*IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)*
*e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 16, Issue 4 Ver. IV (April. 2017), PP 125-129*
*www.iosrjournals.org*

# Splenic Cystic Lymphangioma, A Rare Cause Of Massive Splenomegaly In Adults: A Case Report and review of Literature.

Dr Promise N Wichendu, Dr Emmanuel O Afolabi
*Department Of Surgery, University Of Port Harcourt Teaching Hospital, Choba, Port Harcourt, Nigeria.*

***Abstract:***
***Introduction:*** *Lymphangioma of the spleen is a very rare entity in adults, we therefore deemed it necessary to share this finding with colleagues and highlight the diagnostic challenges inherent in arriving at a preoperative diagnosis.*
***Case presentation:*** *A 26 year old female presented to the surgical outpatient department of the hospital with a history of left upper abdominal swelling of 14 years duration. The spleen was markedly enlarged on abdominal examination extending from the left hypochondrium to the right iliac fossa. An abdominal ultrasound also revealed a massively enlarged and echogenic spleen with multiple lobulations and multicystic areas, extending from the left hypochondrium, to just above the uterus with no calcification. In view of the long standing duration of the symptoms, the persistent complain of a nagging abdominal pain,and financial constraints in carrying out further investigations, a Splenectomy was done and histology confirmed it to be a splenic lymphangioma.*
***Conclusion:*** *In spite of the rarity of splenic lymphangioma as a cause of massive splenomegaly in adults, it should be considered as a possible differential especially in resource poor settings.*
***Keywords****: splenomegaly, lymphangioma, splenectomy, hypochondrium, swelling*.

## I. Introduction

Lymphangiomas are benign lesions due to congenital malformation of the lymphatic system [1].They were first described by Rodenber in 1828 [2].They occur mainly in children being very rare in adults [2]. The common locations of lymphangiomas are the neck and the axillary region, where the loose connective tissues allow for easy expansion of the lymphatic channels [3]. Lymphangioma of the spleen is a very rare entity especially in adults. The first case of lymphangioma of the spleen was reported by Frink in 1885 [4], and between 1939 and 2010, only 189 cases of splenic lymphangiomas were reported in the literature [5]. Therefore, splenic lymphangiomas are considered uncommon benign tumors, occurring mainly in childhood, with only a few cases reported in adults.Despite its rarity it should be considered in the differential diagnosis of cystic splenic lesions in adults. Therefore the goal of this report is to bring this possibility into sharp focus by sharing our experience in the management of this rare condition.

## II. Case presentation

A 26 year old married house wife presented to us via the surgical outpatient department with complaints of an abdominal swelling of 14 years duration, and a dragging pain in the abdomen of two year duration. The swelling was initially painless and started from the left upper abdomen. Few months prior to presentation it rapidly increased in size, growing from left upper abdomen across the midline to the right lower abdomen. There was associated persistent abdominal pain that was dragging in nature as well as a burning sensation over the swelling which was aggravated by food. There was a history of easy satiety, and weight loss, but no history of trauma or change in bowel habit. She had a caesarean delivery two years prior to presentation. She was neither hypertensive nor diabetic and did not have sickle cell disease.

On examination she was not in any obvious distress, and was not pale or jaundiced. She looked emaciated otherwise her general condition was satisfactory. Abdominal examination revealed a huge, tender mass extending from the left hypochondrium to the right iliac fossa; we could not get above it and had a notch on the medial border. It was firm in consistency, and lobulated. It measured 25cm below the left costal margin. There was no other palpably enlarged organ in the abdomen. A clinical diagnosis of massive splenomegaly secondary to Tropical splenomegaly syndrome was made. Fig 1.

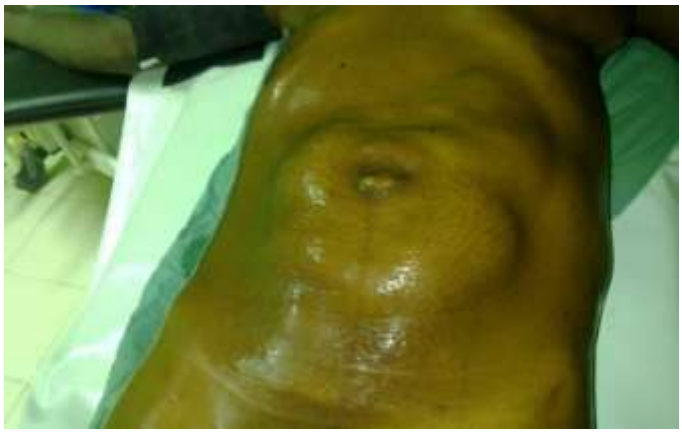


**Fig. 1:** Massive spleen on abdominal examination

Investigations requested include – Haemogram, clotting profile, Igm antibody, Mantoux test, urinalysis and electrolytes urea and creatinine. The results are stated below. Imaging investigations include a chest radiograph, and an abdominopelvic ultrasound.

**Investigations results**:
Mantoux test: 13mm (Positive). Serum IgM: elevated,
**Chest x-ray:** normal
**Abdominal USS:** The spleen was massively enlarged and echogenic with multiple lobulations and cystic areas
Packed cell Volume (PCV) 34%
White Blood Cells: $3.2 \times 10^9/L$
DIFFERENTIALS: Neutrophils 50%, Lymphocytes: 48%
Monocytes: 02%. Platelets 100,000/microliter
HIV I & II: Sero-negative
HBsAg: Negative
HCV Antibody: Negative

While the chest radiograph was normal the ultrasound revealed a massively enlarged echogenic spleen with lobulations as well as multicystic areas. It also showed that the spleen extended from the epigastrium downwards to just above the uterus. There were no calcifications. An abdominal CT and MRI were not done due to financial constraints. A therapeutic trial of antimalarial prophylaxis for one year yielded no benefit. Left with no option and in view of these findings as well as the persistent abdominal pain over the swelling, an elective splenectomy was done figures 2 and 3.

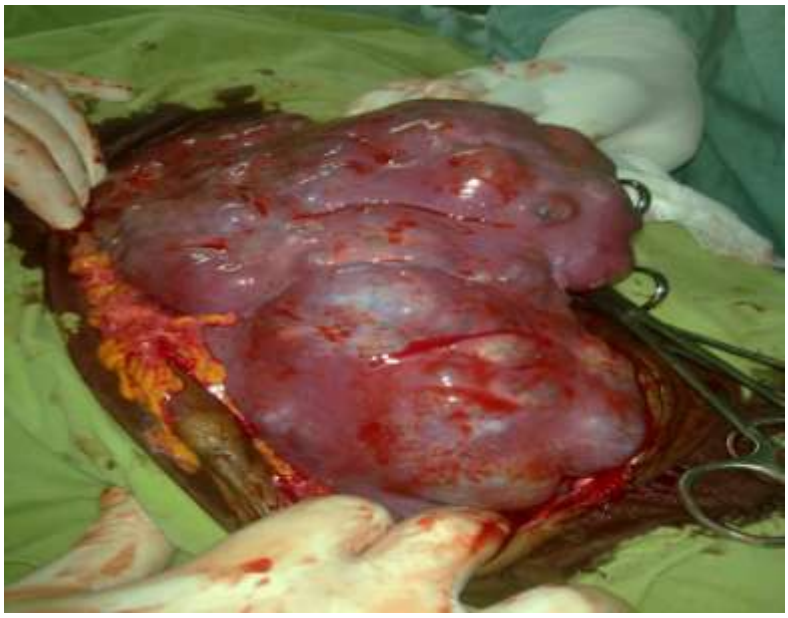


**Fig. 2** The massively enlarged in situ

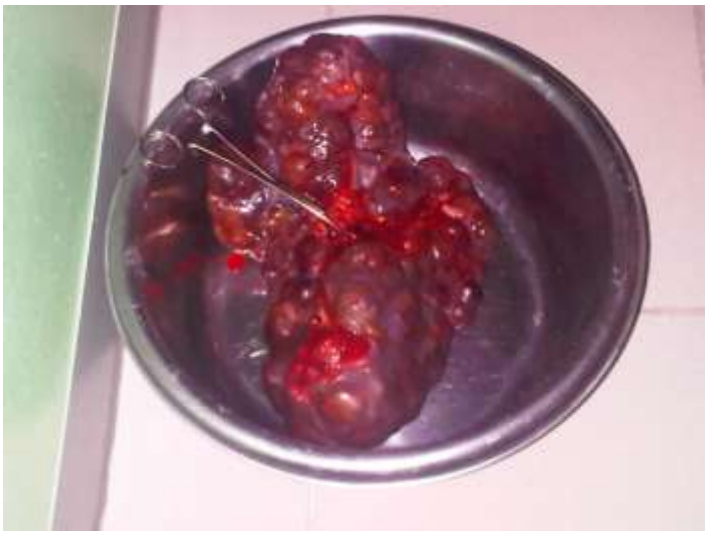
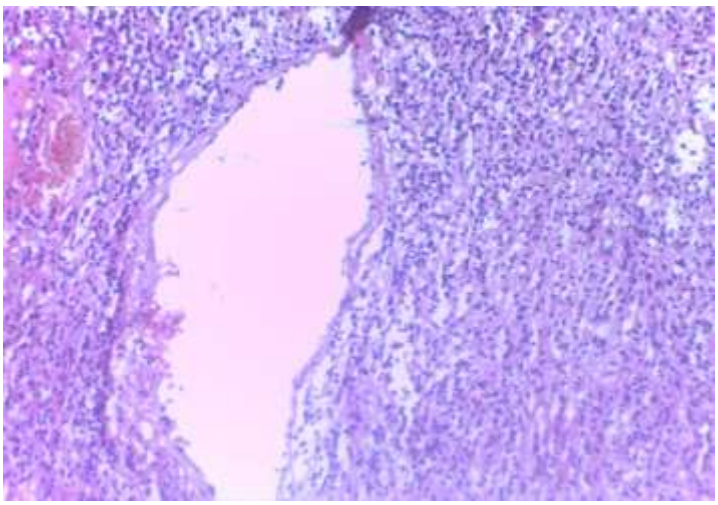


**Fig 3.** The spleen in a basin

The histopathology report revealed the specimen to be a huge multinodular spleen which weighed 2.5kg and measured 25cm x23cm x15cm. The cut surface showed a multinodular and multicystic tissue; some of the cysts contain gelatinous material.

**Microscopy**: The sections of the spleen show numerous large vascular channels that are lined by a single layer of flattened cells. For most part their lumina are filled by eosinophilic fluid; a few contain red blood cells. The splenic parenchyma shows areas of haemorrhage otherwise it is unremarkable. There is no evidence of malignancy. A diagnosis of lymphangioma was made figure 4.



Photomicrograph of the spleen H&E X 100 Vascular space lined by endothelium

## III. Discussion

Lymphangiomas are benign tumours that develop from congenital malformations of lymphatic vessels. They are found to be composed of lymphatic spaces lined by attenuated endothelium[4].Although no consensus has yet been reached on whether splenic lymphangioma is a neoplasm or a hamartoma, most researchers support the latter opinion [4]. Histologically, lymphangiomas are classified into three groups: capillary, cavernous and cystic lymphangiomas [5,6].This nomenclature can be confusing as it is reminiscent of the classification of haemangiomas. The pathogenesis of lymphangioma still remains largely unclear but congenital developmental abnormality of the lymphatic tissue, dilatation of abnormal channels, and localized lymphatic blockage are thought to be important causes [1,7]. Other factors implicated in the pathogenesis include, trauma, vascular endothelial permeability disorders, inflammatory and fibrotic processes [7]. Lymphagiomas are usually found in the neck, axilla, mediastinum, retroperitoneum and other areas rich in lymphoid tissues [8]. About 75% of lymphangiomas are found in the cervical region, 20% in the axillary region, and the remaining 5% are found in

other areas like kidney, bones, and mediastinum [9,10,11]. Abdominal lymphangiomas rarely occur in pancreas, liver, spleen, retroperitoneal and gastrointestinal tract [12, 13, 14]. However ours occurred in the spleen.

Lymphangioma of the spleen can occur sporadically as in our case or as part of a generalized lymphangiomatosis, a condition in which lymphangiomas simultaneously occur in many organs [15]. Splenic lymphangiomas are commoner in females as in the index case and 80-90% are discovered in the second decade of life [16]. Howeverour patient was in the third decade of life. Clinically, the presentation depends on the size of the lesion. Small lesions are asymptomatic while large ones produce symptoms by compressing the adjacent structures like the kidney, duodenum and stomach [17]. Our patient had vague abdominal pains and had even had treatment for dyspepsia in a peripheral health facility. They can present with acute abdominal pain due to rupture or intracystic haemorrhage. Compression of renal artery leading to renal hypertension, have also been reported[18].Rarely, splenic lymphangiomas can be part of Klippel-Trenaunay syndrome (characterized by varicose veins, bony and soft tissue hypertrophy, cutaneous hemangiomas, and or malformations of the lymphatic system) [19].Cases have been reported of larger lymphangiomas complicated by consumptive coagulopathy, bleeding, hypersplenism, and portal hypertension [20]. The pathophysiological consequences of a lymphangioma exceeding 3 to 4 kg can be diaphragmatic immobility and lung atelectasis or pneumonia [21]. Fortunately our patient never had these symptoms probably because it did not attain the requisite weight.

On ultrasound, splenic lymphangioma may appear as a cystic or multicystic lesion with internal septations as in our case. It may be difficult to differentiate from a lymphoma. Colour Doppler interrogation can demonstrate the vasculature of the mass (including the intrasplenic arteries and veins along the cyst walls). It may assist in determining the splenic origin by demonstrating the vessels at the splenic hilum [22].

Computed tomography usually shows low-density single or multiple thin-walled sharply marginated subcapsular cysts.The presence of curvilinear peripheral mural calcifications is suggestive of cystic lymphangiomas but is not a specific finding because it can also be seen in hydatid cyst [23]. On T1-weighted magnetic resonance imaging, the cystic lesions can appear hypointense relative to the surrounding viscera or hyper intense when filled with haemorrhagic or proteinaceous material. On T2-weighted images, the mass is characterized by multiloculated hyper intense areas that correspond to the dilated lymphatic channels [22]. PET scan can confirm that the lesion is benign by revealing no fludeoxyglucose uptake [23]. Sadly our patient could not afford these two imaging modalities that would have been very useful in making a preoperative diagnosis.

On histology it typically shows as vascular spaces lined by endothelium as in our case (figure 4). When the histologic characteristics are not clear, the endothelial origin of the cyst may be established with immunohistochemical techniques to demonstrate reactivity for CD31, CD34, factor VIII, podoplanin (D2-40), or vascular endothelial growth factor receptor 3 (VEGFR-3). D2-40 is a monoclonal antibody against dysgerminoma, observed to selectively stain lymphatic endothelium. The D2-40 antibody was also found to positively stain lymphangiomas but not benign tumours of blood vessels [24].These were not indicated in our patient as the histologic findings were unequivocal.

In resource poor settings such as ours the preoperative diagnostic dilemma is even more apt as in our case who could not afford further diagnostic tests like magnetic resonance imaging or computerised axial tomography that could have help refine the preoperative diagnosis. We had to resort to splenectomy and histopathology as the only way to identify the nature of the splenic mass while relieving her symptoms. Percutaneous biopsy was not an option because of the risk of haemorrhage, poor sample and peritoneal seeding if the lesion is malignant.

The treatment of choice for splenic lymphangiomas is complete surgical resection because other therapeutic modalities (aspiration, drainage, sclerosis and irradiation) have shown unsatisfactory results [25]. This is in line with our treatment option. Some investigators prefer conservative treatment in the case of small asymptomatic lesions detected incidentally, reserving splenectomy for large, multiple, or symptomatic lesions like ours [26]. Partial resection techniques have also been applied in cases of limited disease; however, leaving a splenic remnant in cases of diffuse involvement increases the risk of further growth and enlargement, sometimes requiring a second operation. Laparoscopic splenectomy is emerging as the procedure of choice in patients with a normal to moderately enlarged spleen but is considered a contraindication in patients like ours with massive splenomegaly[26]. The most important aspect when considering treatment options is that surgery should be recommended immediately after the diagnosis has been established to avoid complications such as infection, haemorrhage, rupture, intestinal obstruction, and tumour enlargement that may eventually prevent complete removal [27].

The prognosis of splenic lymphangioma after resection is favourable. The main complication is recurrence, which is demonstrated in 9.5% of patients, frequently after incomplete resection [22]. Our patient was in good health as regards the tumour until she was lost to follow up after one year.

## IV. Conclusion

This is a case with limited preoperative diagnostic work up, however splenectomy did resolve all the patient's symptoms, and provided specimen for histopathological which revealed the exact nature of the mass. Therefore, splenic lymphangiomas should be considered in the differential diagnosis of splenomegaly or left upper quadrant pain even among adults because they are amenable to curative treatment with good prognosis. Lack of modern diagnostic equipment should not delay or deny the patient surgical therapy especially in resource limited settings such as ours. Lessons in this wise should be learnt from our experience. Delay in surgical intervention may lead to severe complications [27].

## References


[1]. Stavropoulus M, Vagianos C, Scopa C D, Dragotis C, Androulakis J. Solitary hepatic lymphangioma. A rare benign tumour: a case report. HPB surgery8, 1994, 33-36.
[2]. Avigad S, Jaffe R, Frand M, Izhak Y, Rotem Y. Lymphangiomatosis with splenic involvement. JAMA,236, 1976, 2315–2317.
[3]. Anadol AZ, Oguz M, Bayramoglu H, Edali MN. Cystic lymphangioma of the spleen mimicking hydatid disease. J. clinical Gastroenterol, 26,1998, 309-411.
[4]. Chung SH, Park YS, Jo YJ, Kim SH, Jun DW, Son BK, Jung JY et al. Asymptomatic lymphangioma involving the spleen and retroperitoneum in adults. World JGastroenterol, 15, 2009, 620–623.
[5]. Alerci M, Dore R. Computed tomography of cystic lymphangioma in a wandering spleen. Acta Radiol, 31, 1990, 589-590.
[6]. Asch MJ, Cohen AH, Moore TC. Hepatic and splenic lymphangiomatosis of the skeletal involvement: report of a case and review of literature. Surgery, 76, 1974; 334-335.Pubmed abstract.
[7]. Enzinger FM, Weiss SW, Soft tissue tumours. (Mosby6th Edition, St. Louis, 1998) 614-637.
[8]. Losanoff JE, Richman BW, El-Sheriff A, Rider KD, Jones JW. Mesenteric cystic lymphangioma. J Am coll surg,196, 2003: 598-603.
[9]. Chang CH, Hsieh CB, Yu JC, Jan CI. A case of lymphangioma of the spleen. J med Sci,24, 2004, 109-112.
[10]. Levy AD, Cantisani V, Miettinen M. Abdominal lymphangiomas: imaging features with pathologic correlation. AJR AM J Roentgenol,182, 2004,1485-1491
[11]. Pistoia F, Markowitz SK. Splenic lymphangiomatosis: CT diagnosis. AJR Am JRoentgenol, 150, 1988, 121-122.
[12]. Bliss DP Jr, Coffin CM, Bower RJ, Stockman PT, Ternberg JL. Mesenteric cysts in children. Surgery, 115(5), 1994. 571-577.
[13]. Shigematsu A, I'ida M, Hatanaka M, Kehrogi N, Matsui T, Fujishima M, Itoh, etal. Endoscopic diagnosis of lymphangioma of the small intestine. Am J Gastroenterol, 83,1998, 1289-1293
[14]. Koshy A, Tandon RK, Kapur BM, Rao KV, Joshi K. Retroperitoneal lymphangioma. A case report with review of the literature. Am J Gastroenterol, 64, 1978, 485- 490
[15]. Vezzoli M, Ottini E, Montagna M, La fianza A, Paulli M, Rosso R, Mazzone A. Lymphangioma of the spleen in an elderly patient. Haematologica, 85,2000, 314-317.
[16]. Morgenstern L, Bello JM, Fischer BL, Verham RP.The Clinical spectrum of lymphangiomas and lymphangiomatosis of the spleen. Am Surg, 58(10), 1992, 599-604.
[17]. Dachman AH, Ros PR, Murari PJ, Olmsted WW, Lichtenstein JR. Nonparasitic splenic cysts. A report of 52 cases with radiologic-pathologic correlation. Am J Roentgenol, 147, 1986, 537-542
[18]. Jindal R, Sullivan R, Rodda B, Arun D, Hamady M, Cheshire NJ. Splenic malformation in a patient with Klippel-Trenaunay syndrome: a case report. J Vasc Surg, 43, 2006, 848–850.
[19]. Dietz WH Jr, Stuart MJ. Splenic consumptive coagulopathy in a patient with disseminated lymphangiomatosis. J Pediatr, 90, 1977, 421–423.
[20]. Moir C, Guttman F, Jequier S, Sonnino R, Youssef S. Splenic cysts: aspiration, sclerosis, or resection. J Pediatr Surg, 24, 1989, 646–648.
[21]. Abbott RM, Levy AD, Aguilera NS, Gorospe L, Thompson WM. From the archives of the AFIP: primary vascular neoplasms of the spleen: radiologic pathologic correlation. Radiographics, 24, 2004, 1137–1163.
[22]. Eghtedari M, Sicklick J, Kono Y, Peterson MR, Santillan CS. Unusual
[23]. Imaging profile of a solitary splenic lymphangioma. Acta Radiol Short Rep, 1(8), 2012, Pii: arsr 2012.120033.
[24]. Debelenko LV, Perez-Atayde AR, Mulliken JB, Liang MG, Archibald TH, Kozakewich HP. D2-40 immunohistochemical analysis of pediatric vascular tumors reveals positivity in kaposiform hemangioendothelioma. Mod Patho, 18(11), 2005, 1454– 1460.
[25]. Makrin V, Avital S, White I, Sagie B, Szold A. Laparoscopic splenectomy for solitary splenic tumors. Surg Endosc, 22(90), 2008, 2009–2012.
[26]. Maurus CF, Schafer M, Muller MK, Clavien PA, Weber M. Laparoscopic versus open splenectomy for nontraumatic diseases. World J Surg, 32, 2008, 2444–2449.
[27]. Yu X, Yu J, Liang P, Liu F. Real-time contrast-enhanced ultrasound in diagnosing of focal spleen lesions. Eur J Radiol, 81, 2012, 430–436.
[28]. Roisman .I, Manny .J, Fields .S. Shilon .E. Intra-abdominal lymphangioma. Br. J. Surg,76, 1989, 485-489