Cavernous Lymphangioma of the Spleen
— A case report —

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Introduction

Lymphangiomas are benign congenital malformations of the lymphatic system usually affecting the neck and axilla\(^1\). Involvement of the visceral organ is less common\(^2\). Splenic involvement is quite rare.

Approximately 90 cases have been reported\(^6\). The lesion is characterized by a single or classically by multiple macroscopic and microscopic cysts\(^3\).

We report a case of splenic lymphangioma in which CT appearance suggested the correct diagnosis.

Case report

A 52 year old women admitted with seven month's history of a large left upper quadrant mass and epigastric pain. Egg sized mass was initially developed after first delivery 30 years ago. This mass has progressively grown during this interval.

Physical examination revealed a large mass in left upper quadrant which was hard, slightly tender and fixed. Ultrasonogram revealed huge echogenic mass with multiple small, moderate and large sized anechoic rounded lesions(Fig. 1). CT demonstrated marked splenomegaly in which multiple small, moderate and large sized low density changes were seen. Several small calcifications were also noted within cysts(Fig. 2.A, B.)

Splenectomy was performed. Gross specimen re-
Fig. 1. Ultrasonogram
Longitudinal sonogram reveals huge echogenic mass in the spleen with multiple small, moderate and large sized anechoic rounded areas.

revealed huge spleen which measured $24 \times 14 \times 10$ cm and weighted 1780 gm.

The histologic diagnosis was cavernous lymphangioma (Fig. 3)

Discussion

Lymphangioma of the spleen was first reported by Fink in 1885[5]. Fowler reported 27 cases in his review of splenic tumors in 1953[5]. In 1981, pyatt reported 90 cases of splenic lymphangioma in the literature review[6]. Clinical symptoms are usually nonspecific due to compression of adjacent viscera. Pain, discomfort and mass sensation are most common. Pathologically, lymphangioma are classified as simple, cavernous and cystic. The simple type is composed of capillary-sized, thin walled channels, the cavernous type consists of dilated lymphatic channels often with fibrous adventitial coats, and the cystic type is composed of cysts ranging from a few millimeters to several centimeters in diameter. The space must be lined by flattened endothelial cells or the diagnosis is suspect. Even when a large cyst is present, there often are

Fig. 2 CT Scan
a. Higher level CT scan. CT reveals multiple small, moderate and large sized cystic lesions in the spleen with scattered calcifications.
b. Lower level CT scan. CT reveals marked splenomegaly with multiple variable-sized cystic lesions scattered in the entire spleen.

Fig. 3. Photomicrograph of the tumor reveals large, cavernous spaces lined by flattened or attenuated endothelial cells and filled with lymph fluid. The cavernous spaces are separated by thin or slightly thick fibrous tissue with a few areas of remnant splenic tissue (H & E, ×100).
other small cysts in the splenic parenchyma or in the subcapsular region\(^3\). Plain abdomen film may disclose splenomegaly, curvilinear calcification in the cyst wall, and mass effect upon adjacent viscer\(a\)\(^7\).

Radionuclide scans show splenomegaly with multiple focal defects\(^9\). Angiographic findings include well defined avascular lesions scattered throughout the spleen with absence of neovascularity, arteriovenous shunting, and venous pooling. A classic “Swiss-cheese appearance” has been described\(^9\). Sonogram may reveal the presence of splenomegaly.

Depending on the size and macroscopic morphology of the spleen, its sonographic appearance may vary. A single well defined cyst similar to a post-traumatic cyst can be seen. In diffuse lymphangioma multiple variably sized sonolucent areas are depicted.

CT will show single or multiple low density cystic areas, some of which may be subcapsular in location, and deform the splenic contour. CT numbers range from 15 to 33 HU, depending mostly on the proteinous content of the fluid. The thin wall and sharp margination the lesion should suggest lymphangioma rather than other neoplasms which may be of low density due to necrosis or lipid content\(^5,11,12\).

The differential diagnosis of multifocal splenic disease is extensive and includes lymphoma, infarction, splenic emboli, metastasis (melanoma, breast, ovary and the lung), and splenic cyst. Determination of the cystic nature of the lesions is extremely helpful and narrows the differential diagnosis considerably. The clinical classification of splenic cysts by Pearl and Nassar is useful\(^3\): the three categories are\(^1\) parasitic cyst (taenia echinococcus)\(^2\), primary cyst with true cellular lining, such as those that are endothelial-lined (hemangiom, lymphangioma) and those that are epitheli-lined (dermoid, epidermoid) and secondary or traumatic cysts without a true cellular lining.

The presence of thin walled, well defined, nonehancing splenic cysts with contains mural califications may suggest the diagnosis of lymphangioma.

REFERENCES

10. 김재현, 원정연, 김영순 둥=임파판종의 초음파 소견, 대한방사선의학회지 21=969-974, 1985
11. 김순용, 임재훈, 고영태 둥=유마액을 함유하는 장간막임파종 1례. 대한방사선학회 20=148-151, 1984