Pseudomyxoma peritonei - case report

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Summary

The syndrome pseudomyxoma peritonei is rare, present in only 2/10,000 laparotomies. We report the case of a 58-year-old woman with a primary tumor of the appendix, and secondary involvement of other structures and organs of the abdominal cavity. In our case, we performed maximal surgical reduction of the tumor, with remaining implants on diaphragmatic domes and liver, as we did not have technical conditions to safely perform prolonged surgery which would have included a surgical procedure on the liver and administration of intraoperative chemotherapy. The patient underwent six series of parenteral chemotherapy, but refused the second-look surgery. Even though our patient did not receive intraperitoneal chemotherapy, maximal surgical tumor reduction, and refused second-look surgery, she is still alive and without any major complaints two years after the surgery.

Key words: Pseudomyxoma peritonei; Appendix; Ovary; Diagnosis; Treatment.

Introduction

Pseudomyxoma peritonei is characterized by the presence of gelatinous ascites and mucinous implants on the peritoneum and omentum. It is present in 2/10,000 laparotomies and more often in women (ratio 3:1) [1]. In multiorgan involvement it is not possible to determine the organ of primary origin only by imaging and intraoperative findings. The most reliable method is immunohistological staining.

Case Report

A female patient (58 years old) came for a gynecological examination for lower abdominal discomfort. Endovaginal ultrasound (US) examination revealed an enlarged left ovary (diameter 55 mm) with tumor mass consisting of solid-cystic components and papillary proliferations suspicious for malignancy. There was ascites in the Douglas pouch. The uterus and right ovary had a normal morphology. During preparation for elective surgery, apart from standard laboratory blood and urine analysis, chest X-ray, ECG and internist examination, abdominal and pelvic computed tomography (CT) were performed. Moreover, blood concentrations of tumor marker CA 19.9 and CEA were elevated (CA 19.9: 263.4 IU/ml, CEA: 146.8 ng/ml). CT examination showed: 1) a left ovarian cystic mass with internal septa and solid components (size 32 x 55 x 48 mm); 2) moderate ascites in the Douglas pouch; 3) diffuse omental implants - omental caking; 4) a cystic mass (4 cm) with ring postcontrast enhancement in the ileocoecal region; and 5) subdiaphragmatic, perihepatic and perisplenic peritoneal implants. After preoperative preparation, surgery was performed. The operation revealed: 1) a mucinous malignant left ovarian tumor (ex tempore histopathology - malignant); 2) peritoneal tumor implants in the Douglas pouch, left paracolic region, bladder peritoneum; 3) tumor of the appendix; 4) diffuse great omentum carcinomatosis; and 5) gelatinous tumor tissue in the region of the diaphragm domes, spleen and liver, which indicated the syndrome of pseudomyxoma peritonei. During surgery total abdominal hysterectomy with bilateral salpingo-oophorectomy, resection of the bladder and Douglas pouch peritoneum with the removal of tumor implants, appendectomy, total omentectomy and splenectomy were performed. Postoperative recovery was satisfactory, and the patient was discharged ten days after the surgery. Histopathology examination included the following methods: paraffin blocks of specimen fixed in pulverized formalin were made, then cut to the thickness of 5 μm and finally dyed with the methods H&E, PAS, CK 7, CK 20, estrogen receptors (ER) and CA125. In the specimen, all organs showed the same biphasic histological appearance - part of the material

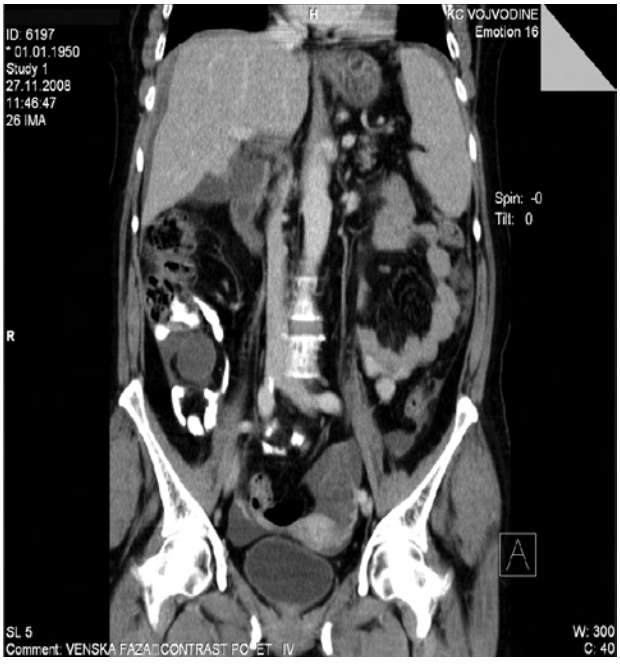

Figure 1. — Cystic appendiceal lesion, omental caking, perihepatic, perisplenic peritoneal implants and ascites, left ovarian cyst.

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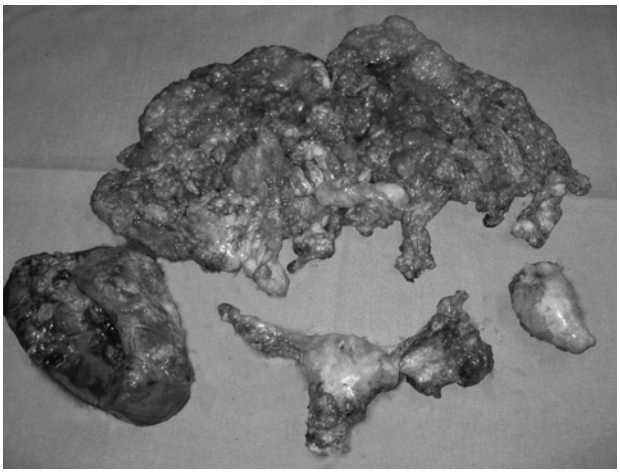

Figure 2. — Omentum, uterus, adnexa, spleen and appendix.

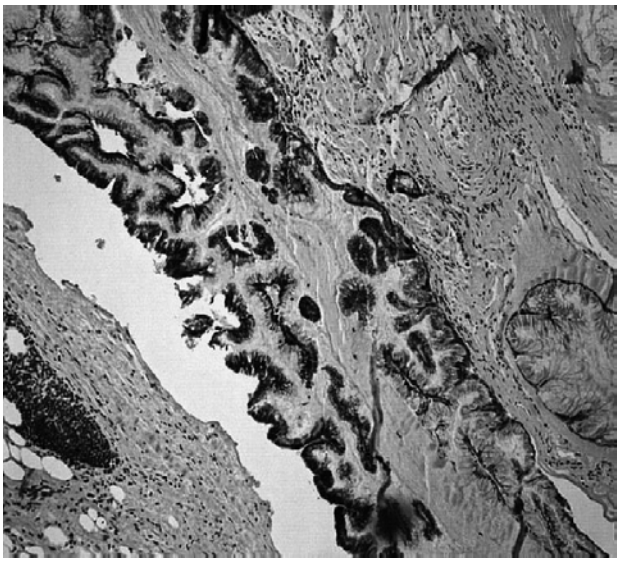

Figure 3. — CK20 strong positivity of epithelium.

had a benign mucinous character, while in adjacent parts stratified, clearly dysplastic malignant mucoproduction characteristic of epithelial cells with stromal infiltration by individual cells and smaller atypical adenoid formations were seen. In all regions of the tumor a significant production of immature mucus with the formation of bigger mucus lakes was depicted. The immunohistochemical profile showed epithelial membranous positivity to CK 20, while negativity was registered to CK 7, ER and CA125, based on which we concluded that the lesion was a primary tumor of the appendix with secondary involvement of other structures and organs of the abdominal cavity. The clinical term of peritoneal pseudomyxoma in pathological terms in our case became peritoneal mucinous carcinomatosis with intermediate or discordant features (PMCA-I/D). At oncological consilium, adjuvant chemotherapy was ordered (taxol and carboplatin - 6 series), with control of tumor markers CA 19.9 and CEA after the 3rd and 6th administration. Six months after the chemotherapy levels of CA 19.9 were 46.7 IU/ml and CEA was 11.3 ng/ml, while 12 months after the therapy CA 19.9 was 29.6 IU/ml while CEA 7.5 ng/ml. At control CT examinations after six and 12 months, persistant diffuse peritoneal implants were seen, without the presence of ascites. Two years after surgery, the patient is still alive and without major complaints.

Discussion

Mucinous lesions can be found in different locations in the body, but the most common are bowel (appendix) and ovarian tumors [2, 3]. These lesions can be seen in benign, intermediate and malignant variants, but the most serious complication is clinically called peritoneal pseudomyxoma. It is simple if the tumor lesion is limited to the single organ no matter if the pseudomyxoma has developed i.e., diagnosis of the localization of the primary neoplastic lesion is made easier. A basic problem arises when more organs, which can all have primary mucinous lesions, are involved, or in other words if, as in our patient, tumor with a substantial production of mucus has spread to many organs [4-6]. In our case, the left ovary, appendix, omentum and peritoneum were involved, while implants were also found in appendice epiploicae.

In such cases of multiple organ involvement, it is of a great importance to apply immunohistochemical methods of staining, mostly CK-7 and CK-20 which aid in differentiation of organs from which a mucoproductive lesion had arisen. Expression of CK-7 indicates that the lesion originates from ovarian tissue, while expression of CK20, i.e. membranous and cytoplasmatic positive reaction with antibodies to CK-20, indicates that the lesion originates from the digestive system [7]. In cases of pseudomyxoma peritonei syndrome maximal surgical reduction of tumor including total peritonectomy is performed. Additional intraperitoneal chemotherapy, which gives the best chances for success, is also administered. In such cases, appendectomy should always be performed, as the appendix in this syndrome is very often malignantly altered. From a histological point of view, several similar but different histological entities should be distinguished: disseminated peritoneal adenomucinosis (DPAM), peritoneal mucinous carcinomatosis (PMCA), and peritoneal mucinous carcinomatosis (PMCA-I/D) with a different microscopic appearance [8]. In our case, we performed maximal surgical reduction of the tumor, with remaining implants on the diaphragmatic domes and liver, as we did not have technical conditions to safely perform prolonged surgery which would have included a surgical procedure on the liver and administration of intraoperative chemotherapy. After completing parenteral chemotherapy and a control CT examination, second-look surgery should have been performed with the aim of reducing any eventual residual tumor tissue on the diaphragm domes and liver, as part of a protocol in such cases [9, 10]. The patient underwent six series of parenteral chemotherapy, but refused the second-look surgery. Even though our patient did not receive intraperitoneal chemotherapy and maximal surgical tumor reduction, she is still alive and without major complaints two years after the surgery. Moreover, control CT examinations have not shown any

signs of disease progression (peritoneal implants were the same in size and number, and no ascites was present). Regarding the fact that for our patient maximal tumor resection and second-look surgery were not performed, and that control CT showed no progression of the disease, while the general condition of the patient 24 months after the surgery was rather good, we expect that the prognosis of the disease in this case will correlate with the available references. Ronnett reports that the 5- and 10-year survival rates for all patients with carcinomatosis (PMCA-I/D+PMCA) are 26% and 9%, respectively. The mean and median survival times for all patients with carcinomatosis were 35 and 22 months [6].

References

[1] Djurdjevic S., Kesic V.: "Ginekoloska onkologija.Udruzenje za ginekolosku onkologiju Srbije (UGOS) i Medicinski fakultet Novi Sad". S*can Graphics Novi Sad*., 2009, 230.

[2] Mukherjee A., Moran B.: "Pseudomyxoma peritonei-symptoms, signs and clinical differential diagnosis". *CME J. Gynecol. Oncol*., 2003, *8,* 199.

[3] Smeenk R.M., Verwaal Vj, Zoetmulder F.A.: "Pseudomyxoma peritonei". *Cancer Treat. Rev*., 2007, *33,* 138.

[4] Ronnett B.M., Shmookler B.M., Diener-West M., Sugarbaker P.H., Kurman R.J.: "Immunohistochemical evidence supporting the appendical origin of pseudomyxoma peritonei in women". *Int. J. Gynecol. Pathol*., 1997, *16*, 1.

[5] Lee K.R., Scully R.E.: "Mucinous tumors of the ovary: a clinicopathologic study of 196 borderline tumors (of intestinal type) and carcinoma, including an evaluation of 11 cases with pseudomyxoma peritonei". *Am. J. Surg. Pathol*., 2000, *24,* 1447.

[6] Ronnett B.M.: "Pseudomyxoma peritonei: pathologic features, site of origin and prognosis". *CME J. Gynecol. Oncol*., 2003, *8,* 192.

[7] Guerrieri C., Franlund B., Fristedt S., Gillooley J.F., Boeryd B.: "Mucinous tumors of the vermiform appendix and ovary, and pseudomyxoma peritonei: histogenic implications of cytokeratin 7 expression". *Hum. Pathol*., 1997, *28,* 1039.

[8] Bryant J., Clegg A.J., Sidhu M.K., Brodin H., Davidson P.: "Systematic review of the Sugarbaker procedure for pseudomyxoma peritonei". *Br. J. Surg*., 2005, *92,* 153.

[9] Sugarbaker P. "Disseminated appendiceal mucinous tumors and the pseudomyxoma peritonei sydrome". *Gynecol. Oncol*., 2003, *8,* 204.

[10] Esquivel J., Sugarbaker P.H.: "Clinical presentation of the pseudomyxoma peritonei syndrome". *Br. J. Surg*., 2000, *87*, 1414.

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