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A unique case of splenic tumor exhibiting a serous carcinoma phenotype

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Abstract

The spleen has no epithelial element; thus, primary carcinoma of the spleen is quite rare. We present the case of a patient with serous carcinoma of the spleen. A 76-year-old woman with no significant medical history presented with a huge lesion in the spleen. Except this lesion, clinical examination, including imaging examination, revealed no remarkable findings. She underwent excision of the spleen for treatment and diagnosis. Postoperative pathological examination revealed neoplastic cells with pleomorphic and hyperchromatic nuclei, prominent nucleoli, and frequent mitotic activity. The neoplastic cells exhibited a papillary pattern with psammoma bodies. Immunohistochemistry showed positivity for cytokeratin 7, PAX-8, WT-1, p16, p53, and Ber-EP4 and negativity for cytokeratin 20, thyroid transcription factor-1, carcinoembryonic antigen, CD10, estrogen receptor, calretinin, D2-40, intelectin-1, and sialylated HEG1. We inferred that this tumor was a primary splenic serous carcinoma. Serous tubal intraepithelial carcinoma is the plausible origin of most pelvic serous carcinomas. However, the origin of serous carcinoma of the spleen remains unknown. We speculated that endosalpingiosis might be the origin of the tumor.

Keywords

Spleen, splenic adenocarcinoma, serous carcinoma, immunohistochemistry, splenectomy

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Introduction

The spleen is believed to have no epithelial element, and most primary splenic malignancies have lymphatic or vascular origins, like lymphoma or angiosarcoma.[1] Few cases of primary splenic carcinoma have been reported, but their pathogeneses have been unknown. Herein, we report a case of serous carcinoma of the spleen and discuss its probable origin.

Case report

A 76-year-old woman with no significant medical history presented with a left upper abdominal mass at a routine medical checkup. Upper gastrointestinal endoscopy demonstrated extrinsic compression in the middle of the gastric body. Computed tomography and magnetic resonance imaging indicated a huge lesion in the spleen (Figure 1). There was no abnormal increase in glucose metabolism in parts other than the spleen according to whole-body ^{18}F-fluorodeoxyglucose positron emission tomography (Figure 2). No other abnormality was revealed on clinical examination; thus, the lesion was thought to be a primary splenic tumor, such as an angiosarcoma. She underwent excision of the spleen for treatment and diagnosis.

The tumor was a white-gray, well-defined mass containing necrotic areas and measured approximately 130 × 120 × 90 mm with an expansive growth pattern (Figure 3). It showed infiltration into the surrounding lymph nodes, left lung, diaphragm, and stomach. Histopathological examination of the resected tissue specimens showed pleomorphic and

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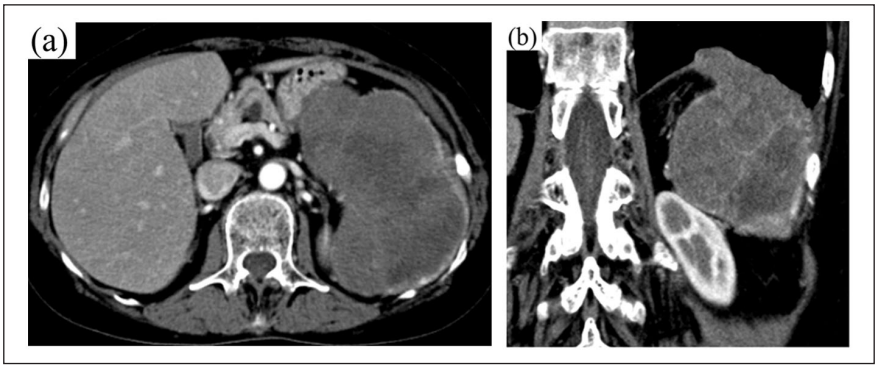

Figure 1. Contrast-enhanced computed tomography showing a lobulated hypodense mass in the spleen: (a) transaxial image; (b) coronal image.

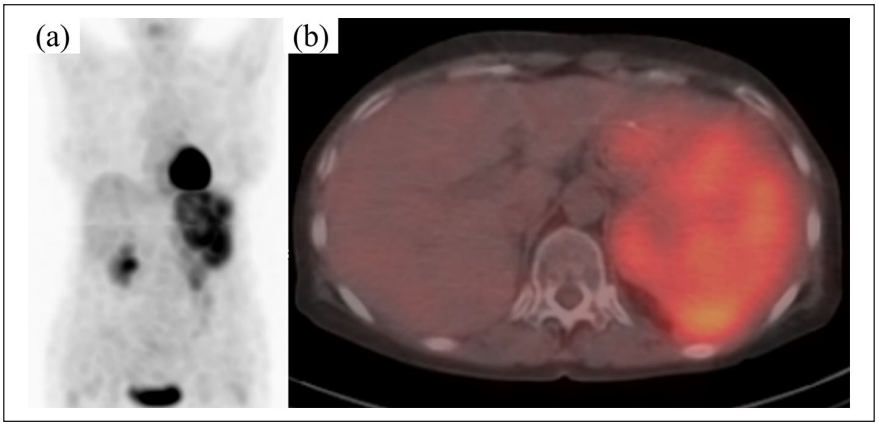

Figure 2. (a) Whole-body ^{18}F-fluorodeoxyglucose positron emission tomography demonstrating high tracer accumulation only in the spleen. (b) Fused positron emission tomography/computed tomography images of the cross section of the spleen showed hot spot (maximum standard uptake value = 5.5).

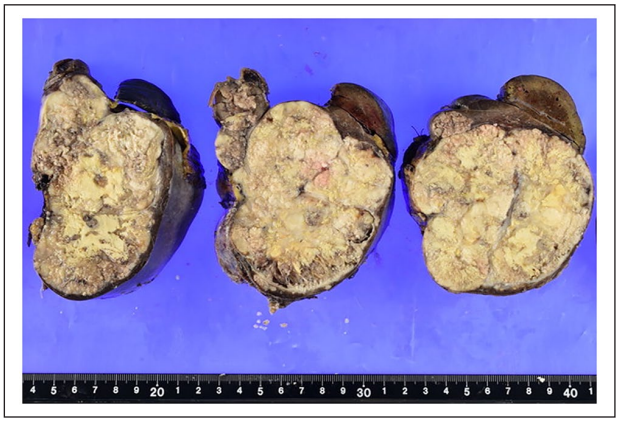

Figure 3. Macroscopic view of the resected spleen. A white-gray, well-defined mass containing necrotic areas, 130 × 120 × 90 mm in size, showing an expansive growth pattern in the spleen.

hyperchromatic nuclei, prominent nucleoli, and frequent mitotic activity. The neoplastic cells exhibited a papillary pattern with psammoma bodies. The representative features are shown in Figure 4. On immunohistochemical analysis, the tumor cells were positive for cytokeratin (CK) 7 (Figure 5(a)), PAX-8 (Figure 5(c)), WT-1 (Figure 5(d)), p16, p53 (Figure 5(e)), and Ber-EP4 (Figure 5(f)) but negative for CK20 (Figure 5(b)), thyroid transcription factor-1, carcinoembryonic antigen, CD10, estrogen receptor, calretinin, D2-40, intelectin-1, and sialylated HEG1. Based on these findings, we diagnosed the tumor as a serous carcinoma.

Discussion

Splenic tumors account for only 0.03% of all tumors types in human.[2] Typically, primary malignant neoplasms of the spleen are lymphomas arising from the white pulp and angiosarcomas arising from the red pulp.[1] One of the differential diagnoses in this case was epithelioid-type malignant mesothelioma, but neoplastic cells showed a papillary pattern with multiple psammoma bodies and showed pleomorphic nuclei and frequent mitotic activity. Furthermore, immunohistochemistry showed that the tumor cells were positive for Ber-EP4, PAX-8, and WT-1 but negative for calretinin,

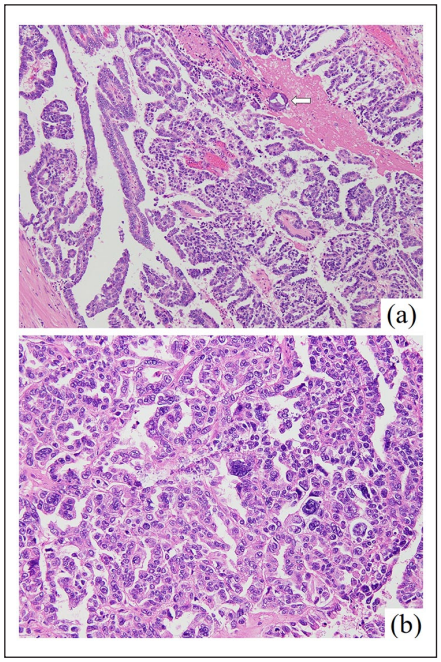

Figure 4. Hematoxylin and eosin staining of the splenic tumor neoplastic cells showing a papillary pattern with necrotic tissue and a psammoma body (arrow) (a, ×100). Pleomorphic and hyperchromatic nuclei were observed (b, ×200).

D2-40, intelectin-1, and sialylated HEG1. These findings could help distinguish serous carcinoma from epithelioid-type malignant mesothelioma.[3]

The spleen has no epithelial element or afferent lymphatics;[4] most splenic carcinomas are considered to be hematogenous metastases from other organs.[1] The main primary sites are the lung (21%), stomach (16%), pancreas (12%), liver (10%), colon (9%), esophagus (8%), nasopharynx (6%), breast (4%), and ovary (4%).[4] Based on the immunohistochemical findings of positivity for PAX-8 in this tumor, we should have considered splenic metastasis from uterine corpus, ovarian, fallopian tubal, or peritoneal serous carcinoma.[5] However, apart from the lesion in the spleen, no remarkable findings of tumors in the uterine corpus, ovary, fallopian tubes, or peritoneum were observed intraoperatively, on clinical examination, or on imaging. Besides, the patient had no family history of ovarian or breast cancer. Salpingectomy and adjuvant chemotherapy as in ovarian, fallopian tubal, and peritoneal serous carcinoma were considered, but strict follow-up alone was decided upon considering the patient's wish. The patient has remained disease free after 8 months without undergoing adjuvant chemotherapy or radiotherapy. Thus, we finally inferred that the tumor was a primary serous carcinoma of the spleen. Primary serous carcinomas of the spleen are extremely rare, and we

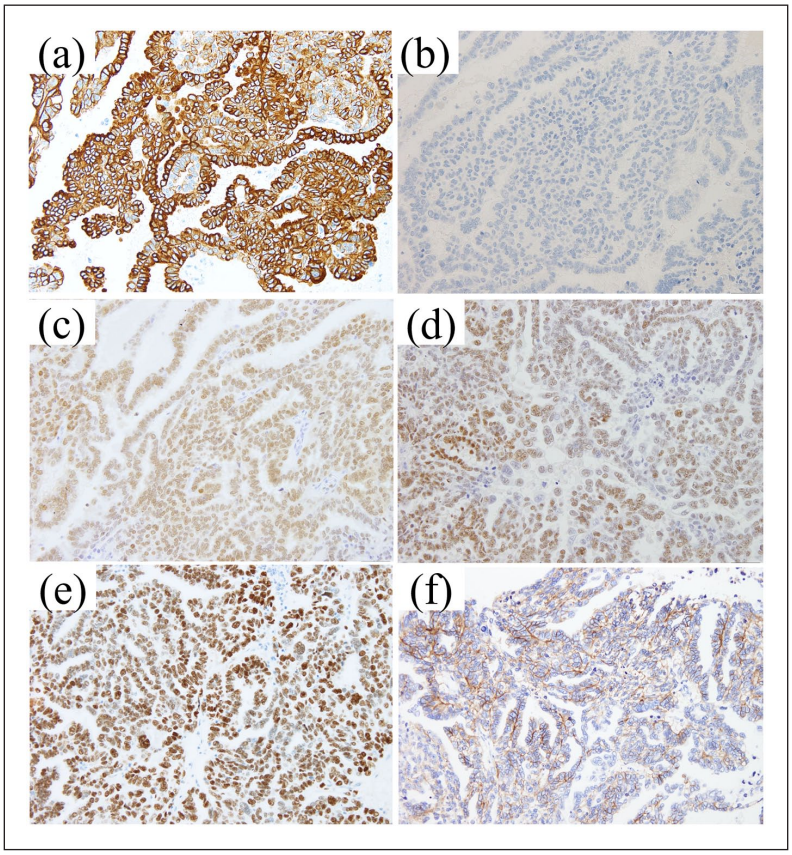

Figure 5. Immunohistochemical staining of the tumor cells for cytokeratin 7 (a, ×200), cytokeratin 20 (b, ×200), PAX-8 (c, ×200), WT-1 (d, ×200), p53 (e, ×200), and Ber-EP4 (f, ×200).

could find only one reported case.[6] Serous tubal intraepithelial carcinoma is the plausible origin of most pelvic serous carcinomas.[7,8] However, the origin of splenic serous carcinoma remains unknown.[6] In our case, metastasis from latent serous tubal intraepithelial carcinoma could not be ruled out, but it was possible that the tumor developed from an ectopic fallopian tube-like epithelium, that is, endosalpingiosis. Endosalpingiosis is a müllerianosis and is frequently observed in organs such as the lymph nodes, ovaries, or peritoneum where müllerian epithelium is normally absent.[9] Few cases of splenic endosalpingiosis have been reported in the literature.[10–12] According to some reports, mesothelial cells have the potential to differentiate into fallopian tube-like epithelium, and splenic endosalpingiosis arises from the invaginated splenic capsule or peritoneum.[6,10,12] Another hypothesis is that aberrant remnants of the proximal portion of the müllerian ducts develop into fallopian tube-like epithelium.[9,10] The term "secondary müllerian system" is used to explain these theories,[9,10] but the mechanism remains unproven. The secondary müllerian system can be a clue to reveal the origin of this tumor.

In conclusion, we reported a unique case of splenic tumor exhibiting a serous carcinoma phenotype. In this case, the secondary müllerian system and endosalpingiosis are likely to be related to the occurrence of the tumor. The accumulation of similar cases can be the key to unraveling the pathogenesis of pelvic and extrapelvic serous carcinomas.

Declaration of conflicting interests

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Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

Informed consent

Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

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References

1. Kaza RK, Azar S, Al-Hawary MM, et al. Primary and secondary neoplasms of the spleen. *Cancer Imaging* 2010; 10: 173–182.
2. Jiang Z, Ma F and Liu X. Squamous cell carcinoma of the spleen: a case report. *Oncol Lett* 2018; 16: 3973–3975.
3. Baker PM, Clement PB and Young RH. Malignant peritoneal mesothelioma in women: a study of 75 cases with emphasis on their morphologic spectrum and differential diagnosis. *Am J Clin Pathol* 2005; 123(5): 724–737.
4. Lam KY and Tang V. Metastatic tumors to the spleen: a 25-year clinicopathologic study. *Arch Pathol Lab Med* 2000; 124: 526–530.
5. Tong GX, Devaraj K, Hamele-Bena D, et al. PAX8: a marker for carcinoma of Mullerian Origin in serous effusions. *Diagn Cytopathol* 2011; 39: 567–574.
6. Chou CK and Chou SC. Serous cystadenocarcinoma of the spleen. *Radiol Case Rep* 2017; 12: 273–277.
7. Przybycin CG, Kurman RJ, Ronnett BM, et al. Are all pelvic (nonuterine) serous carcinoma of tubal origin? *Am J Surg Pathol* 2010; 34: 1407–1416.
8. Kindelberger DW, Lee Y, Miron A, et al. Intraepithelial carcinoma of the fimbria and pelvic serous carcinoma: evidence for a causal relationship. *Am J Surg Pathol* 2007; 31(2): 161–169.
9. Dubeau L. The cell of origin of ovarian epithelial tumors and the ovarian surface epithelium dogma: does the emperor have no clothes? *Gynecol Oncol* 1999; 72(3): 437–442.
10. Tanahashi J, Kashima K, Daa T, et al. Florid cystic endosalpingiosis of the spleen. *APMIS* 2006; 114(5): 393–398.
11. Nguyen BD and McCullough AE. Gastrointestinal: cystic endosalpingiosis of the spleen: CT, MR, and US imaging. *J Gastroenterol Hepatol* 2017; 32(12): 1911.
12. Lee YS and Teh M. Histogenesis of true splenic cysts: a histological and immunohistochemical study. *Ann Acad Med Singap* 1993; 22(3): 372–376.