the genitourinary tract; 15-30%, from unknown sites; and 3-6%, from the lung and breast.[12] Barrow. in his study had reported that cancer of the stomach accounted for 25%, ovary 12.4%, colon and rectum 10.0%, pancreas 7.4%, gallbladder 3.4%, uterus, 3.0%, liver 2.4%, endometrium 1.4%, sarcoma 1.5% and small bowel 1.0%.[13] Rarely, it can be a metastatic nodule from malignant mesothelioma.[14]

The mechanism of metastasis of the primary tumor to umbilicus is not clear; however, several hypotheses have been proposed. First hypothesis is that spread is via peritoneal infiltration (the most common route) or through arteries, veins, or lymphatic channels. Another is that spread occurs through embryonic structures such as the urachus, round ligament of liver, vitello intestinal duct remnant, or obliterated vitelline artery. There is a connection between liver and umbilicus via venous and lymphatic channels and these channels seems to be means of mutual seedlings but which organ is affected first is not clear.[12]

Generally, SMJ nodule presents as a firm irregular nodule, size range from 1 cm to 1.5 cm, and may reach up to 10 cm in diameter.[12] It can be a fissured or ulcerated nodule, and depending from origin, it can secrete serous, mucinous, purulent, or bloody discharge. The lesion may be of different colors such as white, bluish violet, brownish red with or without itching.[15]

Tumors such as Paget's disease, angioma, and other ailments such as umbilical hernia, endometriosis, hypertrophic scar, keloid, mycosis, psoriasis, pyogenic granuloma, melanocytic nevi, epithelial inclusion cysts, epidermoid cysts, omphaliths, eczema, squamous cell carcinoma and basal cell carcinoma are differential diagnoses of an umbilical nodule.[16]

When a patient presents with an umbilical nodule, to establish an easy and early diagnosis, the following

diagnostic tests like: Culture and special microbiology staining of ulcer specimen, FNA of the umbilicus for cytology, umbilical biopsy for histopathological examination, immunohistochemistry, imaging modalities (ultrasound, computed tomography, magnetic resonance imaging scan of the abdomen) and tumor markers such as carcinoembryonic antigen, prostate-specific antigen, and cancer antigen are essential.[10,16]

The presence of SMJ nodule means disseminated disease and it means that it is not amenable to cure. Without treatment, life expectancy is about 2-11 months. However, some issues seem to control the prognosis of such patients like direct invasion by the peritoneum is associated with a worse prognosis. The prognosis is good when the metastasis is discovered before the primary tumor.[10]

Recent reports have suggested that prognosis is better with aggressive management, the mean survival being 17.6-21 months with surgery and adjuvant chemotherapy instead of surgery alone (7.4 months) or chemotherapy alone (10.3 months). However, surgery is usually recommended only in patients with a solitary umbilical metastasis. Surgery should be avoided in cases with widespread disseminated disease. In case of widespread disease, effective palliation can be attained with adjuvant chemoradiotherapy. Prognosis depends on the diseased organ. It has been noted that patients with ovarian cancer have a better survival rate than do patients with other malignancies.[10,12]

CONCLUSION

Sister Mary Joseph nodule is an uncommon manifestation of visceral and other malignancies. It can be a presenting symptom or sign of undiagnosed underlying malignancy, or an alarming symptom or sign of widespread disease. Although similar cases have been reported in the literature, this manifestation of the transitional carcinoma of urinary bladder is rare and clinician must be conscious of this rare clinical entity so that they can diagnose the primary cancer in early stage and thus can stop its progression and avoid its recurrence.

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