Giant Splenic Lymphangiomas in Adult: a Diagnostic Dilemma

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Abstract Cystic lymphangiomas of the spleen occurs secondary to developmental malformation of the lymphatic system. It is one of the rare entities generally seen in children. It is usually seen in children. We report a case of cystic lymphangiomas of the spleen occurring in an adult woman presenting with massive splenomegaly. Total splenectomy was considered to be the treatment of choice for making definitive diagnosis and to exclude the presence of malignancy or other causes of massive splenomegaly

Keywords Splenomegaly · Giant · Cystic · Lymphangiomas · Splenectomy

Introduction

Lymphangiomas are benign lymphatic system malformation of unknown etiology. It can be either solitary or multiple. They principally occur in children and are rarely seen in the older age [1]. The most common locations of lymphangiomas are the neck and the axillary region [2]. Cystic lymphangiomas of the spleen is a very rare entity and is usually solitary and asymptomatic. This is a rare condition, with only nine cases being reported between 1990 and 2010. The only presentation is massive splenomegaly. Even though rare, it is very difficult to diagnose. Cystic lymphangiomas should be considered in the differential diagnosis of massive splenomegaly by excluding other common causes.

Case Presentation

A 35-year-old female presented with complaints of heaviness, vague pain, and swelling in the upper abdomen along with early satiety for about 1 year. She also complains of occasional nausea and vomiting. There was no associated fever, dyspepsia, or weight loss. There was no history of gum bleeding, hematemesis, melena, or bleeding per rectum. There was no history of jaundice. Physical examination showed no pallor, no icterus, or significant lymphadenopathy. On abdominal examination, the whole abdomen was diffusely distended with minor vascular marking. On palpation, the spleen was grossly enlarged, 20 cm below the costal margin at midclavicular line, extending up to the right iliac fossa and covering the whole abdomen. The surface is smooth without nodularity. The margin was not well defined. The splenic notch was not felt. There was no free fluid in the peritoneal cavity, and no other findings were noted. Initial diagnostic possibilities considered were malaria, kala-azar, chronic myeloid leukemia, low-grade lymphoma, and other diagnoses. Complete hemogram, bleeding profile results were within normal limits. The peripheral blood smear examination was within normal limit. The chest X-ray result was not significant. On ultrasound, there was huge splenomegaly with multiple cysts divided by septa, and CT scan of the abdomen showed splenomegaly with a large well-defined cystic mass surrounded by multiple peripheral cysts, all divided by thin septa with focal lesions which were highly suggestive of lymphoma or granulomatous disease (Fig. 1). As a diagnostic and therapeutic option, she underwent elective splenectomy without any complications. The spleen on gross pathological examination was measuring 36×22×18 cm and weighing 2100 g (Fig. 2). The spleen was sent for histopathological examination which revealed splenic lymphangiomas.

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Fig. 1 CT scan shows splenomegaly with a large well-defined hypodense cystic mass surrounded by multiple peripheral cysts, all divided by thin septa

Discussion

Lymphangiomatosis is a cystic, benign, slow-growing tumor and a very rare condition, usually seen in children, where it is discovered incidentally [3]. These tumors occur more frequently in females, and 80–90 % of these tumors are detected before the end of the second year of life. Lymphangiomas are generally considered to be congenital malformations of the lymphatic system, and they occur mostly in the neck, mediastinum, and retroperitoneum and are rarely found in the spleen [4].

Lymphangiomas of the spleen are rare and can present as solitary nodules, multiple nodules, or diffuse growth. More often, they consist of multiple cysts of varying sizes lined by flat endothelial cells, which contain eosinophilic serous fluid. The septa are made of fibrous connective tissue with rare

vessels [5]. Splenic lymphangiomas are mostly asymptomatic; therefore, the final diagnosis should be based on a combination of clinical, radiological, and histopathological findings. Histologically, lymphangiomas are classified into three subtypes according to the congenital dilated lymphatic channels: capillary (super-microcystic), cavernous (microcystic), or cystic (macroscopic) [5].

At ultrasonography, the cysts are hypoechoic with internal debris. The septa are hyperechoic and may show detectable vasculature at color Doppler study. In most cases, splenic lymphangiomas present with thin-walled cystic masses without enhancement or with only slight enhancement of the thin septa in the CT studies. In the current scenario, a mention of magnetic resonance lymphangiography would be worthwhile. Multiorgan involvement such as the liver, bone, and spleen is known to occur. The differential diagnosis of cystic lymphangiomatosis includes true splenic cysts, mesothelial cysts, echinococcus, and very rarely, cystic hamartoma [2]. True splenic cysts will have definite epithelial lining. Parasitic cysts most commonly due to *Echinococcus granulosus* may be indistinguishable from any other multicystic masses; therefore, they may resemble cystic lymphangiomatosis. Serological test is required for echinococcus. Cystic hamartomas are very rare in the spleen and occur more frequently in the liver in the pediatric age group [7]. In our case, it was initially treated for its common cause—splenomegaly—with antitubercular and antimalarial drugs, but the spleen progressively increased in size. There was difficulty to make a final diagnosis even after ultrasound and CT scan. Although cystic lymphangiomatosis of the spleen is a benign entity, one case of malignant lymphangioma was also reported [8]. The malignant degeneration was suggested by the increased proportion of solid areas. Lymphangiomatosis involves all the splenic parenchyma; therefore, a total splenectomy is the treatment needed. Laparoscopic splenectomy is considered as the procedure of choice for normal or moderately enlarged spleens, whereas in the case of severe splenomegaly, open splenectomy is preferred [6]. Pazopanib, a tyrosine kinase inhibitor, may cause stabilization of lymphangiomatosis through VEGF. Splenic lymphangiomatosis may occur concurrently with Proteus syndrome [9].

Conclusion

Cystic lymphangiomatosis of the spleen should be considered in the differential diagnosis of massive splenomegaly even in adult, and its final diagnosis is made by histopathological examination.

Conflict of Interest None

Fig. 2 Gross resected massive splenomegaly with nodularity

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