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# Splenic hamartoma associated with thrombocytopenia: A case report

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ABSTRACT

*INTRODUCTION:* Hamartomas are rare, benign tumors of the spleen. Few cases of splenic hamartomas associated with thrombocytopenia have been reported.
*PRESENTATION OF CASE:* An asymptomatic 64-year-old man with myelodysplastic syndrome was found to have a splenic tumor. Laboratory tests were significant for thrombocytopenia, with a platelet count of $7.8 \times 10^4/\mu L$. Ultrasonography showed splenomegaly ($10.8 \times 6.6$ cm), and a hypoechoic splenic mass ($8.0 \times 7.0$ cm). Color doppler ultrasound revealed blood flow within the mass, and the mass density was homogeneous on abdominal computed tomography (CT). Contrast-enhanced CT showed heterogeneous enhancement of the splenic mass during the arterial phase. Positron emission tomography (PET)-CT showed no significant fludeoxyglucose (FDG) accumulation within the mass. The differential diagnosis included splenic hamartoma, splenic hemangioma, splenomegaly associated with extramedullary hematopoiesis, and malignant tumor, including solitary splenic metastasis. A laparoscopic splenectomy was performed due to the possibility of malignancy, the presence of thrombocytopenia, and the risk of splenic rupture. The resected specimen showed a localized, well-demarcated, $8.0 \times 7.0$ cm splenic mass. Histological examination revealed abnormal red pulp proliferation and the absence of normal splenic structures. The patient's post-operative course was uneventful. His platelet count improved on post-operative day 1 and he was discharged on post-operative day 9. He remained in good health with a normal platelet count one month after surgery.
*DISCUSSION:* Making definitive preoperative diagnosis is difficult in splenic hamartomas. Surgery is necessary for diagnosis when malignancy cannot be ruled out.
*CONCLUSIONS:* Surgery may also improve symptoms of hypersplenism, including thrombocytopenia.


## 1. Introduction

The present work has been reported in line with the SCAREcriteria [1].

Hamartomas are rare, benign tumors of the spleen. Most patients are asymptomatic and splenic hamartomas are usually identified incidentally on imaging. However, a minority of patients have symptoms of hypersplenism, including thrombocytopenia, anemia, and pancytopenia. Few cases of splenic hamartomas associated with thrombocytopenia have been reported. We report herein a case of splenic hamartoma associated with thrombocytopenia.

## 2. Presentation of case

An asymptomatic 64-year-old man with myelodysplastic syndrome and hypertension was referred to our department for evaluation and treatment of a newly identified splenic tumor that was discovered by the ultrasonography accidentally. Laboratory tests showed the following: hemoglobin 15.0 g/dl; white blood cell count $5.54 \times 10^3/\mu L$; platelets $7.8 \times 10^4/\mu L$; serum total protein 7.0 g/dL; serum albumin 4.3 g/dL; total bilirubin 0.9 mg/dL; aspartate aminotransferase 38 IU/L; alanine aminotransferase 79 IU/L; alkaline phosphatase 291 IU/L; and serum glutamyltransferase 441 IU/l. Soluble interleukin-2 receptor was within normal limits (352 U/ml).

Ultrasonography revealed splenomegaly ($10.8 \times 6.6$ cm), and a solid, hypoechoic mass ($8.0 \times 7.0$ cm) in the spleen (Fig. 1a, b). Color doppler ultrasound demonstrated blood flow within the mass (Fig. 1c). Abdominal computed tomography (CT) showed an isodense splenic mass (Fig. 2a). Contrast- enhanced CT showed het-

*Abbreviations:* CT, computed tomography.

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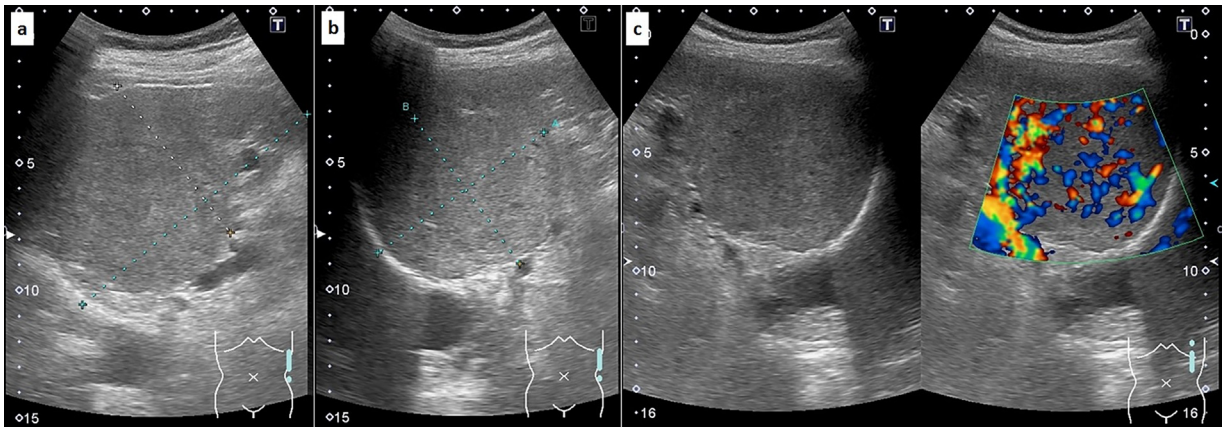


**Fig. 1.** Ultrasonography showed splenomegaly (10.8 × 6.6 cm), with a solid, hypoechoic splenic mass (8.0 × 7.0 cm) (Fig. 1a, b). Color doppler ultrasound showed blood flow within the mass (Fig. 1c).

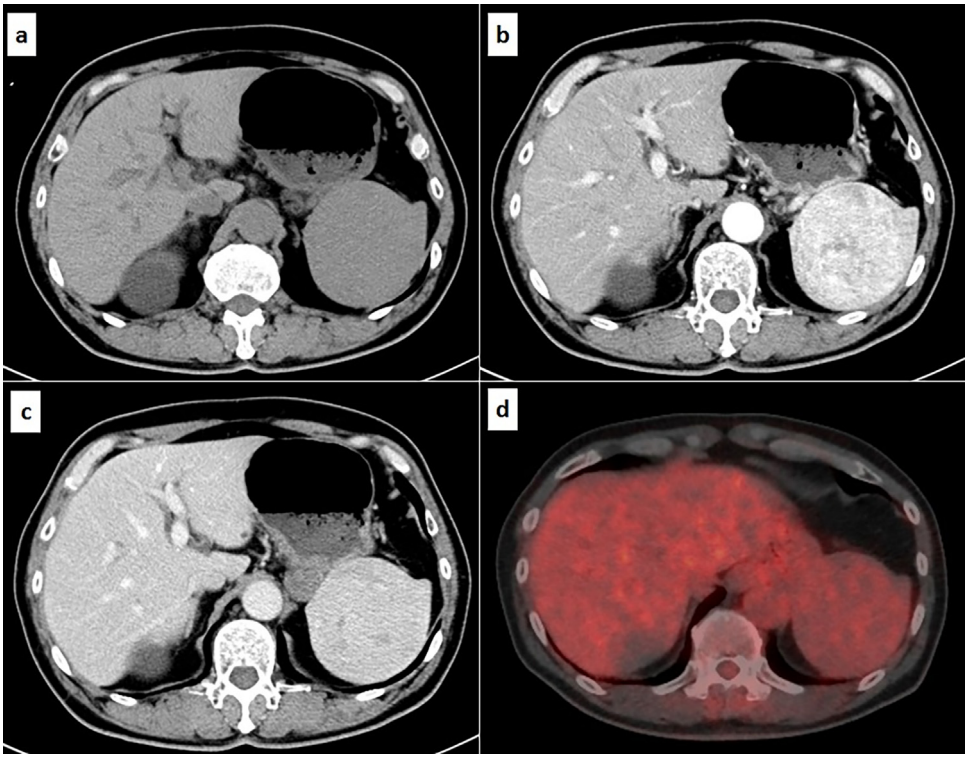


**Fig. 2.** Abdominal computed tomography (CT) revealed an isodense splenic mass (a). Contrast- enhanced CT showed heterogeneous enhancement of a solid splenic mass (8.0 cm) during the arterial phase (b). The mass was isodense compared to normal splenic parenchyma in the portal phase (c). PET-CT showed no significant FDG accumulation within the mass (d).

erogeneous enhancement of the mass in the arterial phase (Fig. 2b). The mass was isodense compared to normal splenic parenchyma in the portal phase (Fig. 2c). Positron emission tomography (PET)-CT showed no significant fludeoxyglucose (FDG) accumulation within the mass (Fig. 2d). The differential diagnosis included splenic hamartoma, splenic hemangioma, splenomegaly due to extramedullary hematopoiesis in the context of myelodysplastic syndrome, and malignant tumor, including solitary splenic metastasis. A laparoscopic splenectomy was performed given the possibility of a malignant tumor, the presence of thrombocytopenia, and the risk of splenic rupture.

The resected specimen showed a localized, well-demarcated, 8.0 × 7.0 cm splenic mass (Fig. 3a,b). Histological examination revealed abnormal red pulp proliferation and the absence of normal splenic structures. No extramedullary hematopoiesis was observed (Fig. 3c,d).

The patient's postoperative course was unremarkable and he developed no complications. His platelet count improved on post-operative day 1, and he was discharged on post-operative day 9. He remained in good heath with a normal platelet count one month after surgery.

## 3. Discussion

Splenic hamartomas were first reported by Rokitansky in 1861 [2]. They are non-capsulated, single or multiple nodules in the spleen and consist of grossly disproportionate native splenic elements. Splenic hamartomas are rare, benign tumors with a reported incidence of 3 per 200,000 splenectomies in a single center series

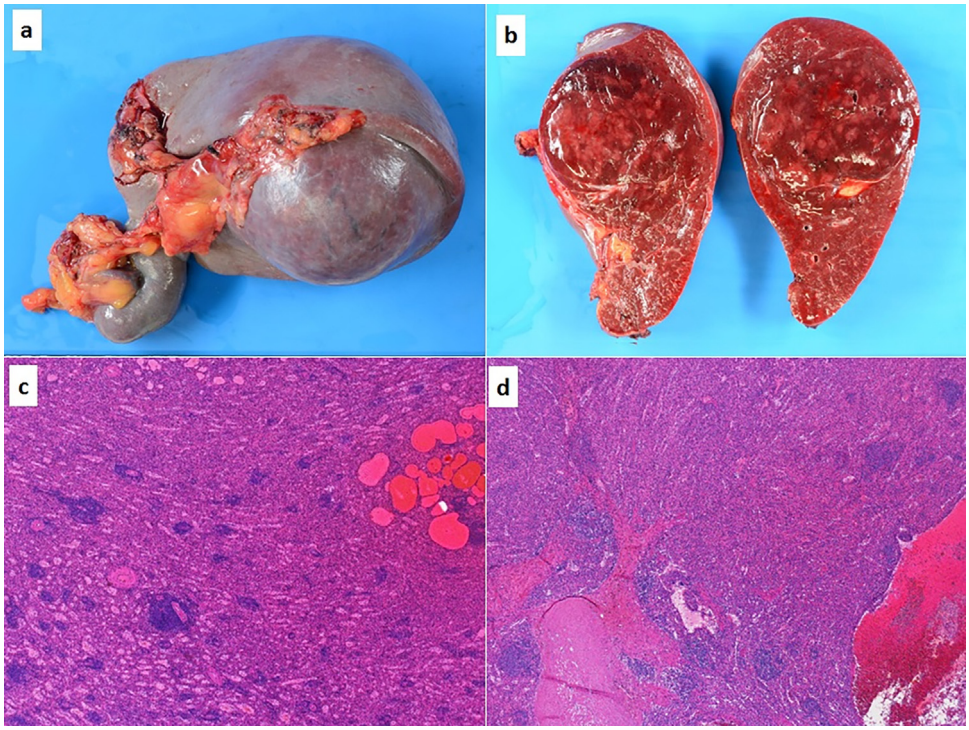


**Fig. 3.** The resected specimen showed a localized, well-demarcated splenic mass (8.0 × 7.0 cm) (Fig. 3a,b). Hematoxylin- eosin stain revealed abnormal red pulp proliferation and the absence of normal splenic structures. No extramedullary hematopoiesis was observed (Fig. 3c,d).

[3], and 0.024%–0.13% in an autopsy review [4]. They can occur in any age group (11 months to 86 years) and they occur with equal frequency in males and females. Women tend to have larger lesions, possibly due to hormonal influences [5]. Most patients with splenic hamartomas are asymptomatic and the lesions are usually identified incidentally on imaging [6]. A minority of patients have symptoms such as pain, palpable mass, or spontaneous splenic rupture (generally associated with large lesions). Hypersplenism leading to thrombocytopenia, anemia, pancytopenia, or malignant hematological conditions, has also been reported [7].

Splenic hamartomas usually appear as hypoechoic lesions on ultrasonography, sometimes with multiple anechoic cystic changes or with an inhomogeneous appearance [6]. They can also appear as an isodense mass on CT. After contrast medium administration, these masses show early and sustained enhancement during the delayed phase of contrast- enhanced CT [6]. However, imaging findings are nonspecific and variable, making definitive preoperative diagnosis difficult [8].

Splenic hamartomas must be differentiated from other vascular tumors of the spleen, including hemangioma, littoral cell angioma, lymphangioma, hemangioendothelioma, sclerosing angiomatoid nodular transformation of the spleen, and angiosarcoma. Solid lesions of the spleen, such as inflammatory myofibroblastic tumor, lymphoma, metastatic disease, disseminated fungal or mycobacterial infections, and sarcoidosis, are also included in the radiologic differential diagnosis [5]. Diagnosis is confirmed by histopathological examination.

There are two types of splenic hamartoma: the pulposal type, resembling the splenic red pulp, and the lymphoid type, resembling the splenic white pulp [3]. Immunohistochemistry may reveal CD8 positive cells lining the vascular channels [9]. These cells are also positive for CD31, factor VIII–related antigen, and vimentin. Immunostaining for CD34 has led to inconsistent results, and the endothelial cells are negative for CD21. CD68 is positive in scattered stromal macrophages but negative in the cells lining the vascular channels [5].

In recent years, laparoscopic splenectomy has become a standard procedure for most benign and malignant hematologic diseases [10]. Nevertheless, massive splenomegaly, defined as a maximum diameter exceeding 20 cm, represents a contraindication for laparoscopy due to the difficulty of the procedure. Complications following laparoscopic splenectomy with splenomegaly generally relate to the size of the spleen, regardless of the underlying disease [11]. The prognosis of splenic hamartomas is good, with a low incidence of postoperative recurrence or metastasis [7]. Surgical indications include possibility of malignancy, high risk of spontaneous splenic rupture, rapidly increasing or symptomatic hypersplenism, and Kasabach-Merritt syndrome [12]. In the present case, the patient underwent splenectomy due to the possibility of malignancy, the presence of thrombocytopenia, and the risk of spontaneous splenic rupture.

Splenic hamartomas associated with thrombocytopenia have rarely been reported. For example, in PubMed, only 19 cases have been reported, and only 6 cases have been reported in the Japanese literature. Watanabe et al. [13] reported that the mean diameter of splenic hamartomas was 5.3 cm, and the diameter of those associated with thrombocytopenia was 9.9 cm. Furthermore, they reported that thrombocytopenia was caused by hypersplenism due to splenomegaly. Histopathological examination in the present case revealed no evidence of splenic (extramedullary) hematopoiesis, though splenomegaly caused by splenic hematopoiesis in the context of myelodysplastic syndrome has been reported [14]. In the present case, the mass measured 8.0 × 7.0 cm, hence, thrombocytopenia may have been due to hypersplenism caused by splenomegaly in the context of splenic hamartoma.

## 4. Conclusions

We have reported a case of splenic hamartoma associated with thrombocytopenia. Most patients with splenic hamartomas are asymptomatic, and it can be difficult to make a definitive preoperative diagnosis. Surgery can confirm the final diagnosis, grant

cure and may also improve symptoms of hypersplenism, including thrombocytopenia

## Conflicts of interest

The authors declare that they have no Conflicts of interest.

## Funding

The authors declare that this study was not funded externally.

## Ethical approval

Not applicable.

## Consent

When obtaining informed consent for surgical procedures, general consent for publication and presentation was obtained from the patients.

## Authors' contributions

TK drafted the manuscript. TK, JH, MK and NH participated in the care of the patients. AN, MM and IY performed the literature search. MK provided the histopathological examination and diagnosis. JH, MK,TK, IO, MY, TI, HM, NH participated in critical revision of the manuscript. All authors read and approved the final manuscript.

## Guarantor

Jun hihara.

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