Online Journal of Health and Allied Sciences
Peer Reviewed, Open Access, Free Online Journal
Published Quarterly : Mangalore, South India : ISSN0972-5997
Volume 13, Issue 1; Jan-Mar 2014

Case Report:

Umbilical Metastasis as the Manifestation of an Asymptomatic Gallbladder Carcinoma

Authors
Pampanagouda SKM, Post Graduate Student in Surgical Oncology,
Sailaja Suryadevara, Post Graduate Student in Surgical Oncology,
Veerendra Kumar, Professor, Department of Surgical Oncology,
Vijayalakshmi Deshmane, Professor, Department of Surgical Oncology,
Malathi Mukunda, Professor, Department of Pathology,
Jaipal, Professor, Department of Radiodiagnosis,
Kidwai Memorial Institute of Oncology, Bangalore, India.

Address for Correspondence
Dr. Pampanagouda SKM,
212/2-1, Balaji Road,
Thyagarajnagar,
Bangalore -560028,
India.
E-mail: drpampanagouda@yahoo.co.in

Citation
Pampanagouda SKM, Suryadevara S, Kumar V, Deshmane V, Mukunda M, Jaipal. Umbilical Metastasis as the Manifestation of an Asymptomatic Gallbladder Carcinoma. *Online J Health Allied Scs.* 2014;13(1):7. Available at URL: http://www.ojhas.org/issue49/2014-1-7.html
Open Access Archives
http://cogprints.org/view/subjects/OJHAS.html
http://openmed.nic.in/view/subjects/ojhas.html

Submitted: Mar 28, 2014; Accepted: Apr 24, 2014; Published: May 15, 2014

Abstract: Sister Mary Joseph's nodule (SMJN) is an eponym used to describe the metastatic lesion of the umbilicus. Umbilical metastases are rare and its occurrence as a first manifestation of an asymptomatic primary malignancy is very rare. The gallbladder is the source of SMJN in 2%-3% of cases. SMJN can present in any one of the following ways; first manifestation of an occult primary malignancy, an indication of recurrence in a patient with a previous malignancy, or as progression of an underlying symptomatic primary disease. Our study emphasizes the importance of identifying this very useful and easily applicable clinical sign by careful physical examination of the abdomen, and also the need for proper clinical evaluation of any umbilical lesion, and the histological diagnosis. In the present study we report on a case of Sister Mary Joseph's Nodule as the presentation of an asymptomatic gallbladder carcinoma, and review the relevant literature.

Key Words: Sister Mary Joseph's Nodule (SMJN); Umbilicus mass; Gall bladder carcinoma

Introduction:

SMJN has been reported mainly in association with primary abdomino-pelvic malignancies, and it is estimated that 1% to 3% of gastrointestinal and gynecological tumors metastasize to the umbilicus. Umbilical metastasis from Gallbladder Carcinoma is usually found following laparoscopic cholecystectomy for an apparently benign gallbladder disease. Umbilical metastases were first reported by Walshe in 1846.(1) Sister Mary Joseph (1856-1939), was the surgical assistant for Dr William J Mayo, at St Mary's Hospital, Minnesota, between 1890-1915. She was the first to note the presence of a nodule at the umbilical region as being a sign of an advanced intra-abdominal neoplasm with unfavorable prognosis, and brought it to the attention of Dr William Mayo. She made this observation while preparing the patient's abdomen for surgery. Dr. Mayo reported this condition as "pants button umbilicus" in 1928.(2) The eponym was coined in her recognition by the British Surgeon Hamilton Bailey in 1949 in the 11th edition of his textbook entitled "Demonstration of Physical signs in Clinical Surgery".(3)

Case Report:

A 60 years old female presented with a swelling at the site of umbilicus since 1.5 months, and mild dull aching pain at that region since one month. There was no history of any variation in the size of the swelling. She had hysterectomy done 20 years ago and didn't have any other co-morbidities. On examination, a 4x3 cm, non tender nodule present at the site of umbilicus.(Figure 1) There was no palpable intra abdominal mass or organomegaly. USG of abdomen and pelvis revealed Gall bladder mass with calculi and a porta hepatis LN (1.6x1 cm) with hypoechoic lesion (3x2 cm) at the umbilicus. CT scan of the abdomen showed heterogeneous mass in the gall bladder with calculi,(Figure 2) with few necrotic LNs in the porta hepatis and para aortic region, and a soft tissue mass (metastatic) in the umbilical region.(Figure 3) Complete blood count, liver function test and renal function test were within normal limits. Serum CA 19-9 was 29.94 U/ml (Normal- up to 27 U/ml). Esophago gastro duodenoscopy was normal. Fine needle aspiration cytology of umbilical lesion was positive for carcinoma, (Figure 4) and gall bladder mass showed features of adenocarcinoma. Umbilical lesion was positive for CK 7 and negative for CK 20. Immunohistochemistry of umbilical nodule was positive for CA 19-9.(Figure 5) After explaining the prognosis, patient opted for symptomatic treatment due to financial constraints, and was lost for follow up after 4 months.

Figure 1: Umbilical nodule.

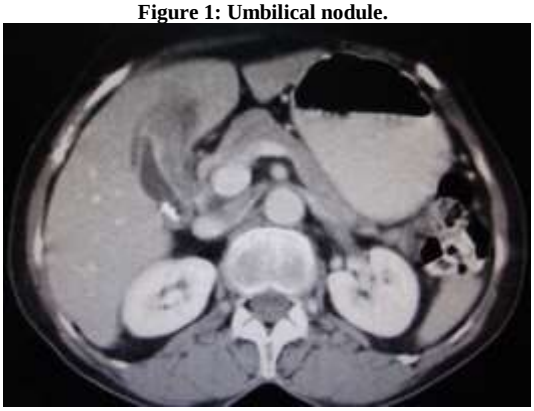

Figure 2: CT Abdomen showing Gall Bladder mass with calculi.

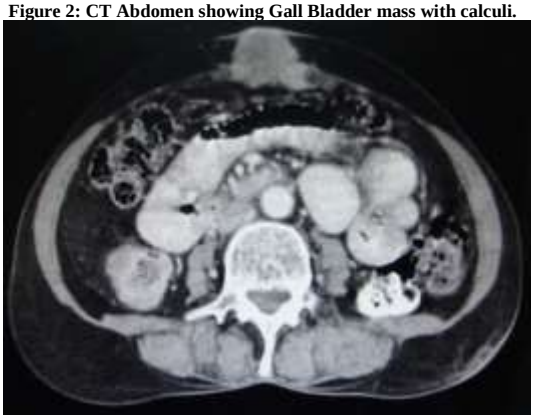

Figure 3: CT abdomen showing Umbilical nodule.

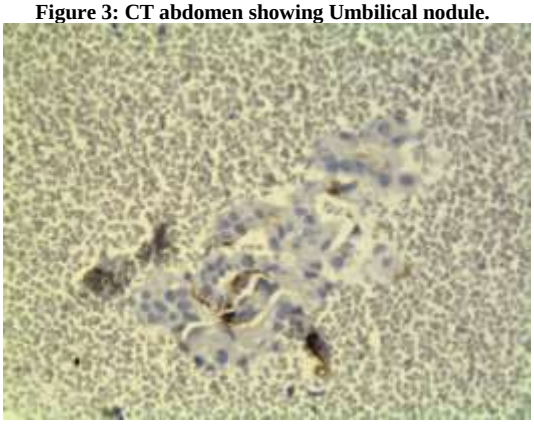

Figure 4: Fine needle aspiration cytology of SMJN stained with Papinicolaou stain.

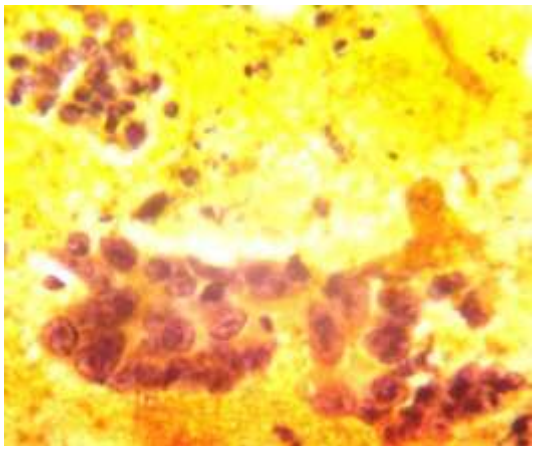

Figure 5: Umbilical nodule immunohistochemical staining positive for CA 19-9.

Discussion:

Umbilicus consists of a puckered scar through the linea alba at the former attachment of the umbilical cord. Umbilical region shows a rich arterial supply that includes the inferior epigastric artery and deep circumflex iliac artery: branches of the external iliac artery, and the superficial epigastric artery: branch of the internal mammary artery. Venous drainage from the umbilicus occurs to axillary vein via the lateral thoracic vein and to femoral vein through superficial epigastric vein. Small para umbilical veins connect with the portal system along the ligamentum teres. There is documented lymphatic communication between the umbilicus and axillary, para aortic, internal mammary, external iliac and inguinal lymph nodes. Tumor spread to the umbilical region has been postulated to occur through several ways. The most common route is contiguous spread from the peritoneal surface, and is considered to be the most important route for tumor spread from visceral malignancies. Other probable methods of tumor spread are through the hematogenous, lymphatic route, or along the remnants of embryological origin which are associated with the umbilical region such as urachus, ligamentum teres, vitello-intestinal duct remnant and the medial umbilical ligaments.

The mean age of diagnosis of cases with SMJN is 50.6 years (range 18-87 years). SMJN could present as a painless or painful, firm to hard indurated lesion with irregular margins, or even necrotic, sometimes fissured or ulcerated with serous, mucinous, purulent, or bloody discharge, and occasionally is pruritic. The nodule has been described as white, bluish violet and brownish red. The nodule may be seen or felt deep in the sub cutis at the umbilical region. Patients may present with other clinical signs or symptoms consistent with the primary disease, and guide in identifying the source. SMJN is an initial presenting sign in up to 14-33% of the cases, where as in 40% of patients with a known neoplasm the nodule is an early sign of relapse.(4) If umbilical disease is the initial presentation of a metastatic tumor, then the chances of identifying the primary is less than 50%, and these patients are more likely to be women.(5)

An umbilical lesion can be benign (57%) or malignant (43%). Benign lesions to be considered are umbilical hernia, pilonidal sinus, pyogenic granuloma, endometriosis, mycosis, urachal duct cyst, omphalitis, fibroma, foreign body granuloma, hypertrophic scar, lymphangioma, melanocytic nevi, papilloma, cutaneous epithelial inclusion cysts, keloid and myxoma. Primary umbilical neoplasms are melanoma, squamous cell carcinoma, basal cell carcinoma, adenocarcinoma and sarcoma. Known primary sites of origin for SMJN are stomach, colorectal, pancreas, lung, esophagus, liver, gallbladder, prostate, kidney, penis, bladder,

peritoneum, ovary, endometrium, breast, cervix, vagina, fallopian tube and lymphoma.

Fine needle aspiration (FNA) is an easy and reliable minimally invasive method for diagnosis, and should be considered as an initial diagnostic test to evaluate an umbilical neoplasm. Once umbilical nodule is discovered, FNA or biopsy is mandatory to establish the diagnosis, and to guide search for the possible site of origin with appropriate investigations. Excision of the umbilical mass to confirm the diagnosis may be required as a guide for further management. Immunohistochemical analysis may define the cellular origin in 72 % of cases of unknown primary tumor. Sometimes the nodule may not be evident clinically and identified only on imaging studies, especially in obese patients.

Cutaneous metastasis occurs in 5%-9% of all cases of malignant disease. Metastasis to the umbilicus is rare and represents only 10% of tumors which metastasize to the skin.(6) Primary umbilical tumors account for 17% of cases, whereas metastatic lesions constitute 83% of all malignant umbilical tumors. Malignant melanoma is the most common primary umbilical malignancy. Among all reported cases of umbilical metastases, 35-65% metastasize from gastrointestinal malignancies; 12-35% from the genitourinary tract; 15-30% from unknown sites; and 3-6% from other malignancies such as those of the lung and breast.(7) In men, the commonest primary site is the digestive tract, of which stomach is the most common site, whereas gynecological malignancies, particularly epithelial ovarian tumors, are the most common primary sites in women. Among gastrointestinal malignancies the common sites in decreasing order of frequency are: stomach (25%), colorectal (10%), and pancreas (7%). Histologically, the most common tumor type is adenocarcinoma (75%). SMJN is more commonly seen in females, and is probably due to a large number of gynecological malignancies that might involve the umbilicus in addition to the gastrointestinal cancers.

The average time between manifestation of skin lesion and diagnosis of cancer is 3 months, and the mean life expectancy is 2-11 months without the treatment, and less than 15% of the patients survive more than 2 years. Some of the studies have shown better survival for patients if they are treated aggressively with a combination of surgery and adjunctive therapy (21 months) instead of surgery alone (7.4 months), chemotherapy alone (10.3 months), or no treatment (2.3 months).(8) However, appropriateness of such therapy is determined by the clinical state of the patient and the treatment should be individualized. It is known that the survival is better in patients who detected such a metastasis before definitive treatment of the primary tumor rather than as a recurrence of it. A better survival rate is possible in patients with primary ovarian carcinoma and in patients with metastases confined only to the umbilicus.

Summary:

Even though, SMJN is a rare clinical sign, the ability to identify this lesion may save a patient an unnecessary diagnostic and therapeutic intervention. Given the variable appearance of the lesion, high index of suspicion is necessary to make an accurate diagnosis. In this era of advanced technology, SMJN still remains as an interesting and useful diagnostic clinical sign.

References:

1. Walshe WH. The nature and treatment of cancer. *The British and Foreign medical review.* 1846:293-311.
2. Mayo WJ. Metastases in cancer. *Proceedings of the Staff Meetings of the Mayo Clinic* 1928;3:327.
3. Bailey H. Demonstration of Physical Signs in Clinical Surgery, 11th edition. Baltimore, Williams and Wilkins, 1949, pp 227.
4. Srinivasan R. Metastatic cutaneous and subcutaneous deposits from internal carcinoma. An analysis of cases diagnosed by fine needle aspiration. *Acta Cytol*. 1993;37:894-898.
5. Papala JA, Selim MA. Metastatic vs primary malignant neoplasms affecting the umbilicus : clinicopathologic features of 77 tumors. *Annals of Diagnostic Pathology*. 2011;15:237-242.
6. Lookingbill D, Spangler N, Sexton FM. Skin involvement as the presenting sign of internal carcinoma. A retrospective study of 7316 cancer patients. *J Am Acad Dermatol* 1990;22:19-26.
7. Falchi M, Cecchini G, Derchi LE. Umbilical metastasis as first sign of cecal carcinoma in a cirrhotic patient (Sister Mary Joseph nodule). Report of a case [Italian] *Radiol Med.* 1999;98:94-96.
8. Gabriele R, Conte M, Egidi F, Borghese M. Umbilical metastases: current view point. *World J Surg Oncol* 2005;3:13.