CASE REPORT

Pancreatic cancer presenting as a Sister Mary Joseph's nodule

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SUMMARY

A 69-year-old man presented with a painful umbilical nodule of 1 month duration. On examination the nodule was blackish in colour with a serous discharging fluid and was 2×2 cm in size, tender and fixed to the skin. There were no scars or sinuses at the umbilicus and no palpable mass or organomegaly on systemic examination. The patient underwent wide local excision of the skin nodule and on histopathology was reported as metastatic adenocarcinoma of the skin. A CT of the abdomen was performed to look for the primary site, which showed a 5×4 cm lesion in the tail of the pancreas. A biopsy from the pancreatic lesion was taken which was reported as an adenocarcinoma. CA19–9 was more than 1000 U/mL. The patient was advised palliative chemotherapy with gemcitabine. He was unwilling to take any further treatment in view of the advanced nature of the disease.

BACKGROUND

Sister Mary Joseph's (SMJ) nodule is a common term given to the metastatic umbilical nodules. Malignant umbilical nodules may be the presenting feature in a variety of malignancies. About half of the cases of umbilical metastasis originate from intra-abdominal or pelvic structures, the majority being from the gastrointestinal tract (GIT). It is important to know about this clinical condition, its aetiology and the associated prognosis.

CASE PRESENTATION

A 69-year-old man presented with a painful umbilical nodule of 1 month duration. The nodule was blackish in colour with a serous discharging fluid (figure 1). On examination, the nodule was 2×2 cm in size, tender and fixed to the skin. There

were no scars or sinuses at the umbilicus. There was no palpable mass or organomegaly on systemic examination. The patient underwent excision of the nodule under spinal anaesthesia. The histopathology was reported as a metastatic adenocarcinoma of the skin (figure 2). A CT of the abdomen was performed to look for the primary site, which showed a 5×4 cm lesion in the tail of the pancreas (figure 3). A biopsy from the pancreatic lesion was taken which was reported as adenocarcinoma (figure 4).

INVESTIGATIONS

A CT of the abdomen showed an ill-defined heterogeneously enhancing lesion involving the tail of the pancreas, measuring 5×4 cm with a loss of adjacent peripancreatic fat stranding and multiple deposits in the peritoneum. Mild ascites was noted. There is a peripherally enhancing hypodense lesion in the midline indenting the rectus sheath with nodular stranding along the periumbilical region. A few air pockets are seen (postoperative changes).

CA19–9: more than 1000 U/mL.

TREATMENT

He underwent wide local excision of the umbilical nodule, which showed a metastatic adenocarcinoma of the skin.

OUTCOME AND FOLLOW-UP

The patient was advised palliative chemotherapy with gemcitabine. He was unwilling to take any further treatment in view of the advanced disease. The umbilical scar healed by primary intention.

Figure 1 Clinical photograph showing the dark pigmented umbilical nodule.

Figure 2 CT showing the heterogeneously enhancing tumour in the tail of the pancreas measuring 5×4 cm.

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Figure 3 Histopathology from the skin biopsy showing infiltration of the dermis with malignant glands suggestive of metastatic adenocarcinoma to the skin.

DISCUSSION

SMJ nodule is a common term given to the metastatic umbilical nodules. The term was first used by Hamilton Bailey in the 11th edition of 'Physical Signs in Clinical Surgery'.¹ It tends to present as a painful lump with fibrotic consistency and irregular margins. It also represents relatively poor prognosis.²

Malignant umbilical nodules may be the presenting feature in a variety of malignancies. There are many possible mechanisms of the spread to the umbilicus, including direct extension, haematogenous spread and lymphatic spread. The most common from primary sites are intra-abdominal in origin.³ Retrograde flux from superficial and deep lymphatics may also be an important mode of spread.³

Forty-two per cent of the cases of umbilical metastasis originate from the intra-abdominal or pelvic structures, the majority being from the GIT. Six per cent of these cases are pancreatic in origin. Histologically, the majority of these nodules are caused by adenocarcinomas (75%).⁴

Malignant umbilical nodules may also be seen in gynaecological malignancies (12–35%).⁵

As mentioned earlier, SMJ nodule presents as a painful lump with fibrotic-to-hard consistency and irregular margins. The surface may be ulcerated and necrotic with blood and serous or purulent discharge.^{2,3}

Immunohistochemical staining for CK7 and 19 are very important in diagnosing adenocarcinomas of pancreas. Serum tumour markers such as CA19–9 should be examined and elevation of CA19–9 is considered as a strong evidence of the disease.⁶

Figure 4 Histopathology taken from the pancreatic tissue showing malignant cells arranged in clusters, sheets and acinar pattern with high nucleus:cytoplasm ratio suggestive of adenocarcinoma.

Imaging modalities have a role. Ultrasonography, CT and MRI may be used to locate the primary lesion in the abdomen. However, for their cost, they are only relatively effective diagnostic tools for this condition. Contrast-enhanced multidetector CT can show the pancreatic mass. Positron emission tomography-CT is useful in detecting distal metastasis, but is not sensitive enough to reveal seeding molecules.⁷

Surprisingly, despite its poor prognosis, it makes diagnosis of such a malignancy easier, as we can perform fine needle aspiration cytology of the umbilical nodule without having to take the unnecessary step of diagnostic laparotomy.⁸

There is still debate as to whether or not the presence of the SMJ nodule by itself implies inoperability of the patient. However, there are arguments to the contrary stating that the surgery may still be a viable treatment modality.⁹

Most often, a wide excision as a palliative treatment followed by search for primary lesion and radiotherapy is performed. Then a second surgery is performed for the excision of primary tumour, and adjuvant therapy is given. This is the standard modality of treatment, especially in cases of gynaecological tumours presenting with umbilical nodules.¹⁰

Survival rates are very poor, and there is very little scope for prognosis when a patient presents with tumours at such a late stage.^{2,3}

However, certain authors argue that the type of treatment may be able to influence the patient's prognosis. A combination of surgery and adjuvant therapy as opposed to surgery alone can produce longer survival periods.⁵

Learning points

- ▶ Sister Mary Joseph's nodule (SMJN) is a common term given to the metastatic umbilical nodules.
- ▶ Metastasis to the umbilicus originates from the intra-abdominal or pelvic organs.
- ▶ Pancreas is the source of an SMJN in 7–9% of cases.
- ▶ Fine needle aspiration cytology from the umbilical nodule can clinch the diagnosis.
- ▶ CT scan is ideal to look for the primary site.

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Competing interests None.

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