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Case Report

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An Accidental Treatment for A Rare Disease

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1. Abstract

1.1. Introduction: Lymphangioma is an uncommon benign tumor believed to be the result of diffuse proliferation of lymphatic tissues. Abdominal lymphangiomatosis is a rare disease characterized by infiltration of the intraperitoneal organs with lymphangiomas. Clinical symptoms of this condition are not clear. Pathological, radiological, and clinical evidence are limited for diagnosis. During operation, it can be observed accidentally. Hereby, it may be an inconvenient instance for surgeons.

1.2. Patients and Methods: In this study, we reported a 42-year-old female who underwent surgery with the diagnosis of primary malignant peritoneal mesothelioma (at the same time we performed HIPEC). However, the histopathological examination revealed lymphangiomatosis unexpectedly.

1.3. Results: In long-term follow-up, we had a good result on this patient. We also want to provide a new point of view on the diagnosis and treatment of this disease.

1.4. Conclusion: Especially, there is not enough evidence in the literature on how to prevent the recurrences of lymphangiomatosis. In our study, we want to take attention to this subject. Hyperthermic Intraperitoneal Chemotherapy (HIPEC) can be an alternative treatment modality for patients who had lymphangiomatosis.

2. Introduction

Lymphangiomatosis is an uncommon clinical entity that presents with the diffuse proliferation of lymphatic vessels. It usually involves bone, dermis, parenchymal organs, and soft tissue [1]. Abdominal lymphangiomatosis is an extremely rare disease that is usually found in adulthood, and the clinical symptoms of this condition are complicated and atypical [2]. The pathogenesis of abdominal lymphangiomas remains unclear. According to a common consideration, it consists due to the obstruction of regional lymph drainage during the embryogenesis of the lymphoid system [3]. Cytoreductive Surgery (CRS) and Hyperthermic Intraperitoneal Chemotherapy (HIPEC) are complex procedures used for gynecologic and gastrointestinal cancer. These treatment modalities are recognized as standard care of susceptible patients with peritoneal carcinomatosis. Combined CRS and HIPEC aim to remove all macroscopic tumors in the abdominal cavity with visceral resections followed by targeting any residual cancer cells with intraperitoneal chemotherapy [4]. It is actually used for treating peritoneal carcinomatosis of ovarian, colorectal, or gastric cancer besides pseudomyxoma peritonei and peritoneal primitive origin tumors (mesothelioma) [5]. To our knowledge, in literature, there is no example of lymphangiomatosis treated with these procedures which have not previously been reported. In this study, we aim to indicate the case suffering from lymphangiomatosis who was treated by using CRS and HIPEC.

3. Case Presentation

Herein, we report a 42-years-old female who underwent surgery with the diagnosis of primary malignant peritoneal mesothelioma. She reported intermittent rough lower abdominal tenderness with-

out nausea, vomiting, constipation, or any other gastroıntestinal symptoms. She had no family history of cancer, trauma, surgery, or asbestos exposure reported. She was a non-smoker. She was submitted to gynecological evaluation with these complaints. A gynecologic Ultrasound (US) revealed free fluid collection around the uterus. Inside of fluid, there were 3-7 cm diameters multıple unilocular cystic masses without enhancement. The cysts were thin-walled and homogenous sectioned. She was referred to our hospital's general surgery department. Physical examination revealed a normal temperature and the abdominal examination was unremarkable. Pelvic Magnetic Resonance Imaging (MRI) was performed. It also showed intraperitoneal septated fluid collections filled the pelvic cavity totally and nodular cystic lesions that their largest dimension measured 6 cm were seen in liquid (Figure 1). Recent US-guided aspiration from liquid was performed by an interventional radiologist. The histopathological analysis showed the sampling was transudate which contained normal peritoneal cells. No significant sign was observed with contrast-enhanced Computed Tomography (CT) of the thorax. Additionally, colonoscopy and upper gastrointestinal endoscopy were normal. At this stage, the diagnoses of primary peritoneal mesothelioma were considered. Laboratory investigations revealed hemoglobin level was 13.0 g/dl [ref. 12-16 g/dl]. Blood chemistry parameters and the tumor markers were normal also. The serum CA 15-3 level was 27.4 U/mL [ref. 0-31 U/mL] and the CA 19-9 level was 11.2 U/mL [ref. 0-34 U/mL]. CA 72-4 <1.5 U/ mL [ref. <6.9 U/mL]. CEA level was 1.6 ng/ml [ref. 0-10 ng/mL].

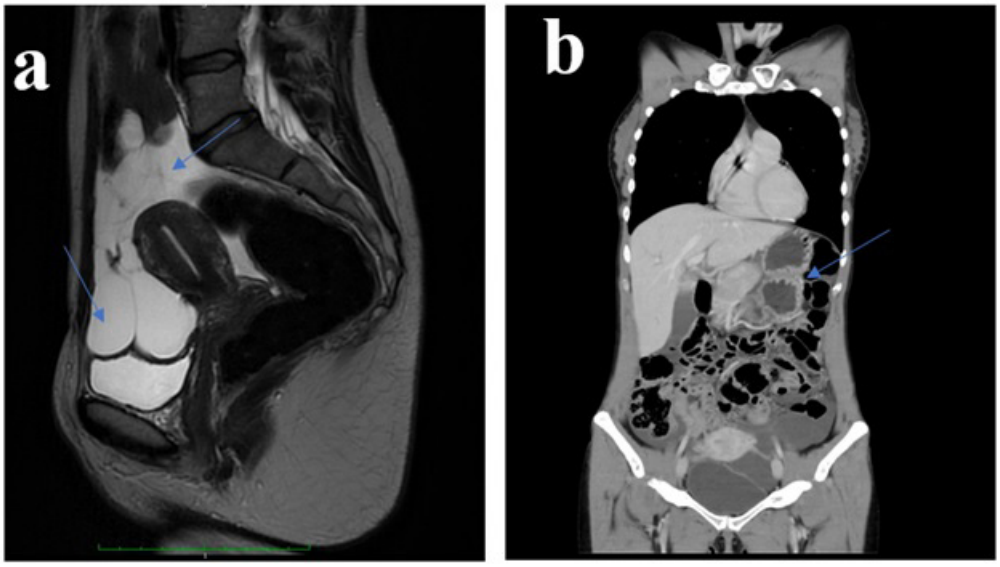

Figure 1: Magnetic resonance imaging reveals multiple cystic lesions in the abdominal cavity.

A: Sagittal images; **B**: Coronal images

After multidisciplinary discussions, we decided to perform surgery with the pre-operative diagnosis of peritoneal malignant mesothelioma. After essential preparations, the patient underwent the operation. The laparotomy was performed. Ascitic fluid accumulation (nearly 3 liters) was revealed. The largest one was 5cm diameters, soft yellowish, lucent multiple cystic masses were seen in pelvic and intraperitoneal cavity. They were located on (with a weak adherence) colon, intestine, omentum, and pelvic organs in four-quadrant and caused inflammatory- hypertrophic adhesions with these organs in patches (Figure 2). Frozen section samples were taken from the cystic lesions and their hypertrophic tissues. Benign features mesothelium cysts and no signs of malignancy were reported. Appendectomy was performed and also on frozen section examination of the appendectomy material showed there was no malignant sign. With a prediagnosis of benign multicystic diffuse mesothelioma, all peritoneal organs with cystic invasions were excised. We performed a partially peritonectomy and complete excision of neoformation, with a free margin of omental fat. In this wise, the epiploic appendices of the colon and the left tuba ovarian excised totally. And also, a short segment of the ileum was excised. The anastomosis was performed. (Because of the extensive cystic involvements of left tuba ovarian and ileum, the lesions couldn't be removed separately). After all, cystic and nodular lesions removed from the patient underwent HIPEC. Inflow and outflow drains were placed. Mitomycin–C intraperitoneal regimen 15 mg/m^2 for a total of 90 minutes a temperature of 41-42° was used for HIPEC bath (4 liters isotonic) with CO_2 agitation. After control of bleeding, drains were removed. Subsequently, standard surgical drains were placed. The abdomen was closed with Polydioxanone (PDS) loop NO:1 suture. There was no complication regarded in the post-operative period. She was discharged 6 days later after the operation in a good condition. The histopathological examination revealed lymphangiomatosis for all excited tissues. No malignancy was reported. The immunohistochemical staining confirmed the case by using antibodies of D2-40 (Podoplanin), calretinin, and PAN-CK (pan-cytokeratin) (Figure 3). It surprised us. However,

the interestingly post-operative course of the patient was uneventful. After two months of discharge first abdominal CT was performed as a control imaging to check the recurrence of the tumor. There was shown no presence of the pathology. She had regular scans, further and further (Figure 4) shows the last CT scan carried out 1 year after the operation.

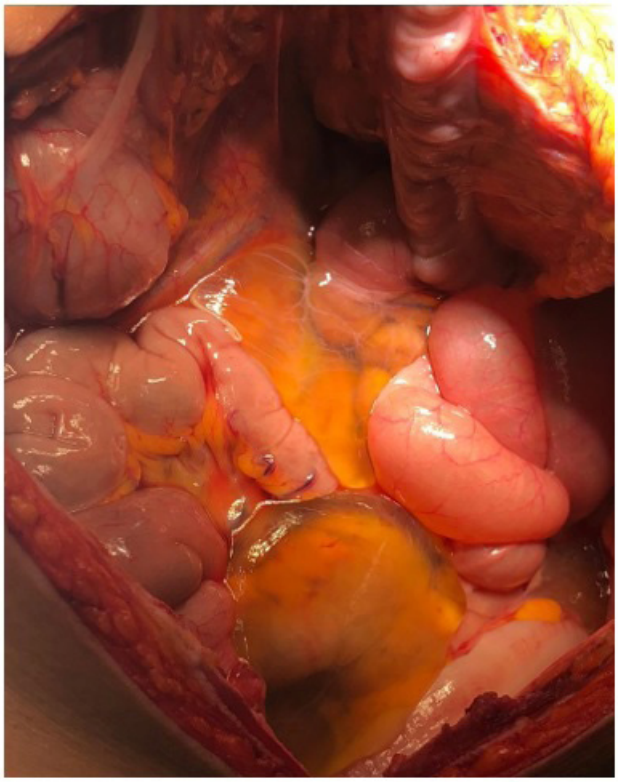

Figure 2*:* Intraoperative view of soft yellowish, lucent multiple cyctic masses in the pelvic and intraperitoneal cavity.

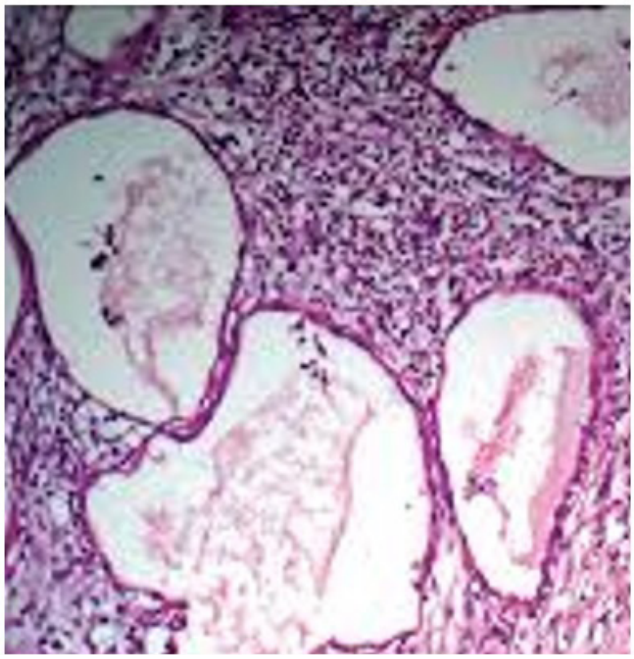

Figure 3: Microscopic view of the abdominal lymphangiomatosis showing the cells lining the cysts. (Hematoxylin and eosin stain, original magnification × 400)

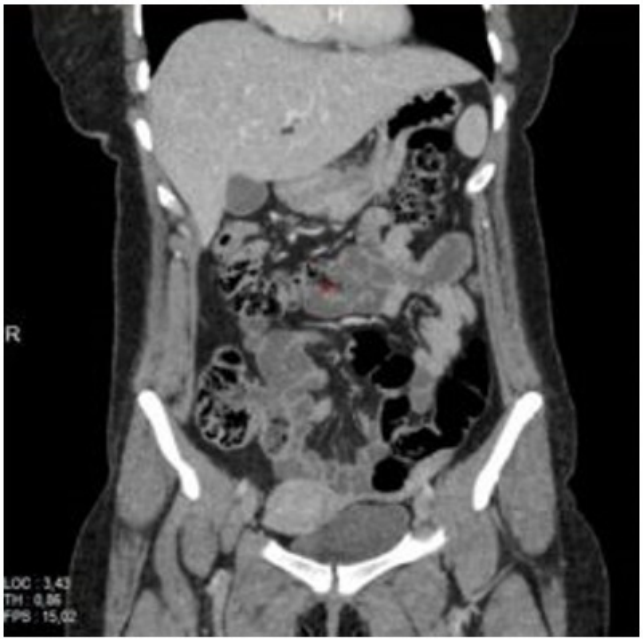

Figure 4*:* CT scans of abdomen and pelvis. (Coronal image). It carried out 1 year after the operation. No recurrent mass formation was seen.

4. Discussion

Lymphangiomatosis is a rare type congenital disorder of lymphatics characterized by the development of malformation of lymphatic channels [1]. In 1928, Rodenber described this anomaly first [6]. There is no difference between men and women concerning the incidence of these lesions. Despite it is a congenital disease, it most frequently presents in late childhood [2]. Because of the intra-abdominal location of lesions, generally, they may not be detected before adulthood. Although the pathophysiology remains elusive, this clinical entity is derived from overexpression of lymphangiogenic growth factors. That causes abnormal dilatation and proliferation of lymphatic channels by the obstruction or agenesis of lymphatic tissues. Finally, it turns into a cystic mass. Except for the central nervous system (devoid of lymphatics) the lesions of lymphangiomatosis can occur in any visceral organs or skeletal system [7]. Our case is an abdominal lymphangiomatosis. It is extremely rare in the literature (<5% of cases). Due to unspecific presentation and varying clinic status, it is hard to diagnose abdominal lymphangiomatosis [8]. The clinical presentations of intraabdominal lesions are in large quantity, asymptomatic (sometimes obstructive), adenoma-like polypoid masses. And the potential symptoms are abdominal distension, abdominal pain, loss of appetite, nausea, vomiting, melena, and diarrhea [8,9]. Likewise, in our case, pre-operative diagnosis of intraabdominal lymphangioma is uncommon. US, MRI, and CT are necessary for describing the cystic lesions and providing important information regarding their location, size, and adjacent organs involvement. However, they are not adequate for the diagnosis of lymphangiomatosis pre-operatively. Most lymphatic cysts are usually found incidentally. The final diagnosis is confirmed after histopathology and/or immunochemistry [10]. The unique treatment modality for abdominal lymphangiomatosis is surgery. All macroscopic cystic masses should be removed. Nevertheless, recurrences are still potential problems of lymphangiomatosis. There is no foolproof way to prevent it.

So far, we have had a good result with HIPEC on this patient. At first, we conclude that even if a frozen section cannot help the surgeons, abdominal lymphangiomatosis may create big difficulties in diagnosis. Secondly, in patients who have severe lymphangiomatosis, the lesions may be perforated during the operation accidentally. We believe that HIPEC may be a good option in these cases.

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