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*Case Report*

# Primary splenic amyloidosis: a case report

Wei Dong[1*], Lu Liu[2*], Zhen Wan[1], Weidong Xiao[1], Yong Li[1]

*[1]Department of General Surgery, The First Affiliated Hospital of Nanchang University, Nanchang 330006, Jiangxi Province, China; [2]Department of Gastroenterology, The Putuo District People's Hospital of Zhoushan City, Zhoushan 316100, Zhejiang Province, China. [*]Equal contributors.*

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**Abstract:** Primary splenic amyloidosis (PSA) is an unusual disease, and it could perform as a kind of tumor. Moreover, the research on prognosis of the disease is poor. The etiology, epidemiology, clinical diagnosis and treatment of the disease remain challenging, because case reports of the disease are few in number. To the best of our knowledge, the majority of patients would survive well after the right treatment in accordance with other benign tumors. Here, we report a woman who had complained of upper left bellyache for 1 month and she was confirmed PSA by histology after splenectomy. Computed tomography showed enlargement of the spleen with multiple heterogeneous masses in the upper pole of the spleen. A laparoscopic splenectomy was performed which allowed histopathologic diagnosis. The patient was diagnosed with PSA, and survived for 16 months after surgery. Here, we have a brief discussion about the histopathological features, clinical behavior and treatment of PSA and review the relevant literature.

**Keywords:** Amyloidosis, immunohistochemical, congo red staining, spleen, laparoscopic splenectomy

## Introduction

Primary splenic amyloidosis (PSA) is an unusual disease, and it could perform as a kind of tumor. It was described for the first time by ATKINSON AJ in 1946 [1], and the incidence of PSA has been reported between 0.51 and 0.128 per thousand people [2]. It usually occurs primarily in older female patients, individuals of any age can be affected, the average age is 40 years. The disease has non-specific symptoms and imaging characteristics, and is mainly diagnosed histopathologically and immunohistochemically after surgery. Upper left abdominal pain is the most common clinical manifestation of PSA patients, sometimes. Amyloidosis can occur in various organs like the liver, spleen, kidneys, suprarenals, bone marrow, pancreas, lymph nodes, and blood vessels [1], but location in the spleen region is rare. Here, we present a 45-year-old female patient of PSA and review the literature, also summarize the features and outcomes of PSA.

## Case report

A 45-year-old woman was admitted to our hospital on January, 2015, complaining of upper left bellyache for almost 1 month. Discomfort and pain were not related to meals, defecation or change in position, and could be tolerated. She had not experienced fever or chills since the onset of symptoms. Following the occurrence of these symptoms, the patient reported no weight loss.

She visited our hospital for treatment. On physical examination, it was significant for abdominal tenderness and the abdomen was distended, with a little large, hard, non-mobile mass in the upper left of the abdomen. The patient had no history of excessive alcohol use or obesity, and no history of working with toxic chemicals. She had no prior history of surgery, medical illness, and no known allergies. She was not using any medication. There was no significant family history of splenic disease.

Laboratory examinations on admission revealed: white blood cell (WBC) count 15.40×10^9/L (normal range 4-10×10^9/L), platelet count 213×10^9/L (normal range 100-300×10^9/L), serum tumor markers, including carbohydrate antigen 19-9 (CA19-9), CA-125, α-fetoprotein (AFP), and carcinoembryonic antigen (CEA), were all within normal ranges. Computed

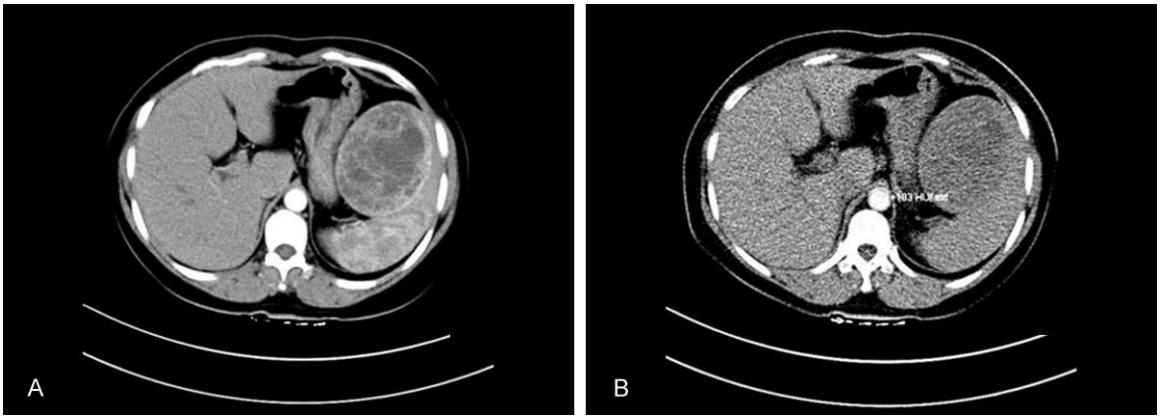


**Figure 1.** CT images of the case of Splenic Amyloidosis. A. CT an upper spleen multiloculated mass measuring 8.2 cm×9.5 cm; B. CT The internal septations were visible and enhanced after intravenous administration of contrast medium.

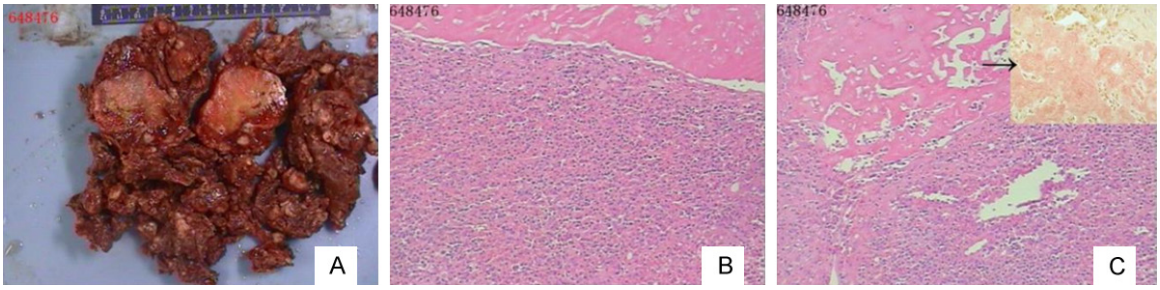


**Figure 2.** Specimens and pathological (HE×100) images of the case of Splenic Amyloidosis. A. A cross-section of the specimen showed an ill-defined reddish lesion with calcification; B. HE×100, the lesion was lined by a small number of red blood cells and lymphocyte whose morphology was normal; C. Congo red staining (arrow indicate).

tomography (CT) imaging of the abdomen at our institution has shown an upper spleen multiloculated mass measuring 8.2 cm×9.5 cm, and normal splenic structure had disappeared. The gallbladder, liver and pancreas were normal (**Figure 1A**). The internal septations were visible and enhanced after intravenous administration of contrast medium (**Figure 1B**). It was considered as a kind of primary splenic tumor: splenic hemangioma? So the diagnosis of the primary splenic tumor was suggested based on imaging findings. A laparoscopic splenectomy was then performed not only for the purpose of curing the disease, but also for histopathologic diagnosis. We observed a crumby tumor mass 8 cm×10 cm in size in the upper pole of the spleen.

A cross-section of the specimen showed an ill-defined reddish lesion with calcification (**Figure 2A**). Then, a histopathologic biopsy was carried out. The pathologic diagnosis of the excised spleen was originating from the spleen, which was confirmed by pathologic examination of the biopsied specimen. Microscopically, the lesion was lined by a small number of red blood cells and lymphocyte whose morphology was normal (**Figure 2B**). The excised spleen tissues are positive for Congo red (+) (**Figure 2C**). Immunohistochemical analysis showed that the tumor cells were positive for CD31+++, CD34+++, and were negative for D2-40.

The patient left the hospital after 12 days with no complications, and she was monitored with abdominal repeat CT after operation. She survived well for 16 months after surgery with no abdominal uncomfortable.

## Discussion

As we all know that the spleen is an important immune organ in our body and it plays an important role in our life, it was rarely reported as the beginning of tumors. Amyloidosis is usually considered as a systemic disease, 10%-

20% cases can be localized [3]. Amyloidosis can occur in various organs like the liver, spleen, kidneys, suprarenals, bone marrow, pancreas, lymph nodes, and blood vessels [1]. To our knowledge primarily isolated spleen has rarely been described in the literature. Though it is possible that the patient has yet exhibited the evidence of systemic disease, spleen involvement can occur in both primary and secondary types of amyloidosis, primary splenic amyloidosis (PSA) is a rare disease caused by a variety of reasons especially spleen cells are infiltrated by special glycoprotein fiber [4, 5]. ATKINSON AJ first described the disease in 1946.

The incidence of PSA has been reported between 0.51 and 0.128 per thousand people [2]. It usually occurs primarily in older female patient, individuals of any age can be affected, the average age is 40 years. The disease has non-specific symptoms and imaging characteristics, and is mainly diagnosed histopathologically and immunohistochemically after surgery. Upper left abdominal pain is the most common clinical manifestation of PSA patients, sometimes. Splenic rupture secondary to PSA was reported [6], the disease could also present with the splenomegaly identified, and quadrant abdominal mass When the patients do the physical examination.

Laboratory findings may have a small link to the disease, and Tumor markers (AFP, CEA, CA-125 and CA19-9) are always within normal ranges or only mildly elevated. While there is still a lack of standardization, the majority of the diagnosis of PSA is suggested at the imaging of the patients. The most common ultrasonographic findings are represented by splenomegaly, ill-defined mass. Computed tomography (CT) imaging could show the enlarged spleen with solitary or multiple nodular masses of heterogeneous low attenuation. The internal septations were visible and enhanced after intravenous administration of contrast medium [7, 8]. In our case, only the CT was performed and the CT findings matched the result reported in previous literature. Nonspecific clinical presentation and laboratory test results emphasize the essential role of imaging in the diagnosis of PSA [9]. PSA should be identified with the splenic benign hemangioma. Our case represents a diagnostic challenge where specific type of amyloid deposit in spleen was difficult to determine, and PSA is mainly diagnosed histopathologically and immunohistochemically after surgery.

The therapeutic strategies for PSA are limited as the disease is extremely rare. Splenectomy is the main treatment method for PSA when it performed as tumor [10]. A laparoscopic splenectomy was performed which allowed histopathologic diagnosis. Due to the immune system of infants is not developed very well, so that a complete splenectomy may have negative effects on system. A complete splenectomy may increase postoperative risks, including the possibility of overwhelming post-splenectomy infection (OPSI). OPSI occurs at an estimated incidence of 0.23%-0.42% per year, with a mortality of 38%-69% [11]. Splenic rupture may occur in PSA [12], thus early diagnosis of PSA followed by splenectomy before rupture may yield a more favorable survival rate. To the best of our knowledge, the majority of patients would survive well after the right treatment in accordance with other benign tumors. In our case, the patient left the hospital after 12 days with no complications, and she was monitored with abdominal repeat CT after operation. She survived well for 16 months after surgery with no abdominal uncomfortable.

## Summary

This case report together with the previous literature review provide an alert for clinicians that primary splenic amyloidosis should be considered when the clinical presentation includes symptoms such as upper left abdominal pain, hematology abnormalities anemia, leukocytosis, early diagnosis showing as tumor, early diagnosis should be a hot topic in the treatment of this disease. The right treatment should be acted, and laparoscopic splenectomy is the first choice for the patients.

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## Disclosure of conflict of interest

None.

**Address correspondence to:** Dr. Yong Li, Department of General Surgery, The First Affiliated Hospital of Nanchang University, 17 Yongwai Zhengjie, Nanchang 330006, Jiangxi Province, China. Tel: +86-791-88694131; Fax: +86-791-88694131; E-mail: yfyly@163.com

## References

[1] Atkinson AJ. Primary amyloidosis of liver, spleen, kidneys, suprarenals, bone marrow, pancreas, lymph nodes, and blood vessels in lungs. Gastroenterology 1946; 7: 477-482.

[2] Cohen AD and Comenzo RL. Systemic light-chain amyloidosis: advances in diagnosis, prognosis, and therapy. Hematology Am Soc Hematol Educ Program 2010; 2010: 287-294.

[3] Urban BA, Fishman EK, Goldman SM, Scott WW Jr, Jones B, Humphrey RL and Hruban RH. CT evaluation of amyloidosis: spectrum of disease. Radiographics 1993; 13: 1295-1308.

[4] Utz JP, Swensen SJ and Gertz MA. Pulmonary amyloidosis. The mayo clinic experience from 1980 to 1993. Ann Intern Med 1996; 124: 407-413.

[5] Kennedy TL and Patel NM. Surgical management of localized amyloidosis. Laryngoscope 2000; 110: 918-923.

[6] Roh J and Huh J. Splenic rupture in primary amyloidosis with chronic neutrophilic leukemia. Blood Res 2015; 50: 5.

[7] Monzawa S, Tsukamoto T, Omata K, Hosoda K, Araki T and Sugimura K. A case with primary amyloidosis of the liver and spleen: radiologic findings. Eur J Radiol 2002; 41: 237-241.

[8] Mainenti PP, Camera L, Nicotra S, Cantalupo T, Soscia E, Di Vizio D, Insabato G and Salvatore M. Splenic hypoperfusion as a sign of systemic amyloidosis. Abdom Imaging 2005; 30: 768-772.

[9] Uchiyama K, Tsukada N, Miyazaki K, Abe Y, Sekine R, Nakagawa Y and Suzuki K. Primary systemic AL amyloidosis with remarkable calcification in the spleen. Rinsho Ketsueki 2014; 55: 334-339.

[10] Liu TY, Chen SC, Wang LY, Chuang WL, Chang WY and Liu CS. Systemic amyloidosis presenting as splenic tumor. Gastrointest Radiol 1991; 16: 137-138.

[11] RN D and RA W. Prevention and management of infections in patients without a spleen. Clin Microbiol Infect 2001; 7: 657-660.

[12] Skok P, Knehtl M, Ceranic D and Glumbic I. Splenic rupture in systemic amyloidosis - case presentation and review of the literature. Z Gastroenterol 2009; 47: 292-295.