Pathology – Research and Practice 211 (2015) 550–555

Contents lists available at ScienceDirect

Pathology – Research and Practice

journal homepage: www.elsevier.com/locate/prp

Teaching cases

Low-grade serous primary peritoneal carcinoma incidentally found in a hernia sac

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ARTICLE INFO

Article history:
Received 11 November 2014
Received in revised form 23 February 2015
Accepted 8 April 2015

Keywords:
Peritoneum
Hernia sac
Primary peritoneal low-grade serous carcinoma
Immunohistochemistry

ABSTRACT

Very rarely, a primary peritoneal serous carcinoma can be observed in a hernia sac. We herein describe a low-grade serous primary peritoneal carcinoma incidentally found in a postmenopausal woman following examination of the femoral hernia repair sac. Our case is significant for its unusual presentation. The lesion initially appeared as a 0.3-cm tumor that disseminated in the peritoneum, persisted, and progressed for 75 months. The absence of ovarian disease indicated a primary peritoneal origin. Tumor cells were immunohistochemically positive for PAX8, claudin-4, and VE1, excluding the possibility of being of mesothelial origin. Recognition that a low-grade serous primary peritoneal carcinoma can be incidentally found in a hernia sac should simplify future diagnoses. Immunohistochemistry is helpful in making the correct diagnosis.

Introduction

Primary peritoneal serous carcinoma is an uncommon lesion histologically indistinguishable from its ovarian counterpart [15]. The majority of reported primary peritoneal serous carcinomas have been high-grade tumors [15,18]. On the contrary, low-grade serous primary peritoneal carcinomas (LGSPCs) are reported much less frequently [13,14,17].

We describe herein a LGSPC incidentally found in a postmenopausal woman following examination of the femoral hernia repair sac. To the best of our knowledge, there are no documented cases of this lesion incidentally found in a hernia sac. Our case is significant for its insidious onset, its peritoneal spread, persistence and progression.

Case report

A 62-year-old woman presented with a right irreducible femoral hernia. This had been present for 3 years, and she had been under investigation for right lower abdominal pain for more than 3 years. Previous CT scan revealed an effusion in the right periovarian area and in Douglas cul-de-sac. Uterus and mesentery were normal. Hernia repair yielded an irregular gray white, thickened, membranous tissue fragment measuring 3.5 cm × 3.2 cm × 1.5 cm. Microscopically, the hernia sac showed a 0.3-cm neoplasm composed of two nodules that had limited submesothelial tissue invasion (Fig. 1A). They were characterized by a combination of a papillary pattern, cord-like pattern, single cells, and cellular nests. The fibrovascular cores were thin and covered by a single layer of uniform cuboidal cells with focal cell detachment (Fig. 1B). There were no mitoses or necrotic areas. The cells showed evident but not prominent nucleoli. There was a sharp boundary between the stroma and the adjacent adipose tissue. The immunohistochemical study revealed tumor cell positivity for cytokeratin AE1/AE3, cytokeratin 7, calretinin (Fig. 1C), and claudin-4 (Fig. 1D), and negativity for cytokeratin 20, thrombomodulin, PAX2, estrogen receptor alpha, and progesterone receptor. PAX8 showed focal positivity. The initial pathologic interpretation was that of primary peritoneal serous borderline tumor. The neoplasm was apparently located in the hernia sac, and the patient received no treatment.

After a latent period of 32 months, she presented with a recent onset of abdominal pain associated with nausea, vomiting and moderate ascites. An exploratory laparoscopy revealed multiple, discrete, ≤1 mm nodules in the omentum, pelvic peritoneum and Douglas cul-de-sac that were biopsied. Later on, a total abdominal hysterectomy and bilateral salpingectomy and oophorectomy were carried out for suspected ovarian carcinoma.

The tiny nodules in the omentum and pelvic peritoneum showed a trabecular pattern of branching cords of tumor cells (Fig. 2A). These cells showed frequent small nucleoli. Cellular atypia was

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http://dx.doi.org/10.1016/j.prp.2015.04.001
0344-0338/

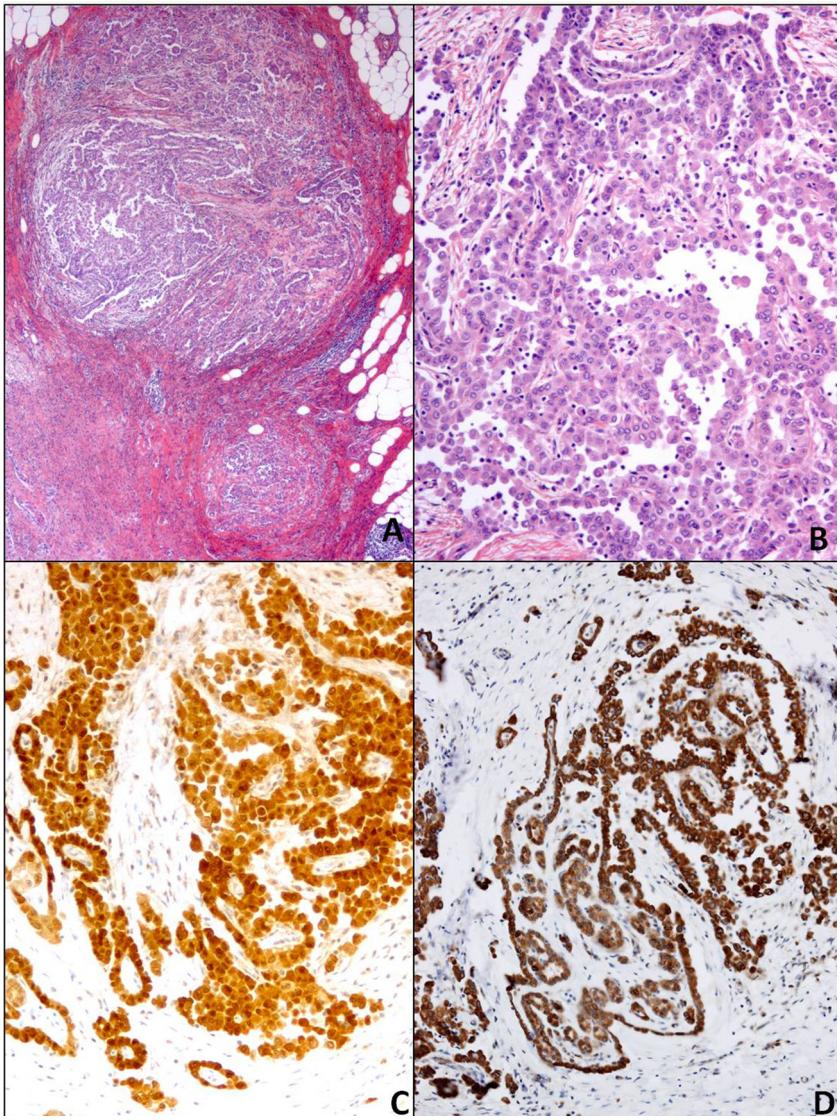

Fig. 1. Low-grade serous primary peritoneal carcinoma in the hernia sac. (A) Panoramic view of two micropapillary smoothly contoured clusters of serous epithelial cells in the superficial submesothelial space. (B) The papillae are lined by one layer of cuboidal epithelial cells with uniform cytologic appearance. Detached cells are present. The epithelial component is positive for calretinin (C) and claudin-4 (D).

low-grade. Mitoses or necrotic areas were absent (Fig. 2B). The surface of the uterus, tubes, and both ovaries showed extensive fibrosis. The ovaries did not show any neoplasm. Fallopian tubes showed no conspicuities. Endosalpingiosis was not observed. Chemotherapy or whole abdomen irradiation was not established.

A PET scan scintigraphy performed 22 months later showed increased metabolic activity in the right femoral region and in the cecum area. The patient was alive and well with stable residual disease. She had no adjuvant treatment.

Nine months later (63 months after herniorrhaphy), she presented with dull pain in the lower abdomen, ascites and increased CA 125 level. At laparotomy, an omental and peritoneal disseminated low-grade tumor was found. Macroscopic tumor nodules measured $\leq$2 cm in greatest dimension. Treatment consisted of cytoreductive surgery, removing the greater omentum, all peritoneal tumors, segments of small and large intestine, gall bladder, and spleen, combined with hyperthermic intraperitoneal perfusion with mitomycin. Microscopic examination disclosed confluence of tumor groups (Fig. 3A), glandular or cystic structures, branching papillae, small group and isolated cells with mild to moderate atypia (Fig. 3B), and invasive tumor clusters of solid architecture and infrequent mitoses (Fig. 3C). A figure suggestive of vascular invasion was not confirmed after staining with CD31, CD34, D2-40 and Verhoeff's elastic stain (Fig. 3D). Necrosis was absent. Cells showed vesicular nuclei with small patent nucleoli (Fig. 3C). Tumor cells were intensely positive for PAX8 (Fig. 4A), claudin-4 (Fig. 4B), VE1 (Fig. 4C), calretinin, D2-40, and WT1; and negative for PAX2, MOC-31, Ber-Ep4, estrogen receptor alpha, and progesterone receptor. There was heavy diffuse mononuclear inflammatory infiltrate with the presence of occasional lymphoid follicles. A retrospective study of the initial lesion in the hernia sac showed positivity for VE1 (Fig. 4D). Antibodies used in this case study are included in Table 1.

The tumor was definitely diagnosed as LGSPC including the initial lesion involving the hernia sac.

The patient suffered from a prolonged postoperative ileus that was resolved.

Six months later, she had residual disease and iron deficiency anemia that was treated with ferric carboxymaltose injection/iron sucrose injection. Residual disease persisted in a control conducted 6 months later (75 months after herniorrhaphy).