Intraabdominal lymphangiomyoma in an infant with protein-losing enteropathy and hemihypertrophy

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SUMMARY: Koltuksuz U, Özgen Ü, Özen S, Saraç K, Gürsoy MH. Intraabdominal lymphangiomyoma in an infant with protein-losing enteropathy and hemihypertrophy. Turk J Pediatr 2000; 42: 341-343.

Lymphangiomyoma is an extremely rare tumor occurring exclusively in women of reproductive age. The tumor is characterized by proliferation of immature smooth muscle along the lymphatic vessels of the abdomen, thorax and lung. Although lymphangiomyoma has been reported in a young girl and a girl infant, none has been reported in boys. We report herein a case of lympangiomyoma in a two-year-old boy. The unusual presentation in this patient was that the tumor arose from the small bowel mesentery without any evidence of lung involvement. The tumor was extirpated and lymphangiomyomatosis was confirmed pathologically.

Key words: lymphangiomyoma, protein-losing enteropathy, hemihypertrophy.

Lymphangiomyoma is an extremely rare tumor occurring exclusively in women of reproductive age. The tumor is characterized by proliferation of immature smooth muscle along the lymphatic vessels of the abdomen, thorax and lung. The most frequent symptoms are usually due to diffuse pulmonary involvement, which may cause severe ventilation/perfusion problems. Progression in the disease may cause death from these conditions[1,2]. Although lymphangiomyoma has been reported in a young girl and a girl infant, none has been reported in males in the pediatric age group[3,4]. We report the first male infant with lymphangiomyoma in the literature, also having protein-losing enteropathy and hemihypertrophy.

Case Report

The patient, a two-year-old boy, was admitted to the Department of Pediatric Surgery with the complaints of abdominal distention, vomiting and diarrhea beginning two months before admission. Physical examination revealed a swelling of the right arm and leg that was present at birth. He previously had undergone a laparotomy, and a mass, 6 cm in diameter, was excised. Pathological examination of the mass revealed a lymphangioma. In addition to the swelling of the extremities, he had right scrotal hydrocele and edema of preputial and scrotal skin. The abdomen was distended. There was neither hepatosplenomegaly nor a palpable intraabdominal mass. There was no evidence of venous dilatation or varicose changes in either the right leg or the arm.

Laboratory studies showed a serum albumin of 1.2 g/dl and total serum protein of 3.4 g/dl. Protein-losing enteropathy was confirmed by α_1-antitrypsin level of 17 mg/g in dry feces which was much higher than normal (normal < 2 mg/g). Chest roentgenogram revealed no abnormal shadow or pleural effusions. The computed tomographic scan (Fig. 1) of the abdomen showed a mass in the left abdomen between the intestinal loops adjacent to the mesentery, measuring 5.5x4 cm. Intravenous urography was normal. On the barium enema, the contrast medium flowed freely through the colon into the distal ileum. The contrast survey of the gastrointestinal tract revealed intestinal irregularity in a few small bowel segments, but isotope lymphangiogram was normal. These physical and laboratory findings led us to a preoperative diagnosis of the mass as intraabdominal lymphangioma.

Since normal diet and formulas were not tolerated, total parenteral nutrition was started.

After the protein levels reached normal levels, the patient was taken to the operating room. At the laparotomy, milky and slightly hemorrhagic fluid was present in the abdominal cavity. A mass measuring about 10x15 cm that arose from the small bowel mesentery was present. Adjacent portions of the small bowel and mesentery were edematous and had a milky appearance. About 15 cm of small bowel segment was resected together with the mass by preserving main mesenteric vessels and an end-to-end anastomosis was performed on both ends. A milky fluid was seen oozing from the mass during the resection. There were multiple lymph nodes on the mesentery. Other intraabdominal and retroperitoneal organs were apparently normal.

The histopathological evaluations of the specimen revealed lymphangiomyoma showing smooth muscle proliferation surrounding lymphatic spaces and lymphoid aggregates (Fig. 2). To investigate cause of the swelling of the right extremities, skin muscle biopsies were obtained from the right leg, but no pathological change was detected.

No postoperative complications were encountered. His diarrhea disappeared and α_1-antitrypsin decreased to normal levels. The serum albumin and total serum protein increased to normal levels. The scrotal and preputial edema resolved. But the swelling of the right extremities remained unchanged during the postoperative period.

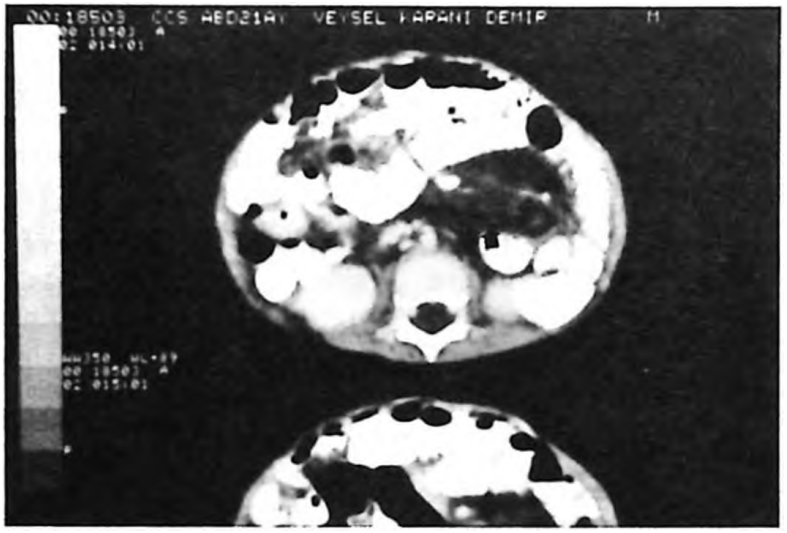

Fig. 1. Lymphangiomyoma showing smooth muscle proliferation (arrow) surrounding lymphatic spaces and lymphoid aggregates (hematoxylin and eosin x 20).

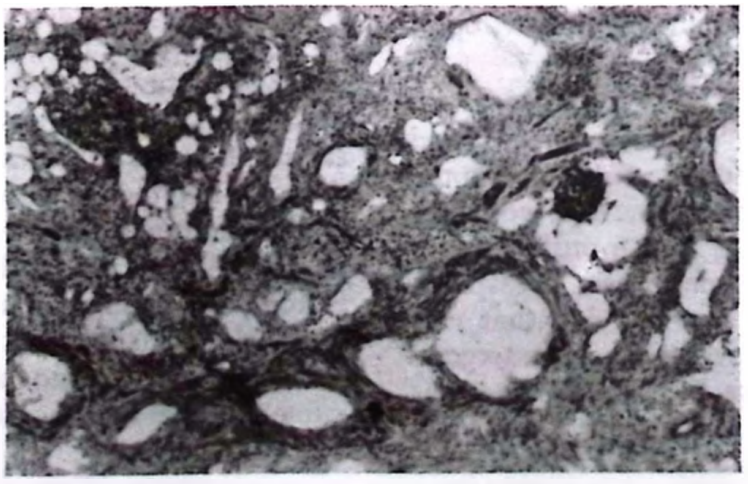

Fig. 2. Abdominal CT scan showing the intraabdominal mass (arrow).

Discussion

Lymphangiomyoma is a rare disease of women occurring almost exclusively during reproductive life and in an age range from 18-69 years. Jacobs et al[5] reported the first case of renal lymphangiomyoma in a male, 79 years old. Although two atients have been previously reported in the pediatric age group, both were female[3,4]. We report a case of intraabdominal lymphangiomyoma in a boy with protein-losing enteropathy. In addition, our patient had no pulmonary symptoms.

Lymphangiomyoma is described as a benign proliferation of smooth muscle involving the abdomen and/or thoracic lymphatics. The site of lymphangiomyoma is usually in the mediastinum and/or the retroperitoneum[6]. The most common complaints refer to the presence of chylous pleural effusion or to pulmonary involvement as well as dyspnea, cyanosis, cough, chest pain, hemoptysis and pneumothorax. Other symptoms include weight loss, abdominal mass, chylous ascites, abdominal pain and abdominal swelling[1]. In the present case, the first symptom was the swelling of the right extremities and face. There was an obvious asymmetry between right and left sides of the body, but without any venous dilatations or varicose changes in organs. Skin and muscle biopsies of the right leg revealed no extra findings. We thus could not find any relation between the swelling and the tumor, and suspected that hemihypertrophy and lymphangiomyoma developed independently.

Although the causes of the vomiting and diarrhea were not clear, we believe that the obsturction which occurred at the tumor level on the small lymphatic branches caused lymphatic leakage and consequently enteric protein loss. This mechanism may have occurred in the present case. Because the tumor invaded mesenteric and lymphatic vessels. There was no sign of mechanical pressure present on the bowels, caused by the tumor. Protein loss disappeared immediately after surgery.

The patient is now four years old, and protein-losing enteropathy did not recur. The follow-up for the intraabdominal mass did not reveal any evident mass. Because of the lack of a similar case reported in the literature, we cannot make any statistical suggestions about the prognosis. However, the regression of the clinical symptoms tends to indicate a favorable outcome.

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