Case Report

A rare case of Sister Marry Joseph's nodule in 45 year old male patient

Harsh Pandya[1*], Vrushali Jadhav[2], Jasmin Jasani[3]

[1,2]2nd Year Resident, [3]Professor,
Pathology Department, SBKSMI & RC, Sumandeep Vidyapeeth, Waghodia, Vadodara, Gujarat, India
[*]Corresponding author email: **harsh.hp003@gmail.com**

International Archives of Integrated Medicine, Vol. 6, Issue 12, December, 2019.

Available online at http://iaimjournal.com/
ISSN: 2394-0026 (P) **ISSN: 2394-0034 (O)**
Received on: 21-11-2019 **Accepted on:** 27-11-2019
Source of support: Nil **Conflict of interest:** None declared.
How to cite this article: Harsh Pandya, Vrushali Jadhav, Jasmin Jasani. A rare case of Sister Marry Joseph's nodule in 45 year old male patient. IAIM, 2019; 6(12): 57-60.

Abstract

Metastasis to umbilicus is very uncommon. When occur, the primary site is usually gastrointestinal tract in men and gynecological organs in women. It is known as Sister Marry Joseph's nodule. Its incidence is 1%–3% of all intra-abdominal or pelvic malignancies. The first and best-known description of umbilical metastasis was published by William James Mayo in 1928. In 1949, the condition was named Sister Mary Joseph's nodule by the British surgeon Hamilton Bailey. Here we present the rarest case of the patient who was having past history of gastric adenocarcinoma followed by Sister Marry Joseph nodule after 11 months. As it is a classical case, we are able to find and document the typical features of Sister Marry Joseph nodule.

Key words

Sister Marry Joseph nodule, Umbilicus, Metastasis, Gastric adenocarcinoma.

Introduction

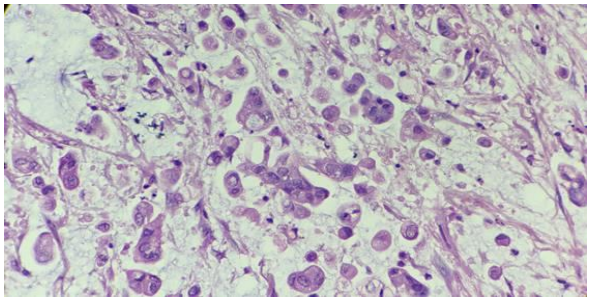
Metastasis to umbilicus is very uncommon [1]. When occur, the primary site is usually gastrointestinal tract in men and gynecological organs in women. Rarely liver, gall bladder, lung, breast, kidney and prostate malignancies may metastasize in the umbilicus [2]. In most cases, umbilical metastasis accompanies the signs and symptoms of the primary malignancy but rarely it is the only manifestation of the underlying disease. Here we present the rarest case of the patient who was having past history of gastric adenocarcinoma followed by Sister Marry Joseph's nodule after 11 months. As it is a classical case, we are able to find and document the typical features of Sister Marry Joseph nodule.

Case report

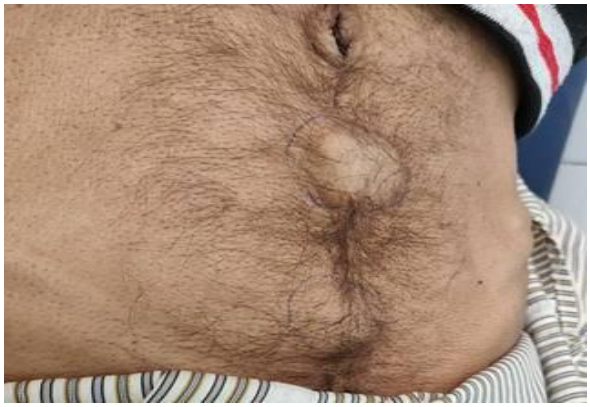
45 year old male came with complain of weight loss and pain in upper abdomen for 20 days to Dhiraj hospital, Vadodara. Patient was non-diabetic, normotensive and vitally stable with no other complain. Upper Gastrointestinal scopy guided biopsy of stomach was done and specimen was sent to Histopathology section (**Photograph – 1**). On histopathological examination multiple sections showed islands of anaplastic cells with cytoplasmic vacuoles. Highly pleomorphic cells with high N:C ratio and hyperchromatism. Final diagnosis was given as moderately differentiated adenocarcinoma (**Photograph – 2**). 11 months later, same patient came to us in cytology department with periumbilical swelling (**Photograph – 3**). FNAC was performed at cytology department by using 22 guage needle [3-11] and smears were prepared. The multiple smears showed high cellularity. There were malignant epithelial cells in clusters, papillary configuration as well as singly scattered, with vacuolated cytoplasm and vesicular nucleus. The cytomorphological findings are that of metastatic adenocarcinoma (**Photograph - 4, 5**). By this we can made the final diagnosis as Sister Marry Joseph's nodule.

Photograph – 1: Gross specimen of total gastrectomy.

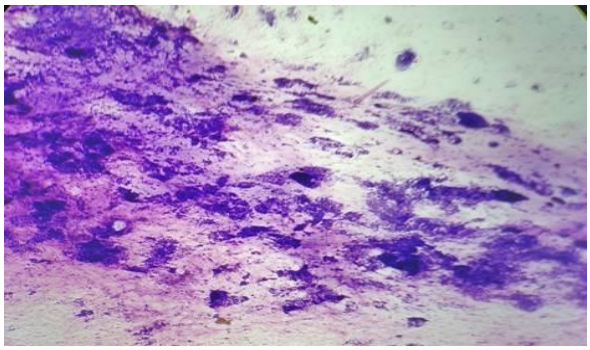
Photograph – 2: Clusters anaplastic cells with cytoplasmic vacuoles (H&E Stain, 40X).



Photograph – 3: Umbilical nodule.



Photograph – 4: Fine needle aspiration cytology showing hypercellular smear (H&E Stain, 10X).

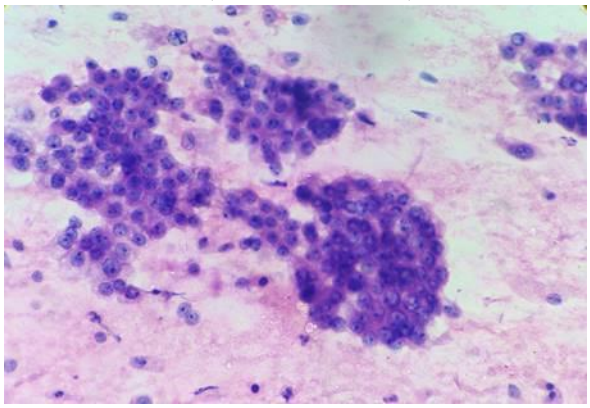


Discussion

Sister Marry Joseph's nodule (SMJN0 is a metastatic umbilical lesion secondary to a primary malignancy of any viscera. It can be a presenting symptom (a sign of undiagnosed malignancy) or a symptom or sign of progression or recurrence in a known case. Its incidence is 1%–3% of all intra-abdominal or pelvic malignancies. The first and best-known description of umbilical metastasis was published by William James Mayo in 1928. In 1949, the condition was named Sister Mary Joseph nodule

by the British surgeon Hamilton Bailey [12]. Typically, Sister Marry Joseph's nodule will be a firm irregular nodule, averaging in size from 1 cm to 1.5 cm, and occasionally reaching a maximum of 10 cm in diameter [13]. Sister Mary Joseph nodule can be a presenting symptom or sign of undiagnosed underlying malignancy, or an alarming symptom or sign of disease progression or recurrence in a known patient [14].

Photograph – 5: FNA smear showed metastatic adenocarcinoma (H&E Stain, 40X).



The most common origin of an umbilical metastasis is an adenocarcinoma from a gastrointestinal or gynecologic primary malignancy. In a review of the world literature on metastatic umbilical tumors, Barrow [13] found that the most common primary tumor identified was carcinoma of the stomach. The second most common was ovarian tumor [13], with colon, rectal, and pancreatic cancers following in descending order of frequency [15]. In very rare cases, the primary tumor involved the breast, cervix, endometrium, small bowel, liver, gallbladder, lung, prostate, kidney, fallopian tube, appendix, and penis. In the present case also primary tumor was Adenocarcinoma of stomach. In approximately 20% to 30% of cases, the primary tumor could not be identified [16].

The mechanism of umbilical seeding from primary tumors is not clearly understood; however, authors worldwide have proposed several hypotheses. One proposal is that a seeding process can occur through one or more routes: contiguous spread of peritoneal infiltration (the most common route) or through arteries, veins, or lymphatic channels. Spread through embryonic structures (such as the urachus, round ligament of liver, vitello intestinal duct remnant, or obliterated vitelline artery) is also proposed as a possible mechanism in certain cases. Some routes are relatively specific to given cancers - for example, pancreatic tumors spread through lymphatic system, and urinary bladder malignancies, through the urachus. It has also been observed that most gastrointestinal malignancies with Sister Marry Joseph's nodule also metastasize to liver. In those cases, it is apparent that the venous and lymphatic channels between liver and umbilicus are the probable means of mutual seeding, but whether the liver spread occurs first and then affects the umbilicus, or vice versa, remains unexplained [17, 18].

Fine needle aspiration cytology of the umbilical nodule is very easy due to its favorable location. Edoute, et al. [19] analyzed the cytological material of 14 patients using fine needle aspiration. Only one case was diagnosed as 'false negative' since 'an inflammatory cell containing aspiration' was obtained. The usual evaluation with Hematoxylin – Eosin may differentiate the primary from metastatic tumors. Moreover, an immuno-histochemical analysis defines the cellular origin in 72% of cases of unknown primary [20]. In the present case also FNAC is the very useful tool for the diagnosis. We have diagnosed this case from the cytomorphological findings.

Sister Mary Joseph's Nodule usually represents an advanced stage of the primary disease and is associated with a grave prognosis. Consequently, palliative management to improve the quality of life is important. In a considerable number of patients, it may be the only presenting sign of a hitherto undiagnosed cancer. Depending upon the site and nature of the primary neoplasm and patients' general condition, combined surgical and medical management may significantly

improve survival (from 2.3 to 17.6 months). Long term survival is uncommon.

Conclusion

Sister Mary Joseph nodule is an uncommon manifestation of visceral and other malignancies. Pathologist need to be aware of this rare clinical condition so that they can promptly diagnose the primary cancer or its progression or recurrence.

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