AN INTERESTING CORE BIOPSY DIAGNOSIS: DIFFUSE LYMPHANGIOMATOSIS OF SPLEEN

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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 a 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 hypersplenism, 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 lymphadenopathy.
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 disorder. 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.
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 (lymphangiomia, metastases, hemangioma) (Fowler, 1953). False cysts may develop secondary to trauma, hemorrhage, infarction-
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degeneration and inflammation (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; Rolves 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 proteinaceous 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 (Schlunkert 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).
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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


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