A 64-year-old female presented with a three-month history of decreased appetite, nausea and progressive abdominal distension. Physical examination revealed a distended abdomen with a fluid wave and a hard periumbilical bluish swelling (Fig. A).

Because intra-abdominal malignancy was suspected, a contrast-enhanced computed tomography of the abdomen was performed which showed a pelvic mass originating from the left ovary, omental and peritoneal metastases, ascites, and a lobulated mass through the umbilicus with infiltration of the surrounding subcutaneous fat (Fig. B, C). Diagnostic laparoscopy and histopathological analysis of the obtained ovarian samples confirmed the diagnosis of metastatic ovarian carcinoma.

A Sister Mary Joseph nodule (SMJN) is a metastatic umbilical lesion originating from intra-abdominal or pelvic malignant disease. It can be a sign of undiagnosed malignancy or a symptom or sign of disease progression or recurrence in a patient with known disease. Its incidence is low, occurring in 1%-3% of all intra-abdominal or pelvic malignancies.

Sister Mary Joseph (1856-1939), a superintendent nurse and surgical assistant of Dr. William Mayo at St. Mary’s hospital (now the Mayo Clinic) in Rochester, Minnesota, USA, was the first to notice the association between the umbilical node and intra-abdominal malignancy while preparing patients for surgery. In 1949, the English surgeon Sir Hamilton Bailey (1894-1961) coined the term ‘Sister Mary Joseph nodule’ in his book ‘Physical Signs in Clinical Surgery’ in honor of its discoverer.

SMJN presents as a firm indurated umbilical nodule. The surface can be fissured or ulcerated, and it may have bloody, mucinous, serous or purulent discharge, depending on the primary tumor. The lesion has been described as variously coloured (white, bluish violet and brownish red) with or without pruritus. It is variable in size with a diameter often less than 5 cm, although some nodules may reach up to 10 cm. Patients with SMJN often present with a number of clinical symptoms consistent with intra-abdominal malignancy like abdominal pain or distension, nausea, anorexia, weight loss, or ascites. However, in up to 30% of cases SMJN is the first and only sign of a malignancy.

In men, the gastrointestinal tract, particularly the stomach (20%), is the most common location of the primary malignancy, whereas in women gynecologic malignancies, especially ovarian and endometrial cancers, are the most common. Primary tumors in many other sites like gall bladder, pancreas, liver, small intestine, lung, breast, kidney, fallopian tubes, prostate and bladder have also been reported to cause SMJN. About 15% to 30% of all cases have an unknown origin. Histology usually reveals adenocarcinoma; however, there have been reports of umbilical metastases from sarcoma, mesothelioma, lymphoma and melanoma.

The exact mode of spread is not known and mechanisms of umbilical seeding from primary tumors remain unclear. Contiguous extension from the anterior peritoneal surface is thought to be the most important method. Hematogenous spread through the extensive arterial and venous networks of the abdominal wall or lymphatic spread to the umbilical region are other probable routes of metastatic spread. Direct extension along the ligaments of embryogenic origin (round ligament of the liver, vitelline duct, urachus) has also been postulated as a method of metastasis.

SMJN represents umbilical metastases; and therefore, both treatment and prognosis depend on the primary tumor. However, the presence of this nodule often indicates an advanced internal malignancy and is generally associated with poor prognosis. Physicians need to be aware of this rare clinical entity so that they can promptly diagnose the primary malignancy or its progression or recurrence.

Reference