Mucinous cystadenocarcinoma of the spleen – a very rare case of a primary splenic MCN

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ABSTRACT: Introduction: We present the case of a primary spleen mucinous cystadenocarcinoma. Several cases of this primary tumor of the spleen have been described worldwide so far. These tumors are classified as mucinous cystic neoplasms (MCN) and occur mainly in the ovaries and pancreas.

Case report: The case concerns a 45-year-old female patient with an accidentally diagnosed splenic tumor with approximately 20 cm in size. Histopathological examinations, following a splenectomy, confirmed the presence of mucinous cystadenocarcinoma.

KEYWORDS: cancer, cystic tumor, spleen

INTRODUCTION

Primary and metastatic tumors in the spleen are rare. Primary tumors are divided into lymphoid and non-lymphoid, and lymphoma and angiosarcoma are the most common [1]. In spite of this fact, splenic lymphomas constitute <1% of all lymphomas [2]. Splenic metastases occur in 2.3–7.1% of all cases [3].

We present the case of a malignant mucinous cystic neoplasm (MCN) of the spleen. Most MCN are in the ovaries and pancreas [4, 5]. They may also be located in the liver [6], the small bowel mesentery and in the appendix as well [7, 8].

Cases involving the spleen are extremely rare [5, 7, 9]. Cystic neoplasms may arise from the heterotopic pancreatic tissue of the spleen [10, 11]. So far, there have been several cases of mucinous cystadenocarcinoma of the spleen reported worldwide [12–15].

CASE REPORT

A 45-year-old female patient was admitted to the Barlicki Hospital in Lodz to the Department of General and Transplant Surgery due to an asymptomatic tumor of the spleen, detected by accident on an ultrasound examination. Based on the patient’s history, we established that the patient had no past history of neoplasms. She was operated on several years ago due to Meckel’s diverticulum, and the only comorbid diagnosis was hypertension.

Approx. a year earlier the patient suffered from abdominal trauma. Contrast-enhanced CT of the abdomen revealed a pathological mass of 174 x 109 x 180 mm in the left epigastrium described as a tumor of the spleen, Fig. 1A.

Before surgical treatment, a parasitic background (a negative result for IgG antibodies to tapeworm from the genus Echinococcus)
case report

In some cases, mucinous cystic adenomas present in places other than the ovaries, such as the pancreas, liver, peritoneal cavity, and spleen, contain ovarian-like stroma (OLS).

was excluded and pneumococcal vaccination was recommended. The patient was operated via the classical method using oblique incision in the left subcostal region. A significantly enlarged spleen with a tumor was found intraoperatively. After the splenic vessels were cut and ligated separately, splenectomy was performed. A drain was placed in the peritoneal cavity. The operation was performed without any complications. Incision of the spleen revealed a large tumor of 200 x 165 x 100 mm, occupying almost the entire organ and consisting of many mucinous cysts (Fig. 1B.). The postoperative period was uneventful. The drain was removed on the 2nd day. On day 5, the patient was discharged without complaints. The platelet blood count upon discharge day was 340,000/mm³ (before surgery 230,000/mm³).

Histopathological examination revealed cysts covered by a glandular epithelium, containing mucinous fluid, and separated by septa of connective tissue (Fig. 1C., 1D.). The Ki-67 proliferation index was 20%. CK7, CK20 and CEA antigens were found in immunoenzymatic tests. The WT1 antigen was negative. Low-grade mucinous cystadenocarcinoma was diagnosed. The blocks with histopathological preparations were re-analyzed by another pathologist from another center, who confirmed the primary diagnosis.

Postoperative recommendations included control of the platelet count in the following weeks, control of the postoperative wound at the Surgical Ambulatory and, due to the presence of a neoplasm, oncological consultation for possible adjuvant treatment and continuous care at the Oncology Clinic. Due to the nature of the tumor, the patient was also recommended to remain under the care of the ambulatory gynecology clinic and undergo PET examination in the postoperative period. The examination showed no other primary foci of MCN, especially in the ovaries.

DISCUSSION

Mucinous cystic neoplasms are most common in the ovaries. Benign adenomas represent about 10–15% of all benign ovarian masses, while mucinous ovarian cancer accounts for about 5% of all ovarian malignant neoplasms [19]. In the pancreas, MCN constitute about 2% of all tumors but about 33% of all cystic pancreatic tumors [18].

In some cases, mucinous cystic adenomas present in places other than the ovaries, such as the pancreas, liver, peritoneal cavity, and spleen, contain ovarian-like stroma (OLS).
This suggests the common origin of these tumors and explains the higher incidence of mucinous cystic neoplasms in younger women at locations other than the ovaries [5].

One study of 15 patients with liver MCN showed the presence of OLS in all female patients (n = 13). The authors conclude that these tumors arise from the epithelium covering the primary gonads in the early period of embryonic development, which are initially located under the diaphragm. The greater prevalence of MCN in women and embryological development of gonads explains the presence of OLS in cystic tumors of the pancreas, liver, spleen and peritoneal cavity [6]. In addition, MCN often contain receptors for estrogens and progesterone [20].

There have been several reports of mucinous cystic neoplasms in the spleen arising from heterotopic pancreatic tissue [10, 11]. Pancreatic heterotopy may explain the extraparenchymal locations of MCN such as colonic polyps [21].

It is noticeable that the diagnosis of ovarian mucinous cystic neoplasms often employs the CEA marker, whose elevated levels are found preoperatively in 88% of patients with ovarian cystic adenocarcinoma [19]. The case is different with the extraovarian location. Elevated CEA levels in malignant pancreatic MCN have very low sensitivity [22]. There is no data on the assessment of serum CEA levels in patients with MCN in the liver, spleen or other locations, although there have been some cases with elevated CEA levels [8].

REFERENCES

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