

Adenocarcinoma of the abdominal wall

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ABSTRACT

We report a rare case of adenocarcinoma confined to the umbilicus. A 60-year-old menopausal woman presented with an umbilical lump of four years. Excision biopsy showed adenocarcinoma with an appearance suggestive of metastasis from an ovarian cystadenocarcinoma. Tumour markers for ovarian malignancy were normal. Computer tomography did not reveal any evidence of ovarian malignancy. She defaulted on follow-up. After five years, she presented with a 6-cm irregular periumbilical mass. She agreed to undergo an excision biopsy of the mass with total abdominal hysterectomy and bilateral salpingo-oophorectomy with omentectomy. The frozen section of the tumour showed papillary adenocarcinoma. Histology showed adenocarcinoma favouring serous cancer. There was no tumour seen in the ovaries, uterus and omentum. On follow-up, there was no disease recurrence. The patient has been disease-free for two years post surgery.

Keywords: adenocarcinoma, extraovarian primary serous carcinoma, primary peritoneal carcinoma, umbilical tumour metastasis

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CASE REPORT

A 60-year-old, para 4, menopausal woman presented with a lump at the umbilicus that was gradually enlarging for four years. All other systems were normal. The lump was excised and histology showed there was extensive invasion of the skin to the deep dermis by papillary adenocarcinoma with psammoma bodies. The tumour extended focally into the superficial dermis but there was no ulceration. The small amount of mucin secretion by tumour cells was less than with mucinous carcinoma. The appearance suggested a metastasis from an ovarian cystadenocarcinoma. Immunohistochemistry indicated reactivity for CK 7 and non-reactivity for CK 20, consistent with serous carcinoma of the ovary. CA 125 was 13.0 U/ml, AFP was 4.0 µg/L, CEA was 3.3 µg/L, and Pap smear was normal.

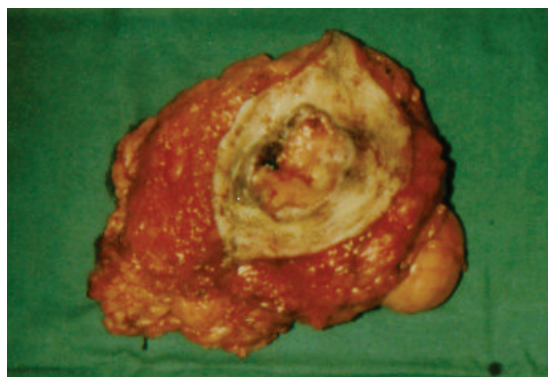


Fig. 1 Photograph of the resected specimen shows the umbilical wall tumour.

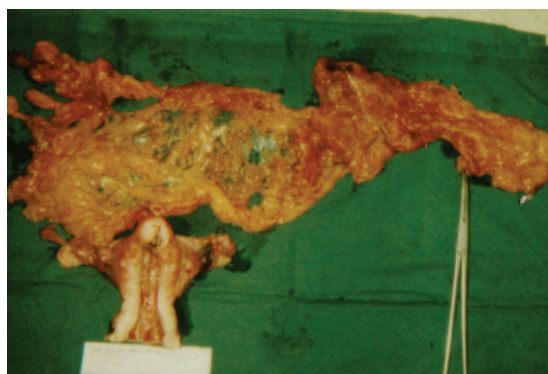


Fig. 2 Photograph of the resected specimen shows the uterus, fallopian tubes, ovaries and omentum.

Computed tomography (CT) showed a midline incisional hernia in the umbilical region. There was no evidence of an ovarian tumour. She was advised to undergo further surgery to ascertain the primary source of the disease. She defaulted on follow-up for five years, after which she presented with a lump over the umbilicus again, discharging pus and blood. There was a 6-cm irregular fleshy periumbilical mass with ulceration and rolled edges. CA 125 was 34.9 U/ml, CEA was 2.9 µg/L, βHCG was 2.3 µg/L, and AFP was 4.0 µg/L. The CT of her abdomen and pelvis showed a large periumbilical hernia with thickening of the right periumbilical skin and an area of calcification within.

She underwent a laparotomy and total abdominal hysterectomy, bilateral salpingo-oophorectomy and infragastric omentectomy. Intraoperatively, there was an

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ulcerative cancer around the umbilicus with a midline incisional hernia. It was resected with wide margins en bloc from the skin to the peritoneum (Figs. 1 & 2). Full intraperitoneal survey was normal with no evidence of cancer. The frozen section of the tumour showed papillary adenocarcinoma. Histology showed adenocarcinoma favouring serous cancer. There was no tumour seen in the ovaries, uterus and omentum. On follow-up, there was no disease recurrence. The patient has been disease-free for two years post surgery.

DISCUSSION

An umbilical tumour as a presenting symptom must be investigated for the possibility of metastasis. 75% of malignant umbilical tumours correspond to a Sister Joseph's nodule.⁽¹⁾ In this case, it is unlikely, as there was no primary carcinoma found elsewhere. The most probable diagnosis is a primary peritoneal carcinoma localised to the umbilicus. Primary peritoneal carcinoma (also termed serous surface papillary carcinoma) is a malignancy that arises primarily from peritoneal cells. The mesothelium of the peritoneum and the germinal epithelium of the ovary arise from the same embryological origin; therefore, the peritoneum may retain the multipotentiality of the müllerian system, allowing the development of a primary carcinoma.⁽²⁾ It is a very rare tumour and usually affects women in the older age group. The average age at diagnosis is 57.4 years.⁽³⁾ Although primary peritoneal carcinoma has a very poor prognosis, our patient had a disease-free interval of two years post surgery.

Clinically, primary peritoneal carcinoma may be difficult to distinguish from ovarian carcinoma. Compared with ovarian carcinoma, primary peritoneal carcinoma has a higher rate of abdominal distension, volume of ascites, malignant cells in the ascitic fluid, lower rate of pelvic palpable mass, and personal breast cancer history.⁽⁴⁾ Primary peritoneal carcinoma is histologically indistinguishable from primary epithelial

ovarian carcinoma. However, primary ovarian cancer can be excluded in this case, based on both ovaries being of normal size with no tumour involvement and a histology that favours serous carcinoma. The disease is normally disseminated throughout the peritoneum. 33% of patients have macroscopical uterine involvement.⁽⁵⁾ Our patient's disease was confined to the umbilicus with no evidence of dissemination.

Treatment is the same as for ovarian carcinoma. The best chance for survival is cytoreductive surgery, followed by adjuvant chemotherapy.^(6,7) The median overall survival period is 23.5 months.⁽⁸⁾ Our patient refused adjuvant chemotherapy and had no recurrence so far. In our literature search, there has not been a case of localised primary peritoneal carcinoma localised to a region where primary resection could render the patient disease-free for a relatively long period of time.

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