Recent Advances in Pathology & Laboratory Medicine
Volume 6, Issue 1&2 - 2020, Pg. No. 5-7
Peer Reviewed & Open Access Journal

Case Report

Multiple Gastro Intestinal Lymphangiomatosis - Case Report

Gurleen Gill[1], Sarandeep Singh Puri[2]

[1]Post Graduate, [2]Associate Professor, Department of Pathology, School of Medical Sciences and Research, Sharda University, Greater Noida, Uttar Pradesh, India.
DOI: https://doi.org/10.24321/2454.8642.202003

I N F O

Corresponding Author:
Sarandeep Singh Puri, Department of Pathology, School of Medical Sciences and Research, Sharda University, Greater Noida, Uttar Pradesh, India.
E-mail Id:
drsarandeep147@gmail.com
Orcid Id:
https://orcid.org/0000-0002-6523-3070
How to cite this article:
Gill G, Puri SS. Multiple Gastro Intestinal Lymphangiomatosis - Case Report. Rec Adv Path Lab Med. 2020;6(1&2):5-7.

Date of Submission: 2020-05-17
Date of Acceptance: 2020-06-21

A B S T R A C T

Lymphangiomatosis is a result of congenital errors of lymphatic development which takes place before 20 weeks of gestation. By definition Lymphangiomatosis is a benign soft tissue tumour of lymphatic channels. We present one such case of lymphangiomatosis which involved multiple gastrointestinal organs. Lymphangiomatosis is a rare entity which is rarely seen and belongs to spectrum of developmental disorders. Recognition of the disease requires a high index of suspicion and an extensive workup. Because of its serious morbidity, lymphangiomatosis must always be considered in the list of our differential diagnosis.

Keywords: Lymphangiomatosis, Multifocal, Abdomen, Prognosis, Diagnosis, Infiltration

Introduction

Lymphangiomatosis is a result of congenital errors of lymphatic development which takes place before 20 weeks of gestation. 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. By definition Lymphangiomatosis is a benign soft tissue tumour of lymphatic channels. Strikingly Lymphangiomatosis presents as a multifocal lesion.[1,2] We present one such case of lymphangiomatosis which involved multiple gastrointestinal organs.

Case Report

A 46 - year - old male patient came to surgery OPD with complaint of a lump in abdomen since last 12 years. The swelling was painless and progressive in nature. The patient was admitted in the male surgery ward for further workup. History Of Present Illness - Patient was apparently asymptomatic 12 years back when he noticed a swelling in the right lower abdomen, gradually increasing in size. No rapid increase in the size of the swelling. No past medical or surgical history. No other associated complaints. On examination the swelling was 15 x 12 cm in size in right side of abdomen with limited mobility, firm to cystic in consistency, non - tender. Patient could reduce the swelling manually by himself.

USG Whole Abdomen Revealed Liver: Enlarged in size (20 cm) with multiple enlarged cystic lesions are seen in liver with variable echogenicity. The largest cyst measuring approximately 18.5 x 14.5 cm in right lobe of liver with thick internal echoes. Pancreas: Cystic lesions seen in the tail region, largest lesion measuring approximately 7.7 x 5.9 cm. Spleen: Enlarged in size (12.7 cm) with few intraparenchymal cystic lesions, largest measuring approximately 3.4 x 2.4 cm. An anechoic cystic lesion measuring approximately 16.9

x 14.7 x 13.1 cm is seen in right lumbar region extending into right iliac fossa. The lesion shows few internal free floating echoes. Impression on USG was suggestive of Multiple cystic lesions in liver, pancreatic tail, spleen as mentioned above with hepatosplenomegaly.

CECT Scan of Abdomen and Pelvis: Revealed Multiple hepatic, splenic, peritoneal and retroperitoneal cystic lesions. Possibility of multiple hydatid cysts to be considered as primary differential.

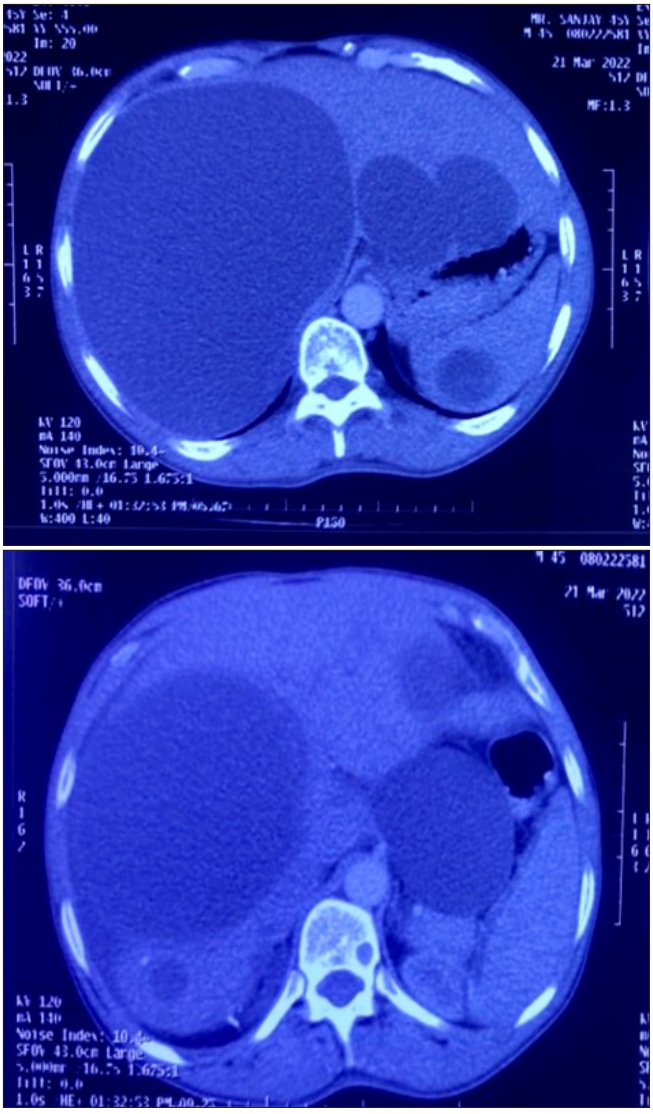

Figure 1-2.CECT Scan of Abdomen and Pelvis

Exploratory Laparotomy: With aspiration and excision of cysts was planned and performed. The **intra - operative findings** were as follows: A large cyst (20 x 20 cm) arising from retroperitoneal region involving aorta, three small mesenteric cysts largest 2 x 2 cm, Left lobe of liver cyst (5 x 5 cm) with septations, Large right lobe of liver cyst (15 x 15 cm), Lesser sac cyst arising from liver (5 x 5 cm). Cysts were drained and cyst wall sent for histopathology examination.

Histopathology section received following biopsies for examination - 1. Retroperitoneal cyst wall, 2. Mesenteric cyst wall, 3. Hepatic cyst wall, 4. Lesser sac cyst wall. Received four containers - Container 1: Retroperitoneal cyst wall - Received cut open cystic structure measuring 10 x 5 x 1.5 cm with smooth polished surface and multi cystic spaces (loculi). Multiple sections show a large cyst along with multiple cysts (loculi). The cyst wall is made up of fibro collagenous tissue and is lined by flattened cuboidal epithelium. There is severe chronic lymphocytic infiltration with focal areas of haemorrhage and rupture. The cysts are filled with lymphatic fluid. No eosinophils or parasite identified. Histological picture is that of lymphangioma. Container 2: mesenteric cyst wall - Received multiple gray tan soft tissue together measuring 1.5 x 1 x 0.5 cm. Multiple sections show multiple multiloculated cysts lined by flattened cuboidal epithelium, some of which are filled with lymphatics. Histological picture is that of lymphangioma. Container 3: Hepatic cyst wall - Received cystic tissue fragments measuring 3.5 x 3 x 0.5 cm. Multiple sections show lymphangioma. Container 4: Lesser sac cyst wall - Received cystic tissue pieces measuring 8 x 3.5 x 0.5 cm. Multiple sections show lymphangioma.

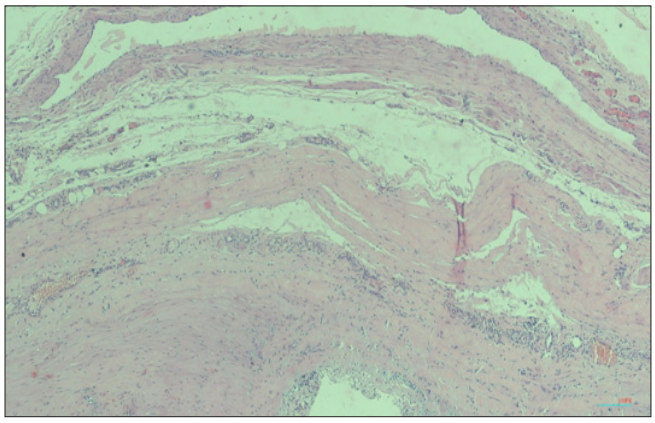

Figure 3.Histopathological Findings

The final histopathological diagnosis made from Multiple fragments from retroperitoneum, mesentery, liver and lesser sac was Lymphangiomatosis. There was no evidence of any parasite.

Discussion

Lymphangiomatosis is a rare benign tumour which is unusually is diffuse or multifocal. Lymphangiomatosis shows diffuse infiltration of various size and shapes of lymphatic channels.[3] One must be able to differentiate this entity from multiple haemangiomas and Angiosarcoma. Lymphangiomatosis will contain acellular fluid instead of RBCs which will be a feature of haemangiomas on histopathological examination.[4] Angiosarcoma is a malignant mesenchymal lesion which will reveal highly atypical spindle cells on biopsy.

ISSN: 2454-8642
DOI: https://doi.org/10.24321/2454.8642.202003

It is important to remember young age of the patient and bland histological features are always seen in Lymphangiomatosis. Multifocal lesions in Lymphangiomatosis can be seen in soft tissue, bones, any other organs except Brain.[5] The prognosis of the patient depends on the type of organ involved. Involvement of soft tissue with or without bone lesions has excellent prognosis whereas lesions can in liver, spleen, intestine, heart, lung carries poor prognosis.[6]

Our case revealed specifically visceral involvement of gastrointestinal organs. Lymphangiomatosis is a rare disease which can be diagnosed by collective effort of diagnostic modalities like radiology and histopathology. The treatment plan includes Surgical intervention mainly. One point to be noted is that it is likely that after removal these lesions may reoccur.[7,8]

Conclusion

Lymphangiomatosis is a rare entity which is rarely seen and belongs to spectrum of developmental disorders. Recognition of the disease requires a high index of suspicion and an extensive workup. Because of its serious morbidity, lymphangiomatosis must always be considered in the list of our differential diagnosis.

Source of Funding: None

Conflict of Interest: None

References

1. Ramani P, Shah A. Lymphangiomatosis. Histologic and immunohistochemical analysis of four cases. Am J Surg Pathol 1993;17:329-35. Back to cited text no.
2. Gomez CS, Calonje E, ferrar DW, Browse NL, Fletcher CD. Lymphangiomatosis of the limbs. Clinicopathologic analysis of a series with a good prognosis. Am J Surg Pathol 1995;19:125-33.
3. Rao VK, Weiss SW. Angiomatosis of soft tissue. An analysis of histologic features and clinical outcome in 51 cases. Am J Surg Pathol 1992;16:764-71.
4. Venkatramani R, Ma NS, Pitukcheewanont P, Malogolowkin MH, Mascarenhas L. Gorham's disease and diffuse lymphangiomatosis in children and adolescents. Pediatr Blood Cancer. 2011;56(4): 667-70.
5. CS Wong, TYC Chu. Clinical and radiological features of generalised lymphangiomatosis. Hong Kong Med J 2008;14:402-4.
6. O'Sullivan DA, Torres VE, de Groen PC, Batts KP, King BF, Vockley J. Hepatic lymphangiomatosis mimicking polycystic liver disease. Mayo Clin Proc. 1998;73(12): 1188-92.
7. Yeager ND, Hammond S, Mahan J, Davis JT, Adler B. Unique diagnostic features and successful management of a patient with disseminated lymphangiomatosis and chylothorax. J Pediatr Hematol Oncol. 2008;30(1):66-9.
8. Zisis C, Spiliotopoulos K, Patronis M, Filippakis G, Stratakos G, Tzelepis G. Diffuse lymphangiomatosis Are there any clinical or therapeutic standards? J Thorac Cardiovasc Surg 2007;133:1664-5. Back to cited text no. 4.

ISSN: 2454-8642
DOI: https://doi.org/10.24321/2454.8642.202003