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Primary peritoneal adenocarcinoma as content of an incarcerated umbilical hernia: A case-report and review of the literature

D. Varga-Szabó^{a,*}, M. Papadakis^a, S. Pröpper^b, H. Zirngibl^a^a Chirurgisches Zentrum, Helios Klinikum Wuppertal, Wuppertal, Germany^b Institut für Pathologie, Helios Klinikum Wuppertal, Heusnerstr. 40, 42283 Wuppertal, Germany

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ABSTRACT

INTRODUCTION: Umbilical hernia is a common finding in many cases, posing potentially life-threatening complications, such as incarceration or strangulation. The presence of malignancy in hernia sacs is, however, rather rare.

PRESENTATION OF CASE: Here we report on a case of primary peritoneal adenocarcinoma found through histological examination of omental tissue, resected due to an incarcerated umbilical hernia of an 84-years-old woman. There was no macroscopic sign of malignancy during operation; only after routine examination of histological sections the diagnosis was found.

DISCUSSION: To our knowledge this is the first report of primary peritoneal cancer as content of an umbilical hernia. This is a rare neoplasm and histologically identical to epithelial ovarian carcinoma. For this reason, the diagnosis is usually based on the histological finding and exclusion of a primary ovarian tumor. Primary peritoneal cancer has a poor outcome in general. Early diagnosis is, therefore, essential for effective treatment.

CONCLUSION: Histological analysis of resected hernia sac or content should be performed routinely to discover malignant diseases in the background of a hernia.

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1. Introduction

Umbilical hernias are common findings on physical examination. In infants, this is the result of the incomplete closure of the umbilical ring. In African children, the incidence can be as high as 40%, however the spontaneous closure is rather high and the incidence decreases to 15% by one year of age [1]. In contrast, umbilical hernias in the adult are less frequent, with an incidence of approximately 5% of all abdominal wall hernias, and mainly acquired [2]. However, higher incidence has also been reported [3]. In Germany, approximately 30 new cases per 100.000 people are reported per year [4].

In general, conditions raising intra-abdominal pressure can lead to the development of an umbilical hernia. Obesity, pregnancy, ascitic fluid and excessive muscular effort are the most well-known causes. Incarceration is a severe complication requiring immediate surgical treatment to avoid tissue damage due to ischemia. The frequency of incarceration of adult umbilical hernias is not well documented in the literature. We found only one large nationwide study dealing with this topic. Helgstrand et al. report that approx-

imately 11% of all umbilical hernia repairs in Denmark, during a 4-year period, were performed as emergency hernia repair [5].

Small bowel and omental tissue are the most common hernia sac contents; however, rarely also tumors can be discovered. Further, in some cases this is the first manifestation of malignant diseases or metastatic cancer, such as hamartomas, endometrial squamous cell carcinomas, ovarian cancer, colonic cancer, bladder carcinoma and primitive neuroectodermal tumors [6–11].

Here, we report – in line with the CARE criteria [12] – the case of an 84-year-old woman with primary peritoneal carcinoma found as content of an incarcerated umbilical hernia. The related literature is discussed.

2. Presentation of case

An 84-years-old woman with a painful umbilical swelling was examined in our emergency room. She complained of progressive umbilical pain for 2–3 days with nausea but no vomiting. Her medical history showed arterial hypertension and a hysterectomy with removal of the right ovary 45 years ago due to uterine fibroids. On physical examination a painful periumbilical mass of approximately 6 cm in diameter was found. On inspection the abdomen was non-distended, on palpation there was slight abdominal pain without guarding. To further assist the clinical diagnosis of incarcerated umbilical hernia, abdominal ultrasonography was

* Corresponding author. Fax: +49 202 896 2930.

E-mail address: david.varga-szabo@helios-kliniken.de (D. Varga-Szabó).

performed. This confirmed the umbilical hernia with an orifice of approximately 15 mm and omental tissue as hernia sac content. There was no sign of small or large bowel in the hernia sac. Laboratory tests showed leukocytosis and an increase of CRP, without elevation of serum lactic acid levels.

The patient was taken to emergency surgery, where a 10 × 6 cm segment of omentum majus with the hernia sac was resected and sent to histological examination. It is important to state that there were no signs of malignancy visible; the excision was performed due to signs of ischemia, however, without necrosis. The hernia defect was closed and a piece of excess skin was resected.

The pathology report described a moderately well differentiated, invasive adenocarcinoma with papillary growth pattern and psammoma bodies. Immunohistochemical staining was strongly positive for cytokeratin (CK) 19, cancer antigen (CA) 125 and CK 7 but negative for thyroid transcription factor (TTF)-1 and CK 20. These results were consistent with primary peritoneal carcinoma (PPC) or primary ovarian carcinoma.

In order to locate the primary tumor, computed tomography of the abdomen was performed. This revealed a suspect enlargement of the left ovary without further signs of malignancy in the abdominal cavity. Further, esophagogastroduodenoscopy and colonoscopy were performed and showed no suspect lesion in the gastrointestinal tract. Laboratory tests showed normal level of the tumor marker CA 125 (22.08 U/ml).

The patient subsequently underwent a diagnostic laparoscopy and open left ovariectomy. There were no macroscopic signs of malignancy; nonetheless, a debulking procedure including the resection of the rest of the omentum majus was performed. The final pathology report revealed a serous cystadenofibroma of the left ovary with no evidence of malignancy but multiple foci of carcinoma in the omentum majus leading to the diagnosis of primary peritoneal carcinoma. The patient received over a period of 3 months 6 cycles of carboplatin chemotherapy; however, unfortunately after 14 months pleural carcinosis has been diagnosed and after 30 months of follow-up the patient died.

3. Discussion

General surgeons often treat patients with umbilical hernias. However, incarceration and strangulation of umbilical hernia content is rather rare and finding a neoplasm in an incarcerated umbilical hernia is even more seldom. There are only a few reports in the literature, which present such cases. Among malignancies found in umbilical hernias ovarian cancer [8], malignant peritoneal mesothelioma [13], primitive neuroectodermal tumor [11] and metastases of various intraabdominal cancers – so called Sister Mary Joseph's nodule – [13] have been reported. Whereas, there is a single case report of a primary peritoneal carcinoma in a femoral hernia sac [14], to our best knowledge, this is the first case of primary malignant peritoneal carcinoma detected within an incarcerated umbilical hernia.

Primary peritoneal carcinoma (PPC) is a rare neoplasm, histologically identical to epithelial ovarian carcinoma (EPOC) [15]. It has an age-standardised incidence rate of 6.78 per one million people in the US, steadily increasing over the last three decades. PPC has a very strong female predominance usually affecting older ages. A mean age of 67 years in a series of 6458 patients from 24 population-based registries in the US during the period 1995–2004 has been reported [16]. In the same study, PPC shows a declining incidence trend in the age group of our patient (80–85). Epidemiological analysis shows a positive correlation between hysterectomy in the medical history, as well as between increasing age at the last pregnancy and the prevalence of PPC or EPOC [17].

The most common symptoms of PPC include gastrointestinal symptoms such as abdominal distension and pain. The main clinical sign is ascites, which appears in approximately 85% of cases [18], but was not observed in our patient. Moreover, our staging was negative for lymphadenopathy, which according to Eltabbakh et al. is clinically or surgically present in 11% of the cases [19]. The CA 125 level in our case was also normal, differing again from the expected elevation of tumor marker levels [18].

PPC can diffusely infiltrate the peritoneum while the ovaries are not or only minimally involved. As it is histologically indistinguishable from EPOC, the diagnosis is usually based on the lack of another primary and is usually made intra- and postoperatively. Clinical differential diagnosis includes not only ovarian and fallopian tube carcinoma but also benign peritoneal tumors and metastatic peritoneal cancer.

Whether PPC and EPOC are distinct entities or share common origins, remains elusive. Comparative epidemiological analyses confirmed, however, significant differences in the risk factor associations between PPC and EPOC suggesting development along divergent pathways [17]. Still, the therapeutic approach is similar for both diseases and is mainly in the hand of gynecologists.

The therapy of PPC includes surgery with the goal of maximal cytoreduction and a platinum-based chemotherapy; the response rate of approximately 60% is similar to that of EPOC [20,21]. Low drug resistance to both platinum and taxane chemotherapy was an independent predictor of improved survival in patients with PPC as determined by an *in vitro* drug resistance assay [22]. The median survival has been reported to vary between 12 and 25 months [21,23,24], which decreases to 8.5 months once brain metastasis occurs [25].

4. Conclusion

We have reported on a patient with a PPC presenting as incarcerated umbilical hernia. Malignancy in a hernia is a rare finding; however there are several reports of ovarian carcinoma as hernia content. PPC is histologically identical to EPOC; therefore, PPC should be considered in the differential diagnosis of incarcerated or strangulated umbilical hernias. Early diagnosis is essential for effective treatment of the tumor; therefore, we recommend routine histological examination of the resected hernia sac or content even in the absence of clinical signs of malignant disease.

Conflict of interest

All authors declare no conflict of interest.

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Ethical approval

The manuscript required no studies on patients.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Author contribution

D. Varga-Szabo, M. Papadakis and H. Zirngibl wrote the paper. S. Pröpper provided histological images for the manuscript.

Guarantor

D. Varga-Szabó.

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