

Lymphangiomatosis in a Case Presenting Duodenal Ulcer Perforation

Duodenal Ülser Perforasyonu Nedeniyle Ameliyat Edilen Olguda Ortaya Çıkan Lenfanjiyomatozis

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Lymphangiomatosis which is a benign tumor consisting of a cluster of dilated lymphatic channels, is very unusual. It is most often found in the neck and head. Less than 5% of cases are diagnosed intraabdominally. Herein a case in the abdomen in a 22-year-old man is reported. The clinical picture was with 7 days progression with constipation and abdominal pain associated with meals. An acute abdomen suggestive of perforated appendicitis was considered. In abdominal exploration, massive purulent fluid which consists of gastric content, pus and also fibrin process was seen among the intestinal loops. An active ulcer on the second part of duodenum and perforated ulcer anterior of this location were determined. There were disseminated multiple nodularities along the small bowel wall and mesentery, and also different number and sized cauliflower-like polypoid masses from the ileo-caecal valve. Histopathological examination was reported as cavernous type lymphangiomatosis plus acute inflammatory reaction. Lymphangioma is difficult to be considered in the differential diagnosis because of the silent clinical course and lack of awareness of the clinical and morphological features. Our case report and review of the literature suggest that lymphangiomatosis is a very rare entity which could be seen with acute abdomen.

Key words: Abdomen, acute; abdominal neoplasms/diagnosis/pathology; lymphangioma, cystic/diagnosis.

Lenfanjiyomatozis, dilate lenfatik kanallardan oluşan ve nadir görülen benign bir tümördür. En sık olarak baş bölgesinde görülür. Olguların %5'den azı intraabdominal yerleşimlidir. Bu yazıda, intraabdominal lenfanjiyomatozis saptanan 22 yaşında bir erkek hasta sunuldu. Klinik tablo, yedi gündür devam eden konstipasyon ve yemeklerle ilişkili karın ağrısı idi. Perfore akut apandisit olarak değerlendirildi. Abdominal eksplorasyonda bağırsak segmentleri arasında mide içeriği, pü ve fibrin ağlarından oluşan pürülan sıvı birikimi görüldü. Duodenum ikinci bölümünde aktif bir ülserle bağlı perforasyon saptandı. İnce bağırsak duvarı ve mezenteri boyunca çok sayıda yaygın nodüller oluşum saptandı. Aynı zamanda ileoçekal valvden itibaren farklı sayı ve büyüklükte, karnabahar tarzında, polipoid lezyonlar görüldü. Histopatolojik inceleme kavernoöz tip lenfanjiyomatozis ve akut enflamatuvar reaksiyon olarak geldi. Morfolojik ve klinik özelliklerinin ayırtedilememesi ve sessiz klinik seyri nedeniyle lenfanjiyomatozis ayırıcı tanıda akla gelmemektedir. Olgu ve literatür bilgileri ışığında, akut batin ile beraber görülebilen son derece nadir bir olgu olarak bildirilmiştir.

Anahtar sözcükler: Batın, akut; karın neoplazisi/tanı/patoloji; lenfanjiyom, kist/tanı.

Lymphangioma as a benign tumor, consisting with a cluster of dilated lymphatic channels is very unusual.⁽¹⁾ The lymphangioma is considered as a benign neoplasm of embryonic origin of the lymphatic vessels and it is more frequently reported in children and yet occasionally in the adult

patients.⁽²⁾ The tumors are usually located in the neck, axilla, and rarely in the mediastinum, lungs, esophagus, diaphragm, duodenum, stomach, small and large intestine, spleen, and liver.⁽³⁾ Five percentage or less of lymphangiomatosis has been discovered in the abdominal cavity.⁽⁴⁾ Herein, the

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case of abdominal widespread lymphangiomas in an adult was reported.

CASE REPORT

A 22-year-old patient representing abdominal pain, nausea, vomiting, postprandial distention and constipation for 7 days was admitted. Because there had been no improvement on his own health condition with therapy in the infirmary, he was transferred to the emergency room in Gulhane Military Medical Academy Haydarpaşa Training Hospital.

On physical examination, the temperature was 37.4°C, the pulse rate was 92/min and the respiration rate was 24/min. The blood pressure was 80/50 mmHg. He had no relevant medical or family history. In the abdomen moderate distention was noticed and bowel sound was increased in auscultation. Rebound tenderness was determined in the right lower quadrant and middle part of the abdomen. Digital rectal examination gained no abnormality. The results of laboratory tests (complete blood count, urine test and routine biochemical tests) were in normal ranges except white blood cells that was 17.500/mm³. Plain X-ray for abdomen showed some loops of gas in the right part of the abdomen and few tiny air-fluid levels as well (Fig. 1). Abdominal ultrasonography showed dilated and inactive bowel loops and fluid sweeping around the intestines. A great deal of fluid was placed around the cecum and retrovesical space. Moreover abdominal ultrasonography was not capable to show any part of



Fig. 1. Plain abdominal X-ray showing suspicious air-fluid level and gas on the right side of the abdomen.

appendix. Finally, an acute abdomen suggestive of perforated appendicitis was considered and abdominal exploration was performed urgently.

In abdominal exploration, significant gastric dilatation extending to the pelvic region, massive purulent fluid which consists of gastric content,



Fig. 2. Multiple nodularities through the whole ileum and mesenterium.



Fig. 3. Different number and sized cauliflower like polypoid masses through the length of the intestine.

pus and also fibrin process was seen between the intestinal loops. An active ulcer on the second part of duodenum and perforated ulcer with a diameter of 3 mm anterior surface of this location were determined. There were multiple nodularities through the whole ileum and mesenterium and also different number and sized cauliflower-like polypoid masses were present through the length of the intestine (Fig. 2, 3).

Other intraabdominal organs were normal. The perforated ulcer was primarily sutured and omentoplasty was performed. Multiple biopsies were taken from the masses of intestinal wall and it's mesentery. In the postoperative period the patient was discharged with 15 days of medication (on a proton pump inhibitor and a gastrokinetic agent).

The cross section of the biopsy specimens microscopically showed variable sized, markedly dilated lymphatic channels in the mesentery and all parts of the bowel wall lined by flat endothelial cells (Fig. 4, 5, 6). The wall of the lymphatic spaces was built up fibroconnective tissue accompanies by aggregates of lymphoid tissue as well as normal arteries and veins. The final histopathologic diagnosis was cavernous type lymphangioma-tosis.

DISCUSSION

Abdominal lymphangioma is rare and often classified with mesenteric and retroperitoneal cysts.⁽⁵⁾ They occur on the mesentery of the small and large bowel but also at retroperitoneum less frequently. Lymphangiomas are divided into three groups; simple capillary, cavernous and cystic lymphangiomas.

The clinical symptoms range from chronic to acute abdominal pain sometimes due to traumatic rupture,⁽⁶⁾ obstruction⁽⁷⁻⁹⁾ or anemia due to hemorrhage.⁽¹⁰⁾ In adults, the most common symptom is chronic abdominal pain whereas in children the onset of symptom is usually acute due to intestinal obstruction.⁽¹¹⁾ Abdominal localization and it's development may produce symptoms and clinical features of acute abdomen leading the problems in the differential diagnosis.⁽¹²⁾ Our patient presented with acute abdominal symptoms due to duodenal ulcer perforation and it was difficult to establish an accurate preoperative diagnosis.

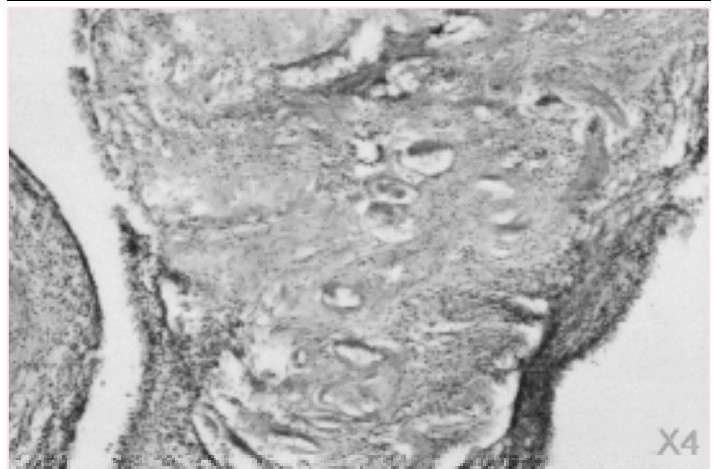


Fig. 4. Low power (4x) view of the cavernous lymphangioma. Multiple dilated lymphatic channels in the connective tissue derived from lymphocytes, smooth muscle and fibrous tissue.

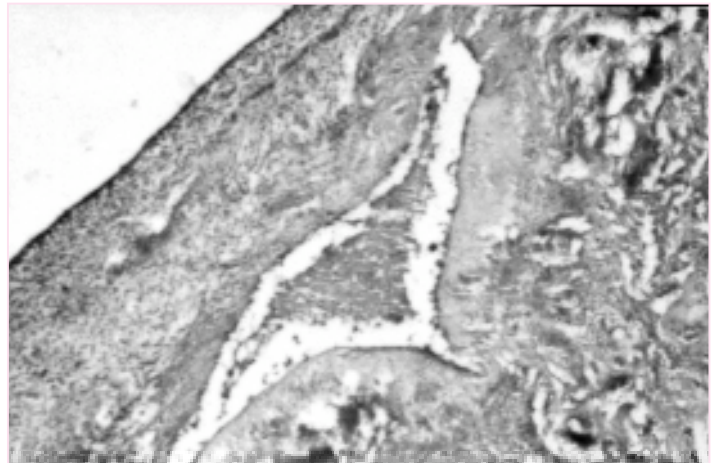


Fig. 5. Photograph showing dilated proliferated lymphatic channels in fibroconnective tissue (10x).

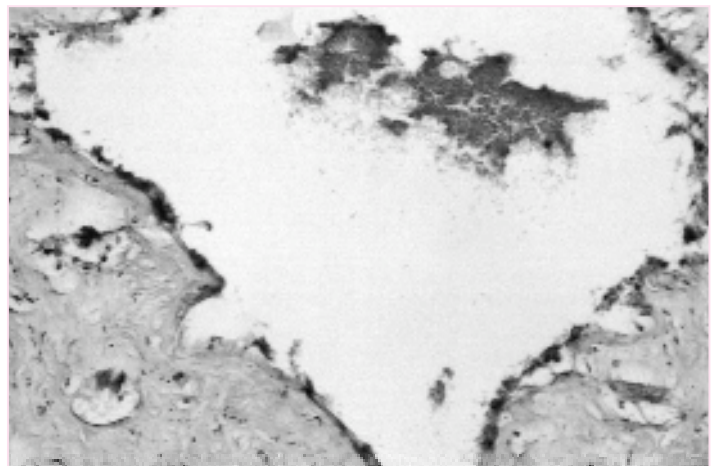


Fig. 6. Dilated lymphatic channel; its wall lined by endothelial cells (40x).

These tumors are benign and must be differentiated from other cystic intraabdominal lesions. The differential diagnosis of intraabdominal lymphangioma includes many benign and malignant tumors. Preoperative diagnosis is difficult due to silent clinical course and lack of awareness of the clinical and morphological features of this disease.⁽¹¹⁾ Therefore, a diagnosis is often first made during operation because of the macroscopic aspect and then definitely because of the histological examination.⁽¹³⁾ In general, most of the patients can be cured of the disease after a to-

tal resection of the tumor. Nevertheless tumor recurrence has been reported.⁽¹⁴⁾ Long term survival can be achieved even in patients receiving no treatment. Resection of the lymphangiomatosis is the most commonly reported therapeutic modality.⁽¹⁾ However it was impossible to perform an urgent total resection in our patient due to double synchronous morbidity (duodenal ulcer perforation and lymphangiomatosis).

In conclusion, lymphangiomatosis is a very rare entity which should be seen with acute abdomen.

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