

Sister Mary Joseph's nodule: a rare cytologic presentation

Kafil Akhtar, Sufian Zaheer, Shamshad Ahmad, Rana K Sherwani

Department of Pathology, Jawaharlal Nehru Medical College, Aligarh Muslim University, Aligarh, (U.P) India

Abstract

Umbilical metastasis is one of the main characteristic signs of extensive neoplastic disease and is universally referred to as Sister Mary Joseph's nodule. It indicates neoplasm of inner organs mostly located in the gastrointestinal tract or the pelvis. However, in approximately 15-30% the primary tumor remains occult. In most cases, Sister Mary Joseph's nodule appears as an early and sometimes the only symptom of the malignancy. Here we report a rare fine needle aspiration (FNA) cytologic diagnosis of umbilical metastasis of an ovarian carcinoma.

Introduction

Umbilical metastasis is one of the main characteristic signs of extensive neoplastic disease and is universally referred to as Sister Mary Joseph's nodule. Metastasis to the umbilicus is rare among skin metastases, which range between 5-9% in neoplasms of inner organs.¹ Only 10% of all tumors which have metastasized to the skin are located at the umbilicus,² predominantly in females,² and 1-3% of the abdominopelvic carcinomas metastasize to the umbilicus. Most of these metastases are from adenocarcinomas (75-93%), followed by squamous cell carcinoma and undifferentiated malignant tumors or carcinoids.¹ In men, the commonest primary site is the gastro-intestinal tract, of which stomach is the single most common entity whereas gynaecological malignancies, particularly epithelial ovarian tumours are the most common primary sites in women.^{3,4} Other possible primary tumors are from the colon, pancreas, gall bladder, and breast.^{4,5}

In up to 30% of cases, Sister Mary Joseph's nodule is the first and only sign of a neoplasm.⁶ On the other hand, the metastasis indicates mostly advanced tumor stages with poor prognosis.⁷ The average time between manifestation of skin lesions and diagnosis of cancer is 3 months and the average survival is 10-11 months.⁷ Here we report on an umbilical metastasis of an ovarian carcinoma which was

diagnosed on fine needle aspiration (FNA) cytology.

Case Report

A 50-year-old female presented with circumferential erythema with mild swelling and pruritis around the umbilicus which manifested a month earlier (Figure 1).

The skin lesion was initially treated for fungal disease, but slowly progressed. Fine needle aspiration cytology (FNAC) of the umbilical lesion was performed which demonstrated cutaneous metastasis of well differentiated adenocarcinoma showing malignant columnar cells in acinar pattern (Figure 2).

Subsequently the patient developed fullness of abdomen with an ill-defined mass in the right adnexa. Extensive clinical staging was performed to search for a primary malignancy, including gynaecologic examination, colonoscopy, ultrasonography and computer tomography scan. The latter demonstrated an ill defined right adnexal mass with focal cystic changes, adhered to the uterus and colonic structures. Subsequently the patient underwent hysterectomy with right salpingo-oophorectomy. Histopathology of the right adnexal mass revealed mucinous cystadenocarcinoma of the ovary, with sheets of columnar cells showing nuclear atypia and abundant intracellular mucin (Figure 3). Consequently Cisplatin 50 mg and 5-Fluoro-uracil 100 mg x 6 cycles was started. After 6 months of follow up, our patient is doing well.

Discussion

The first documentation of umbilical metastasis was made by Baluff in 1854, but the best known description was published in 1928 by William James Mayo. Sister Mary Joseph worked for many decades in the Mayo clinic as nursing superintendent and was the first surgical assistant to William James Mayo. She first observed the combination of umbilical metastasis and abdominal neoplasms while preparing patients for surgery and identified this to Dr. Mayo.¹ In 1949, the British surgeon Hamilton Bailey coined in his textbook *Physical Signs in Clinical Surgery* the term *Sister (Mary) Joseph's nodule* for an umbilical metastasis as reference to her finding.⁸

Metastasis may appear virtually anywhere in the body, however, certain sites are more common than others and umbilical metastasis are unusual site.⁷ A variation in vascularity and embryological development makes the umbilicus easy target for metastasis from an intra-abdominal tumor.⁶

Correspondence: Dr. Kafil Akhtar, Department of Pathology, Jawaharlal Nehru Medical College, Aligarh Muslim University, Aligarh, (U.P) India. E-mail: drkafilakhtar@gmail.com

Key words: umbilical metastasis, adenocarcinoma ovary, FNAC.

Received for publication: 10 March 2011.

Accepted for publication: 25 March 2011

This work is licensed under a Creative Commons Attribution 3.0 License (by-nc 3.0).

©Copyright K. Akhtar et al., 2011
Licensee PAGEPress, Italy
Clinics and Practice 2011; 1:e6
doi:10.4081/cp.2011.e6

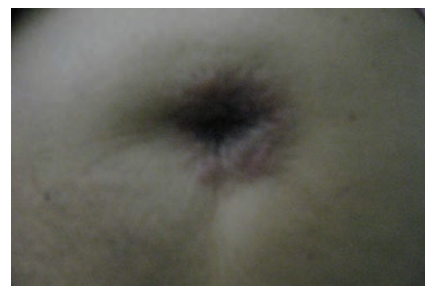


Figure 1. Gross photograph depicting circumferential umbilical erythema.

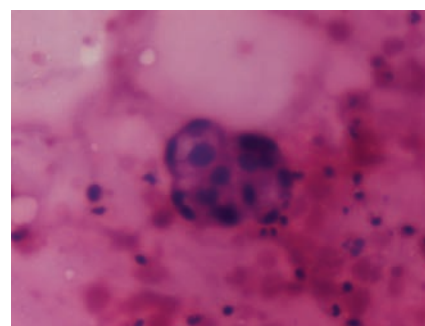


Figure 2. Well-differentiated adenocarcinoma. Fine needle aspiration cytology smear from umbilical site shows malignant columnar cells in acinar pattern. H & E x 400.

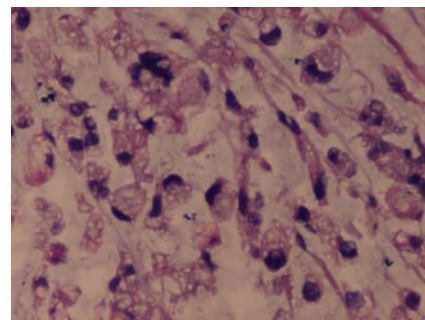


Figure 3. Mucinous cystadenocarcinoma: Tissue section shows sheets of columnar cells showing nuclear atypia and abundant intracellular mucin. H and E x 400.

There are numerous potential routes by which a carcinoma can metastasize to the umbilicus. Malignant carcinomatous cells in the portal venous system may reach the umbilicus by way of a patent umbilical vein.⁹ A pelvic carcinoma may spread via lymph nodes to the umbilical region; as in our present case. In addition, dermal lymphatics are a potential route of spread to the umbilicus.⁷ Furthermore, the spread of malignant cells could have occurred via embolization through the arterial blood supply to the umbilicus.^{7,10}

Umbilical metastasis is one of many characteristic signs of extensive neoplastic disease. It suggests advanced distant metastasis and is associated with poor prognosis.⁷ Histological / cytological evaluations of all umbilical lesions are mandatory, not only to determine its nature but also to guide the clinician to search for the possible primary source, if it is a metastatic one.⁵ Fine needle aspiration cytology of the umbilical nodule is very easy due to its favorable location. Edoute *et al.*¹¹ analyzed the cytological material of 14 patients using fine needle aspiration and only one case was diagnosed as *false negative* since an *inflammatory cell containing aspiration* was obtained. Signet ring cells containing a large amount of mucin, with the nucleus pressed to one side of the cell and appearing crescent-shaped are predominantly seen.¹² It has been shown that the prognosis and average survival is better with primary surgery plus adjuvant chemotherapy (17.6-21 months) compared with surgery only (7.4 months) or with chemotherapy only (10.3 months).¹

Conclusions

Because Sister Mary Joseph's nodule may sometimes be the first and only sign of an internal neoplasm and prognosis is mostly poor, diagnosis has to be confirmed in early stages to improve average survival. We suggest FNAC as an initial diagnostic procedure in cases suspected of umbilical metastasis. It is not only simple, fast, accurate and inexpensive, but can also save the patient from other invasive diagnostic procedures.

References

1. Renner R, Sticherling M. Sister Mary Joseph's nodule as a metastasis of gallbladder carcinoma. *Int J Dermatol* 2007;46:505-7.
2. Touraud JP, Lentz N, Dutron Y, et al. Umbilical cutaneous metastasis (or Sister Mary Joseph's Nodule) disclosing an ovarian adenocarcinoma. *Gynaecol Obstet Fertil* 2000;28:719-21.
3. Dubreuil A, Domp Martin A, Barjot P, et al. Umbilical metastasis or Sister Mary Joseph's nodule. *Int J Dermatol* 1998;37:7-13.
4. Galvan VG. Sister Mary Joseph's Nodule. *Ann Intern Med* 1998;128:410.
5. Ranen KA, Rejaul K, Baronodaya C, et al. Sister Mary Joseph's nodule. *Ind J Med Paediatr Oncol* 2008;29:40-3.

6. Trebing D, Goring HD. Die Nabelmetastase. Sister Mary Joseph's undihere ziet [The umbilical metastasis. Sister Mary Joseph and her time]. *Hautarzt* 2004;55:186-9.
7. Panaro F, Andorno E, Di Domenico S, et al. Sister Joseph's Nodule in a liver transplant recipient: Case report and mini-review of literature. *World J Surg Oncol* 2005;3:4.
8. Bailey H. Demonstrations of physical signs in clinical surgery. 11th ed. Baltimore: Williams and Wilkins; 1949.
9. Raoul JL, Boucher E, Gondier MJ, et al. Metastase ombilicale diun carcinome hepatocellulaire [Umbilical metastasis of an hepatocellular carcinoma]. *Gastroenterol Clin Biol* 1988;22:470-1.
10. Ioannidis O, Cheva A, Stavrakis T, et al. Sister Mary Joseph's nodule as the sole presenting sign of gastric signet ring cell adenocarcinoma. *Gastroenterol Clin Biol* 2010;34:565-8.
11. Edoute Y, Malberger E, Kuten A. Umbilical metastasis diagnosed by fine needle aspiration. *J Surg Oncol* 1990;45:56-8.
12. Zadeh VB, Kadyan R, Al-Abdulrazzaq A, et al. Sister Mary Joseph's nodule: a case of umbilical cutaneous metastasis with signet ring cell histology. *Indian J Dermatol Venereol Leprol* 2009;75:503-5.