An Atypical Case of Splenic Lymphangioma

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AUTHORS' CONTRIBUTIONS
All authors contributed to the design and planning of the manuscript and revised the manuscript. Initial draft written by Daniel Reyes. Dr. Zambrano Tola also processed the pathology specimens, captured and described the pathology images, and contributed to the corresponding sections in the manuscript.

CONFLICT OF INTEREST
None

ETHICAL STATEMENT
No experiments on human or animal subjects

DISCLOSURES
None.

ABSTRACT

Lymphangioma is a rare, benign lymphatic malformation that most commonly occurs in infants and toddlers. It is often asymptomatic and detected incidentally on US or CT, though MRI allows optimal characterization. The spleen is an uncommon location for lymphangiomas, with the neck (75%) and axilla (20%) being the most common locations, followed by orbit, bone, mediastinum, and abdomen less commonly. When the lesions occur in the abdomen, they are most commonly found in the mesentery and omentum, in addition to the adrenal, kidney, gastrointestinal tract, liver, and pancreas. The lesions commonly are cystic with septa/loculations, and no hypermetabolism on PET. This report presents an atypical case of a young adult with a splenic lymphangioma, an already rare finding, which is further unusual in having a solid appearance with lack of septa/loculations, and mild hypermetabolism on PET, though it was confirmed as a lymphangioma with classic findings demonstrated on histopathology. There were also arm and back lesions that likely represented venous malformations based on radiology (no pathology was obtained), signifying a systemic process, and raised suspicion that the splenic lesion was of similar etiology. Genetic analysis identified a c.2740G>A mutation on exon 19 of the PIK3CA gene, in keeping with being on the PIK3CA-related overgrowth spectrum (PROS), which includes Klippel-Trenaunay syndrome, a rare disorder involving vascular and lymphatic malformations. This case highlights that splenic lymphangioma should be considered in cases with splenomegaly or left upper quadrant pain, and in the differential with hemangiomas and other splenic lesions, even in non-classic presentations clinically and on imaging. The presence of venous malformations elsewhere should also raise suspicion for splenic lesions being of similar nature and the possibility of a vascular malformation syndrome

CASE REPORT

BACKGROUND

This report demonstrates an atypical presentation of the already rare lesion, splenic lymphangioma, which should be considered in the evaluation for splenic lesions and/or associated symptoms, particularly in the presence of other vascular/ lymphatic malformations.

CASE REPORT

An otherwise healthy 18-year-old female presented for evaluation of a left forearm mass, which was present since she was in the first grade but had enlarged in recent months, becoming painful. She underwent MRI at a general hospital, where the radiologist was concerned about a soft tissue sarcoma.

Afterwards, the patient was lost to follow-up. In the interim, the patient discovered a painful/tender mass on her back and presented to our facility for further evaluation. During further workup, a large splenic mass was found on a CT scan of her abdomen. Physical examination demonstrated a tender, hard left forearm mass, and another tender round mass on the upper back. The abdomen was soft, non-tender, and nondistended, with no hepatosplenomegaly or masses noted.

Imaging Findings

Abdomen MRI (Figure 1) showed a well-defined focal mass in the anterior spleen measuring 8.1 x 6.4 x 9.0 cm, with progressive peripheral enhancement through delayed imaging and a central non-enhancing stellate component, likely reflecting fibrotic changes. A broad differential was given for this lesion, including hemangioma or sclerosing angiomatoid nodular transformation (SANT).

Sonographic images of the distal lateral left forearm (Figure 2) showed a complex 4.7 x 1.9 x 2.6 cm lesion within the distal lateral soft tissue of the left forearm. The lesion contained focal calcification and Doppler imaging demonstrating slow/venous blood flow within the lesion. The adjacent radial artery and vein were normal. These findings were suggestive of a venous or venolymphatic malformation.

MRI of the upper extremity (Figure 3) demonstrated an intermediate to slightly increased T1 signal and markedly increased T2 signal lesion along the radial aspect of the forearm abutting the dorsal radial diaphysis measuring 2.2 x 1.0 x 4.3 cm. There were scattered areas of vascularity and lesion enhancement. Additionally, a markedly low T1 and low T2 signal component was seen, highly suggestive of a phlebolith. Initial interpretation indicated suspicion of sarcoma, but second-opinion review by a pediatric radiologist determined the lesion was likely a venous or venolymphatic malformation. No pathology was obtained for this lesion.

PET CT (Figure 4) demonstrated mild metabolic activity in left upper back and left arm lesions (Max SUV 1.7 and 1.6, respectively.) The splenic lesion had mild peripheral FDG avidity, slightly above that of normal splenic parenchyma, but showed decreased central FDG avidity, again suggesting necrosis.

Management and Follow-up

Splenectomy was performed due to unclear pathology and large lesion size. Histopathologic examination of the lesion confirmed the diagnosis of splenic lymphangioma, and genetic analysis identified a c.2740G>A mutation of the PIK3CA gene on exon 19 with p.G914R protein alteration. Together, along with extra-splenic vascular malformation, findings were felt to represent Klippel-Trenaunay syndrome.

DISCUSSION

Etiology & demographics

Lymphangiomas, also known as lymphatic malformations, are rare, benign lymphatic fluid-filled sacs that are most commonly seen in children, with approximately 60% diagnosed prior to 1 year of age and 90% under 2 years, and few cases identified in adults [1,2]. The histogenesis is thought to involve abnormal dilation of lymphatic vessels [1]. They are most commonly found in the neck (75%) and axilla (20%) (where they are sometimes called cystic hygromas), with the orbit, bone, mediastinum, and abdomen less commonly affected [1-7]. Abdominal lesions are rare and most commonly found in the mesentery and omentum, in addition to the adrenal, kidney, gastrointestinal tract, liver, and pancreas, but spleen involvement is exceptional [2, 1]. In terms of frequency, 189 cases of splenic lymphangiomas were reportedly identified in the literature between 1939 and 2010 [1].

Splenic lymphangiomas, which generally appear in children (80-90%) [8] and involve congenital or acquired malformation of splenic lymphatic channels, have been proposed to form by abnormally interconnected and dilated thin-walled splenic lymphatic vessels increasing in number and enlarging to form cysts [1]. Their formation may also be attributed to lymphatic inflammation or bleeding that leads to obstruction and lymphangiectasia [1]. Morphologically, splenic lymphangiomas are thin-walled endothelium-lined cysts whose walls and septa are comprised of thin bands of fibrous/fibromuscular tissue with lymphatic tissue and spaces, filled with eosinophilic proteinaceous fluid [8]. Calcification may be present in the cyst walls/septa [6]. They are most frequently subcapsular and least commonly intraparenchymal in location [8].

Clinical findings

In most cases, splenic involvement is not isolated but rather is part of a syndrome of diffuse, multiorgan lymphangiomas, which some have referred to as a lymphangiomatosis syndrome [1, 6]. Some splenic lymphangioma cases have been associated with cystic hygromas of the neck, or rarely as part of Klippel-Trenaunay syndrome, which is characterized by lymphatic malformations, varicose veins, skin hemangiomas, bony and soft tissue hypertrophy, and/or cutaneous hemangiomas [1].

Splenic lymphangiomas are most commonly asymptomatic and found incidentally, but can present with left upper quadrant abdominal pain, splenomegaly, abdominal mass, increasing abdominal girth, or with symptoms related to complications of hypersplenism, portal hypertension, or coagulopathy/ hemorrhage [2,6,7].

Diagnosis involves imaging including ultrasound, computed

tomography (CT), and magnetic resonance imaging (MRI) [2,5], and the lesions typically present as cystic lesions without solid components that are avascular, lobulated, and multiloculated and do not enhance significantly on contrast studies [2,6]. Angiography can be used to differentiate benign spleen tumors [6,7].

Differentiation from hemangioma can be challenging, but lymphangiomas tend to differ in that they involve areas where lymphatics are more concentrated, such as the subcapsular area or larger trabeculae of the spleen, whereas hemangiomas tend to be more randomly localized. Endothelially-lined spaces of lymphangiomas also tend to be filled with proteinaceous material rather than blood [6,7]. Histology shows cystic formation with septa made up of connective tissue stroma/fibromuscular and lymphoid tissue, and lymphatic endothelium. Such findings were present in our case (Figure 1).

Immunohistochemical marker analysis can aid in recognizing the endothelial cells [6,7].

Treatment & prognosis

Treatment of splenic lymphangioma may include aspiration along with incision and drainage, enucleation with marsupialization, and splenectomy, which is regarded as the treatment of choice by some authors [7]. Treatment depends on the size of the lesion and the presence of complications, but most are large and require open or laparoscopic total splenectomy to avoid complications [2]. Accessory spleens should also be sought during surgery and removed since they may be part of the disease process [2,8]. Surgical intervention should not be delayed in the absence of contraindications, such as infection, as delay in intervention can lead to potentially life-threatening complications such as rupture leading to peritonitis, invasive hemorrhage, infection, abscess formation, pleural effusion, or empyema [7,8]. Prognosis is good with low rates of recurrence and malignancy, although some cases of malignant transformation of lymphangiomas to lymphangiosarcomas have been reported [8,9]. Conservative treatment of splenic lymphangioma with interferon-alpha has been performed in a child with success and good tolerance, though the optimal treatment period and dose have not been determined [2,8,10].

Imaging findings

As with most splenic lesions, lymphangiomas are often detected incidentally during CT [6]. CT often demonstrates the cystic, multiloculated nature of these lesions. Occasionally, lesions appear more solid, with heterogenous areas of enhancement, similar to microcystic lymphatic malformations elsewhere [1,11]. The cysts may have peripheral calcifications, although this does not help differentiate from other splenic cysts such as pseudocysts or hydatid cysts [8].

Splenic lymphangiomas may be initially detected by ultrasound (US) performed for other reasons. On US, lesions classically appear as round, well-defined hypoechoic or anechoic cystic lesions of various sizes that may include multiple internal septations and intralocular echogenic debris. Lesions usually show minimal vascularity due to their cystic nature, although this may not be true in cases of numerous microcysts [1,2,8].

Magnetic resonance imaging is the imaging test of choice for lymphangioma evaluation at the authors' institution. T1-weighted imaging demonstrates relatively hypointense cystic lesions, with high T1 intensity possible with large amounts of proteinaceous content or if there is internal bleeding. T2 images demonstrate multiloculated hyperintense dilated lymphatic channels, with intervening fibrous septa that appear as hypointense bands [1]. Lesions typically do not enhance aside from the septa, but lesions may appear to enhance if microcystic.

Positron emission tomography (PET) is not routinely used but will typically demonstrate no uptake to confirm the benign nature of the lesions.

Differential diagnosis

The differential diagnosis of splenic lymphangiomas includes other solid and cystic splenic lesions including hemangiomas, primary cysts with epithelial lining (dermoid, epidermoid, or transitional), mesothelial cysts, post-traumatic pseudocyst, parasitic cysts (such as Echinococcus granulosus), sclerosing angiomatoid nodular transformation (SANT), splenic infarction, septic embolism, lymphoma, and metastasis [1-8].

CONCLUSION

Splenic lymphangioma is a very rare lymphatic lesion that most commonly presents in children, with most occurring in infants and toddlers. Description of the precise nature and genesis of these lesions varies, and the International Society for the Study of Vascular Anomalies (ISSVA) recommends the term "lymphatic vascular malformation" for these lesions [12], whereas the World Health Organization (WHO) recommends the name "lymphangioma" but also considers "lymphatic vascular malformation" as an acceptable alternative [13]. This lesion is often asymptomatic and detected incidentally on US or CT, though MRI allows optimal characterization. Diagnosis is made based on clinical, radiological and histopathological presentation. Splenic lymphangioma should be considered in cases presenting with splenomegaly or left upper quadrant pain, and in the differential with hemangiomas and other splenic lesions, including in atypical cases. Our case differed from typical cases, including presentation at young adult age, location in the spleen, solid appearance with lack of typical septa/loculations, and mild hypermetabolism on PET (presumably related to microcystic morphology). Our case had classical findings on histopathology. Although our case did not fit classical lymphangioma appearance on imaging, appearing more like a hemangioma or SANT, it should be considered in the appropriate clinical setting, such as patients with suspected Klippel-Trenauney syndrome. While generally considered benign lesions, splenectomy is often performed due to the difficulty of splenic biopsy and out of concern for complications due to large lesion size.

TEACHING POINT

Lymphangioma is a rare lymphatic malformation that most commonly presents in infants and toddlers and is often asymptomatic and detected incidentally on US or CT, though MRI allows optimal characterization. Splenic lymphangioma, which comprises a rare location for lymphangiomas (with most occurring in the neck or axillae), should be considered in the differential in cases presenting with splenomegaly or left upper quadrant pain, and with hemangiomas and other splenic lesions, including in atypical cases that do not conform to the typical presentation on imaging.

QUESTIONS

Question 1: Which of the following answer choices is true?

A. Splenic lymphangioma is more commonly seen in adults than children

B. Lymphangiomas most commonly occur in the neck or axillae (applies)

C. Abdominal lymphangiomas most commonly occur in the spleen

D. Lesions most commonly appear as simple, solid, and tumor-like lesions

E. Lymphangiomas are usually identified on work-up of classic symptoms

Explanation: Lymphangiomas most commonly occur in young children and are most commonly located in the neck or axillae, with abdominal lesions occurring mesentery and omentum, in addition to the adrenal, kidney, gastrointestinal tract, liver, and pancreas. The spleen is an unusual location. They usually appear cystic and multiloculated in nature on imaging and are most often identified incidentally or on work-up for non-specific symptoms.

Question 2: Which of the following answer choices is false?

A. Splenic lymphangioma is usually an isolated finding (applies)

B. Splenic lymphangioma is generally a benign lesion

C. MRI is a useful imaging modality for lymphangiomas

D. PET does not typically demonstrate uptake of the lesions

F. Pathology can be used to confirm the nature of the lesions

Explanation: Splenic lymphangioma is not usually an isolated finding.

Question 3: What is the most common location of lymphangiomas?

A. Mediastinum

B. Neck and axilla (applies)

C. Abdomen

D. Spleen

Explanation: The lesions most commonly occur in the neck and axilla.

Question 4: The mainstay of treatment for large or complicated splenic lymphangiomas is which of the following?

A. Surgical resection (applies)

B. Sclerotherapy

C. Interferon therapy

D. Drainage

Explanation: Surgical resection is a mainstay of treatment, in particular for large or complicated lesions.

Question 5: Symptoms that may occur in splenic lymphangiomas include all of the following except:

A. Left upper abdominal pain

B. Splenomegaly

C. Increased abdominal girth

D. Costovertebral angle tenderness (applies)

Explanation: Left upper abdominal pain, splenomegaly, and increased abdominal girth may be presentations associated with the lesions. Costovertebral angle tenderness is not a finding commonly associated with the lesions.

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FIGURES

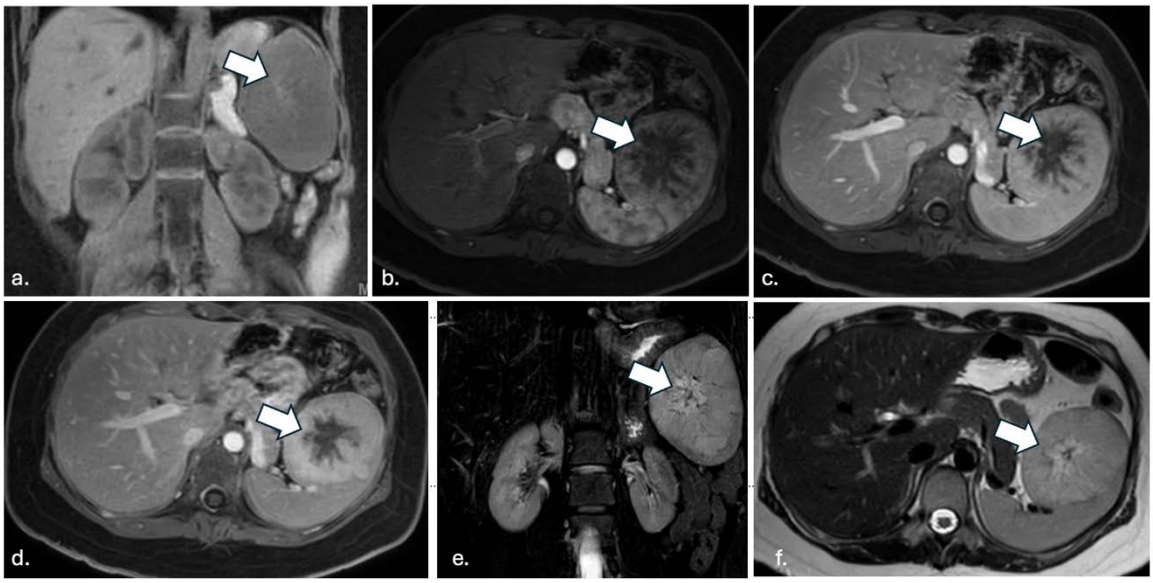

Figure 1: 18-year-old female with splenic lymphangioma. MRI Abdomen showed a well-defined focal mass (arrows) at the anterior spleen measuring 8.1 x 6.4 x 9.0 cm, with progressive peripheral enhancement through delayed imaging with a central non-enhancing stellate component, likely reflecting fibrotic changes. (A) MRI abdomen T1 late arterial phase postcontrast, coronal view; (B) T1 late arterial phase postcontrast, axial view (C) T1 venous phase postcontrast, axial view; (D) T1 5 minutes postcontrast, axial view; (E) T2, coronal view; (F) T2, axial view.

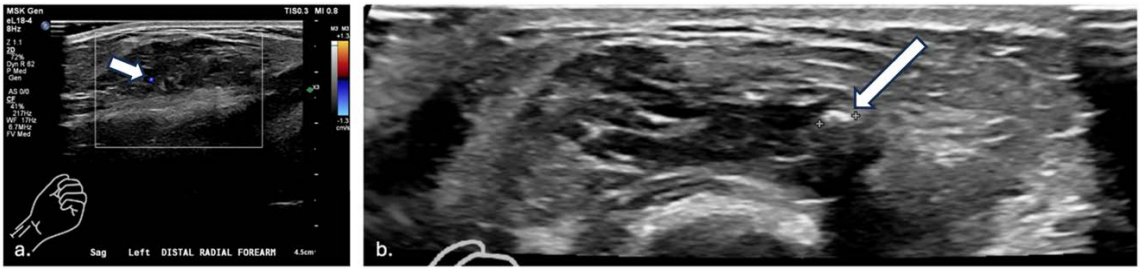

Figure 2: 18-year-old female with splenic lymphangioma. Ultrasound images of the distal lateral left forearm demonstrate a complex 4.7 x 1.9 x 2.6 cm lesion within the distal lateral soft tissue of the left forearm. Doppler imaging of the lesion (A) demonstrates slow/venous blood flow within the lesion (arrow), and on grayscale imaging (B) the lesion contains focal calcification (arrow).

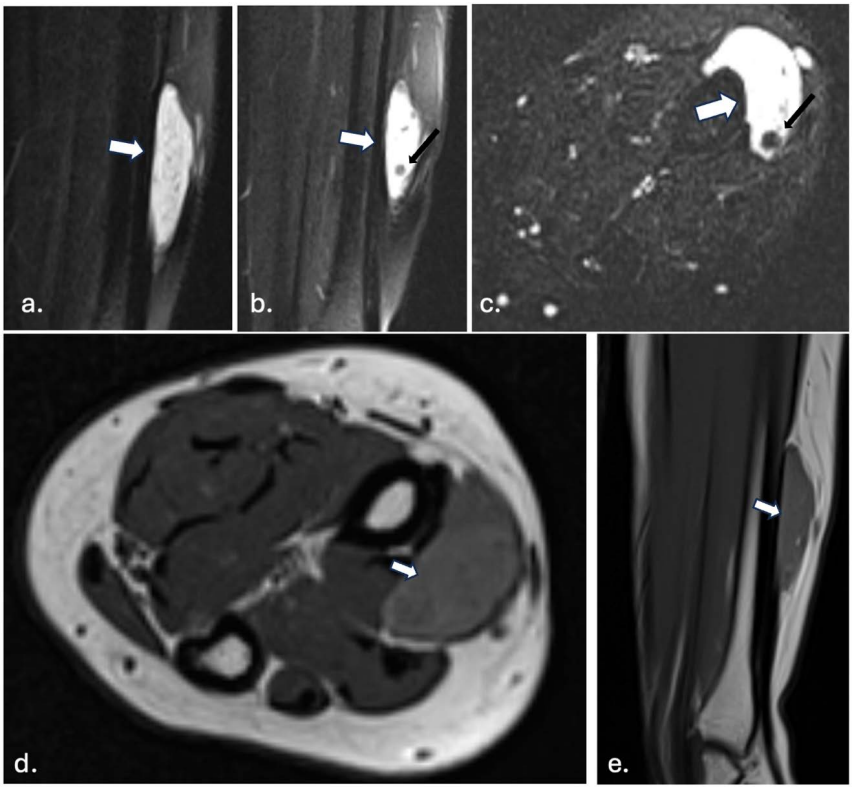

Figure 3: 18-year-old female with splenic lymphangioma. MRI of the upper extremity demonstrated an intermediate to slightly increased T1 signal and markedly increased T2 signal lesion (white arrows) along radial aspect of the forearm abutting the dorsal radial diaphysis measuring 2.2 x 1.0 x 4.3 cm. There were scattered areas of vascularity and lesion enhancement. Additionally, a markedly T1/T2 low signal lesion was seen (black arrows), highly suggestive of a phlebolith. (A) MRI upper extremity T2, coronal view; (B) T2, coronal view, demonstrating phlebolith (black arrow); (C) T2, axial view, demonstrating phlebolith (black arrow); (D) T1 precontrast, coronal view; (E) T1 precontrast, axial view.

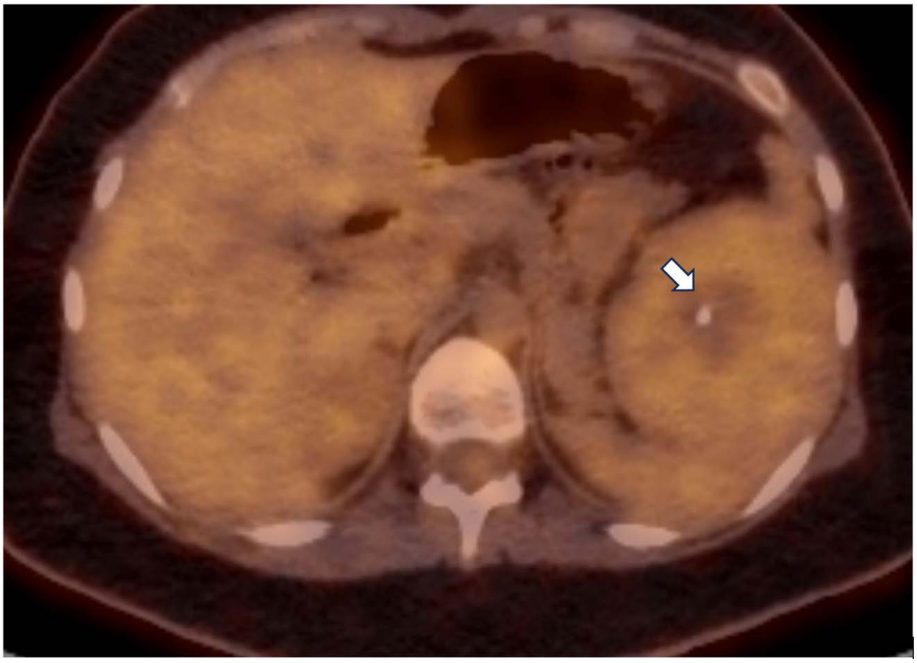

Figure 4: 18-year-old female with splenic lymphangioma. PET CT, axial view demonstrated mild metabolic activity in left upper back and left arm lesions (Max SUV 1.7 and 1.6, respectively.) Splenic lesion had mild peripheral FDG avidity, slightly above that of normal splenic parenchyma, but showed decreased central FDG avidity (arrow), again suggesting necrosis.

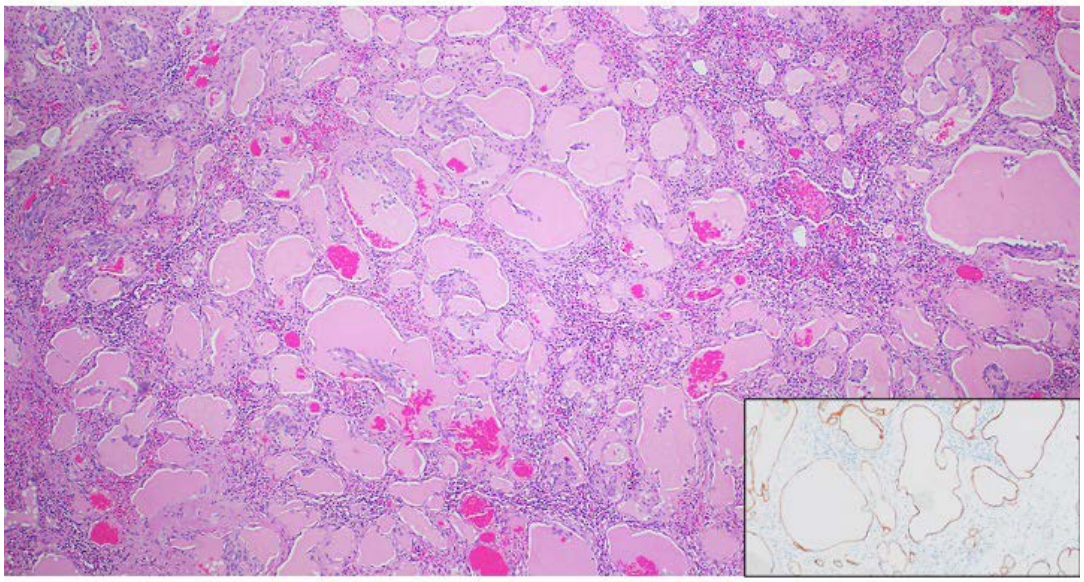

Figure 5: 18-year-old female with splenic lymphangioma. Histology sections showed fragments of benign splenic tissue extensively involved by a vascular lesion composed of dilated vascular spaces lined by cytologically bland endothelial cells and filled with proteinaceous material. Lesional endothelial cells were positive for the lymphatic marker D2-40 on immunostaining (inset).

KEYWORDS

Splenic lymphangioma; lymphangioma; lymphatic malformation; splenic lesion; abdominal lesions; Klippel-Trenauney syndrome

ABBREVIATIONS

CT = Computed Tomography
MRI = Magnetic Resonance Imaging
PET = Positron Emission Tomography

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