ISSN No: -2456-2165 https://doi.org/10.38124/ijisrt/25may2104

Lymphatic Malformation of Spleen in an Adult – A Rare Case Report

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Publication Date: 2025/06/06

Abstract: Splenic lymphangioma is a benign cystic neoplasm. Possible etiopathogenesis could be congenital malformation of spleen or inflammation of lymphatic system causing obstruction. It is slow growing neoplasm accounting for <0.007%. Usually presents in childhood. Rare beyond 20 years of age. Other associated symptoms are nausea, vomitings and pain. Lymphangiomas most commonly involve the neck (75%) and axilla (20%) and are less common in the mediastinum, retroperitoneum, kidney, bone, adrenals, spleen, liver and pancreas. Lymphangiomatosis syndrome involves lymphangiomas at multiple sites.

Keywords: *Splenic Lymphangioma, Adult, Female, Cystic Lesion.*

How To Cite: Dr. Gorle Siva Sai; Dr. Bhamidipaty Kanaka Durga Prasad; Dr. Kesinakurthi Satish Kumar; Dr. Atla Bhagyalaxmi (2025) Lymphatic Malformation of Spleen in an Adult – A Rare Case Report. *International Journal of Innovative Science and Research Technology,* 10(5), 3681-3685. https://doi.org/10.38124/ijisrt/25may2104

I. INTRODUCTION

Splenic diseases and primary tumors are rare which include cysts, benign and malignant tumors [1]. Splenic lymphangioma is slow growing neoplasm common in children [2-4]. Lymphangiomatosis is rare in adulthood [5-9] and exceedingly uncommon in individuals over the age of 20. Patients may present with symptoms of abdominal fullness due to increase in size of spleen.

➢ *Clinical Details:*

A 33-year-old female came with complaints of abdominal distension and mass per abdomen since 10 years. No other associated symptoms. Personal and family history were not significant.

Abdomen is grossly distended with a visible, non-pulsatile mass measuring ~25 x 20cm noted extending from epigastric region to pelvis. No visible peristalsis noted.

Laboratory work up revealed Hemoglobin ~7.5gm%, Hematocrit (PCV) ~28.0 vol%. MCH, MCV, MCHC were also reduced. Thrombocytopenia was noted with count of ~0.9lakhs/cu mm.

II. IMAGING INCLUDES ULTRASONOGRAPHY AND COMPUTED TOMOGRAPHY (CT)

➢ *Ultrasonography:*

An ill-defined multicystic lesion with no internal / peripheral vascularity measuring ~28 x 12cm was noted in abdomen extending from left hypochondrium to pelvis. Spleen was not separately visualized.

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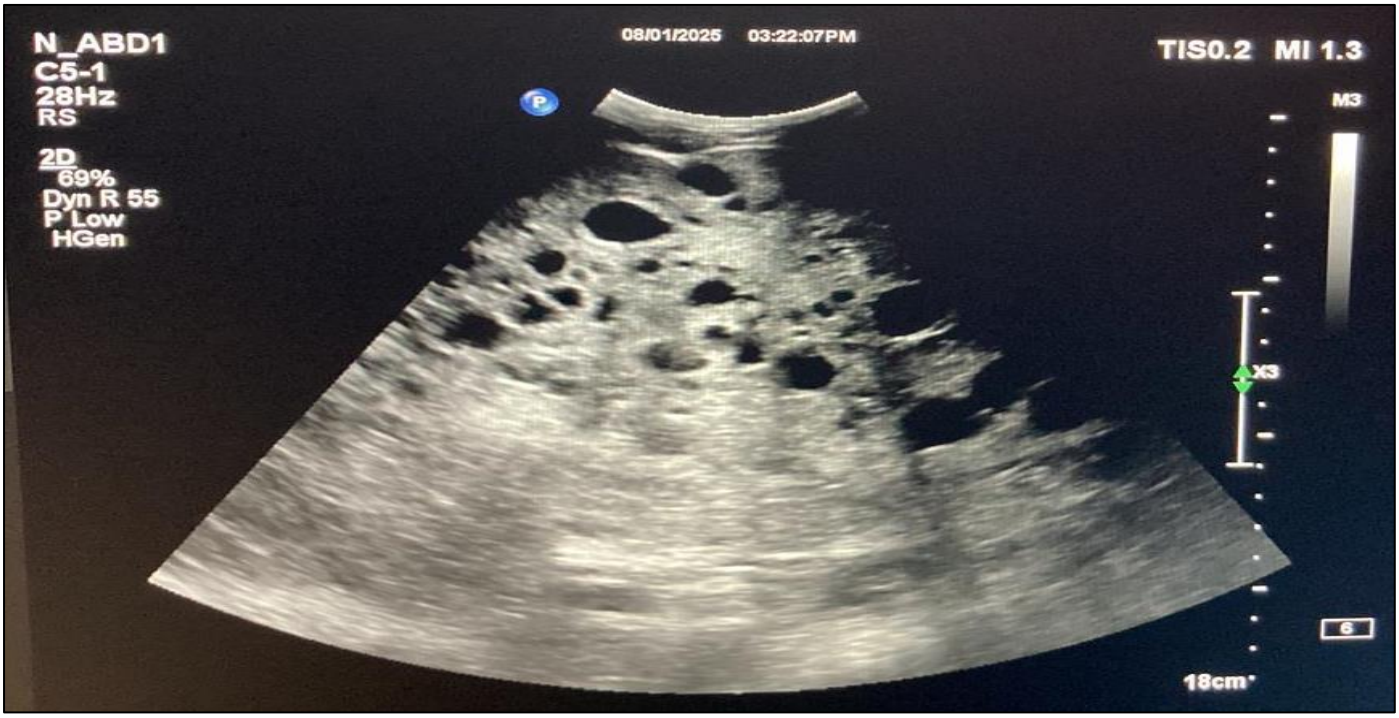

Fig 1 Ultrasonography of Abdomen Showing Multicystic Lesion Extending from Left Hypochondrium to Pelvis.

➢ *Plain CT & Contrast enhanced CT:*

Spleen was grossly increased in size measuring ~34 x 25 x 19cm (CC x TR x AP) extending from left hypochondrium to right iliac fossa. Multiple hypodense lesions with multiple calcifications noted involving entire splenic parenchyma. No obvious solid component was noted. On contrast study, no enhancement was noted. Splenic vein was dilated of diameter measuring ~12mm.

There is compression of superior pole and posteroinferior displacement of left kidney and compression of mid ureter causing upstream dilatation of proximal ureter and pelvicalyceal system noted on left side. However, fat planes with adjacent organs were maintained.

Imaging features were suggestive of Splenic lymphangioma.

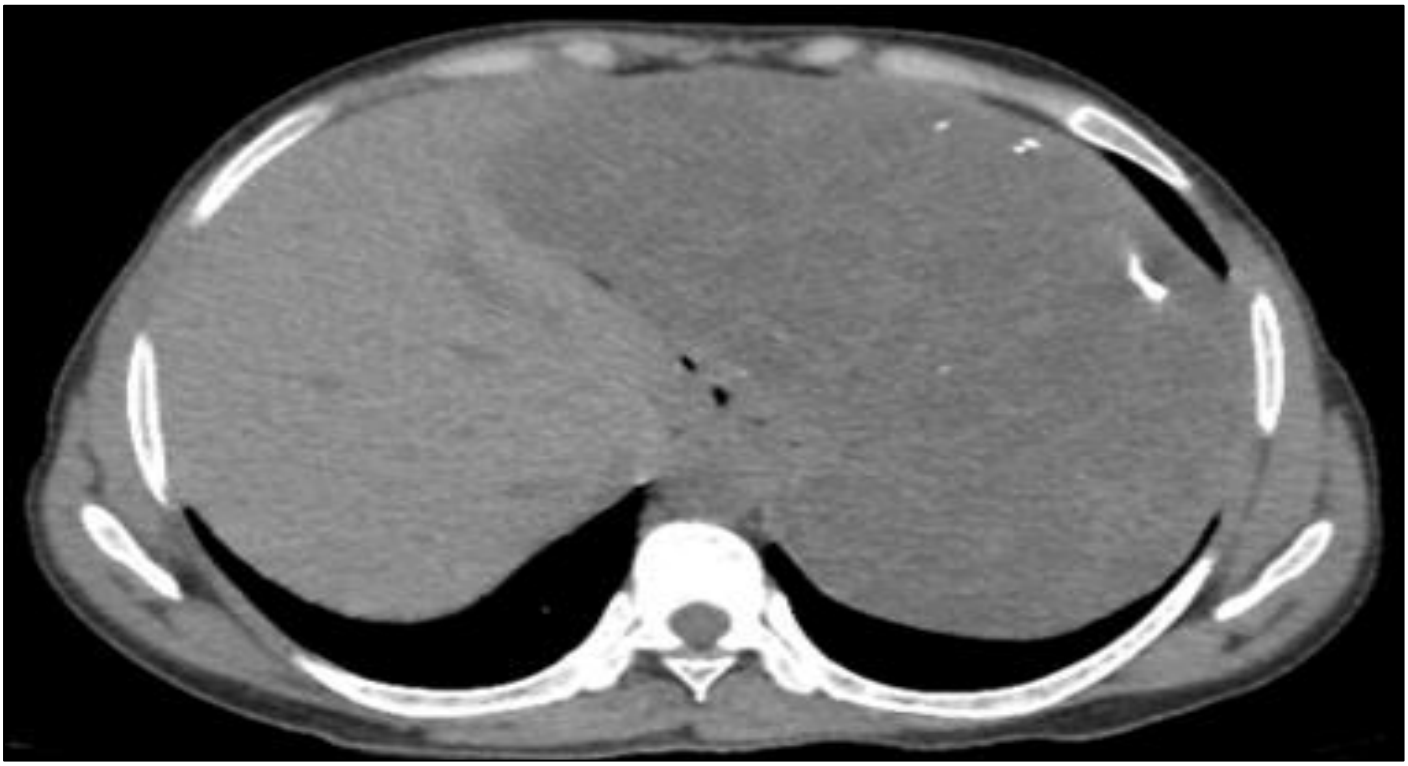

Fig-2 Plain CT Abdomen Showing Enlarged Spleen with Multiple Hypodense Lesions & Calcifications in Splenic Parenchyma.

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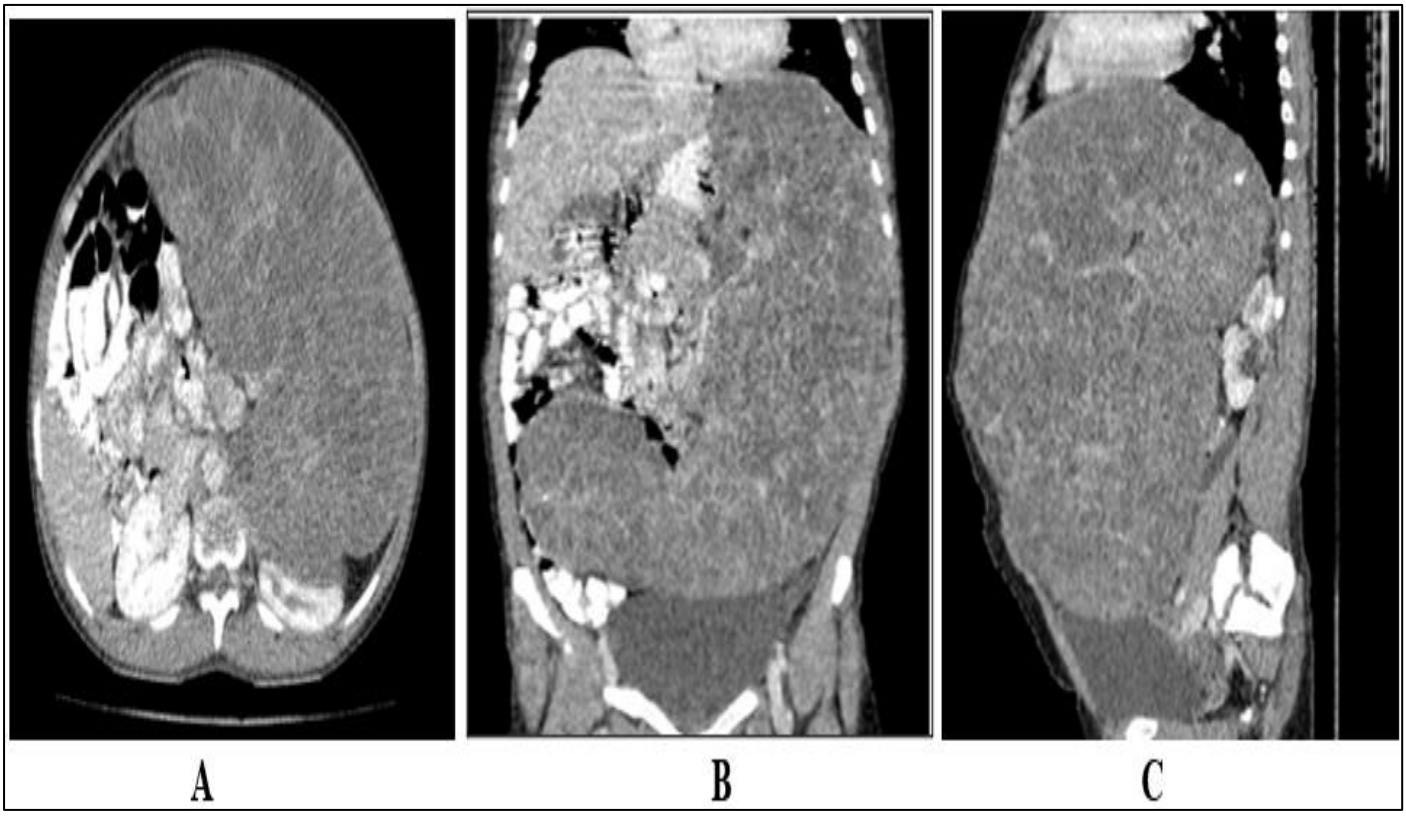

Fig-3 (A) Axial, (B) Coronal, (C) Sagittal sections. Contrast enhanced CT Abdomen showing enlarged spleen with multiple non-enhancing hypodense lesions & calcifications in splenic parenchyma seen compressing left kidney.

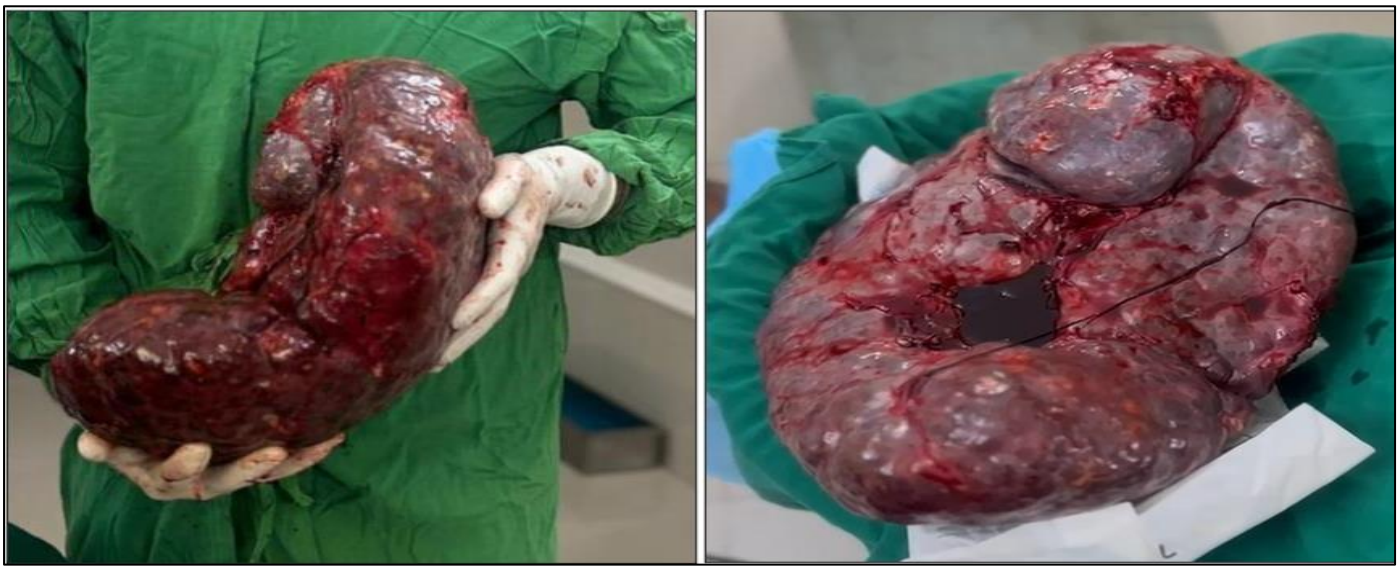

Fig 4 Gross Specimen of Enlarged Spleen with Multiple Nodules in Different Views.

III. HISTOPATHOLOGY OF SPLENECTOMY SPECIMEN

➢ *Macroscopic Features:*

Splenectomy specimen measured ~52 x 36 x 14cm and weighed ~5.6 kgs. Grossly, multiple nodules seen. Spleen was firm in consistency with focal grey white to grey tan areas. Cut section showed multiple cystic areas scattered throughout the spleen ranging from 0.5cms to 4.0cms in diameter. Cysts were filled with serous and gelatinous material.

➢ *Microscopic Features of pathological specimen of spleen:*

Sections studied shown spleen with capsule. Underlying parenchyma showed multiple variable sized, thin-walled cystically dilated lymphatic spaces. These spaces were lined by flattened endothelium and with eosinophilic secretions in lumen. Focal areas of red pulp and white pulp were seen with intervening fibrous tissue. Areas of hemorrhage was seen. Focal areas of microcalcifications were seen.

Features are in favour of "Splenic lymphangioma".

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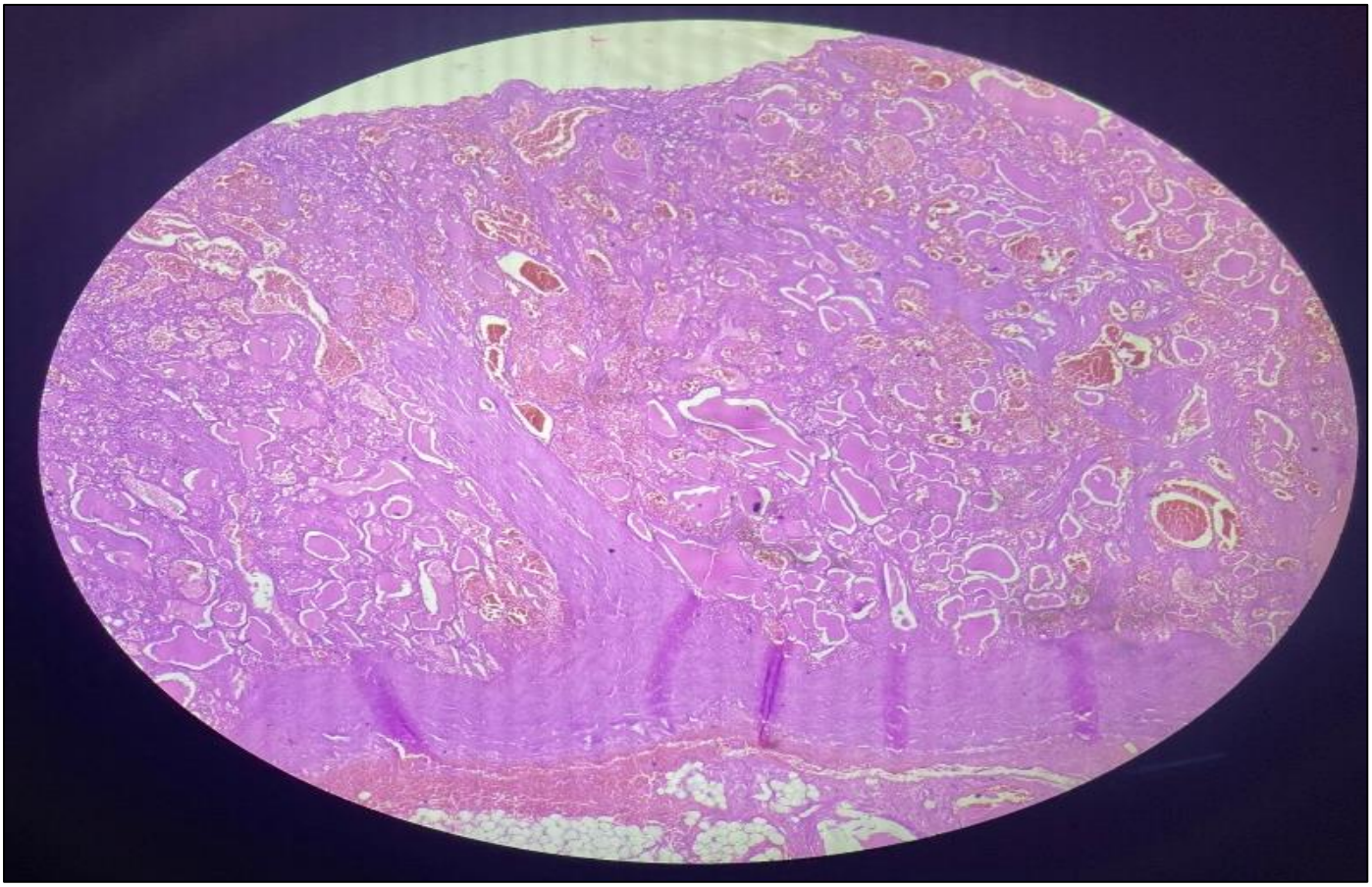

Fig 5 Histopathological Image Showing Thin-Walled Cystically Dilated Lymphatic Spaces.

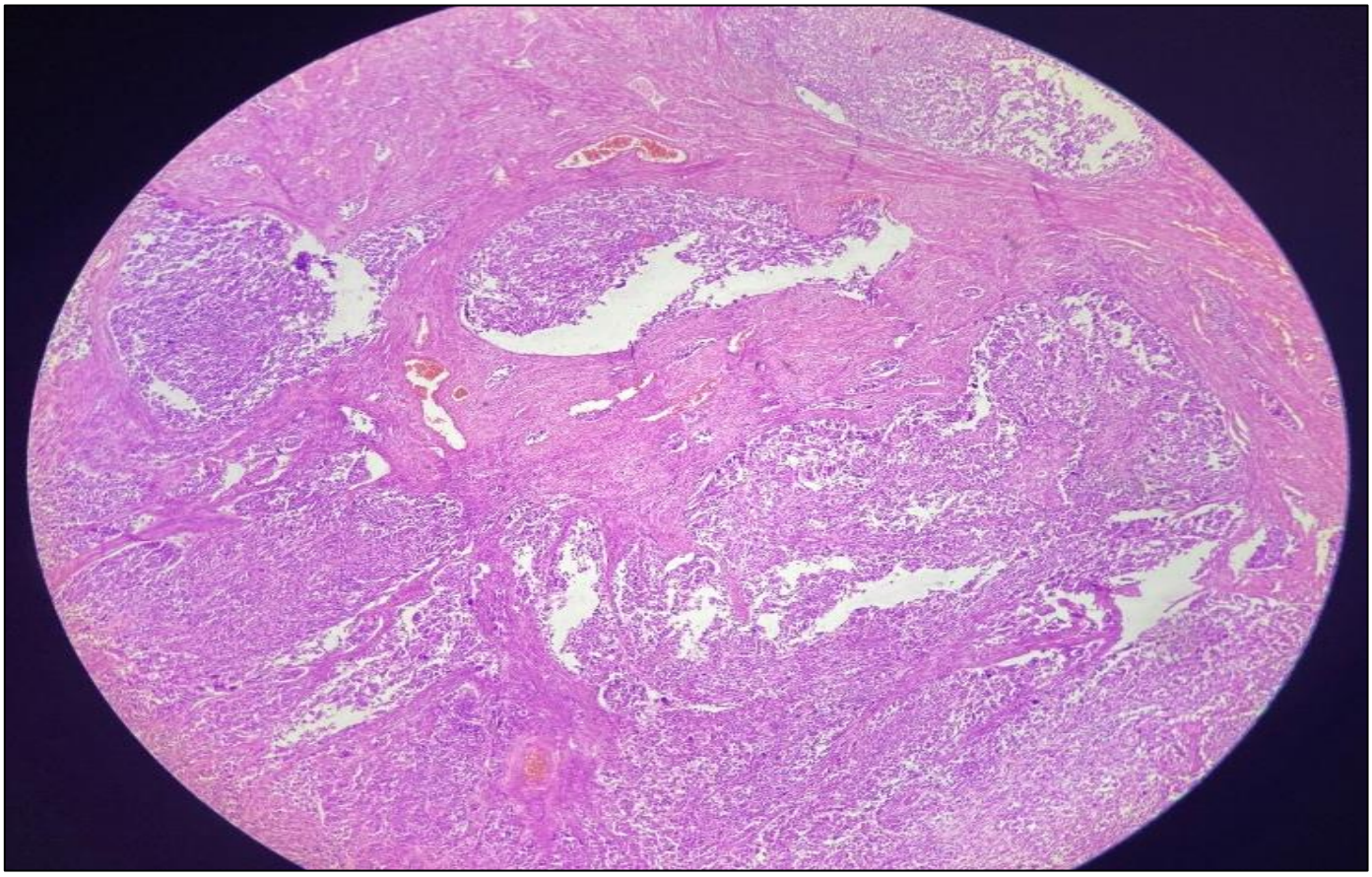

Fig 6 Histopathological Image Showing Red Pulp and White Pulp with Intervening Fibrosis.

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IV. DISCUSSION

Grossly, splenic lymphangioma may present as either solitary or multiple nodular or cystic lesions within the spleen [1,11]. Diffuse lymphangiomatosis [4] may present with very minimal normal splenic parenchyma. Microscopically, these cysts are made up of multiple vascular channels which are lined by single layer of endothelial cells and contain eosinophilic amorphous proteinaceous contents [11]. Lymphangiomas are classified into 3 types according to the size of the dilated lymphatic channels: (1) capillary (supermicrocystic) (2) cavernous (microcystic) (3) cystic (macrocystic) [7,11]. Lymphangioma of cystic type is the most common variant and it appears as a honeycomb of varying sizes of thin-walled cysts containing lymph [5-9]. The remaining parenchyma may or may not show fibrosis, inflammation or congestion.

Selective markers such as podoplanin (D2-40), a monoclonal antibody against dysgerminoma, can be used to distinguish lymphangiomas from hemagiomas as it selectively stains the lymphatic endothelium. Other immunohistochemical techniques such as CD31, CD34 and factor VIII can likewise be utilized [7]. Surgical management by total or partial splenectomy is recommended while conservative treatment is still on debate. As it was a case of massive splenomegaly, an open laparotomy was preferred over laparoscopic surgery. Malignant transformation is very low. Recurrence rate, which is approximately 9.5%, usually after incomplete resection. Other complications of this condition such as infection, torsion, bleeding and rupture may be avoided with immediate surgical management once diagnosis has been established. Its prognosis after complete resection is favourable.

V. CONCLUSION

Splenic lesions, both benign and malignant are uncommon. Among the lesions of spleen, splenic lymphangiomas is common in children and presentation in adulthood is extremely rare. So, diagnosis of lymphangioma before surgery may be difficult in asymptomatic patients. Splenomegaly with or without obstructive symptoms is the most common presentation. Once diagnosed, surgery may be the most effective modality of treatment.

ACKNOWLEDGEMENT

We thank the Departments of Radiology, Pathology, and General Surgery at NRI Institute of Medical Sciences, and the patient, for their cooperation.

- *Conflict of Interest:*
  None declared.

- *Funding:*
  No external funding was received.

- *Institutional details:*
  NRI Institute of Medical Sciences, Visakhapatnam, India.

- *Ethical Committee:*
  Not applicable. Case reports based on de-identified clinical findings do not require IEC approval.

- *Approval Number:*
  N/A

- *Informed Consent:*
  Informed written consent was obtained from the patient for publication of clinical details and images.

- *Data Availability:*
  All data related to this case report are included in the manuscript. Further details available upon request.

- *Use of Artificial Intelligence:*
  No generative AI tools were used in the preparation of this manuscript.

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