International Journal of Basic and Applied Medical Sciences ISSN: 2277-2103 (Online)
An Open Access, Online International Journal Available at http://www.cibtech.org/jms.htm
2017 Vol. 7 (1) January-April, pp. 22-27/Ingle and Chavan

Case Report

AN INTERESTING CORE BIOPSY DIAGNOSIS: DIFFUSE LYMPHANGIOMATOSIS OF SPLEEN

***S. B. Ingle and Kundan Chavan**
Department of Pathology, MIMSR Medical College, Latur, Maharashtra 4132512, India
**Author for Correspondence*

ABSTRACT
Splenic lymphangiomatosis is an unusual, benign entity, characterized by the presence of varying sized multiple cystic lesions replacing all the splenic parenchyma. It is considered to result from developmental failure in the lymphatic system. Herein, we are reporting an interesting case of splenic lymphangiomatosis in 27 years old female with huge Splenomegaly along with review of literature, diagnosis of which was done on core biopsy and confirmed by immunohistochemistry. Elective total splenectomy was performed and patient is doing well on follow up since last 6 months.

Keywords: *Lymphangiomatosis, Spleen, Core Biopsy, Immunohistochemistry*

INTRODUCTION
Lymphangiomatosis of spleen is a unique, rare disorder that occurs predominantly in children and young adults less than 20 years of age (Laverdiere *et al.,* 2000). It is rare in persons more than 20 years of age. It is considered to be a developmental anomaly and can occur alone or be part of multi organ disease. Histologically, the lesion is composed of varying sized cystic spaces filled with lymph lined by flat endothelium and surrounded by fibrous connective tissue (Fisher and Hiller, 1994; Pernick Nat. (online); Patti *et al.,* 2010). In the majority of cases, this disease has vague clinical presentation and, despite the use of modern imaging techniques, often makes preoperative diagnosis difficult. In such scenario of nonspecific features of this disease and absence of signs of hepersplenism, core biopsy can be used as an effective diagnostic tool to arrive at correct pathological diagnosis by exclusion of other malignant lesions and to aid proper treatment.

CASES
A 27 year–old woman presented to surgery department of YCR Hospital Latur, with history of vague pain in the left side of the abdomen. There was no fever, dyspepsia or weight loss. Physical examination showed no pallor or lympadenopathy.
Spleen was grossly enlarged- 10 cms below the costal margin. In view of clinical findings, initial differential diagnoses kept were myelofibrosis or low grade lymphoma. Complete haemogram results were within normal limits.
The platelet count was 175000/cumm. There was no evidence of bleeding, consumptive coagulopathy, hypersplenism and portal hypertension. Bone marrow aspirate and biopsy were normal did not reveal any lymphoproliferative diorder. Ultrasonography and CT scan of the abdomen showed splenomegaly with cystic lesions. In view of normal platelet count and absence of signs of hypersplenism, USG Guided FNAC (Fine Needle Aspiration Cytology) of the mass was tried from the cysts with 22 guage needle on aspiration 1 ml yellowish fluid was obtained**.** The smears were prepared from the centrifuged sediments showed abundant eosinophilic proteinaceous material along with scattered small lymphocytes. No evidence of lymphoma or any malignancy on the aspirate studied (Figure 1).
On the background of FNAC report, immediately core biopsy was planned to confirm the histopathological diagnosis. Histologically, multiple cystic spaces lined by flat endothelial cells were seen. There was no evidence of any lymphoproliferative disorder or of malignancy (Figure 2). On immunohistochemistry the cells were found to be positive for CD 31 (Figure 3) and negative for AE1/AE3 (pan cytokeratin); confirming the endothelial nature of the lining cells. She underwent elective splenectomy without any complications and doing well on follow up since last 6 months.

Case Report

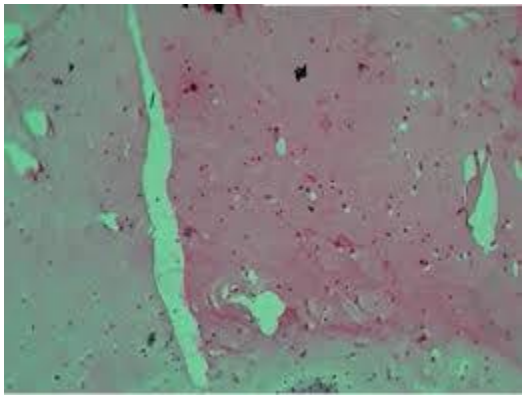

Figure 1: FNAC Showing Abundant Eosinophilic Proteinaceous Material along with Scattered Small Lymphocytes

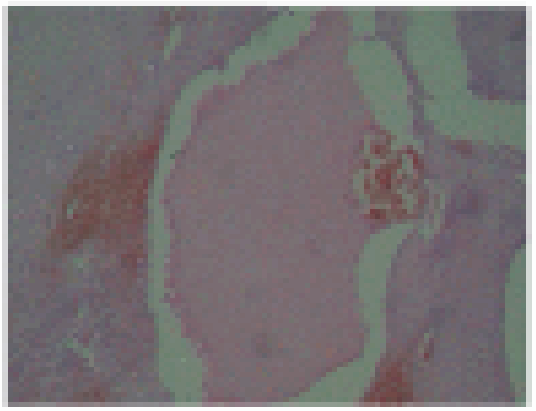

Figure 2: Histologically, Multiple Cystic Spaces Lined by Flat Endothelial Cells were Seen

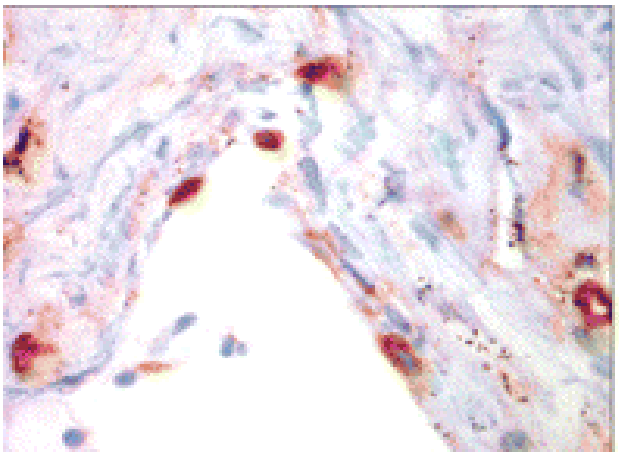

Figure 3: On Immunohistochemistry the Lining Cells were Found to be Positive for CD 31

DISCUSSION

Lymphangiomatosis is a rare, sporadic disorder characterized by cystic lymphangiomas involving multiple organs. Lymphangiomatosis involves bone, soft tissues, and viscera in a diffuse fashion (Huaringa *et al.*, 2005; Marymont and Knight, 1987). Splenic lymphangiomatosis is very rare (Komatsuda *et al.*, 1999; Ros *et al.*, 1986; Morgenstern *et al.*, 1992). It is marked by the presence of cysts of varying size and number. The cysts are thin-walled lymphatic channels that are abnormally interconnected and dilated (Tazelaar *et al.*, 1993). The condition may involve a single organ system and more commonly presents by age 20. Lymphangiomatosis is a benign lesion tends to invade surrounding tissues and cause problems due to invasion and/or compression of adjacent structures (Tazelaar *et al.*, 1993). 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 (Marymont and Knight, 1987).

There are different classifications for cystic lesions of spleen (McClure and Altemeier, 1942; Fowler, 1953; Qureshi and Hafner, 1965; Martin, 1958). The most accepted of these classifications is based on the presence of epithelial lining into primary (true) and secondary (false) cysts (Ingle *et al.*, 2013; Ingle *et al.*, 2014). Primary cysts can be further divided into non parasitic or parasitic (i.e. echinococcal). True nonparasitic cysts include congenital (i.e. epithelial) and neoplastic cysts (lymphangioma, metastases, hemangioma) (Fowler, 1953). False cysts may develop secondary to trauma, hemorrhage, infarction-

degeneration and inammation (Giovagnoni *et al.,* 2005). Histologically, it is classified into three subtypes: Simple (capillary), cavernous and cystic (Takayama *et al.,* 2003).

Isolated splenic lymphangioma can present with different manifestations (Morgenstern *et al.,* 1992). It is asymptomatic in the majority of cases (Chang *et al.,* 2004; Chan and Khoo, 2003). Large cystic lesions may attain sufficient size to cause significant splenomegaly and left-upper quadrant symptoms. Symptoms are usually related to the splenic size (Bevilacqua *et al.,* 1976). The clinical manifestations of lymphangioma are left upper quadrant pain, abdominal distension, loss of appetite, nausea, vomiting and a palpable mass. These are usually nonspecific and are mostly due to compression of adjacent organs, such as stomach, diaphragm or kidney (Ross *et al.,* 1977; Park and Song, 1971; Economides *et al.,* 1980; Konen *et al.,* 2002).

The complications associated with more extensive or larger lymphangiomas of the spleen include bleeding, consumptive coagulopathy, hypersplenism and portal hypertension (Devi and Pillai, 1974; Dietz and Stuart, 1977). The effect of abdominal mass produced by the lymphangioma when it exceeds 3,000 or 4,000 gm can occasionally lead to diaphragmatic immobility and consequent atelectasis or pneumonia (Morgenstern *et al.,* 1985).

Rarely, reversible hypertension due to renal artery compression may be seen (Park and Song, 1971; Economides *et al.,* 1980). Acute abdominal pain, or a rapid increase in size of the cyst, may occur because of infection or rupture of a cyst (Moir *et al.,* 1989; Katkhouda *et al.,* 1998; Hashizume *et al.,* 1998; Targarona *et al.,* 1998).

USG commonly shows hypoechoic spaces which may contain internal echoes (Kawashima and Fishman, 1994; Asch *et al.,* 1974). Although, CT scan is nonspecific, CT scans usually demonstrate low density (Kaza *et al.,* 2010), multiple thin-walled, sharply marginated subcapsular cysts, which may contain mural calcifications, thus suggesting a diagnosis of cystic lymphangioma (Ito *et al.,* 1995; Abbott *et al.,* 2004; Rolfes and Ros, 1990).

On MRI, the mass is shown as multiloculated hyper-intensity areas on the T2-weighted images, thus, corresponding to the dilated lymphatic spaces whereas T1 imaging is only slightly increased (Solomou *et al.,* 2003). This is usually due to protein aceous or hemorrhagic content (Urrutia *et al.,* 1996). The septa are demonstrated as hypointensity bands, corresponding to an abundant amount of fibrous connective tissue. The MR findings typically correlate well with the histologic findings (Pistoia and Markowitz, 1988). A characteristic 'Swiss cheese' appearance of the spleen has been considered pathognomonic (Avigad *et al.,* 1976; Tuttle and Minielly, 1978).

The correct diagnosis depends on histopathologic examination after removal of the spleen. Histochemical staining of the endothelium demonstrates reactivity with CD31, CD34, factor VIII-related antigen and keratin to varying degrees (Alkofer *et al.,* 2005; Qutub *et al.,* 2006).

Lesions that are symptomatic have generally been treated by a splenectomy (Morgenstern *et al.,* 1992) Laparoscopic splenectomy (LS) is emerging as the procedure of choice for splenic removal in patients with a normal to moderately enlarged spleen, benign tumors and hypersplenism (Schlinkert and Teotia, 1999; Caprotti *et al.,* 1998; Klingler *et al.,* 1998). Successful laparoscopic complete excision of a splenic lymphangioma was first reported by Kwan *et al.,* (2001). LS recommended when a splenic tumor is suspected to be either benign or borderline (Maluenda *et al.,* 2004; Sellers and Starker, 1997; Fahel *et al.,* 2000; Comitalo, 2001). However, massive splenomegaly has been considered a contraindication for LS (Targarona *et al.,* 1998; Maurus *et al.,* 2008). During surgery, both open and laparoscopic; the search for accessory spleens is an important step. These must be removed even if they appear macroscopically normal, because they could be involved in the pathological process (Barrier *et al.,* 2002; Khan *et al.,* 1986).

The post operative recurrence rate and the rate of transformation into malignancy is low and the prognosis is good (Witzel *et al.,* 1999). Our case is doing well on follow up since last 6 months. There have been very few reports of a splenic lymphangioma developing into malignant lymphangio-sarcoma (Feigenberg *et al.,* 1983). Complications following surgery may include peritonitis, bleeding, abscess and torsion (Yang *et al.,* 2004).

Case Report

Differential diagnosis that always must be borne in mind include but are not limited to hydatid cysts, pancreatic pseudocysts, splenic cysts, cystic hamartomas and hemangiomas, coelomic cysts and cystic intestinal remnants (Ferrozzi *et al.,* 1996).

Conclusion

Diffuse lymphangiomatosis is an unusual pathological entity having vague clinical presentation. In such scenario core biopsy can be used as an effective diagnostic tool to arrive at correct diagnosis by excluding other malignant lesions and to aid early diagnosis and proper treatment plan of the lesion, thus, preventing untoward complications later in the course of disease i.e. bleeding, consumptive coagulopathy, hypersplenism and portal hypertension.

REFERENCES

Abbott RM, Levy AD, Aguilera NS, Gorospe L and Thompson WM (2004). From the archives of the AFIP: Primary vascular neoplasms of the spleen: Radiologic-pathologic correlation. *Radiographics* **24** 1137-63.

Alkofer B, Lepennec V and Chiche L (2005). Splenic cysts and tumors: Diagnosis and management. *Journal de Chirurgie (Paris)* **142** 6-13.

Asch MJ, Cohen AH and Moore TC (1974). Hepatic and splenic lymphangiomatosis with skeletal involvement: Report of a case and review of the literature. *Surgery* **76** 334-39.

Avigad S, Jaffe R, Frand M, Izhak Y and Rotem Y (1976). Lymphangiomatosis with splenic involvement. *Japan Automobile Manufacturers Association (JAMA)* **236** 2315-17.

Barrier A, Lacaine F, Callard P and Huguier M (2002). Lymphangiomatosis of the spleen and 2 accessory spleens. *Surgery* **131** 114-16.

Bevilacqua G, Toni G and Tuoni M (1976). A case of cavernous haemangioma of the spleen. *Tumori* **62** 485-92.

Caprotti R, Porta G, Franciosi C, Codecasa G, Romano F, Musco F *et al.*, (1998). Laparoscopic splenectomy for hematological disorders. Our experience in adult and pediatric patients. *International Surgery* **83** 303-07.

Chan IYF and Khoo J (2003). Retroperitoneal lymphangioma in an adult. *Journal of Hong Kong College of Radiologists* **6** 94- 96.

Chang CH, Hsieh CB, Yu JC and Jan CI (2004). A case of lymphangioma of the spleen. *Journal of Medical Science* **24** 109-12.

Comitalo JB (2001). Laparoscopic treatment of splenic cysts. *Journal of the Society of Laparoendoscopic Surgeons* **5** 313-3.

Devi NN and Pillai PG (1974). Cystic lymphangioma of the spleen: A case report. *Indian Journal of Pathology & Bacteriology* **17** 61-62.

Dietz WH Jr and Stuart MJ (1977). Splenic consumptive coagulopathy in a patient with disseminated lymphangiomatosis. *Journal of Pediatrics* **90** 421-23.

Economides NG, Benton BF, Fortner TM and Miles RM (1980). Splenic pseudocysts: Report of two cases and review of the literature. *American Surgeon* **46** 644-48.

Fahel E, Amaral PCG and Filho EMA (2000). Videolaparoscopic approach of the splenic cyst: A case report. *Journal of the Society of Laparoendoscopic Surgeons* **4** 23-26.

Feigenberg Z, Wysenbeek A, Avidor E *et al.*, (1983). Malignant lymphangioma of the spleen. *Israel Journal of Medical Sciences* **19** 202-04.

Ferrozzi F, Bova D, Draghi F and Garlaschi G (1996). CT findings in primary vascular tumors of the spleen. *American Journal of Roentgenology (AJR)* **166** 1097-101.

Fisher D and Hiller N (1994). Case report: Giant tuberculous cystic lymphangioma of posterior mediastinum, retroperitoneum and groin. *Clinical Radiology* **49** 215-216 PMID:8143418.

Fowler RH (1953). Collective review: Non parasitic benign cystic tumors of the spleen. *International Abstracts of Surgery* **96** 209-27.

Giovagnoni A, Giorgi C and Goteri G (2005). Tumours of the spleen. *Cancer Imaging* **5**(1) 73-77.

Hashizume M, Tanoue K, Morita M, Ohta M, Tomikawa M and Sugimachi K (1998). Laparoscopic gastric devascularization and splenectomy for sclerotherapy-resistant esophagogastric varice with hypersplenism. *Journal of the American College of Surgeons* **187** 263-70.

Huaringa AJ, Chittari LM, Herzog CE, Fanning CV, Haro M & Eftekhari F (2005). Pleuro-Pulmonary Lymphangiomatosis: Malignant behavior of a benign disease. *The Internet Journal of Pulmonary Medicine* **5**(2). Available: http://ispub.com/IJPM/5/2/9120.

Ingle SB, Hinge CR and Jatal SN (2013). An interesting case of primary epithelial cyst of spleen. *Indian Journal of Pathology and Microbiology* **56** 181–182.

Ingle SB, Hinge Ingle CR and Patrike S (2014). Epithelial cysts of the spleen: a mini review. *World Journal of Gastroenterology* **20**(38) 13899-903.

Ito K, Murata T and Nakanishi T (1995). Cystic lymphangioma of the spleen: MR findings with pathological correlation. *Abdom Imaging* **20** 82-84.

Katkhouda N, Hurwitz MB, Rivera RT *et al.*, (1998). Laparoscopic splenectomy: Outcome and efficacy in 103 consecutive patients. *Annals of Surgery* **228** 568-78.

Kawashima A and Fishman EK (1994). Benign and malignant splenic lesions. In: Gore RM, Levine MS, Laufer I (edition), *Textbook of Gastrointestinal Radiology*, (USA, Philadelphia: Saunders) 2251-8.

Kaza RK, Azar S, Francis IR *et al.*, (2010). Primary and secondary neoplasms of the spleen. *Cancer Imaging* **10** 173-82.

Khan AH, Bensoussan AL, Ouimet A, Blanchard H, Grignon A and Ndoye M (1986). Partial splenectomy for benign cystic lesions of the spleen. *Journal of Pediatric Surgery* **21** 749-52.

Klingler PJ, Tsiotos GG, Glaser KS and Hinder RA (1998). Laparoscopic splenectomy: Evolution and current status. *Surgical Laparoscopy Endoscopy* **9** 1-8.

Komatsuda T, Ishida H, Konno K, Hamashima Y, Naganuma H, Sato M, Ishida J and Masamune O (1999). Splenic lymphangioma: US and CT diagnosis and clinical manifestations. *Abdominal Imaging* **24** 414-417.

Konen O, Rathaus V, Dlugy E *et al.*, (2002). Childhood abdominal cystic lymphangioma. *Pediatric Radiology* **32** 88-94.

Kwon A-H, Inui H, Tsuji K, Takai S, Imamura A and Kamiyama Y (2001). Laparoscopic splenectomy for a lymphangioma of the spleen: Report of a case. *Surgery Today* **31** 258-61.

Laverdiere C, David M, Dubois J, Russo P, Hershon L and Lapierre JG (2000). Improvement of disseminated lymphangiomatosis with recombinant interferon therapy. *Pediatric Pulmonology* **29** 321-324 PMID:10738021.

Maluenda FG, Burdiles PP, Braghetto IM and Csendes AJ (2004). Hematologic diseases laparoscopic splenectomy. *Revista Médica de Chile* **132** 189-94.

Martin JW (1958). Congenital splenic cysts. *The American Journal of Surgery* **96** 302-08.

Marymont JV and Knight PJ (1987). Splenic lymphangiomatosis: A rare cause of splenomegaly. *Journal of Pediatric Surgery* **22** 461-462.

Maurus CF, Schäfer M, Müller MK, Clavien PA and Weber M (2008). Laparoscopic versus open splenectomy for nontraumatic diseases. *World Journal of Surgery* **32** 2444-49.

McClure AD and Altemeier WA (1942). Cysts of the spleen. *Annals of Surgery* **116** 98-102.

Moir C, Guttman F, Jequier S, Sonnino R and Youssef S (1989). Splenic cysts: Aspiration, sclerosis or resection. *Journal of Pediatric Surgery* **24** 646-48.

Morgenstern H, Rosenberg J and Geller SA (1985). Tumors of the spleen. *World Journal of Surgery* **9** 468-76.

Morgenstern L, Bello JM, Fisher BL and Verham RP (1992). The clinical spectrum of lymphangiomas and lymphangiomatosis of the spleen. *American Surgeon* **58** 599-604.

Park JY and Song KT (1971). Splenic cyst: A case report and review of literature. *American Surgeon* **37** 544-47.

Patti R, Iannitto E and Di Vita G (2010). Splenic lymphangiomatosis showing rapid growth during lactation: A Case Report. *World Journal of Gastroenterology* **16**(9) 1155-1157. PMID:20205289.

Pernick Nat. (2009). *Soft Tissue Tumors Part 3 Muscle, Vascular, Nerve, Other Lymphangiomatosis.* Available: PathologyOutlines.com, Inc., 10/17/2009. [Accessed 6 Sep 2011] PMID:20205289.

Pistoia F and Markowitz SK (1988). Splenic lymphangiomatosis: CT diagnosis. *American Journal of Roentgenology (AJR)* **50** 121-22.

Qureshi MA and Hafner CD (1965). Clinical manifestations of splenic cysts: Study of 75 cases. *American Surgeon* **31** 605-08.

Qutub W, Lewis K, Gonzalez R, Quaife R, Russ P and McCarter M (2006). Lymphangiomatosis masquerading as metastatic melanoma. *American Surgeon* **72** 367-70.

Rolfes RJ and Ros PR (1990). The spleen: An integrated imaging approach. *Critical Reviews in Diagnostic Imaging* **30** 41-83.

Ros PR, Goodman ZD, Ishak KG, Dachman AH, Olmsted WW, Hartman DS and Lichtenstein JE (1986). Mesenchymal hamartoma of the liver: radiologic – pathologic correlation. *Radiology* **158** 619-624.

Ross ME, Ellwood A, Yang SS and Lucas RJ (1977). Epidermoid splenic cysts. *Archives of Surgery* **112** 596-99.

Schlinkert RT and Teotia SS (1999). Laparoscopic splenectomy. *Archives of Surgery* **134** 99-103.

Sellers GJ and Starker PM (1997). Laparoscopic treatment of to benign splenic cyst. *Surgical Endoscopy* **11** 766-68.

Solomou EG, Patriarheas GV, Mpadra FA, Karamouzis MV and Dimopoulos I (2003). Asymptomatic adult cystic lymphangioma of the spleen: Case report and review of the literature. *Magnetic Resonance Imaging* **21** 81-84.

Takayama A, Nakashima O, Kobayashi K and Kojiro M (2003). Splenic lymphangioma with papillary endothelial proliferation: A case report and review of the literature. *Pathology International* **53** 483-88.

Targarona EM, Espert JJ, Balague C, Piulachs J, Artigas V and Trias M (1998). Splenomegaly should not be considered a contraindication for laparoscopic splenectomy. *Annals of Surgery* **228** 35-39.

Tazelaar HD, Kerr D, Yousem SA, Saldana MJ, Langston C and Colby TV (1993). Diffuse pulmonary lymphangiomatosis. *Human Pathology* **24**(12) 1313-22.

Tuttle RJ and Minielly JA (1978). Splenic cystic lymphangiomatosis. *Radiology* **126** 47-48.

Urrutia M, Mergo PJ, Ros LH, Torres GM and Ros PR (1996). Cystic masses of the spleen: Radiologic-pathologic correlation. *Radiographics* **16** 107-29.

Witzel K, Kronsbein H, Pleser M, Hunfeld H and Rumpf KD (1999). Intra-abdominal cystic lymphangioma in childhood. Report of 2 cases. *Zentralblatt für Chirurgie* **124** 159-62.

Yang DM, Jung DH, Kim H, Kang JH, Kim SH, Kim JH *et al.*, (2004). Retroperitoneal cystic masses: CT, clinical, and pathologic findings and literature review. *Radiographics* **24** 1353-65.