

Case Report

Isolated Cystic Splenic Lymphangioma in an Adult Female in South-South Nigeria: A Case Report

Ima-Abasi Bassey*¹, Theophilus Ugbem¹, Inyama Marcua² and Ima-Obong Ekanem¹¹Department of Pathology, University of Calabar Teaching Hospital, Calabar, Nigeria²Department of Haematology, University of Calabar Teaching Hospital, Calabar, Nigeria**Article History**

Received: 17.07.2020

Accepted: 27.07.2020

Published: 10.08.2020

Journal homepage:<https://www.easpublisher.com/easjms>**Quick Response Code**

Abstract: Splenic cystic lymphangioma in young adult patients is very rare and should be considered as a differential diagnosis of splenomegaly in malaria-endemic region. We report the case of a 24-year-old female who presented with a 28 cm left sided painless abdominal swelling of 7 years duration. Her physical examination revealed severe anaemia and a massive splenomegaly. An abdominal ultrasound scan was not conclusive. A splenectomy was performed and histopathological analysis led to the diagnosis of cystic splenic lymphangioma. The patient's postoperative course was uneventful, and she was discharged from the hospital. Histopathological analysis of the resected specimen is the gold standard for definitive diagnosis in such cases.

Keywords: Splenomegaly, Lymphangioma, Pathology, Immunohistochemistry, Diagnosis, Tropics.

Copyright @ 2020: This is an open-access article distributed under the terms of the Creative Commons Attribution license which permits unrestricted use, distribution, and reproduction in any medium for non-commercial use (Noncommercial, or CC-BY-NC) provided the original author and source are credited.

INTRODUCTION

Lymphangiomas are uncommon lesions of lymphatic channels that are often present at birth and diagnosed mostly (90%) before the age of two years (Kraus, J. *et al.*, 2008). These primary benign tumours of the spleen account for less than 0.007% of all tumours identified at surgery and autopsy (Raza, M. *et al.*, 2018). They are commonly found in the neck, axillary regions and less frequently, in the mediastinum, retroperitoneum, extremities and visceral organs (Ioannidis, I., & Kahn, A. G. 2015). The condition may be restricted to the spleen, but in most cases it involves multiple organs as part of a systemic lymphangiomatosis. Splenic lymphangioma is a very rare condition and is usually found incidentally especially in adults but manifests mainly in children (Chandrasinghe, P. C. *et al.*, 2012). Isolated splenic lymphangiomas constitute a much rarer type (Efareed, B. *et al.*, 2018).

The clinical presentation of splenic lymphangiomas is usually related to size of the spleen, but they can be asymptomatic or incidental findings during radiological assessment for other reasons. If they are large, they can cause abdominal pain, loss of

appetite, nausea, vomiting and may compress or block adjacent organs (Chang, W. C. *et al.*, 2007). The uncommon solitary localization could pose a challenge to clinicians in making an accurate pre-operative diagnosis.

This is a case report of an isolated splenic lymphangioma in a 24-year-old woman who presented with left sided painless abdominal swelling of seven years duration. The patient underwent splenectomy and diagnosis was confirmed by histopathological and ancillary immunohistochemical studies.

CASE REPORT

A 24 year old female presented with a progressively enlarging left sided painless abdominal swelling of 7 years duration. The swelling was associated with weight loss and dyspnoea which made her seek medical attention in our facility. There was no previous history of intermittent fever, trauma and swelling around the neck region.

Physical examination revealed a young severely pale wasted young female with axillary lymphadenopathy and a prominent left-sided abdominal

mass extending towards the right iliac fossa. The mass measuring about 28cm in maximum dimension had a smooth surface and a medial indentation. Haematological indices were within normal range with exception of the packed cell volume indicating severe anaemia. Electrolyte and urea were normal. However, abdominal ultrasonography was inconclusive. Peripheral blood smear and bone marrow aspirate carried out were normal. Suspected splenic lymphoma was ruled out and the patient had a laparotomy and splenectomy was the choice of treatment carried out by the surgical team. The entire tissue was submitted for thorough histopathological examination.

Macroscopic examination revealed a splenectomy specimen weighing 3.6kg and measuring 28x17x15cm. Its external surface was nodular, greyish-tan to dark-red with a firm consistency. (Fig.1) Cut surfaces showed cystic areas of variable sizes

containing straw-coloured fluid with others displaying firm glistening material (Fig.2).

Microscopic sections of the spleen showed variably sized cystically dilated lymphovascular channels containing amorphous eosinophilic material probably lymph and cellular debris with adjacent congested small-sized blood vessels containing red blood cells. The dilated channels lined by bland endothelial cells lacked atypia. (Fig.3 and 4) The stromal infiltrate was predominantly lymphoplasmacytic inflammatory cells with focal formation of follicles without atypia. A diagnosis of Cystic lymphangioma was made following routine Haematoxylin & Eosin staining and ancillary immunohistochemistry which confirmed CD34 positive cells lining the lympho-vascular channels of varying sizes (Fig. 5 and 6).

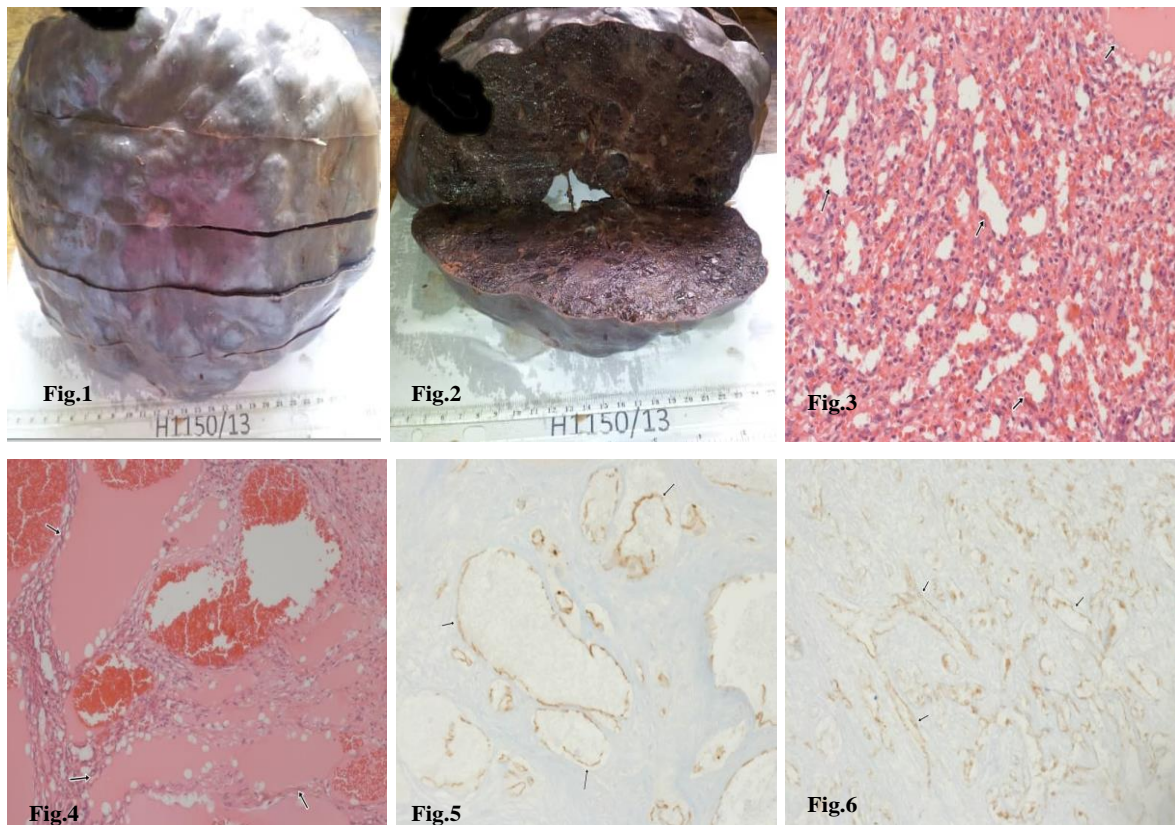


Figure 1&2: Photographs of splenectomy specimen shows nodular, greyish-tan to dark-red external surface with a firm consistency (Fig.1). Cut surfaces showed cystic areas of variable sizes containing straw-coloured fluid (fig.2).

Figure 3&4: Photomicrograph of H&E Sections of the spleen showing variably sized cystically dilated lymphovascular channels containing amorphous eosinophilic material (fig 4x100 and fig 5X400) Figure 5&6: Photomicrograph of immunohistochemistry confirming CD34 positive cells lining the lympho-vascular channels of varying sizes. (x400).

DISCUSSION

Splenic lymphangiomas are relatively rare benign tumours that correspond to abnormal dilatation of lymphatic channels that can be either congenital or acquired. It is characterised by the presence of cysts, resulting from increase in the size and number of thin-

walled lymphatic vessels that are abnormally interconnected and dilated. The cysts tend to be subcapsular and multilocular but can be unilocular. The presentation of cystic lymphangioma in this environment is diverse. There is a high preoperative morbidity with postoperative complications occurring

in about 41.7% and recurrence reported in 4.6% (Kakaje, A. *et al.*, 2020). Cystic lymphangioma has been reported in various sites in Nigeria. Olaoye reported a case of a huge retroperitoneal cystic lymphangioma in a 20 year old lady involving the mesentery, liver and kidney but sparing the spleen (Sowande, O. A. *et al.*, 2003). Recurrent multiloculated genital, thigh, groin and retroperitoneal lymphangioma has also been reported in Nigeria (Olaoye, I. O., & Adesina, M. D. 2018). Interestingly, isolated splenic lymphangioma in adult patients is very rare (Olabanji, J. K. *et al.*, 2009). Available literature search locally, showed that splenic lymphangioma is very rare and in Calabar, Nigeria this happens to be the first reported case in this centre. Because lymphangiomas more commonly involve multiple organs at same time, the diagnostic evaluation should be extended to search for other possible sites affected by the disease (Ioannidis, I., & Kahn, A. G. 2015; & Chandrasinghe, P. C. *et al.*, 2012).

This index patient presented with a huge left-sided painless abdominal mass of 7 year duration and severe anaemia. Considering the duration of presentation and living in a malaria-endemic region a provisional diagnosis of splenic lymphoma and severe malaria was made with a differential diagnosis of Tropical Splenomegaly Syndrome (TSS).

TSS or hyper-immune reactive malarial splenomegaly syndrome (HMSS) is a leading cause of massive splenomegaly in malaria-endemic countries (Olabanji, J. K. *et al.*, 2009). Humaira *et al.*, in a descriptive case study of 100 patients, showed chronic liver disease (64%) to be the most common cause of splenomegaly followed by malaria (16%) and haematological malignancies (14%) (Leoni, S. *et al.*, 2015). Adelusola *et al.*, in a retrospective study of splenectomy specimens in Nigeria, revealed trauma to be the major indication for (62.7%) with road traffic accident (RTA) accounting for 86% of all trauma cases. Haematological malignancy was the next predominant finding in the spleen (16%) with Chronic lymphocytic leukaemia (CLL) being the most common haematological malignancy, which tended to occur in older patients (Humaira, M. *et al.*, 2016). There was no case of lymphangioma in that study.

The blood work-up which included full blood culture, peripheral blood smear analysis and bone marrow aspiration were not conclusive of a lymphoma and there were no palpably enlarged lymph node at the time of presentation for histopathologic assessment in this case. She was referred consequently to the surgical unit for a splenectomy. Total splenectomy is an adequate therapeutic option, especially for large lesions, in order to prevent complications such as splenic rupture, infection, haemorrhage, intestinal obstruction, or recurrence (Adelusola, K. A. *et al.*, 2017; & Yang, F., & Chen, W. X. 2013).

The histopathological assessment was the gold standard for diagnosis in this case as the patient could not afford CT or MR imaging studies due to the cost and these were unavailable in our facility. Solitary lymphangiomas of the spleen are usually found incidentally during ultrasound or CT examinations (Safa A. Al-Shaikh. 2017). In most solitary splenic lymphangiomas, the cystic components predominate, and CT or MRI studies typically show a thin-walled, cystic mass without substantial enhancement (Yang, F., & Chen, W. X. 2013; & Safa A. Al-Shaikh. 2017).

The macroscopic and microscopic findings of cystic lesions varying in number and in size and lined by flattened endothelial cells with cavities containing amorphous acellular eosinophilic secretion are characteristic features. Immunohistochemical analysis confirms the endothelial and lymphatic nature of cells lining the cyst walls, since they stain positive for endothelial markers viz CD34, CD31, factor VIII. Histologically, lymphangiomas are classified into three subtypes according to the congenital dilated lymphatic channels: capillary (super-microcystic), cavernous (microcystic), or macrocystic (Adelusola, K. A. *et al.*, 2017; & Safa A. Al-Shaikh. 2017). Our case was ultimately diagnosed as the macrocystic type.

The aetiology of splenomegaly varies according to the geographical areas and splenic cystic lesions such as pseudocysts caused mainly by trauma, infections such as malaria, schistosomiasis and Kalazar in the tropics or infarction, haemangiomas should be considered as a differential diagnosis although these depend upon the endemic, genetic and haematological diseases in the particular region.

CONCLUSION

Radiologic features of splenic lymphangiomas are not specific and are often misleading (Ioannidis, I., & Kahn, A. G. 2015; Marwah, N. *et al.*, 2014; & Patti, R. *et al.*, 2010). Therefore, splenic lymphangioma, though very rare, should be considered as a differential diagnosis of splenomegaly in malaria-endemic region. Solitary cystic lymphangioma, though a benign tumour, can present with complications such as consumptive coagulopathy, hypersplenism or portal hypertension in patients with larger lesions ((Ioannidis, I., & Kahn, A. G. 2015; Roman, A. *et al.*, 2016; & Abbott, R. M.) and therefore requires therapeutic surgical removal with histopathologic evaluation being the gold standard for definitive diagnosis.

REFERENCES

1. Abbott, R. M., Levy, A. D., Aguilera, N. S., Gorospe, L., & Thompson, W. M. (2004). From the archives of the AFIP: primary vascular neoplasms of the spleen: radiologic-pathologic correlation. *Radiographics*, 24(4), 1137-1163.

2. Adelusola, K. A., Osasan, S. A., & Afolabi, O. A. (2007). Histopathological study and audit of the spleen in Nigerians. *African Journal of Health Sciences*, 14(3), 195-200.
3. Chandrasinghe, P. C., Abeyranthe, I. G. K. P., & Fernando, R. F. (2012). Lymphangioma of the spleen in an asymptomatic patient. *Sri Lanka Journal of Surgery*, 29(2), 100-102
4. Chang, W. C., Liou, C. H., Kao, H. W., Hsu, C. C., Chen, C. Y., & Yu, C. Y. (2007). Solitary lymphangioma of the spleen: dynamic MR findings with pathological correlation. *The British journal of radiology*, 80(949), e4-e6.
5. Efareed, B., Atsame-Ebang, G., Zabeirou, A., Hammam, N., Mazaz, K., El Fatemi, H., & Chbani, L. (2018). Isolated splenic lymphangioma presenting as a huge mass causing anemia and abdominal distension in an adult patient: a case report. *Journal of Medical Case Reports*, 12(1), 97.
6. Humaira, M., Ahmed, A., Memon, A., Siddique, S. A., Hayee, P. A., Ghani, M. H., & Ghouri, R. (2016). Etiological Pattern of Splenomegaly in Adults at Tertiary Care Hospital Jamshoro Sindh. *Journal Of The Liaquat University Of Medical And Health Sciences*, 15(1), 40-45.
7. Ioannidis, I., & Kahn, A. G. (2015). Splenic lymphangioma. *Archives of Pathology and Laboratory Medicine*, 139(2), 278-282.
8. Ise, M., Takenouchi, T., & Sakai, C. (2004). Splenic lymphangiomatosis with inflammatory signs and elevated serum interleukin-6. *Journal of Clinical and Experimental Hematopathology*, 44(2), 77-80.
9. Kakaje, A., Mujahed, R., & Hamdan, O. (2020). Isolated Splenic Lymphangiomas Presenting in an Infant with Isolated Anaemia. *Case Reports in Medicine*, 2020.
10. Kraus, J., Plzák, J., Bruschini, R., Renne, G., Andrlé, J., Ansarin, M., & Betka, J. (2008). Cystic lymphangioma of the neck in adults: a report of three cases. *Wiener klinische Wochenschrift*, 120(7-8), 242-245.
11. Leoni, S., Buonfrate, D., Angheben, A., Gobbi, F., & Bisoffi, Z. (2015). The hyper-reactive malarial splenomegaly: a systematic review of the literature. *Malaria journal*, 14(1), 185.
12. Marwah, N., Sangwan, M., Ralli, M., Verma, R., & Samal, S. (2014). Cystic Lymphangioma of Spleen: A Case Report. *International Journal of Scientific Study*, 2(1), 41-42.
13. Olabanji, J. K., Oladele, O. A., Famurewa, O. C., Adejuyigbe, O., & Ademola, S. A. (2009). Retroperitoneal and Genital Lymphangioma Therapeutic Challenges in a Developing Country. *Libyan Journal of Medicine*, 4(1), 44-45.
14. Olaoye, I. O., & Adesina, M. D. (2018). Rare huge retroperitoneal cystic lymphangioma presenting as acute abdomen in an adult. *BJR/ case reports*, 4(3), 20170120.
15. Patti, R., Iannitto, E., & Di Vita, G. (2010). Splenic lymphangiomatosis showing rapid growth during lactation: a case report. *World journal of gastroenterology: WJG*, 16(9), 1155.
16. Raza, M., Preethi, S. P., Kumar, H., Ali, M. Z., & Jasuja, P. (2018). Splenic lymphangioma: a rare benign tumor in adults. *International Surgery Journal*, 5(10), 3403-3406.
17. Roman, A., Iancu, C., Andreica, V., Socaciu, M., Anton, O., Sechel, R., & Badea, R. (2016). Splenic cystic lymphangioma with atypical ultrasound findings. *Journal of Medical Ultrasonics*, 43(1), 99-105.
18. Safa A. Al-Shaikh. (2017). Splenic lymphangioma in an adult: Case Report. *Saudi Med J*, 38 (11), 1148-1152.
19. Sowande, O. A., Adejuyigbe, O., & Abubakar, A. M. (2003). Management of cystic lymphangiomas in Ile-Ife, Nigeria. *Nigerian Journal of Surgical Research*, 5(1), 32-37.
20. Yang, F., & Chen, W. X. (2013). Splenic lymphangioma that manifested as a solid-cystic mass: a case report. *World Journal of Gastroenterology: WJG*, 19(5), 781-783.