Int. j. pathol 2013; 11(2): 70-72

Case Report

Spleen, Non-Systemic Diffuse Lymphangiomatosis

Sarosh Majid*, Samina Zaman, Ambreen Anwar***, Omer Chughtai*****

* AJK Medical College, Muzaffarabad, **Institute of Child Health Lahore, ***PGMI Lahore, ****Chagatai's Lahore Lab

Abstract

Lymphangiomatosisis a rare disorder believed to be result from a developmental malformation of lymphatic system. Lymphangiomatosis involves bone, soft tissues, and viscera in a diffuse fashion. We describe an unusual case of splenic lymphangiomatosis in 41 years old female.

Keywords: Spleen, Lymphangiomatosis.

Introduction

Lymphangiomatosis is a unique, rare disorder that occurs predominantly in children and young adults less than 20 years of age.[1] It is considered to be developmental anomaly, can occur alone or be part of multi organ disease. This is characterized by abnormal proliferation of lymphatic channels, regarded as a congenital malformation of the lymphatic system. Histologically, the lesion is composed of cystic spaces of various sizes lined by endothelium and surrounded by connective tissue.[2,10,11]

Case Report

41 years old female presented with abdominal distension and pain in left hypochoindrium. On examination spleen was enlarged ultrasonography revealed cystic lesions in the spleen with echinococcal disease as the most likely diagnosis. Other organs were found essentially normal. Laparotomy was performed and specimen of spleen sent for histology. Spleen was2.7kg in weight and measured 30x20x15cm. External surface was brownish with multiple small swellings. Cut surface showed numerous variable sized cysts like spaces containing yellowish fluid with intervening splenic tissue (fegure1). Microscopic examination revealed many dilated vascular channels that are devoid of blood and contain pink proteinaceous material (lymph). A few dilated blood vessels are also present. Intervening splenic parenchyma is unremarkable (figure 2).

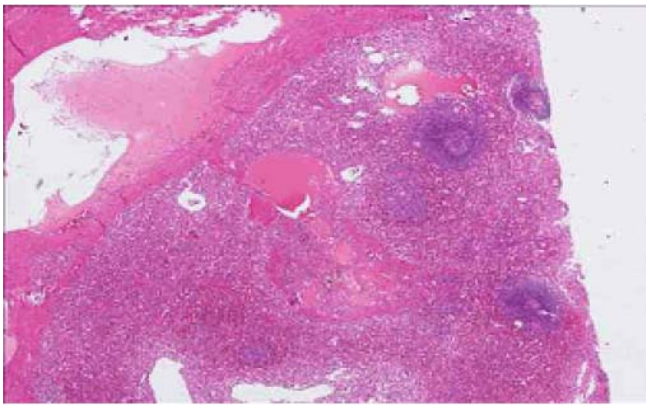

Figure 1: Cut Section of Spleen Showing Dilated Lymph Spaces

Correspondence: Dr. Sarosh Majid,
AJK Medical College, Muzaffarabad

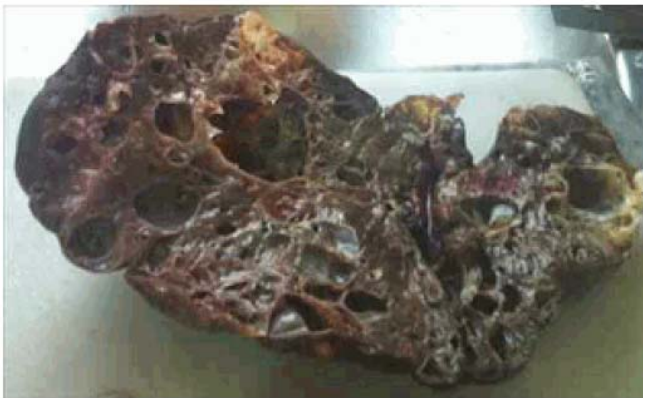

Figure 2: Dilated Lymphatic Channel with Splenic Tissue (H&E X100)

Discussion

Lymphangiomatosis is a rare disorder that is believed to be a developmental malformation of the lymphatic system in which obstruction or agenesis of lymphatic tissue results in lymphangiectasia secondary to lack of normal communication of the lymphatic system.

Lymphangiomatosis involves bone, softtissues, and viscera in a diffuse fashion.[4,5] Splenic lymphangiomatosis is very rare.[7-,9] This is marked by the presence of cysts that result from an increase both in the size and number of thin-walled lymphatic channels that are abnormally interconnected and dilated. Lymphangiomatosis is a condition marked by the presence of cysts that result from an increase both in the size and number of thin-walled lymphatic channels that are abnormally interconnected and dilated.[6] The condition may involve a single organ system and more commonly presents by age 20. Lymphangiomatosis isa benign lesion tend to invade surrounding tissues and cause problems due to invasion and/or compression of adjacent structures.[6] Lymphangiomatosis is usually seen in children in whom it is frequently discovered incidentally.These tumors occur more frequently in female and 80 to 90% are detected before the end of the second year of life.[5]

Cystic lesions of the spleen include parasitic and non-parasitic cysts. Parasitic cysts, almost exclusivelycaused by echinococcal disease, represent 50 to 80% of splenic cysts, although splenic echinococcosis represents only 3.5% of echinococcal diseases. In addition, encephalic localization of echinococcosis has been described.[3] Non-parasitic cysts are classified as primary or true cysts, which have an epithelial or endothelial lining, and pseudocysts which may be classified as post-traumatic, degenerative or inflammatory. Endothelial true cysts may be lymphangiomas or hemangiomas.[1] The neck (75%) and maxillary regions (20%) are the most common locations of lymphangioma, but itcan occur in the retroperitoneum, mediastinum,mesentery, omentum, colon, pelvis, groin, bone, skin, scrotum and spleen. Lymphangioma of the spleen can involve the spleen alone, or it can be a part of multi visceral involvement; when diffuse it is termedsystemic cystic angiomatosis. Patients with splenic lymphangioma may be asymptomatic or symptomatic at diagnosis. Upper left quadrant pain is the most common symptom, frequently followed by fever, nausea, vomiting, and weight loss, i.e. the symptoms reported by our patient, hypertension, hypersplenism and consumptive coagulation disorders. Because of the similarity of symptoms and signs, it is often confused with hydatid disease which a negative E. granulosus agglutination test result cannot always exclude. The right diagnosis depends on histopathologic examination after removal of the spleen. Ultrasonography and computed tomography are the most helpful imaging techniques in the diagnosis of splenic cystic lesions. In our case it was not diagnostic so surgical excision was planned.

Splenic lymphangiomamay involve the spleen in the form of solitary nodules, multiple nodules or diffuse growth. Histologic examination of the nodules promptly excluded the clinical suspicion

of parasitic cysts and established the vascular origin of the lesions and an eosinophilic proteinaceous material filling the endothelium-lined spaces favored the diagnosis of splenic lymphangioma instead of hemangioma, in which the endothelium-lined spaces are filled by blood. Although benign, abdominal lymphangioma may become locally invasive, so treatment remains surgical and splenectomy seems to be the best choice.The prognosis is excellent.

Conclusion

Splenic lymphangiomatosis is a rare condition. Splenic lymphangiomamay involve the spleen in the form of solitary nodules, multiple nodules or diffuse growth.

References

1. Laverdiere C, David M, Dubois J, Russo P,Hershon L, Lapierre JG : Improvement of disseminated lymphangiomatosis with recombinantinterferon therapy. Pediatr Pulmonol 2000;29: 321-324.
2. Fisher D, Hiller N : Case report : Giant tuberculouscystic lymphangioma of posterior mediastinum, retroperitoneum and groin. Clin Radiol 1994;49: 215-216.
3. Faul, JL, Berry GJ, Colby TV, Ruoss SJ, Walter MB, Rosen GD, and Raffin, TA. Thoracic Lymphangiomas, Lymphangiectasis, Lymphangiomatosis, and Lymphatic Dysplasia Syndrome. Am. J. Respir. Crit. Care Med. 2000; 161(3): 1037-1046.
4. Huaringa AJ, Chittari LM, Herzog, CE, Fanning, CV, Haro, M & Eftekhari, F. Pleuro-Pulmonary Lymphangiomatosis: Malignant behavior of a benign disease. The Internet Journal of Pulmonary Medicine. 2005; 5(2). http://ispub.com/IJPM/5/2/9120
5. Marymont JV, Knight PJ : Splenic lymphangiomatosis: A rare cause of splenomegaly. J Pediatr Surg 1987;22: 461-462
6. Tazelaar HD, Kerr D, Yousem SA, Saldana MJ, Langston C, Colby TV. Diffuse pulmonary lymphangiomatosis. Hum Pathol. 1993;24(12):1313-22..
7. Komatsuda T, Ishida H, Konno K, Hamashima Y, Naganuma H, Sato M, Ishida J and Masamune O. Splenic lymphangioma: US and CT diagnosis and clinical manifestations. Abdominal Imaging 1999; 24:414-417
8. Ros PR, Goodman ZD, Ishak KG, Dachman AH, Olmsted WW, Hartman DS and Lichtenstein JE. Mesenchymal hamartoma of the liver: radiologic – pathologic correlation. Radiology 1986; 158: 619-624
9. Morgenstern L, Bello JM, Fisher BL and Verham RP. The clinical spectrum of lymphangiomas and lymphangiomatosis of the spleen. Am Surg 1992;58:599-604
10. Pernick, Nat. “Soft Tissue Tumors Part 3 Muscle, Vascular, Nerve, Other Lymphangiomatosis.” PathologyOutlines.com, Inc., 10/17/2009. Web. 6 Sep 2011
11. Patti R, Iannitto E, and Di Vita G Splenic lymphangiomatosis showing rapid growth during lactation: A Case Report World J Gastroenterol. 2010;7;16(9): 1155-1157.