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SISTER MARY JOSEPH’S NODULE AS A METASTASIS FROM GASTRIC CANCER: A CASE REPORT

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ABSTRACT:

Sister Mary Joseph’s nodule is a well-known umbilical sign that may indicate an underlying abdominal or pelvic malignancy. The patients with this uncommon sign usually have poor outcome. We present a case of a 58-year-old woman with an umbilical firm nodule that originated from a gastric cancer. The patient underwent laparoscopic subtotal gastrectomy successfully, however, her prognosis still remains a question. Careful examination of a suspected umbilical lesion is the key that can identify the presence of the malignancy early and may improve the prognosis for the patient.

Keyword: *Sister Mary Joseph’s nodule – gastric cancer – umbilical metastasis.*

CASE REPORT

Mrs. Dang Thi Hai, a 58-year-old woman, was admitted to the Can Tho University of Medicine and Pharmacy Hospital due to severe epigastric colicky pain on April 23, 2018. She had a history of dull ache of the epigastric region and anorexia for 3 months. The pain worsened after each meal accompanying with nausea and she did not get any treatment. After several weeks, the patient’s condition deteriorated as her pain appeared more often and she vomitted about 5 times a day without eating anything. Then she was taken to a local clinic and was prescribed some kinds of medicine (including colloidal aluminium phosphate). However, after taking the medicines, her symptoms did not reduce. Then she came to our hospital for esophagogastroscopy. A fungating tumor was identified

at her pyloric antrum and she was admitted to the hospital for further investigation.

The patient had a history of hypertension and did not have any hereditary disorder. After 3 months suffering the disease, she had lost 2 kilograms.

At the time she presented to the Department of General Surgery, she was not in acute condition and her vital signs were stable with blood pressure of 110/70 mmHg, pulse of 92 beats per minute and the temperature of 37 degrees centigrade. The pain increased markedly on palpation. A 1 x 1 cm erythematous protruding nodule was identified at her umbilicus. Her laboratory findings revealed a mild anemia with hemoglobin concentration of 10.4 g/dL and the CEA level was 4.9 ng/mL. The CT scan showed the thickening of greater curvature's wall near the pyloric antrum accompanying with a suspected mass with arterial hyperintensity located at the umbilicus.



Figure 2: The suspected umbilical nodule

The patient was diagnosed with distal gastric cancer and the umbilical mass was identified as a Sister Mary Joseph's nodule as the result of the disease's extension. Then she underwent laparoscopic distal subtotal gastrectomy with D1+ lymphadenectomy. The continuity of GI tract was restored by a Billroth II gastrojejunostomy. The umbilical mass was also removed for biopsy. The post-operative pathological results showed a poorly differentiated gastric adenocarcinoma with 12 of 13 nodules retrieved was found locally metastatic. The pathological nature of the umbilical mass had the same characteristics as the gastric tumor. Therefore it could be concluded that the umbilical nodule was the extension of gastric adenocarcinoma or Sister Mary Joseph's nodule. The patient was discharged from the hospital on post-operative day 8 without any complication. She was then appointed for adjuvant chemotherapy.

DISCUSSION

a. Name and history

Sister Mary Joseph's nodule (SMJN) is a firm umbilical mass usually associated with abdominal and pelvic malignancies. The presence of SMJN not only indicates malignancy but may also suggest poor prognosis [1]

The lesion was named after Sister Mary Joseph, the first one who noticed about the existence of it. Sister Mary Joseph Dempsey, who had an Irish origin, was born as Julia Dempsey in Salamanca, New York. In 1878, she entered the third order regular of St. Francis of the congregation of Lady of Lourdes in Rochester, Minnesota taking the name Sister Mary Joseph [1].

After years of teaching at some schools, in 1889, she was assigned to St. Mary's Hospital (now known as the famous Mayo Clinic). That was a hospital that came to activity since 1887 and was run by the Mayo family included William Worrall Mayo (1819-1911), who had planned the hospital and his two sons, Charles Horace Mayo (1865-1939) and William James Mayo (1861-1939). In 1892, she became the nursing superintendent of the hospital until she died in March 29, 1939. Sister Mary Joseph



Figure 3: Sister Mary Joseph Dempsey (1856 – 1939) – from Mayo Clinic

contributed a lot in the developing of the nursing department as she opened St. Mary's Hospital School for Nurses. She also helped organizing the Catholic Hospital Association of the United States and Canada and was chosen its first vice president. During the time working as Dr. William James Mayo's first surgical assistant (from 1890 to 1915), while preparing for gastric cancer patients before surgery, Sister Mary Joseph noted the presence of firm umbilical masses on some individuals. She also found that patients with that kind of nodules usually had poorer outcome and died relatively early post-operatively. She drew William James Mayo's attention to this sign and its prognostic indicator. In 1928, Dr. William James Mayo first mentioned about this kind of nodule in his

lecture at Cincinnati Academy of Medicine, as he called it "pants-button umbilicus", without credit to Sister Mary Joseph [18]. The term "Sister Mary Joseph's nodule" did not exist until 1949, when the English surgeon Hamilton Bailey (1894-1961), in the 11th edition of his textbook "Physical Signs in Clinical Surgery," suggested that the lesion should be called by that name in honor of her discovery [3].

Approximately 43% of all umbilical nodules are malignant in nature [27]. The majority (83%) of malignant umbilical lesions are secondary (metastatic) tumors. The most common histological variant of secondary umbilical tumors is adenocarcinoma although they can be originated from sarcoma, mesothelioma and melanoma [9]. In approximately 50% of cases, the Sister Mary Joseph's nodule is associated with gastrointestinal malignancies include gastric, colonic and pancreatic (mainly body and tail) carcinoma [30]. About 25% of cases originate from gynecological malignancies, primarily ovarian (of which serous papillary cystadenocarcinoma is the most frequent [12] [20]), but also uterine cancer. Occasionally, nodules may relate to malignancies of urinary or respiratory tract, or the appendix (Pseudomyxoma peritonei) [13]. Rarely, a Sister Mary Joseph's nodule may either arise from a benign disease such as abdominal tuberculosis [26]. In some cases, the primary tumor remains unknown [13].

The mechanism of umbilical metastases remains largely unknown but there can be several theories include: direct spread of intra-abdominal malignancies to the anterior aspect of the peritoneum; lymphatic spread as the umbilical region is connected with axillary, inguinal and para-aortic lymph nodes; hematogenous spread through the arterial and venous systems or direct spread via remnant structures such as the falciform ligament, median umbilical ligament and remnants of the vitelline duct.

The nodule is typically described as as a firm, indurate often vascular swelling. It may be fissured or ulcerated and may have serous, mucinous, purulent or bloody discharge. Color can vary from white or bluish violet to brownish red and is occasionally pruritic. It is usually irregular in shape, generally painless when palpated except if the overlying skin has ulcerated. It is usually less than 5 cm in diameter but occasionally enlarges enough to form a protruding tumor [8].

The majority of patients with Sister Mary Joseph's nodule often have symptoms suggestive of internal malignancy like nausea, vomiting, dyspepsia, early satiety,

abdominal pain or distention, anorexia, weight loss, or ascites. However, it is important to note that Sister Mary Joseph's nodule may be the first and only presenting complaint in an otherwise well patient, up to 30% of cases [29]. Therefore, when facing an umbilical nodule, careful clinical assessment is required to distinguish between benign and malignant lesions, and between primary and secondary ones. Acquiring a detailed history, especially focussing on non-specific symptoms such as nausea, anorexia, change in bowel habit, rectal bleeding or weight loss... is also the key. Previous past medical history, smoking and family history should be ascertained, too [25].

This should be followed by a thorough clinical examination noting the presence or absence of cachexia, lymphadenopathy, digital clubbing, and abnormal findings on cardiorespiratory and abdominal examination. A digital rectal examination should be documented. Relevant blood tests and tumor markers should be undertaken [25].

Various imaging modalities can aid in establishing the diagnosis such as Ultrasonography (US), Computed tomography (CT), Magnetic resonance imaging (MRI), and Positron emission tomography (PET). Those facilities can also help evaluating the stage of a potential tumor if existed. However, CT scan is often the first step to characterize the extent of the umbilical mass and to look for other lesions and a primary malignancy [6]. Magnetic Resonance Diffusion-Weighted Image (MR-DWI) is not only help providing assistance in establishing the diagnosis of Sister Mary Joseph's nodule but also and help understanding a part of the metastatic mechanism because of its excellent ability to reveal disseminated lesions [23].

The histopathologic features of the nodule should be acquired, either by fine needle aspiration (FNA) or excision to establish diagnosis or suggest the possible primary site [2]. Even in this era of modern technology, fine needle aspiration is still considered as a simple and time-saving way to establishing the diagnosis of Sister Mary Joseph's nodule [15]. The sensitivity of method in diagnosing a malignant nodule has been reported as 98.2% while the positive predictive value was 100% [10]. Fine needle aspiration and cytological examination is advocated as the first diagnostic procedure, as it is simple, fast, inexpensive and relatively non-invasive, and may avoid the need for more invasive percutaneous biopsy [14].

In the situation of an anaplastic tumor, immunohistochemical marker studies and ultrastructural examination may help delineate the tissue of origin [24].

The presence of Sister Mary Joseph's nodule usually indicates an advanced metastasis and may be unable to radically resection. Therefore, the prognosis is poor and the feasible treatment is commonly palliative. The prognosis is depent of several factors include the origin of primary tumor, the stage of disease and available therapies... Certain data has shown a better survival (mean 9.7 months) in patients who detect an umbilical metastasis before definitive treatment of the primary tumor. In contrast, when the lesion appears after the primary tumor has been treated then the survival for these patients does not exceed the 7.6 months [4] [16]. However, with early diagnosis and surgical intervention, a prolonged survival of 18 years was reported. In a recent study that reviewed charts from 1992 to 2017 that included descriptions of the Sister Mary Joseph's nodule has shown that the overall mortality was 79.6%, with follow up ranging from 0.4 to 231.5 months. Of those followed for at least 2 years and 5 years, two year mortality was 61.6%, and 5 year mortality was 87.4%. Of those that died, median survival was 14.6 months.

Moreover, the etiology of the primary malignancy determines the prognosis. For example, improved average survival was associated with gynecologic cancer compared to primary GI neoplasia (38.8 months vs 19.0 months; $p < 0.01$) [5].

Finally, the type of treatment seems able to influence the patient's prognosis. Despite some authors proposing only palliative treatment because of these patients' poor prognosis [11] [16], recent studies have demonstrated that there is a better survival (21 months) for patients if they are treated with a combination of surgery and adjuvant therapy instead of surgery alone (7.4 months) or chemotherapy alone (10.3 months) or no treatment (2.3 months). Aggressive surgical approach combined with chemotherapy may improve survival [12].

CONCLUSIONS

In conclusion, Sister Mary Joseph's nodule usually represents metastasis of an advanced visceral cancer accompany with poor prognosis. Therefore, when facing an umbilical lesion, careful examination is recommended. In addition, this nodule should be biopsied in order to look into its histopathological nature. Diagnosing Sister Mary Joseph's nodule early and properly with such clinical attitude is considered highly important in identifying the presence of intraperitoneal malignant tumors early and improving prognosis by treatment [19].

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