

Retroperitoneal Lymphangiomyomatosis with Lymphedema of the Legs

Rikiya Abe, M.D., Michio Kimura, M.D.*, Akio Hirotsaki, M.D.*, Hiroshi Ishii, M.D., Takashi Nakamura, M.D.*, Morio Kasai, M.D.*, Haruki Wakasa, M.D.***, Hiroo Tateno, M.D.**

*Department of Surgery, **Department of Pathology, Tohoku University School of Medicine, Sendai, Japan

Summary

Retroperitoneal tumor is demonstrated by lymphangiography in a 28 year old woman with lymphedema of both legs. Retroperitoneal tumor was subtotally extirpated, with subsequent subsidence of the swelling of both legs. According to the pathologic examinations of the extirpated specimens, lymphangiomyomatosis was confirmed. This is the first case reported in the literature of lymphangiomyomatosis presenting with lymphedema of the leg in the absence of pulmonary involvement, pleural or abdominal effusion.

Lymphangiomyomatosis is a disease of the lymphatic system within the lung and near the skeletal axis, which is clinically manifested by progressive dyspnea and frequently accompanied with pleural and abdominal chylous effusions. The most frequent symptom of lymphangiomyomatosis is, therefore, related to the lung. It is usually progressive and unresponsive to surgery, chemotherapy or radiotherapy. Histologically, however, lymphangiomyomatosis is a benign disease which occurs exclusively in women. The most prominent pathologic finding is the proliferation of atypical smooth muscle, most frequently within the wall of the thoracic duct, but also within lymph vessels and lymph nodes in the mediastinal and retroperitoneal spaces.

Vazquez (12) reported the 58th case of lymphangiomyomatosis with pulmonary involvement in 1976. Some authors (6, 12) have reported lymphedema of the leg during the late stage of lymphangiomyomatosis. In the present case, symptoms of the chest were not observed,

and the first complaint was the swelling of the right leg. From the clinical view point this complaint without common symptoms of lymphangiomyomatosis makes our case unique among other reported cases.

Report of a Case

A 28 year old Japanese woman was admitted to the Department of Surgery, Tohoku University Hospital on January 1978 with a chief complaint of swelling of both legs. The patient first noticed the swelling of the right leg at the age of 26. For the duration of pregnancy at the age 27, the swelling of the leg subsided almost entirely, but swelling of both thighs gradually increased after delivery in July 1977. No history of hereditary or familial disease was given by the patient.

Positive physical findings at the present admission were limited to both legs. The right leg was entirely swollen and the left thigh was also swollen (Fig. 1). There were no venous dilatations or varicose changes in either leg. The abdomen was soft and flat. There was no hepatomegaly, palpable mass or ascites in the abdominal cavity. No heart murmur was audible. Respiratory sounds were normal. Chest roentgenogram showed no abnormal shadows or pleural effusions.

Routine laboratory studies revealed normal limits. Venography was carried out by injecting contrast medium into the saphenous vein, no obstructions or collateral flows in the superficial and deep veins of the leg were re-

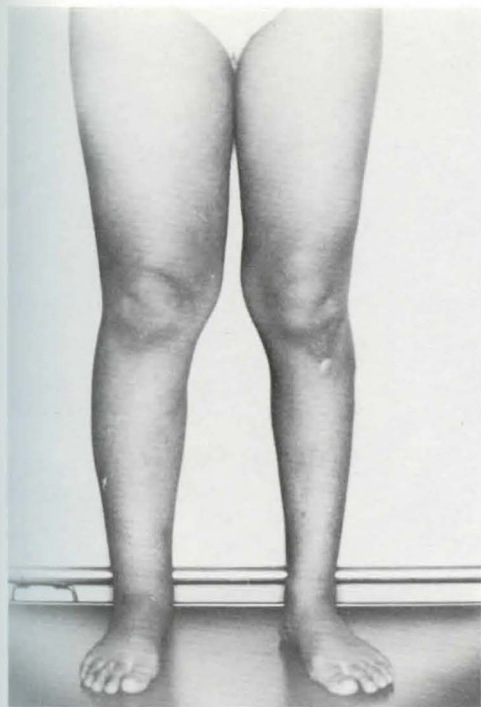


Fig. 1 Lymphedema of the right leg in a 28-year-old woman.

vealed. Lymphangiography was performed exclusively in the right leg. The contrast medium was injected into the superficial lymph vessel at the dorsal side of the foot. Divisions of the lymph flow into the region of popliteal lymph nodes were seen. Dermal backflow or varicose lymph vessels were, however, not seen in the entire leg. The inguinal and external iliac lymph nodes were also observed, but normal lymph vessels in the upper part of inguinal and iliac lymph nodes could not be identified at the termination of injection of the contrast medium. In the roentgenograms taken 24 hours later, the contrast medium could be observed outside of the normal lymphatic pathways and showed irregular figures (Fig. 2). Paraortic lymph nodes and the thoracic duct were not visualized at any time after injection of the contrast medium. These lymphangiographic investigations strongly suggested the existence of a space occupying lesion in the retroperi-



Fig. 2 The 24-hour lymphangiogram. Inguinal lymph node is enlarged and the irregular shadows in the pelvis do not show the contour of lymph node.

toneum of the pelvis. Intravenous pyelography was performed, which revealed abnormalities of the bilateral ureters and the bladder. These abnormalities were characterized by forward and lateral deviation of both ureters and the urinary bladder was compressed from both sides (Fig. 3). Although lymphangiography was not made in the left leg, the findings of pyelography indicated the existence of the space occupying lesions in the left side of the pelvis. On radiological survey of the gastrointestinal tract there was no hypertrophy or irregularity of gastric and intestinal mucosa. On the barium enema the contrast medium flowed freely through the entire colon into the terminal ileum. In the prone position, however, the rectum was found to be slightly compressed laterally. A cystoscopic examination revealed normal mucosa with a deformity due to external pressure. Preoperative diagnosis was made as retroperitoneal lymphangioma.

Operative findings: A midline incision was made and the peritoneal cavity was entered. No ascites were seen in the abdominal cavity. The most striking finding at the beginning of the exploration was large bilateral tumors in the retroperitoneal space of the abdomen and

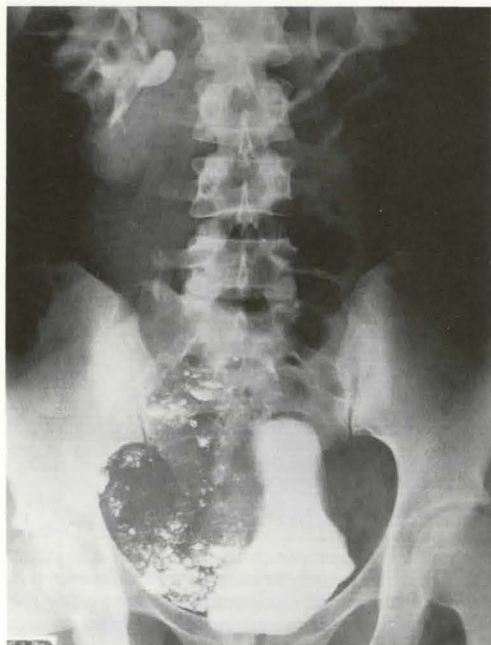


Fig. 3 Intravenous pyelography. Both ureters are displaced laterally and the urinary bladder compressed from both sides.

the pelvis. A contour of the tumors looked like a linkage of multiple balloons having a pinkish color and elastic softness. Bilateral ureters were running over these tumors. The caecum, sigmoid colon, both ovaries and adnexes were displaced forward (Fig. 4). No adhesions was observed between the tumor and these organs. Fibrous adhesion was, however, observed between the bladder and the right side tumor. Retroperitoneum was easily opened by the sharp dissection. The tumor was carefully dissected free from the surrounding tissue, great care being taken to dissect the internal, external and common iliac arteries and veins. The tumor was subtotally excised except the most upper portion which was located at the hilum of both kidneys. During the process of dissection, a large amount of chylous fluids flowed out from the tumor, so that the tumor volume had decreased considerably by the end of the excision.

Postoperative course: Although the degree of

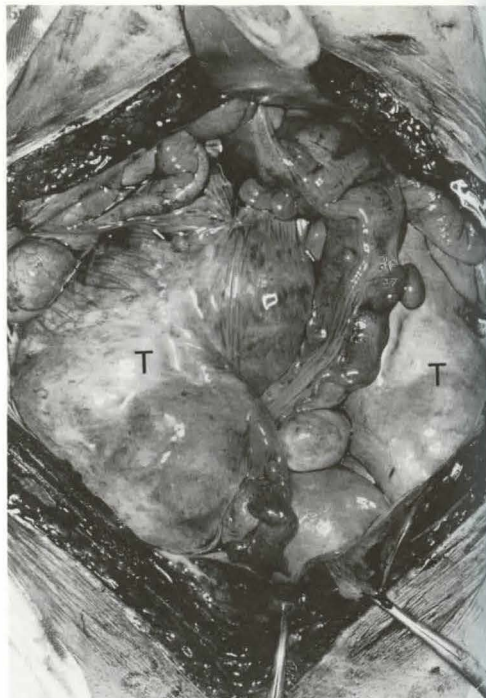
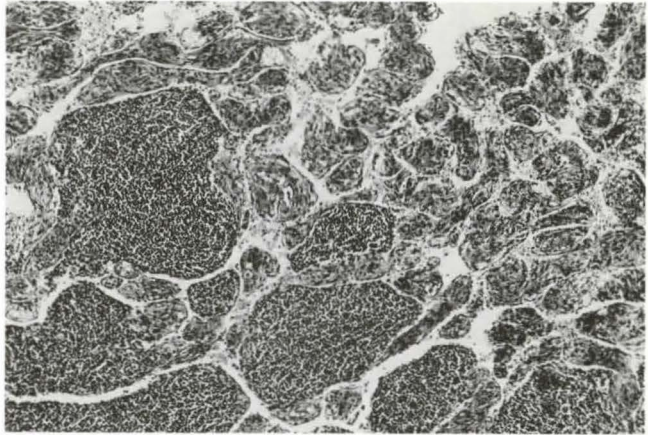


Fig. 4 Operative findings. Bilateral retroperitoneal tumors are covered by peritoneum. Caecum, appendix and sigmoid colon are displaced forwards and upwards. T: Tumor.

the swelling of both legs had not changed during the preoperative hospitalization it decreased markedly after excision of the tumor. No postoperative complications were encountered and she was discharged using elastic stockings for both legs. The patient had been following up during two years. There had been no notable changes in the clinical and various laboratory findings.

Microscopic findings: The tumor was encapsulated by a thin fibrous capsule. Several foci of lymphocytic aggregation were observed in the tumor tissue (Fig. 5). Lymphatics showing marked dilatation were present in the capsule and communicated with the channels of tumor tissue. The tumor was composed of communicating vascular channels and cord-like tissue covered by thin basal lamina with a covering of endothelial cells. The lumen of channels was usually empty with occasional red cells.

Fig. 5 Aggregation of the lymphocytes suggests the tumor tissue to be lymphatic tissue originally. Tumor tissue is composed of vascular channels and band-like stromal tissue. H.E. x 70.

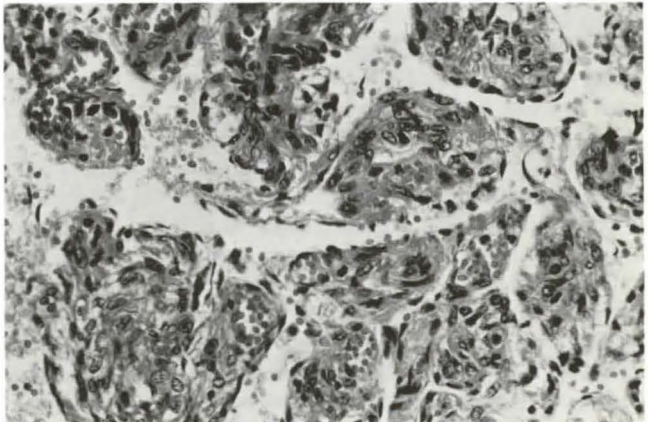


The lumen of channels was usually empty with occasional red cells. The cords were mainly formed by spindle-shaped cells with elongated cytoplasm and blood capillaries (Fig. 6). The cells revealed smooth muscle characteristics by Masson's trichrome staining.

Electronmicroscopic findings: Electronmicroscopic observation was performed on the blocks made from formalin-fixed tissue. The tissue was refixed by a mixture of 4% glutaraldehyde and 4% formalin, and processed in the usual way. Thin sections were cut by LKB microtome and observed by J.E.M. 100 B with original magnification from 2,600 to 3,000.

Tumor cells had vesicular nuclei containing prominent small nucleoli, sparse chromatin and abundant cytoplasm. The tumor tissue was separated into cell groups by vascular channels with covering of endothelial cells, which had cytoplasmic elongation covering basal lamina. The tumor cells were usually elongated in shape and contained numerous mitochondria and small vesicles. The strands of fibrillar structure encircled the nucleus or were located along the cell membrane. There was no evident periodicity in these bundles, but some densities were observed regularly in bundles. The basal lamina-like amorphous structures separated or encircled each tumor cells. The structures sometimes contained a

Fig. 6 Stromal tissue is covered by endothelial cells and contains blood capillaries. Tumor cell has vesicular nucleus containing small nucleoli and elongated cytoplasm. H.E. x 600.



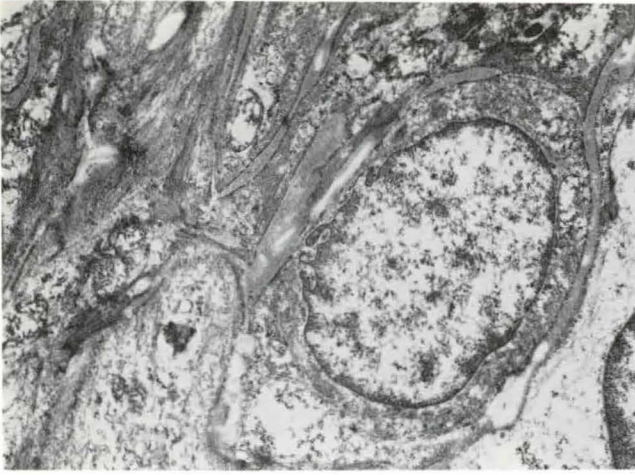


Fig. 7 Tumor cells containing vesicular nucleus with prominent small nucleoli have fine fibrillar structures *myofibrillar. $\times 3\ 700$.

fine fibrillar matrix of varying width and irregularity. Even though the intracytoplasmic bundles did not show remarkable periodicity, these tumor cells appeared to be similar to muscle cells (Fig. 7).

Discussion

Lymphangiography of the present case was chosen for the first diagnostic procedure for lymphedema of the leg. In the 24 hour roentgenogram of the pelvis, an irregular shadow was seen on the outside of the normal lymphatic pathways. This finding suggested a space occupying lesion in the pelvis, and communication between the lymphatic system and this lesion. This retroperitoneal lesion was, therefore, thought to be lymphangioma as a preoperative diagnosis. Lymphangiography has been employed as a diagnostic procedure of lymphangiomyomatosis in several reported cases (3, 6, 7, 9, 11, 14). Frack (6) reported a case with lymphangiomyomatosis in which a lesion was demonstrated on lymphangiograms, extending from the left inguinal region to the lower paraortic zone. This finding is quite similar to that of our case. In our case, however, no symptoms referred to the lung, while Frack's case entered into progressively worsening dyspnea. In the patients with lymphangiomyomatosis the most frequent complaints are referable to the chest, such as dyspnea and pleural effu-

sion. Furthermore, chest pain, cough and hemoptysis are complained of in a small number of patients. Gray (8) collected 36 cases of lymphangiomyomatosis from the literature between 1958 and 1973. Twenty nine of 36 cases had roentgenographic changes in the lung. This disease follows a course of slowly deteriorating respiratory function. Symptoms which are not referred to the chest are weight loss, abdominal pain, abdominal mass and abdominal swelling. In the present case, symptoms referred to the chest and abdomen were not observed, and chest roentgenogram did not show any abnormalities. The first symptom of this patient was the swelling of the right leg. From the clinical view point this complaint without common symptoms of lymphangiomyomatosis makes our case unique among the reported cases.

The site of lymphangiomyomatosis is usually located in the mediastinum and/or the retroperitoneum. In thirty cases collected by Wolff (13), the location of lymphangiomyoma was intrathoracic in 20 cases, retroperitoneal in 16 and supraclavicular lymphnodes in 1. Of twenty patients with intrathoracic lesions, the thoracic duct was involved in 6 patients. If the obstruction of the lymphatic pathway above the portion of the cisterna chyli had been found in the present case, a stasis of the mesenteric lymph flow could occur and lymphangiectasia of the intestinal

tract might develop in the future. A biopsy of the intestinal wall was carried out at the time of the surgery, which revealed a slight dilatation of the lymphatic vessels in the submucosal spaces. With regard to the operative findings of this case, the retroperitoneal tumor firmly adhered to the wall of the urinary bladder. It suggested that communication between the tumor and the bladder may occur and chyluria may appear in the future. Gray (8) reported a case of lymphangiomyomatosis with chyluria, in which a ureterolymphatic backflow was demonstrated by the retrograde cystogram.

The prognosis of the lymphangiomyoma patient is highly variable. Namely, many progress rapidly, while some may stabilize. The progressive course of this disease is mainly due to symptoms related to the lung (honey-comb lung) and chylous effusions. In the present case, the swelling of both legs markedly subsided after excision of the retroperitoneal tumor. Since the pulmonary lesions and/or dietary disturbances may become manifest in the future, a follow-up study is thought to be mandatory.

Wolff (13) first showed ultrastructural evidence of the muscular nature of the tumor cell in mediastinal lymphangioma, in which the presence of myofilament, marginal densities and relative paucity of mitochondria were revealed. He thought that the spindle-shaped tumor cells were in fact smooth muscle cells. Ultrastructural studies justify the use of the name „lymphangiomyoma“ for these lesions (1, 12). On the basis of the presence of strands of fibrillar structure encircling the nucleus or located along the membrane and some densities, we also feel that the spindle-shaped tumor cells were structurally similar to smooth muscle.

It has been postulated by other authors (2,6) that lymphangiomyomatosis is a malformation arising from the lymphatic system and most frequently involving the lung, mediastinum and retroperitoneum. Other theories of the pathogenesis include reactive muscular proliferation (4), trauma (10) and neoplasia (5).

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Rikiya Abe, M.D., F.I.S.L., Department of Surgery, Tohoku University, School of Medicine, Seiryochō 1-1, Sendai 980 Japan