Splenic Lymphangioma
- A Report of Three Cases -

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Lymphangioma is a benign neoplasm most commonly involving the neck and axilla. They can occur sporadically in the mediastinum, retroperitoneum, and internal organs.1 Splenic lymphangioma is a very rare condition and is usually found incidentally. A few cases with abdominal pain, abdominal mass, or increasing abdominal girth have been presented.2 Although the presence of a lymphangioma is well known, the histogenesis is controversial whether it is a true neoplasm or not. Because lymphangioma usually occurs in children and can be presented as a multicentric lesion, the possibility of developmental failure has been suggested.3,4 We report three cases of splenic lymphangioma in elderly women, and discuss the differential diagnoses.

CASE REPORT

Case 1

A 71-year-old female with an incidentally found splenic mass is presented. Computed tomography showed a multicystic splenic mass with peripheral rim calcification (Fig. 1A). A splenectomy was done, and the enlarged spleen weighed 208 g and showed a $10 \times 8 \times 4$ cm sized well demarcated multicystic mass with variable-sized cysts separated by thick fibrous walls with focal calcification (Fig. 1B). Histologically, the cysts were lined by flat endothelial cells and filled with acellular pinkish fluid. The cyst walls mainly consisted of hypocellular fibrous tissue with occasional smooth muscle components. The walls were shared by neighboring cysts, or intervened by interspersed splenic parenchymal components. On immunohistochemical stainings, the lining cells were diffusely positive for CD31 and factor VIII-related antigen (FVIII-RAg), while focally positive for CD34 in all cases.

Case 2

A 50-year-old female who had an abdominal distension for 6
months is presented. Computed tomography showed a huge pelvic cystic mass with thick irregular septation and solid portion and a separate cystic mass in the spleen. A radical hysterectomy with splenectomy was done. Grossly, the left ovary showed a 25 × 22 × 10 cm sized mass and the right ovary showed a 1.2 × 1 × 0.5 cm sized mass. The spleen, weighing 120 g, showed a 3.5 × 3 × 3 cm sized well demarcated multiseptate cystic mass (Fig. 2A). Histologically, both ovarian masses were mucinous cystadenocarcinomas. The splenic cysts were lined by flat endothelial cells and filled with acellular pinkish fluid. The cyst walls consisted of fibrous tissue with intervening splenic parenchyma (Fig. 2B). The lining endothelial cells showed similar immunoprofiles comparable to those of Case 1 (Fig. 2C).

Fig. 1. (A) Computed tomography of case 1 reveals a multicystic splenic mass with peripheral rim calcification, measuring 10 × 8 × 4 cm. (B) The enlarged spleen in case 1 shows a well-demarcated multiseptate cyst occupying subtotal splenic parenchyma. (C) The cysts consist of fibrous wall shared by neighboring cysts or intervened by splenic parenchyma, and flat endothelial linings. (D) The lining cells show diffuse positivity for CD31 on immunostaining.
Case 3

A 46-year-old female who had an epigastric pain for 1 month is presented. Ultrasonography and computed tomography showed a large multilocular cystic mass in the spleen. A splenectomy was done, and the enlarged spleen weighed 510 g, containing a 10 × 9.5 × 7 cm sized well demarcated multicystic mass. Histologically, the cysts were lined by flat endothelial cells and filled with acellular proteinaceous fluid. The cyst walls mainly consisted of hypocellular fibrous tissue with occasional calcification. The cyst walls were shared by neighboring cysts, or intervened by interspersed splenic parenchymal components as in the former cases. The lining endothelial cells showed similar immunoprofiles to those of Case 1.

DISCUSSION

Most of non-parasitic benign cystic lesions of the spleen are posttraumatic pseudocysts. The remainder of cysts are true cystic tumors which include hemangioma, lymphangioma, epidermoid and dermoid cysts. Hemangiomas are the most com-
Splen, and dermoid cysts are the least common. Lymphangiomas of the spleen may occur as a solitary lesion or may be presented as a part of lymphangiomatosis.

Splenic lymphangioma is a benign tumor or tumor-like hamartoma which is composed of a cavernous or cystic vascular formation of lymph vessel origin. Opinions regarding to the histogenesis of splenic lymphangiomas vary (i.e. opinions vary from hamartoma, true neoplasm, posttraumatic lesion, to persistent developmental defect), and a conclusive consensus has not been achieved. Incidences are quite rare and about one hundred cases have been reported in the world and only a few cases have been reported in this country.

The diagnostic differential includes hemangioma. Distinguishing lymphangioma from hemangioma is not always easy. Unlike the random localization of hemangioma, lymphangioma often involves the subcapsular area or larger trabeculae of the spleen, where lymphatics are normally concentrated. Also, the endothelium-lined spaces are filled with eosinophilic proteinaceous material instead of blood. Angiograms can be used to differentiate benign tumors of the spleen. The most common finding of the lymphangioma is avascular mass in the spleen. Immunohistochemical stainings should be necessary for the recognition of lining endothelial cells. Splenic mesothelial cysts also should be included in the differential diagnosis. Mesothelial cysts present subcapsular multicystic proliferations lined by a single layer of cells. Immunohistochemically, the lining cells are positive for keratin and HBME-1 but negative for FVIII-RAg, CD31, and CD34, supporting a mesothelial origin of the lining cells.

Splen, lymphangioma can be presented as an incidental finding or a symptomatic lesion. Most of these cysts are large and require splenectomy for the patient to avoid various complications associated with such lesions. Complications of splenic cysts include rupture with peritonitis, rupture with invasive hemorrhage, infec-

REFERENCE