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Case Report

Surgery Section

Umbilical Metastasis as Primary Manifestation of Cancer: A Small Series and Review of the Literature

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ABSTRACT

Umbilical metastasis is a rare manifestation of intra-abdominal cancer. It appears either as the first sign of a primary malignancy or as metastatic site of an already diagnosed cancer, representing an ominous prognostic finding. We report three cases of umbilical metastasis as the first sign of an underlying malignancy. Hypotheses about pathophysiology of umbilical metastasis are based on the embryological origin of the umbilicus and its residual communication with systematic, portal and lymphatic circulation.

Keywords: Ovarian cancer, Rectal cancer, Sister mary Joseph's nodule, Umbilical metastasis, Urinary bladder cancer

CASE REPORTS

Case 1

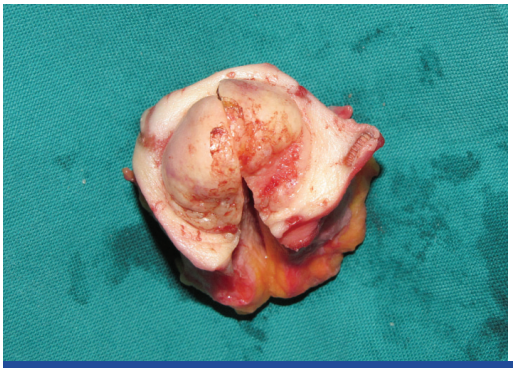
An 85-year-old was admitted with an umbilical mass, which initially caused a slight discomfort for a month prior to presentation but became painful during the last two days. The mass was tender and irreducible upon physical examination, with the overlying skin ulcerated and ischemic. Lacking other symptoms or signs, the diagnosis of incarcerated, strangulated umbilical hernia was presumed. A dense fibrous mass was found instead [Table/Fig-1], which was excised. Histology revealed a low-grade metastatic carcinoma infiltrating the dermis and the subcutaneous fat [Table/Fig-2]. Further diagnostic evaluation included upper and lower gastrointestinal tract endoscopies which were normal and abdominal magnetic resonance imaging (MRI) which revealed a large tumour (6×7cm) in the left lateral wall of the urinary bladder along with an lymph node mass along the course of the left common iliac vessels causing ureteral obstruction and dilatation of the left ureter and hydronephrosis [Table/Fig-3]. Thoracic computed tomography (CT) revealed a 3cm metastatic node, in the lower lobe of the left lung. The patient underwent cystoscopy and the biopsy showed invasive carcinoma with characteristics of both squamous and transitional epithelial origin. The patient was transferred to the urology department for management. The tumour was considered inoperable, and due to the old age of the patient and significant co-morbidities no palliative medical treatment was administered. The patient died six months later.

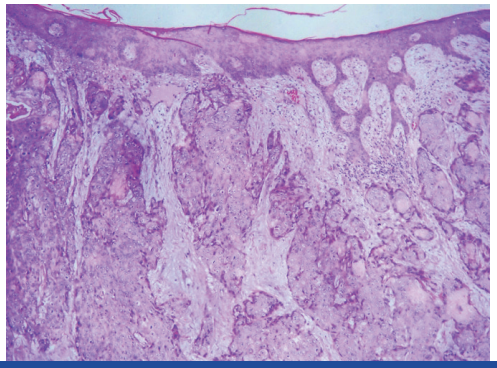
Case 2

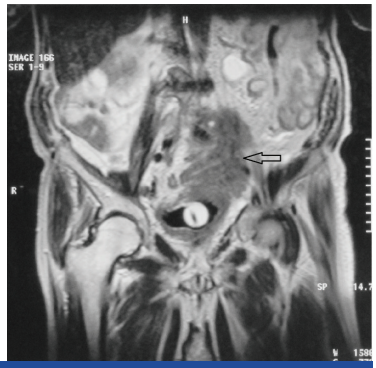
A 66-year-old woman was referred to our department because of a painless umbilical nodule with serous discharge and mild, vague abdominal pain in the lower abdomen. Initially, a sinogram was performed, which revealed a short blind fistula. At operation the fistula was enclosed into a thick, dense fibrous mass, which was firmly adherent to surrounding tissues. The histological examination revealed a medium-grade adenocarcinoma with papillary architecture and psammoma bodies, estrogen receptor positive, highly suggestive for papillary serous ovarian adenocarcinoma [Table/Fig-4]. Consequently, the patient underwent abdominal CT, which confirmed the results of the pathological examination, showing a large multilocular tumour sized 13×11×12cm in the anatomic region of the left ovary with cystic and papilliform characteristics. CT also revealed intra-peritoneal dissemination in the greater omentum and ascites was present throughout the peritoneal cavity. Laboratory tests revealed an increased level of CA-125 (1205 U/ml) and CA 15-3 (115 U/ml). The patient was referred to the oncology department for further management.

Case 3

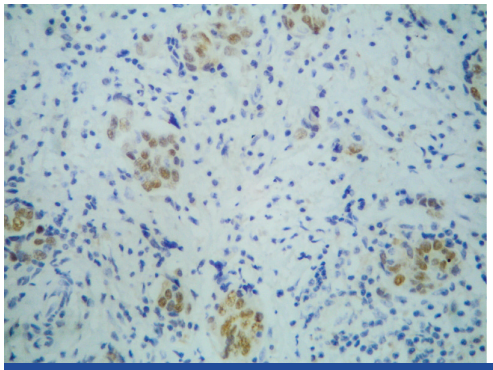
A 78-year-old woman presented with a three month history of progressively increasing fatigue. Physical examination revealed a painless umbilical mass and an annular palpable mass in the middle third of the rectum. No other symptoms or signs were noted. Laboratory tests showed anaemia (Hct: 25%, Hb: 8.2gr/dl), upper gastrointestinal endoscopy was normal and colonoscopy

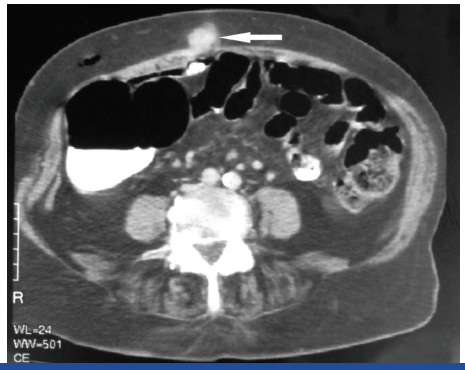


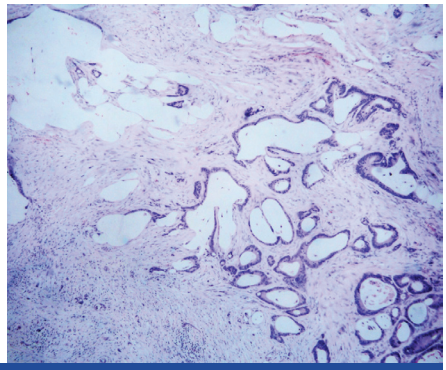




[Table/Fig-1]: Resected and divided sister mary jogeph's nodule (metastasis from urinary bladder carcinoma) **[Table/Fig-2]:** Hematoxylin-Eosin stain ×40 magnification, showing intact epidermis and underneath diffuse infiltration by low-differentiation carcinoma **[Table/Fig-3]:** MRI showing the urinary bladder carcinoma accompanied with an extensive lymphadenous mass along the route of the left common iliac vessels (marker indicating the mass)







[Table/Fig-4]: Immunohistochemical stain (estrogen receptor) ×200 magnification showing nuclear positivity **[Table/Fig-5]:** Sister Mary Joseph's nodule detected in the preoperative CT (marker indicating the nodule)., **[Table/Fig-6]:** Hematoxylin Eosin stain ×40 magnification: metastatic mucinous adenocarcinoma

revealed a mass with cauliflower appearance at a distance of 6-7 cm from the dentate line. Histology confirmed the diagnosis of rectal adenocarcinoma. Abdominal CT showed thickening of the rectum and the distal sigmoid colon, infiltration of the perirectal fat and the uterus, enlarged mesorectal lymph nodes, free fluid in the pelvis as well as an umbilical mass [Table/Fig-5]. Intraoperatively, no macroscopic peritoneal infiltrations were observed and the palpable umbilical mass, viewed from inside, was completely covered by peritoneum. End colostomy and excision of the umbilical mass were performed. Histology of the umbilical mass revealed metastatic colonic adenocarcinoma [Table/Fig-6].

DISCUSSION

The term "Sister Mary Joseph's nodule" was first introduced by Hamilton Bailey in 1949 in recognition of the superintendent nurse of St. Mary Hospital in Rochester, Minnesota, who predicted the presence of intra-abdominal malignancy by palpating a firm nodule in the umbilical region [1].

Umbilical metastasis is a rare finding with the vast majority of cases involving intra-abdominal cancer. Endometriosis, hypertrophic scar, pyogenic granuloma, mycosis, psoriasis inversa, eczema, Paget's disease and angioma are other less common aetiologies of an umbilical nodule [2]. The metastatic nodule size ranges usually from 0.5 to 2cm, reaching in some cases the size of 10cm. Tumours of the gastrointestinal tract and female internal genitalia are the more frequently reported primary lesions, whereas other malignancies are rarely associated with such kind of metastasis [3-5].

The incidence of cutaneous metastasis from internal primary tumours has been estimated about 1-9% at autopsy. Umbilical metastasis is uncommon representing only 10% of these lesions [6]. Sister Mary Joseph's nodule may appear from one month to 10 years after the diagnosis of the primary cancer (average 22 months), while, sometimes, it is the initial presentation of the disease. Rarely, the primary site is undetectable at the time of nodule appearance [7]. The majority of patients complain about a painful, indurated, irregular protrusion in the umbilical region with progressive enlargement. Occasionally, the umbilical skin is ulcerated, ischemic or necrotic and various kind of discharge (blood, pus, serous, mucous) may be noticed.

The mechanisms and pathways leading to umbilical metastasis are still obscure, although several, rather convincing hypotheses have been proposed. Hematogenous metastasis could take place through the rich anastomotic plexus (superior and inferior superficial epigastric vessels, branches of internal mammary and iliac vessels respectively) or through the portal circulation, which is anastomosed with small umbilical veins. The lymphatic drainage of umbilical region follows the venous circulation with final "destination" to the axillary, inguinal and para-aortic lymph nodes. Another possible explanation is the presence of advanced intra-abdominal disease with peritoneal infiltrations and their extension to the umbilical region, which produces the clinical manifestation of the Sister Mary Joseph's nodule. Finally, iatrogenic umbilical dissemination is sometimes encountered after diagnostic laparoscopy [4,8].

Umbilical metastasis from urinary bladder is extremely rare. Only a few cases have been reported in the literature [9,10]. The possible mechanism specifically for bladder carcinoma seems to be dissemination of neoplastic cells through the urachus [4]. Interesting parameters in our first case in which urinary bladder carcinoma was involved were the absence of macroscopic hematuria or dysuria in spite of the advanced disease and the fact that the disease progressed with simultaneous spread in both the iliac lymph nodes and the umbilicus.

Sister Mary Joseph's nodule as a metastatic lesion from ovarian cancer is a more common finding and almost always indicative of advanced disease. Papillary serous adenocarcinomas (as in our second case) have an aggressive clinical behavior and a high metastatic potential [11]. Umbilical metastasis from ovarian cancer may appear during the follow-up period in patients already treated with surgery plus adjuvant chemotherapy. Tsai et al., reported two cases of Sister Mary Joseph's nodules noted accidentally three and eight years after initial management, without other signs of recurrence either from imaging studies or from CA-125 levels [8].

Umbilical metastasis from adenocarcinoma of the large intestine is not unusual. However, the fact that our third patient complained only for the umbilical mass without having the typical symptoms of the disease was also interesting and rare.

Management of umbilical metastasis depends on the site, the histological type and the stage of the underlying malignant disease. Debulking surgery including the umbilical nodule (combined with chemotherapy) or palliative treatment are both possible options. In case that the nodule represents the first clinical finding of an assumed malignancy or a possible recurrence, FNA is a safe, rapid and reliable method for the differential diagnosis of the lesion (sensitivity 98.2%, positive predictive value 100%) [12,13]. However, in equivocal cases umbilical ultrasonography is necessary to rule out the possibility of a strangulated umbilical hernia.

Umbilical metastasis is combined with advanced intra-abdominal malignancies. Patient prognosis, although dependent on the type and site of the primary tumour, remains rather poor. If the umbilical nodule is detected before treatment, the mean survival is 9.7 months, but when it appears as a sign of recurrence the mean survival falls to 7.6 months [14,15]. Nevertheless, treatment should be individualized and include surgery, chemotherapy and/or radiation therapy aiming to an improved prognosis. Clinical doctors must be aware that umbilical lesions that do not undoubtedly fit to the diagnosis of hernia could be a manifestation of malignancy should be thoroughly investigated.

REFERENCES

[1] Bailey H. Demonstrations of physical signs in clinical surgery. 11th ed. Baltimore: Williams & Wilkins; 1949:227.
[2] Chaffins ML, Altman DA, Balle MR, et al. Metastatic umbilical carcinoma. *Cutis.* 1993;52: 37–39.

[3] Galvan VG. Sister Mary Joseph's nodule. *Ann Intern Med*. 1998;128:410.
[4] Gabriele R, Conte M, Egidi F, Borghese M. Umbilical metastases: current viewpoint. *World Journal of Surgical Oncology.* 2005;3:13.
[5] Shetty MR. Metastatic Tumours of the Umbilicus: A Review 1830-1989. *Journal of Surgical Oncology.* 1990;45:212-15.
[6] Lookingbill D, Spangler N, Sexton FM. Skin involvement as the presenting sign of internal carcinoma. A retrosp cancer patients. *J Am Acad Dermatol.* 1990;22:19-26.
[7] Dubreuil A, Dompmartin A, Barjot P, Louvet S, Leroy D. Umbilical metastasis or Sister Mary Joseph's nodule. *Int J Dermatol.* 1998;37:7-13.
[8] Tsai HW, Yuan CC, Wang PH. Umbilicus as the only site of metastasis in recurrent ovarian cancer. *J Chin Med Assoc.* 2006;69:233-35.
[9] Larentzakis A, Theodorou D, Fili K, Manataki A, Bizimi V et al. Sister Mary Joseph's nodule: three case reports. *Cases Journal.* 2008;1:182.
[10] Davar S, Hanna D. Sister Mary Joseph's nodule. *J Cutan Med Surg.* 2012;16:201-04.
[11] Kim YW, Srodon M, Bristow R. Stage IV micropapillary serous ovarian carcinoma: a report of three cases. *Int J Gynecol Pathol.* 2005;24:235-38.
[12] Edoute Y, Malberger E, Kuten A. Umbilical metastasis diagnosed by fine needle aspiration. *J Surg Oncol.* 1990;45:56-58.
[13] Handa U, Garg S, Mohan H. Fine-needle aspiration cytology of Sister Mary Joseph's (paraumbilical) nodules. *Diagn Cytopathol.* 2008;36:348-50.
[14] Chen P, Middlebrook MR, Goldman SM, Sandler CM. Sister Mary Joseph's nodule from metastatic renal cell carcinoma. *J Comput Assist Tomogr*. 1998;22:732-34.
[15] Khan A, Cook B. Metastatic carcinoma of the umbilicus. Sister Mary Joseph's nodule. *Cutis.* 1997;60:297-98.

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