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CASE REPORT

Isolated Diffuse Splenic Hemangiomatosis Arising in an Adolescent: A Rare Case Report and Literature Review

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Background: Diffuse splenic hemangiomatosis (DSH) is an extremely rare benign vascular disorder characterized by the proliferation of multiple blood vessels within the spleen. It is even rarer in pediatric and adolescent patients, with only a few cases reported in medical literature, which was performed as scientific literature review.

Case Presentation: We observed a 14-year-old male who presented with intermittent abdominal discomfort and slight splenomegaly. Laboratory tests revealed no abnormalities. Computed tomography (CT) revealed diffuse splenomegaly with round or oval low-density lesions, suggestive of hemangiomatosis or lymphoma. Magnetic resonance imaging (MRI) revealed multiple lesions with slightly prolonged T1 and T2 signals. A CT-guided percutaneous biopsy of a splenic lesion was performed to address splenomegaly and diagnostic uncertainty. Histopathological examination confirmed diffuse splenic hemangiomatosis with lymphocytic infiltration.

Conclusion: A rare case of DSH in a childhood was presented. This case underscores the importance of integrating imaging and histopathology for an accurate diagnosis. While the condition is typically benign, tissue biopsy remains the definitive diagnostic method when malignancy cannot be excluded.

Keywords: diffuse splenic hemangiomatosis, lymphoma, splenomegaly, tissue biopsy

Introduction

Diffuse hemangiomatosis is a rare vascular anomaly characterized by diffuse involvement of an entire organ or multiple organ systems with hemangioma-like lesions. Diffuse splenic hemangiomatosis (DSH), a specific form predominantly confined to the spleen, was first described by Langhans in 1879.[1] It is histopathologically defined by the diffuse replacement of splenic parenchyma with variably sized cavernous vascular channels that typically do not communicate with normal splenic sinuses. This condition involves extensive arterial capillary proliferation throughout the spleen.

While DSH can rarely coexist with hemangiomatosis of other organs such as the liver, skin, or bone,[2] isolated splenic involvement is exceedingly uncommon, with fewer than 20 well-documented cases reported to date in the medical literature.[1] Its occurrence in pediatric populations is even more exceptional, usually documented only as isolated case reports due to the inherent lack of large-scale epidemiological data.

The clinical picture of isolated DSH is primarily characterized by marked splenomegaly and secondary coagulopathy due to massive platelet sequestration.[3] Without timely diagnosis and intervention, patients face a significant risk of serious complications, including spontaneous hemorrhage or malnutrition. The prognosis and outcomes for isolated DSH are generally favorable following appropriate management. Treatment of choice is typically splenectomy, after which platelet counts and coagulation parameters usually normalize rapidly, leading to an effective cure of the disease. We report an adolescent case of isolated DSH presenting with splenomegaly, notably without the typical accompanying

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hematologic symptoms. This case was incidentally detected during a routine examination for abdominal pain and confirmed through splenic biopsy.

Case Presentation

A 14-year-old boy presented to our hospital with intermittent left upper abdominal pain persisting for one year. The patient reported no significant past medical history, family history of hereditary conditions, or current medication use. Physical examination was unremarkable, and laboratory tests revealed no significant abnormalities. CT imaging revealed a slightly enlarged spleen with multiple round low-density lesions, the largest measuring approximately 2.2×1.9 cm^2 (Figure 1A). Contrast-enhanced scans showed marked enhancement of the lesion during the arterial phase, with sustained enhancement during the venous phase, suggesting that it could be lymphoma or hemangioma (Figure 1B). MRI revealed multiple splenic lesions with slightly prolonged T1 and T2 signals (Figure 1C and D). Diffusion-weighted imaging revealed hypo-intensity. Contrast-enhanced scans showed mild ring enhancement during the arterial phase and higher signal intensity than the spleen during the delayed phase (Figure 1E and F). No enlarged retroperitoneal lymph nodes were identified. No enlarged retroperitoneal lymph nodes were identified. Following a CT-guided percutaneous biopsy using an 18G needle, the collected specimens were formalin-fixed and processed for Hematoxylin and Eosin (H&E) staining. Histopathological examination revealed multiple vascular channels lined by flattened endothelial cells, along with signs of vascular proliferation (Figure 2A). Immunohistochemical staining demonstrated positive expression of CD3, CD20, CD31, CD34, and CD68 in tumor cells (Figure 2B–F). The final pathological diagnosis was DSH with prominent lymphocytic infiltration. Given the benign nature of the lesion and the absence of systemic symptoms, a decision was made to adopt a watchful waiting approach, with regular follow-up consultations after thorough discussions with the patient's family. The patient will be monitored every three months during the first year using ultrasound and CT scans to assess lesion progression, as no other therapeutic interventions have been implemented. At the 14-month follow-up, the patient remained in good health.

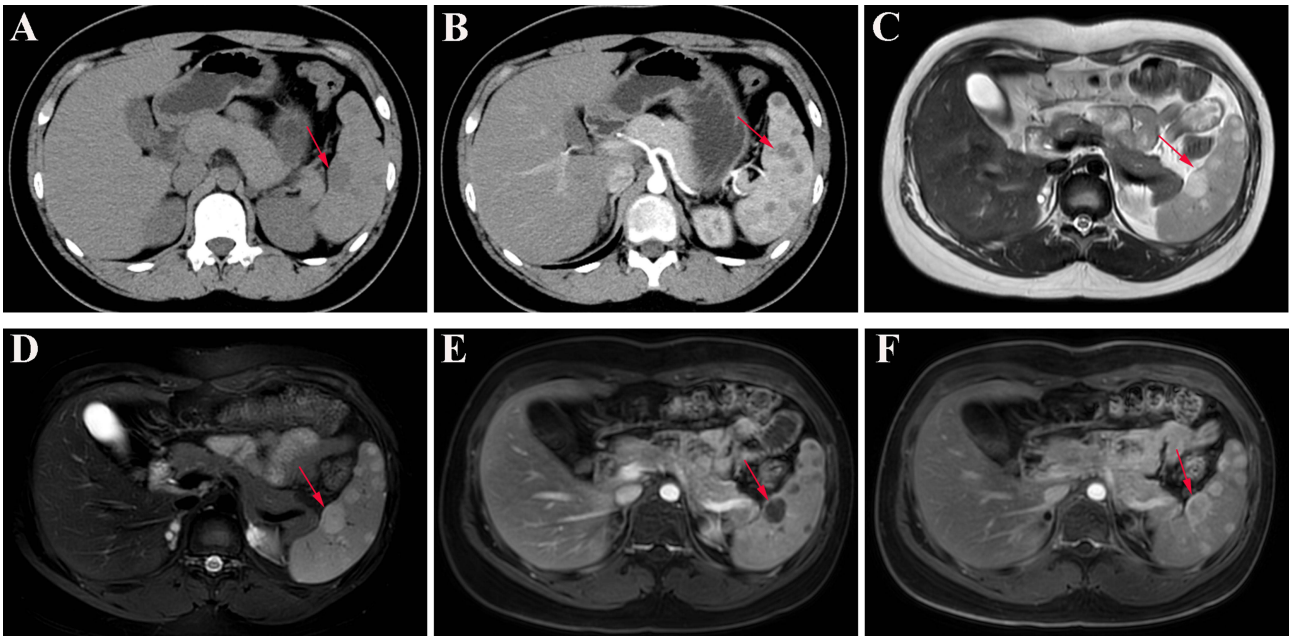

Figure 1 The imaging examinations of the patient. (**A** and **B**) Abdominal CT revealed multiple round or oval low-density lesions in the spleen (**A**), showing progressive enhancement during the contrast-enhanced phase (**B**). (**C**–**F**) MRI revealed multiple slightly hyperintense signals on both T1- (**C**) and T2-weighted (**D**) images in the spleen; Contrast-enhanced MRI demonstrated mild ring-shaped enhancement during the arterial phase (**E**), with delayed-phase enhancement surpassing that of normal splenic tissue (**F**). The lesions are indicated by the red arrows.

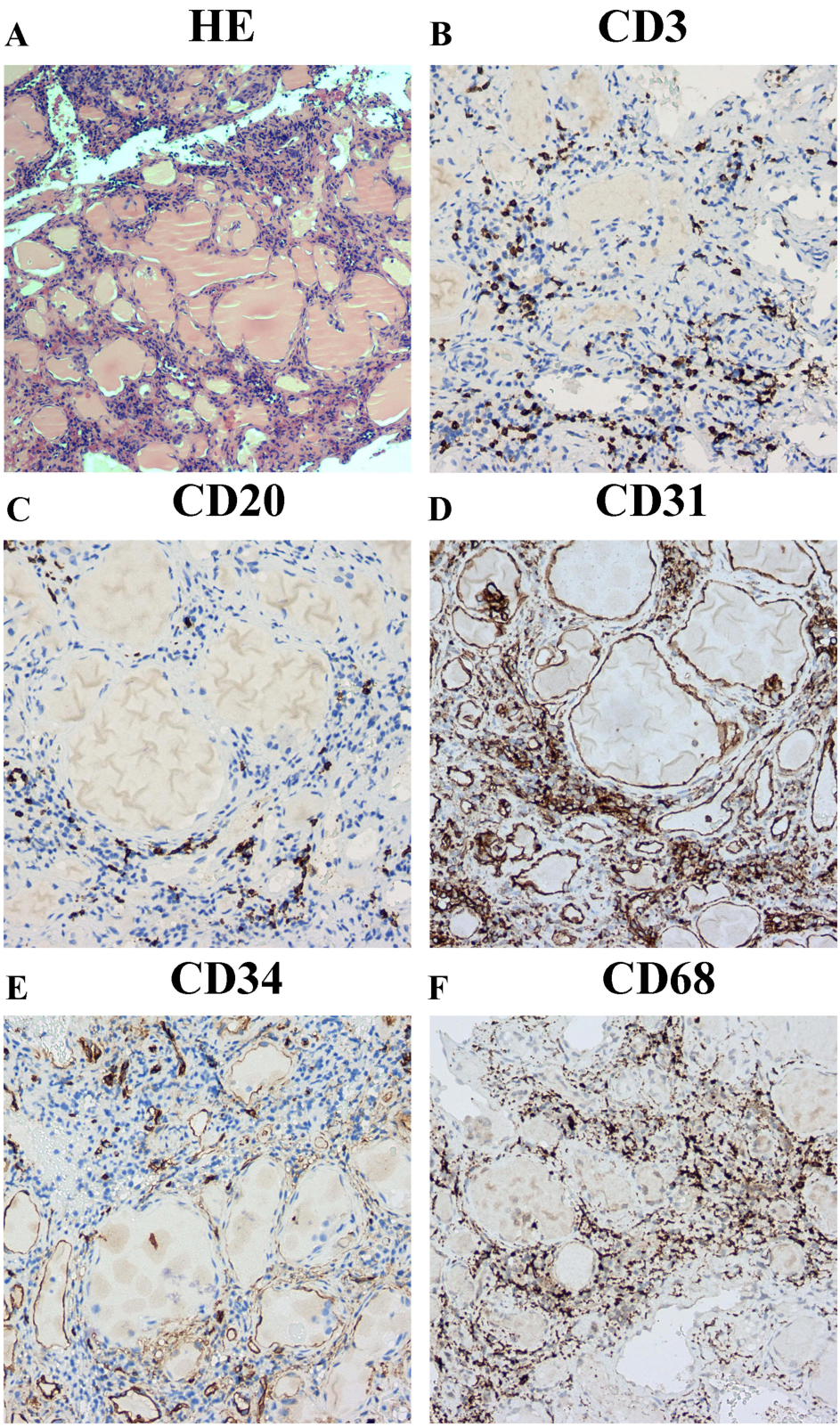

Figure 2 Pathological findings of the tumor mass in the spleen. (**A**) The HE staining revealed that the tumor was composed of multiple vascular channels lined with flat endothelial cells. (**B-F**) Immunohistochemistry showed that the tumor cells are positive for expression of CD3 (**B**), CD20 (**C**), CD31 (**D**), CD34 (**E**) and CD68 (**F**) at the magnification of ×200.

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Discussion

DSH is a rare benign vascular disorder, often considered a manifestation of systemic hemangiomatosis, such as Klippel-Trenaunay syndrome (KTS) and Kasabach-Merritt syndrome (KMS).[4,5] In this study, we report a rare case of isolated DSH, discovered incidentally in a patient presenting with intermittent abdominal pain but no associated symptoms. Previous reports suggest that DSH can occur at any age, with a higher prevalence observed in middle-aged and elderly individuals.[6] However, cases in children and adolescents remain exceptionally rare. A comprehensive review of PubMed and Medline from 2000 to 2025 revealed that literature on isolated DSH in adolescents is extremely limited. We present an exceptionally rare clinical case, with highly limited literature available to date, underscoring the uniqueness of our report (Table 1).[7–12]

Immunohistochemical analysis demonstrated tumor cell expression of CD3, CD20, CD31, CD34, and CD68. Positivity for endothelial markers CD31 and CD34 indicates vascular endothelial origin, supporting aberrant splenic vascular proliferation. CD68 positivity in normal histiocytes of the red and white pulp indicates its conventional role as a macrophage/histiocyte marker; its expression in tumor cells may implicate immune response or inflammatory processes. CD3 and CD20 positivity reflects T and B lymphocyte infiltration, suggesting immune cell involvement in disease pathogenesis. These findings collectively demonstrate the involvement of both vascular components and the immune system in the pathological process of DSH, providing valuable insights into its underlying mechanisms and guiding clinical management.

The etiology and pathogenesis of DSH remain unknown, with its malformative or neoplastic origin still under debate. Ruck et al suggested that partial CD8 expression in tumor cells might indicate an origin from splenic sinus lining cells.[16] However, studies by Arber and Steininger found no CD8 expression in the endothelial cells of pathological vessels.[2,17] Dufau proposed that DSH may be a vascular malformation, with angiomatous regions potentially arising from distorted splenic microcirculation due to malformations of the postsinusoidal venous system.[6] Among the 14 cases reported by Dufau, the condition was predominantly observed in middle-aged and elderly individuals. If DSH truly represents a malformation, the reason for its late-life manifestation remains an unresolved question.

DSH is typically asymptomatic; however, in some cases, splenic hemangiomatosis may present with hypersplenism symptoms such as splenomegaly, thrombocytopenia, anemia, and coagulation disorders like disseminated intravascular coagulation (DIC), especially when associated with KMS.[5,13] In cases where DSH is accompanied by KTS, additional features may include venous malformations, varicosities, red birthmarks, and overgrowth of soft tissues and bones.[4,14] Although rare, complications such as high-output heart failure, portal hypertension, and splenic rupture have been reported in isolated cases.[18]

The nonspecific symptoms of DSH often make early definitive diagnosis challenging. CT and MRI are preferred imaging modalities for screening splenic tumors, as they effectively delineate morphology, size, number, blood supply characteristics, and relationships with surrounding structures.[19] However, imaging alone is insufficient for diagnosing hemangiomatosis, as DSH can easily be mistaken for other vascular tumors or tumor-like lesions, such as lymphangiomas, hemangioendotheliomas, or angiosarcomas.[2,15] Thus, histopathological diagnosis remains the gold standard.

Currently, no consensus exists regarding the treatment of splenic diffuse hemangiomatosis. A "wait-and-watch" strategy is recommended for asymptomatic patients with normal spleen function. Splenectomy is advised when significant splenomegaly, severe anemia, thrombocytopenia, or consumptive coagulopathy occurs.[20] Vascular embolization can also be considered as a temporary alternative for patients with thrombocytopenia who are unsuitable for surgery or have surgical contraindications.[21] For patients with DSH combined with KTS or KMS, an individualized, multidisciplinary management strategy with targeted treatment for complications is recommended.[22] Anticoagulant therapy is recommended for patients at risk of thrombosis. Local injection of sclerosing agents or interventional therapy may be performed in patients with varicose veins or vascular malformations. If thrombocytopenia and coagulopathy are present with contraindications to surgery, immediate correction of coagulation abnormalities, platelet transfusion, and infection prevention should be initiated. Vincristine, an anti-angiogenic drug, has demonstrated significant efficacy in treating KMS and is often combined with corticosteroids.[23] Additionally, mTOR inhibitors have shown promising results in recent years for treating KMS, especially in patients unresponsive to corticosteroids and vincristine.[24] Small molecule VEGFR inhibitors and PD-1/PD-L1 inhibitors have demonstrated potential efficacy in certain vascular tumors, but their

Table 1 A Literature Review of Diffuse Splenic Hemangioma from 2000 to 2025

References	Age/Sex	Laboratory Test	Symptoms	Comorbidities	Immunohistochemistry	Treatment	Survival (Months)
Present case	14/male	Normal	Intermittent pain	None	CD3+, CD20+, CD31+, CD34+, CD68+	Follow-up	>5
Lanjewar DN[4]	63/female	Prolonged prothrombin time	Intermittent pain	KTS	CD31+, D2-40-	Splenectomy	ND
He X[13]	59/female	Anemia, thrombocytopenia, abnormal liver function, elevated D-dimer	Abdominal distension, fatigue	KMS	ND	Combined Regimen of PRED, THD, SRL	>4
Nakano Y[3]	1/male	ND	Abdominal distension, HF	DIC	fVIII+, CD34+, CD31f+, CD8f+	PO: PDN, PPL, VCR, PT, FFP; Partial embolization, splenectomy	ND
Haque PD[9]	38/female	Anemia, prolonged prothrombin time and APTT, hypoproteinemia	Fullness and progressive distension of left upper abdomen	KMS	ND	PO: PCV, IV, CRBCT; IO: FFP; splenectomy	ND
Misawa T[11]	40/female	Anemia, thrombocytopenia	Severe dull abdominal pain	KTS	α-SMA+, CD31+, D2-40-	Splenectomy; post-op: heparin	ND
Dekeyzer S[14]	45/female	Normal	Dyspnea, abdominal pain	KTS	ND	Splenectomy	ND
Ambrosio MR[7]	26/male	Normal	None	HL	fVIII+, CD34+, CD31+, CD8-, D2-40-	Splenectomy	>26
Choi YJ[8]	29/female	Anemia	Intermittent hematochezia	KTS	ND	Oral iron supplementation	ND
Tang JY[5]	2.8/male	Anemia, elevated WBC, thrombocytopenia	Recurrent petechiae	KMS, DIC	ND	PO: LMWH, FIB, PCC; splenectomy	>2
Steininger H[2]	42 /male	Normal	Asymptomatic	None	CD31+, Vim+, fVIII+, CD43f+, CD68f+, CD8f+, CD34f+, D2-40-, D21-	Splenectomy; post-op: Aspirin	>6
Mirali H[12]	48/ female	Normal	Left hypochondrium chronic pain, portal hypertension	None	ND	Follow-up	>36
Patel T[15]	63/female	Normal	Abdominal lump and pain	None	CD34+, fVIII+, CD31f+, CD8-	Splenectomy	ND
C. Langner[10]	62/male	Thrombocytopenia	Jaundice, weight gain, abdominal discomfort	Hepatic Diffuse Hemangiomatosis	fVIII+, CD34+, CD31+, SMA+	ND	Died one week after hepatic coma

Abbreviations: ND, not described; fVIII, factor VIII related antigen; f+, focally positive; Vim, vimentin; HF, heart failure; PT, platelet transfusion; FFP, fresh frozen plasma transfusion; PDN, prednisolone; PPL, propranolol; VCR, vincristine; HL, Hodgkin's lymphoma; DIC, Disseminated Intravascular Coagulation; KTS, Klippel-Trenaunay syndrome; KMS, Kasabach-Merritt syndrome; PCV, pneumococcal vaccine; IV, influenza vaccine; PO, preoperative; IO, intraoperative; post-op, post-operative; CRBCT, concentrated red blood cells transfusion; a-SMA, alpha-smooth muscle actin; LMWH, low molecular weight heparin; FIB, fibrinogen; PCC, prothrombin complex; WBC, white blood count; APTT, activated partial thromboplastin time.

specific effects on KMS require further validation.[25,26] For patients unresponsive to drug therapy or with localized tumors, low-dose radiotherapy can be considered an adjunctive treatment option.[27]

This case report has certain limitations. First, being a single case report, the rarity of isolated splenic diffuse hemangioma in adolescents limits its ability to offer comprehensive insights into imaging and diagnostic characteristics. Second, previous studies have indicated a potential association between diffuse hemangiomas of the liver and spleen and mutations in the *CTC1* gene.[13] However, genetic testing was not conducted in this case due to the benign nature of the lesion. Genetic analysis could offer valuable insights into the potential genetic basis of this condition and guide diagnostic strategies in similar cases. Further research is required to better understand the genetic and pathophysiological mechanisms underlying DSH.

Conclusion

We present a rare case of isolated DSH in an adolescent, without any associated complications. This case contributes to the limited literature on splenic vascular tumors by providing valuable insights into the clinical course, diagnostic considerations, and management strategies. Furthermore, it opens up potential avenues for research into the role of immune factors in the mechanisms and etiology of splenic vascular tumors, particularly in the pediatric and adolescent populations.

Data Sharing Statement

The clinical data supporting the conclusions of this manuscript will be made available by the authors.

Ethics Committee Approval

This study was approved by the Ethics and Scientific Committee of Hubei University of Medicine (XYYYE20240074) and was performed according to the Good Clinical Practice Guidelines and the Helsinki Declaration. Institutional approval was obtained from Xiangyang No.1 hospital for the publication of the case detail.

Consent for Publication

Informed consent for the publication of identifying information/images in an online open-access publication was obtained from the patient's father, who acted as the authorized representative.

Author Contributions

All authors made a significant contribution to the work reported, whether that is in the conception, study design, execution, acquisition of data, analysis and interpretation, or in all these areas; took part in drafting, revising or critically reviewing the article; gave final approval of the version to be published; have agreed on the journal to which the article has been submitted; and agree to be accountable for all aspects of the work.

Disclosure

The authors declare no conflicts of interest in this work.

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