

Primary umbilical adenocarcinoma

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ABSTRACT

Primary umbilical tumours are extremely rare. We report primary serous adenocarcinoma arising from the coelomic mesothelium of the hernial sac. A 60-year-old woman presented with an umbilical swelling of six months duration that became painful in the last three days. Examination revealed a tender umbilical swelling diagnosed as obstructive hernia that needed surgery. When dissecting the sac during surgery, a small subcutaneous abscess was encountered. The sac contained an omentum with a hard nodule at the surface which was excised. Umbilical hernia repair was performed. Histology of the omental nodule revealed serous papillary adenocarcinoma. Chest and abdomen computed tomography, pelvic magnetic resonance imaging, gastroscopy, colonoscopy and laparotomy did not reveal the primary site of the tumour.

Keywords: hernia sac tumour, omental nodule, papillary carcinoma, primary umbilical adenocarcinoma, umbilical tumour

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INTRODUCTION

Most adenocarcinomas of the umbilicus are secondary, while primary umbilical tumours are extremely rare. Primary umbilical adenocarcinoma could arise from a pre-existing endometrioma, the coelomic mesothelium, or an embryological remnant of the umbilicus.⁽¹⁾ The two umbilical embryological remnants from which primary umbilical tumours can arise are the vitello-intestinal (omphalomesenteric) tract and the urachus.⁽²⁾ We report a case of primary serous adenocarcinoma arising from a coelomic mesothelium of the hernial sac.

CASE REPORT

A 60-year-old woman presented with a painful umbilical swelling. The swelling started six months before but became painful in the last three days. The patient did not have any discharge or bleeding from her umbilicus. She was postmenopausal and had seven full-term, normal vaginal deliveries. She also

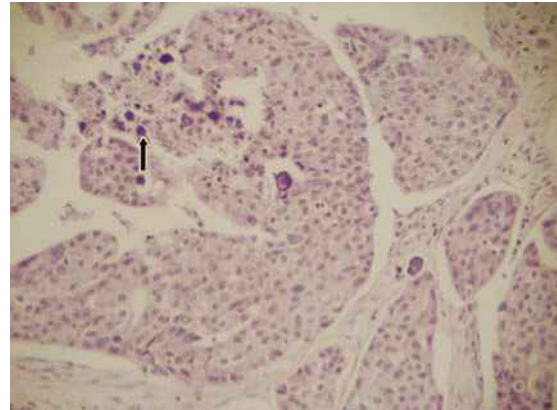


Fig. 1 Photomicrograph of the nodule consisted of a tumour having variable differentiation and serous papillary pattern. Psammoma bodies were prominent (arrow) (Haematoxylin & eosin, × 10).

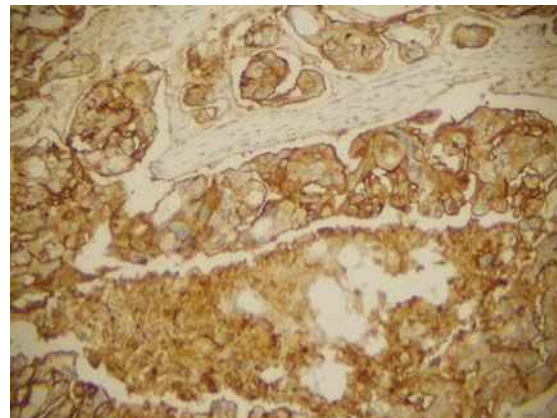


Fig. 2 Immunohistochemistry CA-125 shows a positive reaction (× 10).

had bilateral total hip replacement. Upon physical examination, she was found to be obese. Her blood pressure was 150/100 mmHg. There was a swelling in the periumbilical area having a length of 7 cm transversely and 5 cm longitudinally. The swelling was tender, red, and irreducible with no cough impulse. The abdomen was soft and lax. The swelling was clinically diagnosed as an obstructed umbilical hernia. White blood cell count, serum electrolytes, renal function, and liver function tests were all normal. The patient underwent an emergency repair of her umbilical hernia.

When dissecting the sac, a small subcutaneous abscess was encountered. The sac had only omentum

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with a hard nodule at the surface. The grossly pathological omentum and the hard nodule were excised. The postoperative period was uneventful. Histology of the nodule consisted of a tumour having variable differentiation and serous papillary pattern. Psammoma bodies were prominent (Fig. 1). Immunohistochemistry CA-125 (Fig. 2), oestrogen, and CK7 were positive. Carcinoembryonic antigen (CEA), progesterone, and calretinin were negative. A diagnosis of serous papillary adenocarcinoma was made. We considered the possibility of this lesion being a metastasis from the ovaries. However, ultrasonography of the pelvis was normal. Her serum CA 125 was 13 µg/L (normal range 0–35 µg/L). In addition, computed tomography of the chest and abdomen showed no abnormality, apart from staghorn calculi and hydronephrosis of left kidney, which was confirmed by intravenous pyelography. Magnetic resonance imaging of the pelvis, gastroscopy and colonoscopy were all normal.

The patient underwent exploratory laparotomy. Stomach, colon, small intestine, liver and pancreas were normal. The vermiform appendix was normal in size and shape, and was therefore not removed. The pelvic organs looked normal except for some omental nodules on the right ovary and tube. Peritoneal washing was performed and sent for cytology. Partial omentectomy, sub-total hysterectomy, and bilateral salpingo-oophorectomy were performed. Histopathology of the nodule showed fat and connective tissue of completely undifferentiated large cell malignant tumour with large amount of atypical mitotic figures. No glandular structures or characteristic features could be seen. In addition, the specimen showed normal ovary and tubal structures as well as uterus with a slightly haemorrhagic, mildly hyperplastic corpus endometrium without dysplasia. The patient had a smooth postoperative recovery and was discharged. She received chemotherapy treatment. Follow-up one year after surgery did not show any recurrence of the tumour.

DISCUSSION

Umbilical tumours may present as a nodule of variable size, which may be painful and sometimes, ulcerated. There may also be an abscess underlying the tumour, such as in our patient.⁽³⁻⁶⁾ This can be primary or secondary cancer. The most frequently-encountered histological type is adenocarcinoma, followed by epidermoid, undifferentiated or carcinoid tumours.⁽⁶⁾ Secondary metastasis to the umbilicus in males can originate from the stomach, colorectum, pancreas, lung or prostate.⁽⁷⁾ Rarely, adenocarcinoma of the Mickel's diverticulum may spread to the umbilicus.⁽⁸⁾ In females,

secondary metastasis can originate from the ovary, endometrium, colorectum, stomach, pancreas, cervix or breast.⁽⁷⁾ Other possibilities are primary serous carcinoma of the peritoneum with spread to the umbilicus. Primary umbilical tumours are extremely rare.⁽⁹⁾ This includes primary melanoma of the umbilicus.⁽²⁾ In our patient, the absence of any other tumour, which could be the primary source, and the histological findings have indicated that the lesion is a primary serous adenocarcinoma arising from a coelomic mesothelium of the hernial sac and it is not a metastasis from the ovary. We have excluded the presence of pre-existing endometrioma or gynaecological origin of the tumour. The patient was postmenopausal with no history of gynaecological symptoms.

The morphology and immune profile of the tumour indicates that it was coelomic in origin and not from the omentum, because CA-125 and oestrogen were positive, similar to coelomic epithelium. Whether the tumour has originated from the hernia sac or the visceral peritoneum within the sac cannot be answered, but the localised nature of the tumour within the umbilicus supports that it is from the hernial sac. Tumours originating from the urachus usually have transitional epithelium, but occasionally, glandular epithelium may be observed making the definition of the origin of the tumour difficult.⁽¹⁰⁾ When the tumour arises from the urachus, it will have a distinctive clinical picture. It occurs predominantly in men who present with suprapubic mass and micturition problems.⁽¹¹⁾ In our patient, there was no bladder tumour. Other primary tumours include papillary serous carcinoma of the peritoneum. This resembles papillary serous carcinoma of the ovary.⁽¹²⁻¹⁴⁾ As there was no identifiable source of the tumour during surgery, we think that the umbilical tumour is primary in origin. When it comes to treatment, the treatment is exactly the same as ovarian cancer (i.e. debulking surgery followed by adjuvant chemotherapy with taxitère and platinum-based chemotherapy). Surgical cytoreduction by omentectomy is an essential part of surgery for ovarian carcinoma and primary peritoneal papillary carcinoma.^(3,12,14) Omentectomy was not performed for primary umbilical tumours.^(1,2,10) The partial omentectomy, which was performed in our patient, was to remove two small nodules which were residual from the first operation.

Intra-abdominal malignancy may present as an umbilical mass that is usually missed as an umbilical hernia. When there is intraoperative doubt of such a finding, the excised tissue should be sent for histopathology to confirm the diagnosis. If histology confirms adenocarcinoma, then other sources of primary malignancy should be excluded, in particular,

gynaecological and gastrointestinal origins. Abdominal and pelvic computed tomography and endoscopy will help to define the primary origin of the tumour. Exploratory laparotomy may be indicated when the origin of the tumour cannot be identified. Diagnosis of primary umbilical carcinoma is reached only by exclusion.

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