Case report

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Umbilical metastasis: a report case and review of the literature

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Abstract

Sister Mary Joseph's nodule 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 sign of progression or recurrence in a known case and is usually associated with a poor prognosis. Its incidence is 1%–3% of all intra-abdominal or pelvic malignancies. We report an umbilical metastasis in a case of rectal carcinoma. The clinicopathologic features are discussed, and the related literature is briefly reviewed.

Keywords: Sister marie joseph's nodule, umbilical metastasis, rectal cancer, primary malignancy, pelvic malignancies

Background

Cutaneous metastases localised to the umbilicus are named "Sister Mary Joseph's nodules". In 1949 Sir Hamilton Bailey initially used this eponym in his book "Physical Signs in Clinical Surgery" to describe umbilical metastases, in honour of Sister Mary Joseph, who was the first to note the link between umbilical nodules and intra abdominal malignancy [1,2].

It is an inconspicuous and uncommon clinical sign, which not only shows the presence of visceral malignancy but also reveals the poor prognosis of these malignancies [3,4]. Stomach and colon being most common in men, and ovary, in women. Cases of metastasis from organs such as the cecum, endometrium, pancreas, gallbladder, prostate, and lung, and from unknown primaries have also been reported. The lesion is seen more often in women [5,6].

Case report

A 28-year-old man without any history of colon cancer or any other predisposing factors, had consulted for a burgeoning lesion of the umbilic, rectal bleeding and weight loss for 2 months duration. Rectal exam did not reveal any palpable masses or firmness. Physical examination revealed an erythematous, painful and firm subcutaneous umbilical nodule, that measures 2 cm in diameter. The rest of the physical examination was unremarkable, including freedom of peripheral lymph nodes. Colonoscopy showed an ulceroburgeoning process localized to the medium portion of the rectum. Abdominal CT demonstrated an irregular rectal thickening with a subcutaneous umbilical nodule without lymphadenopathy, peritoneal implant or liver metastasis. Upper fibroscopy was normal. Routine blood chemistry investigations and chest radiography were normal.

Rectal biopsy confirmed the diagnosis of well differentiated rectal adenocarcinoma. The histological study of nodule's biopsy showed, in the dermis, a carcinomatous proliferation of glandular architecture (Figure 1). The tubes were of variable size, lined by cubical and basophilic epithelium, demonstrating

moderate nuclear atypia and rare mitoses (Figure 2). Based on these data, the diagnosis of Sister Mary Joseph's nodule was confirmed.

Considering the advanced stage of the disease, patient was referred to the department of oncology for palliative chemotherapy. The patient is currently receiving treatment.

Discussion

The occurrence of cutaneous metastases from malignant neoplasms ranges in from 1% to up to 9% of individuals, as determined at autopsy. Those metastases to the umbilicus are uncommon and represent only 10% of all secondary tumors which have spread to the skin [1]. Epidemiological studies showed that this condition predominates in females [7].

In the majority of cases, the finding of an umbilical nodule represents recurrence or dissemination of known malignancies. Occasionally, however, this may be the initial presentation of the primary disease in up to 30% of the cases [1,8,9].

Most often, the umbilical metastasis is an adenocarcinoma, rarely a squamous carcinoma, melanoma or sarcoma [4,10,11]. If these nodules are secondary tumors deposits then the source of the primary tumors may be from the gastrointestinal (35–65%) and genitourinary (12–35%) tract. In addition, in 3–6% of cases it originates from haematological malignancies, or lung and breast cancers. In 15% to 30% of patients the source of the primary site of the tumors remains unknown [1,9,10,11].

A full understanding of the mechanism where by the tumor spread to the umbilicus remains unclear. However, several anatomical criteria and several hypothesis have been proposed, including direct invasion, through continuous extension, of the anterior peritoneum, which is the most common; lymphatic spread via the axillary, inguinal, para-aortic, internal mammary and external iliac lymph nodes; hematogenous spread through arterial and venous seeding; and finally direct extension along the embryonic ligaments [1,4,8].

Clinically, Sister Mary Joseph's nodules usually present as a

Figure 1. Carcinomatous proliferation of glandular architecture in the dermis (hematoxylin-eosin, original magnification: x 50).

Figure 2. The tubes were of variable size, lined by cubical and basophilic epithelium, containing moderate nuclear atypia and rare mitoses (hematoxylin-eosin, original magnification: x 100).

painful lump on the anterior abdominal wall. It has irregular margins and a hard fibrous consistency. The surface may be ulcerated and necrotic, with either blood, serous, purulent, or mucous discharge from it. The size of the nodule usually ranges from 0.5 to 2 cm, although some nodules may reach up to 10 cm in size. High-resolution ultrasound (US) helps to clarify the clinical findings by detecting solid umbilical nodules, even if the diagnosis is difficult to make clinically. Moreover, careful examination and imaging of the abdominal contents may also point to the diagnosis [1,4,10,12].

Its unspectacular appearance can lead to many misdiagnosis, and this lesion can discuss the differential diagnosis with umbilical hernia, hypertrophic scar, umbilical endometriosis, pilonidal sinus, pyogenic granuloma, lymphangioma, and primary carcinoma [3,4,5]. Histopathology including

immunohistochemistry plays a vital role in confirming or suggesting possible sites of primary tumors.

Usually, the presence of an umbilical metastasis indicates a poor prognosis, is the sign of advanced neoplastic disease. The survival of these patients without treatment has been reported to range from 2 to 11 months from the time of initial diagnosis [1,3,4].

However, some factors seem to influence the prognosis of such patients, so direct invasion by the peritoneum is associated with a worse prognosis, the nature of the primary tumor, the time of onset of umbilical metastasis, the prognosis seem more favorable when the metastasis is discovered before the primary tumor only when it is discovered after treatment. Finally, the type of treatment, despite some authors proposing only palliative treatment, 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) [1,6].

Conclusion

A careful examination of all umbilical lesions is recommended, especially in those patients with gastrointestinal and genitourinary tract malignancies. All umbilical mass lesions should be biopsied to determinate the pathological nature of the lesion and to offer the patient the best survival probability with a combination of surgery and adjuvant therapy.

Competing interests

The authors declare that they have no competing interests.

Authors' contributions

All authors read and approved the final manuscript.

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