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CLINICAL VIGNETTE

Sister Mary Joseph's Nodule in a Patient Presenting with Diffuse Large B-Cell Lymphoma

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A 70-year-old man with a history of coronary artery disease, atrial fibrillation on warfarin, diabetes mellitus, and hypertension, presented to his outpatient clinic with one week of abdominal pain, bloating, and nausea. He described the pain as pressure-like, localized to his periumbilical area and without radiation. He was prescribed simethicone without relief and he presented to the emergency department one week later due to persistent abdominal discomfort. He also reported several months of generalized fatigue and decreased appetite. Physical examination revealed a palpable erythematous nodule bulging into the umbilicus (Figure A), as well as an enlarged left cervical lymph node. He had mild epigastric tenderness to palpation. The rest of his abdominal exam was unremarkable. Laboratory studies revealed hypercalcemia (Calcium of 13.0), as well as thrombocytopenia (Platelets of 85) and normocytic anemia (Hemoglobin of 13.4). Abdominal ultrasound was notable for confluent lesions at the root of the mesentery and splenomegaly with multiple new splenic lesions.

Computed tomographic (CT) scan of the chest/abdomen/pelvis revealed widespread supraclavicular, axillary, mesenteric, and peri-aortic lymphadenopathy, as well as splenomegaly with multiple ill-defined masses concerning for lymphomatous involvement (Figure B). Core needle biopsy of the left cervical lymph node was consistent with malignant high-grade B cell lymphoma. He was diagnosed with diffuse large B cell lymphoma (DLBCL). The patient's abdominal discomfort rapidly improved after treatment of his hypercalcemia with intravenous fluids and pamidronate. Full body Positron Emission Tomography (PET)/CT scan which confirmed extensive hypermetabolic lymphadenopathy involving the neck, chest, abdomen, and pelvis and hypermetabolic splenomegaly, as well as an intense fluorodeoxyglucose (FDG) avid soft tissue umbilical mass (Figure C). Bone marrow biopsy and lumbar puncture did not show evidence of lymphoma. He completed 6 cycles of R-EPOCH therapy and follow-up PET/CT scan revealed complete resolution of hypermetabolic lesions in the neck, chest, abdomen, and pelvis, suggestive of complete metabolic response to treatment.

Sister Mary Joseph's nodule (SMJN) is a palpable nodule in or around the umbilicus secondary to metastatic malignancy. Mary Joseph Dempsey first described it in the early 1900s during her work with Dr. William James Mayo. SMJN usually presents as a painful lump on the anterior abdominal wall with irregular margins. The surface may be ulcerated and necrotic,

with serous, purulent, or mucous discharge. The size usually ranges from 0.5 to 2 cm, although some nodules may reach up to 10 cm.¹ SMJN has been reported in 1-3% of patients with intra-abdominal or pelvic malignancy, most commonly gastric cancer in men and ovarian cancer in women.¹ Considered a sign of advanced malignancy, the finding is associated with a poor prognosis. We report a case of SMJN as a presenting sign of diffuse large B cell lymphoma (DLBCL).

Diffuse large B cell lymphoma (DLBCL) is the most common subtype of non-Hodgkin lymphoma, accounting for approximately 30% of cases.² Though diffuse nodal enlargement – as in the case of our patient – is a common initial presentation of DLBCL, metastatic involvement of the umbilical region is rare. Up to 88% of cases of umbilical nodules originate outside the umbilicus while the remainder are primary skin tumors.³ The mechanism of metastasis to the umbilicus in SMJ nodules remains unknown but has been proposed to involve the lymphatic, venous, and arterial systems. Direct extension from the anterior peritoneal surface via embryologic remnants has also been considered.^{1,2} The presence of umbilical involvement usually represents advanced metastatic disease and has been associated with a poor prognosis, with average survival of 2 to 11 months.^{1,2} Recent studies report combined surgery and chemotherapy significantly improve survival (from 2 to 17 months).⁴ As in our patient's case, 14% to 33% of patients with SMJN, umbilical metastasis leads to the diagnosis of previously occult neoplasms. One review of cases with SMJN, reported was the only initial presentation of internal malignancy in 64%.²

The most common primary malignancies are gastrointestinal (35-65%) and genitourinary (12-35%), with gastric cancers being the most common in men and ovarian cancers the most common in women.^{1,2} The primary site remains unknown in 15-30% of cases.^{1,3} Hematologic, lung, and breast malignancies are identified in only 3–6% of cases.¹ Only six cases of SMJN secondary to metastatic lymphoma have been reported in the literature, with 5 of 6 identified as non-Hodgkin lymphoma with 3 specifically large B cell lymphoma.^{5,6} Importantly, all previously reported patients with non-Hodgkin's lymphoma responded to systemic chemotherapy, two achieved complete remissions, and one is known to be clinically disease free four years post-treatment.⁵⁻⁸ This case illustrates a rare example of SMJN as a presenting sign of DLBCL and underscores the importance of this exam finding in the initial evaluation of a

patient with undifferentiated abdominal pain. Moreover, it affirms that not all SMJN are attributable to incurable intra-abdominal or intra-pelvic malignancies and that the traditional poor prognosis associated with SMJN does not apply to all cases.



Figure A: Erythematous umbilical nodule on initial presentation



Figure B: CT abdomen/pelvis with widespread lymphadenopathy and an umbilical mass

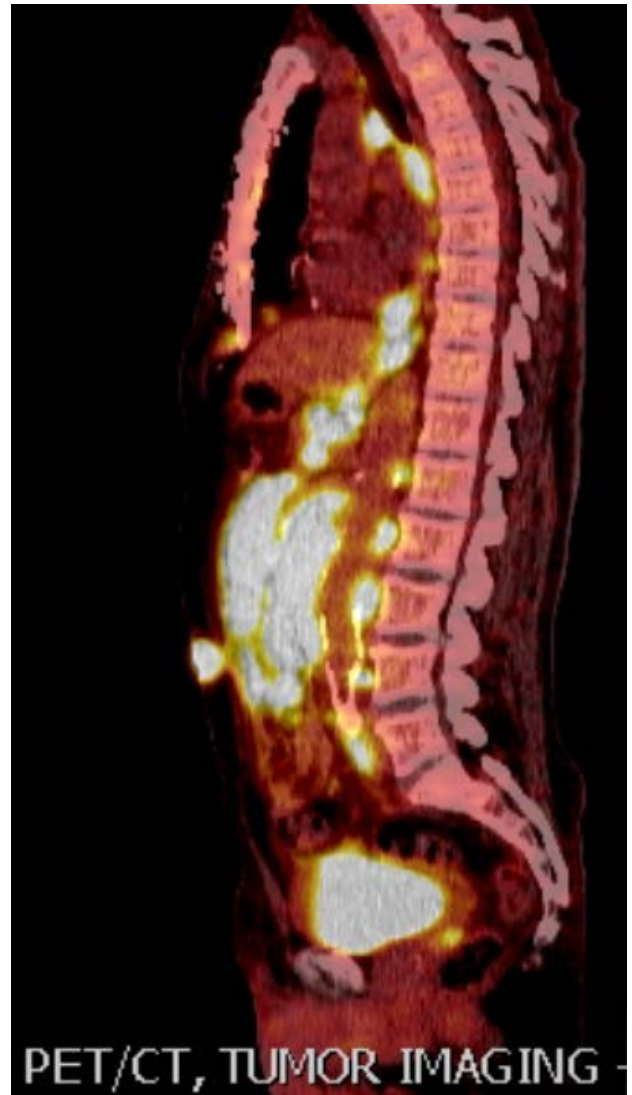


Figure C: PET/CT showing extensive hypermetabolic lymphadenopathy and soft tissue umbilical mass

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