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A Case of Giant Cystic Lymphangioma of the Retroperitoneal Origin

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Lymphangioma is the rarest of tumors in the retroperitoneal region. A reviw of the medical literatures reveals reports of not more than 23 cases of cavernous and cystic lymphangioma until 1961. We experienced a female case, whose initial clinical diagnosis was a cystic tumor of pancreatic origin, which was finally diagnosed as a giant cystic lymphangioma of the retroperitoneal origin on histological examination. We are presenting this case in regard to the rarity of the lesion.

Report of a Case

A 27 year old house wife first noticed a tumor in the left upper quadrant of the abdomen in August 1962, approximately two weeks after her first normal delivery. She visited the out-patient clinic of Sapporo Medical College Hospital for a thorough medical survey concerning the abdominal mass because of its gradual increase in size.

Routine check-up during the last pragnancy showed no abnormality, including negative urinalysis and normal blood pressure values.

The major subjective symptom at the time of admission to our clinic was an intermittent pain in the left upper quadrant of the abdomen felt mainly by activity. There were, however, no gastrointestinal symptoms, chill nor fever.

Family history: The patient's paternal grandfather died of gastric carcionma and maternal grandmother died of cerebral hemorrhage. Otherwise not contributory.

Patient's past history: She enjoyed full health with no remarkable illness in the past.

Physical examination: The patient was a well developed and well nourished woman in no acute distress. Pulse rate was 80 per minute, full and regular. Blood pressure was 134/80 mm Hg. Conjunctiva was neither anemic nor icteric. Cardiovascular and pulmonary systems appeared essentially normal. Upper abdominal wall was slightly distended and a palm-sized elastic soft tumor was palpable in the left subcostal area. The surface was smooth, the edge was dull and the tumor moved vertically at erect position and protruded downward to the umbilicus. Medially it extended approximately three fingerbreadth across midline at right recumbent position. The tumor also showed fluctuation to fillip and pressure.

The gastrointestinal roentogenographic studies were undertaken soon after admission to identify the location of the tumor mass. The upper gastrointestinol tract including the stomach and entire portions of the duodenum showed marked displacement toward

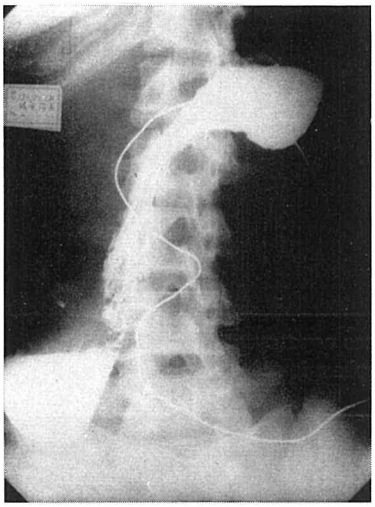

Fig. 1 Barium study roentgenogram of upper gastroinestinal tract showing marked diplacement of the stomach and duodenum due to the tumor mass.

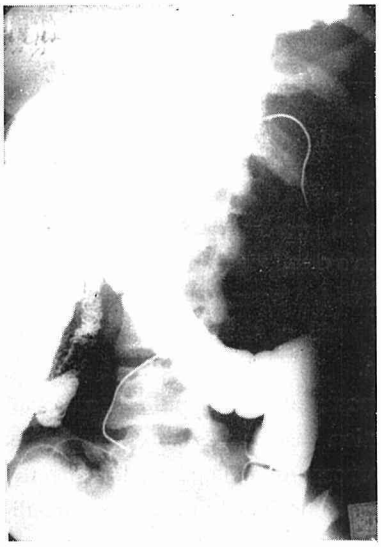

Fig. 2 Fluuroscopic examination of the large bowel with barium enema showing marked displacement of the splenic flexure of the colon by the tumor mass.

Laboratory Tests

Blood Picture	
Erythrocytes	429×10^4
Hemoglobin	86%
Color Index	1.0
Hematocrit	41
Leucocytes	3,900
Seg 77, Eos 5, Mon 4, Lym 14,	
Thrombocytes	143,000

Urinalysis
Light yellowish clear, acid,
Protein (−), Sugar (−),
Urobilinogen (−), Bilirubin (−),

P S. P.	
15 min.	50%
30 min.	20%
60 min.	20%
120 min.	0%

Serum Wassermann Reaction (−),

Serum Electrolytes
Na 151.3 mEq/l, K 5.6 mEq/l,
Ca 4.1 mEq/L, Cl 103.0 mEq/l,

Stool	
Benzidine	(+) (−) (+) (+) (−)
Pyramidon	(−) (−) (−) (−) (−)
Parasite Ova	(−)

Liver Function Study	
Icterus Index	6
Takata Ara's Reaction	(−)
Cobalt Reaction	R_6 (2)
T. T. T.	6
Z. T. T.	14
B. S. P. 30 min.	0%

Serum Electrophoresis	
Total Protein 8.0 g/dl, A/G ratio 1.13	
Albumin	52.8%
Alpha-1 glob.	4.9 »
Alpha-2 glob.	10.6 »
Beta-glob.	11.3 »
Gamma-glob.	20.4 »

Serum Amylase	8 unit
Urine Amylase	16 »

the right due to the left abdominal mass (Fig 1). Fluoroscopic exmination of the large bowel with barium enema also disclosed fronto-caudal displacement of the transversal colon and of oral half of the descending colon to make detour around the central part of the tumor (Fig. 2).

Urological studies with intravenous pyelography and pneumoretro-peritoneography demonstrated incomplete duplication of the left renal pelvis and ureter. However, no other abnormality such as displacement or compression of the kidney with the tumor mass was found. On the 6th week of hospitalization, the patient was seen by gynecologist, who denied relation of the mass with reproductive organs. The possibility of the splenic tumor also could be ruled out by the cystic consistency of the tumor, negative adrenalin test and normal blood and bone marrow pictures with no signs of hypersplenism. Serum and urine amylase activities determined with Wohlgemueth method were 8 and 16 units, respectively. The patient was transferred to surgical ward for an exploratory laparotomy on the 46th day of admission.

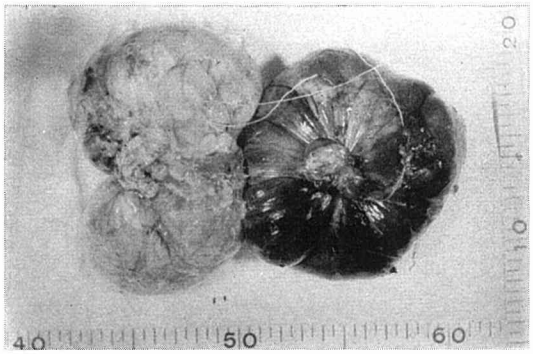

Fig. 3 Gross appearance of excised cystic lymphangioma.

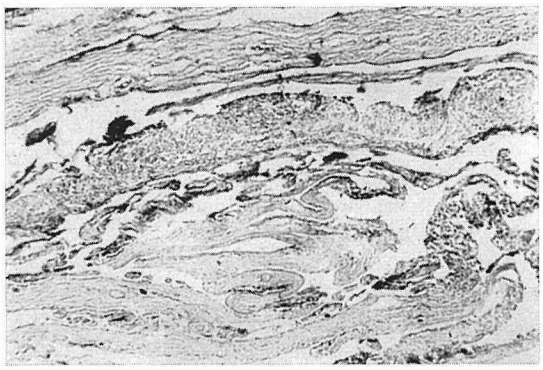

Fig. 4 Hematoxylin-Eosin stain of excised tumor tissue (×100).

Laparotomy findings: The median incision has exposed two huge multilocular cystic masses with sizes of a new born baby's head and of a hen's egg, respectively, which were densely adherent to the medial surface of the spleen and which seemed to originate from the vicinity of the tail of the pancreas (Fig. 3). Serous fluid of the cysts was partially aspirated and the tumor was removed by blunt dissection.

The cystic masses weighed 650 g in total, were composed of thin membraneous wall, and contained yellowish clear fluid. Histological examination disclosed scanty inner space with dilataion of the wall, which microscopically was composed of collagenized connective tissues with numerous lymphocytes and lymphoid follicles. Endothelial lining was partially found on the inner surface of the wall, and among these structures, there were some scattered adipose tissues. The diagnosis was cystic lymphangioma (Fig. 4).

Comment

Retroperitoneal lymphangioma was first reported by Sarway[1] in In 1898. In a review of English literatures, Rauch[2] found 22 cases of the retroperitoneal lymphangioma reported before 1959, of which 18 cases were cystic and 4 were cavernous lymphangioma including

two cases of his own. Barnett[4] added his own case in 1960.

According to Wegner's[4] classification of lymphangiomata, they are divided into 1) simple lymphangioma, 2) cavernous lymphangioma, and 3) cystic lymphangioma, most of which usually originate in the skin, neck, breast, axilla, lower sacral region, thighs and inguinal regions as hygromata[5].

The incidence of intraabdominal cystic lymphangioma, however, is extremely low and among others, retroperitoneal cystic lymphangioma is much rarer than those of mesenteric origin. Location of retroperitoneal lymphangiomata reported in the literatures were as follows; 8 in the right lumbar area, 3 in the left lumbar area and 3 in the region of the head of pancreas. The only case located in the left upper abdomen was that of Barnett. It is interesting to note that the symptoms and signs as well as the clinical course of Barnett's case closely resembled to those of the present case.

Cystic lymphangioma is composed of single or multiple cysts of various sizes containing serous fluid, with practically no connections with adjacent normal lymphatics. Microscopically this type of cysts can be identified from cysts of other type by existence of endothelial lining of the wall with lymphatic space, and abundunt lymphoid tissues found as diffuse collection of lymphocytes or in configuration resembling lymph nodes. According to Barnett, the lining endothelium must not be either cuboidal or columnar endothelium.

Brandsburg enumerated the following four theories on the origin of the lymphangioma: 1) the retention theory, which explains lymphangioma on the basis of mechanical pressure, 2) the disturtance of the secretory function of the endothelium of the lymph vessels, causing hyperproliferation of the lymph, or a disturbance of the permeability of the endothelium, 3) the inflammatory theory, and 4) embryonal theory. Sabin[3] also explained the etiology of lymphangioma on the basis of developmental arrest of the embryonal lymphatic system. The concomittance of the congenital anomaly[6,7] such as incomplete duplication of the left renal pelvis and ureter in the present case may also favour the possibility of the embryonal theory for the causation of the retroperitoneal cystic lymphangioma.

There are no clinical features which serve to differentiate this disease form other abdominal cysts. Patient is aware of this condition by a slowly enlarging soft abdominal mass with or without dragging-like pain. In contrast with lymphatic cysts of mesentery[8,9] which often give rise to acute abdominal symptoms, such as acute intestinal obstruction, and torsion of the intestines accompanied by severe abdominal pain, retroperitoneal lymphangioma rarely presents acute abdominal symptoms. Rauch reported symptoms of partial intestinal obstruction in 40 per cent of the previously reported cases.

As to the prognosis of retroperitoneal lymphangioma, there is no report of the malignant degeneration of this tumor and complete healing is obtained by surgical resection.

Summary

A case of cystic lymphangioma of the retroperitoneal region was presented. Giant cystic mass, which was found in the left upper quadrant of the abdomen, exhibited particular roentogenologic signs simulating a large cyst arising from the tail of the pancreas,

and was subsequently diagnosed as cystic lymphangioma upon histological examination. Presence of another congenital anomaly, duplication of the left renal pelvis and ureter, seemed to favour the embryonal theory for the causation of retroperitoneal cystic lymphangioma.

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