Sister Mary Joseph’s nodule: from the history to the images. A case-based literature review

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Summary
Sister Mary Joseph’s nodule refers to a palpable nodule bulging into the umbilicus as result of a malignant cancer in the abdomen or pelvis. It is associated with multiple peritoneal metastases and usually indicates an advanced stage of disease with a poor prognosis. Most cases are metastatic adenocarcinoma malignancies. The most common primary sites are the gastrointestinal tract (Gastric, Colonic and Pancreas) that accounts for about a half of the underlying sources (52%) and gynecologic (ovarian and endometrial cancer, 28%). This article describes Sister Joseph’s nodule, with a brief overview from history to imaging features in computed tomography.

Key words. Sister Mary Joseph’s nodule, peritoneal neoplasms, neoplasm metastasis, gastrointestinal neoplasms, ovarian neoplasms.

Nódulo de la Hermana María José: de la historia a las imágenes. Una revisión de la literatura basada en casos

Resumen
El nódulo de la Hermana María José, se refiere a una masa palpable en el ombligo que se traduce en una neoplasia avanzada del abdomen o pelvis. Está asociado con la presencia de múltiples metástasis peritoneales y usualmente indica una enfermedad avanzada con mal pronóstico. La mayoría de los casos son neoplasias de tipo adenocarcinoma metastásico. Las causas más comunes son las neoplasias del tracto gastrointestinal (gástrico, colon y páncreas) y representan la mitad de los primarios (52%) y ginecológico (cáncer de ovario y endometrio, 28%). Este reporte busca describir el nódulo de la hermana María José, con una breve reseña de su historia y sus características en la tomografía computada.

Palabras claves. Nódulo de la Hermana María José, neoplasias peritoneales, metástasis, neoplasias gastrointestinales, neoplasias ováricas.

Sister Mary Joseph’s nodule (SMJN) is a palpable periumbilical cutaneous lesion indicative of an underlying advanced intraabdominal neoplastic disease and its presence implies a poor prognosis.1, 2 This review aims to describe the SMJN sign, focusing in its history, clinical and imaging features of multidetector computed tomography (MDCT) images, through cases.
History

The name, was originally coined by Sir Hamilton Bailey, an English surgeon and the first to mention it in its book “Demonstrations of Physical Signs in Clinical Surgery”, published in 1949, honoring the Sister Mary Joseph (1856-1939) (Figure 1), who was superintendent nurse and Dr. William Mayo most frequent first assistant, at the St. Mary’s Hospital (now Mayo Clinic) of Rochester, Minnesota.

The Sister Mary Joseph, was the first person who observed that the patients with advanced abdominal-pelvic malignancies, frequently presented a periumbilical palpable nodule.3, 4 Until now, over 400 cases has being published.5

Clinical features

This nodule refers to a non-sensible palpable, periumbilical nodule bulge which color varies from violaceous to reddish brown that may appear as vascular structure. Occasionally skin ulcerations or fissures with even hematic, serous or purulent discharge can be observed. The lesion diameter rarely exceeds 5 centimeters, even so, it can be larger and simulate an umbilical hernia (Figures 2, 3, 4, 5 and 7).6

The SMJN represents 1-3% of the secondary locations of the gastrointestinal malignancies.7 Despite the majority of the primary lesions that can be identified with imaging studies, in a third of the cases (15-30%) the primary lesion remains unidentified even through tissue samples (molecular biology and immunohistochemistry).8

The most frequent malignancy metastases are gastrointestinal adenocarcinomas (gastric, colon and pancreas) and gynecological (ovary and endometrium), with a 52% and 28% respectively. Gastrointestinal malignancies are, as group, the most frequent neoplasms. Gastric origin can be found as cause of the 25% of the SMJN, followed by colorectal (10%) and pancreas (7%). The types of this tumors are 75% adenocarcinomas, followed by carcinoids and undifferentiated tumors.5

Gynecological origin like epithelial ovary neoplasm and especially, the papillary cystadenocarcinoma that just by itself is the first cause of the SMJN in this group (34%).5, 9

Pathophysiology

The mechanism which these tumors spreads to the umbilicus remains unclear, however, several hypotheses has been developed including 3 possible dissemination pathways: contiguity, lymphatic and hematic spreading (Figure 6). The umbilicus is extensively connected to intraabdominal spaces due to several embryological
Figure 2. Man 56 years old, advanced gastric neoplasm. A) Picture of the umbilical lesion; B) Contrast enhanced MDCT axial slice, in portal-venous phase: Large neoplastic lesion that bulge the anterior wall of the gastric wall (arrow head) with transmural involvement with non-defined borders, that invades the omentum (black asterisk (*)); ascites (white asterisk (*)); metastatic umbilical nodule (SMJN)(arrow).

Figure 3. Man 45 years old, recurrence of surgically treated gastric neoplasm. A) Picture of the umbilical lesion; B) Contrast enhanced MDCT axial slices, in portal-venous phase: Extensive gastric neoplasm recurrence with omental involvement (omental cake) (*); Diffuse nodular peritoneal thickening and ascites (arrow head), suggests peritoneal carcinomatosis; metastatic umbilical nodule (SMJN) (arrow).
Figure 4. Man 48 years old, advanced pancreatic cancer in palliative care. Contrast enhanced MDCT axial slices, in portal-venous phase: Images shows hypovascular liver metastasis and an extensive infiltrative retroperitoneal (*), vascular and peritoneal involvement. A SMJN can be appreciated in the umbilical area (E and F). Clinicians thought that they were leading with a non-complicated umbilical hernia. Left-sided portal hypertension syndrome consists in gastric and esophagus varices with patent portal vein and extensive splenic vein thrombosis secondary tumoral invasion and replacement of splenic vein (arrows). The peritoneal involvement denotes a small amount of loculated ascites and a slightly thickening of the peritoneum with some small nodular areas (arrow head). In this exceptional case, the omentum does not appear to have macroscopic involvement.
Figure 5. Man 50 years old, advanced gastric cancer in palliative care. Contrast enhanced MDCT axial slices, in portal-venous phase: (A, B, C) Huge Gastric neoplastic mass with transmural involvement with non-defined borders that invades the omentum [white asterisk (*)], hepatoduodenal and gastrohepatic ligaments [black asterisk (*)]; hypovascular liver metastasis (yellow arrows). (D, E) Metastatic umbilical nodule SMJN (volume renderings, images, blue arrow and circle).
Figure 6. Illustrative scheme of the possible routes of dissemination of an advanced left colonic neoplasm. Transmural, omental, peritoneal and cutaneous periumbilical metastatic nodule (SMJN) involvement are represented. In this scheme the direct extension mechanism is characterized through the mayor omentum.

Figure 7. Female 81 years old, advanced cirrhosis. Contrast enhanced MDCT axial slices with arterial and portal-venous phase: Images shows a dysmorphic liver with severe portal hypertension and two hepatic lesions in the right lobe with early arterial enhancement with classical washout in portal venous phase, findings consistent with hepatocarcinomas (HCC). Tiny supramesocolic omental nodules (*) and peritoneal pseudonodular thickening that shows enhancement (arrows), findings that represents the peritoneal involvement. Small implant in the hepatic serosal surface in the right lobe (arrow).
remnants structures, and holds a considerable amount of vascular anastomotic structures and peritoneal ligaments as a convergence point.¹⁰

Despite 90% of the neoplastic lesions in the periumbilical area correspond to metastases; a not despicable 10% are primary malignancies of the local tissue being vitelline and uracus remnants adenocarcinomas the most frequent malignancies in this group (Table 1). Less than 1% of this nodule has benign nature like endometrium (Villar’s nodule), epithelial cysts and fibrotic tissue.

**Table 1. Differential diagnosis of the cutaneous periumbilical nodules.**

<table>
<thead>
<tr>
<th>Primary location</th>
<th>Metastases (COPS)</th>
<th>Miscellaneous</th>
</tr>
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<tbody>
<tr>
<td>Melanoma</td>
<td>Gastrointestinal (Stomach, colo-rectal, pancreas)</td>
<td>Endometriosises</td>
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<tr>
<td>Epithelial neoplasm (basocelular and squamous)</td>
<td>Gynecologic (ovary, endometrium)</td>
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<tr>
<td>Miosarcoma</td>
<td>Melanoma</td>
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<tr>
<td>Adenocarcinoma</td>
<td>Sarcoma</td>
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<tr>
<td></td>
<td>Peritoneal Mesothelioma</td>
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**Imaging findings**

SMJN, is usually found in patients between 40 and 70 years old (slightly more frequent in women) with history of weight loss and unspecific general symptoms and laboratory tests. In this context, a cancer of unknown primary is highly suspected. Imaging studies are essential to determine the origin of the primary neoplasm, and generally, at this point an advanced intraabdominal disease can be found.

Ultrasound images can help to clarify the clinical findings by detecting solid umbilical nodules, even if the diagnosis is difficult to make clinically. Thus, by itself the ultrasound images may not lead to the diagnosis of the primary disease and usually require furthermore extensive examination.1

MDCT images allow the visualization of the cutaneous lesion and, in most of the cases; the primary lesion demonstrates its extension. Peritoneal or omental compromise like ascites and diffuse pseudo nodular involvement denotes a peritoneal carcinomatous process or omental cake.

A practical way to remember the main causes of the omental cake and the SMJN is using the acronym “COPS” that can be expanded as: colon, ovary, pancreas and stomach.11, 12

**Discussion**

The SMJN represents a cutaneous metastatic nodule located in the periumbilical cutaneous area. This nodule almost every time is a secondary location of an advanced intraabdominal malignancy and is considered as a very poor prognosis sign with an average life expectancy no greater than 6 months. Careful examination of all umbilical lesions is recommended. Most of the clinical diagnosed SMJNs are metastases of intraabdominal carcinomas that are observables in MDCT studies; 52% of these cases are secondary locations of gastrointestinal neoplasms and a 28% are gynecologic metastases. Although the diagnosis of the SMJN originally translated essentially an advanced cancer of unknown origin, today, its presence may represent up to 40%, recurrence of a previously treated malignancy.

The etiology of the primary malignancy determines the prognosis, and MDCT images and Fine Needle Aspiration Cytology of the tumor are invaluable in the diagnosis and recognition of the primary lesion.

The Sister Joseph’s nodule remains an interesting and valid useful radiologic and clinical sign in our days. Its correct diagnose is invaluable and the prognosis depends on it, radiologists must be aware.

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**Referencias**