Rare cause of spontaneous spleen bleeding: a case report and literature review

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Rupture of the spleen is a serious medical condition manifesting as a sudden abdominal event, potentially life-threatening. Spontaneous spleen rupture is a rare condition. Atraumatic rupture of the spleen is a very unlikely condition. Risk factors include splenomegaly, hemato-oncological diseases, and infections, such as malaria or infectious mononucleosis. Extremely rare is splenic rupture described in autoimmune disease or vasculitis. There has been no reported case of spontaneous splenic rupture as a first manifestation of Churg- Strauss syndrome so far.

Key words: spontaneous spleen rupture, Churg-Strauss, vasculitis.

Zriedkavá príčina spontánneho krvácania sleziny: kazuistika a prehľad literatúry

Ruptúra sleziny je závažný, potencionálne život ohrozujúci stav manifestujúci sa ako náhla brušná príhoda. Spontánna ruptúra sleziny je veľmi zriedkavá príhoda. Atraumatické ruptúry sleziny bývajú asociované s rizikovými faktormi, ako sú: splenomegália, hemato-onkologické ochorenia a infekcie, ako malária a infekčná mononukleóza. Extrémne vzácne sú zaznamenané prípady ruptúry sleziny pri autoimunitných ochoreniach alebo vaskulitídach. Zatiaľ nebol popísaný prípad spontánnej ruptúry sleziny, ako prvej manifestácie Church- Straussového syndrómu, ktorý prezentujeme v tejto kazuistike.

Kľúčové slová: spontánna ruptúra sleziny , Churg-Strauss, vaskulitída.

Introduction

Rupture of the spleen is a serious medical condition manifesting as a sudden abdominal event, potentially life-threatening (1). In most cases, rupture of the spleen is a result of a trauma. Spontaneous spleen rupture is an extremely rare condition and is usually not taken into consideration in the differential diagnosis of acute abdominal pain (1-2). In this report, we present a clinical case of spontaneous spleen rupture in a patient with unknown autoimmune vasculitis, as a first manifestation of disease.

Clinical Case

A 52-year-old woman with previous history of asthma bronchial disease was examined at the emergency department with abdominal pain and repeated vomiting. She gave a history of fatigue, weight loss of 8 kg, pain in the upper abdomen lasting for months. She indicated regular use of prednisone, PPI, and diuretics. Clinical examination pointed pain in epigastrium. Laboratory tests showed serious hyponatraemia (119 mmol/l) and hypochloraemia (85 mmol/l), normocytic anaemia, and elevated level of C-reactive protein (50mg/l). Patient was admitted to

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the internal clinic for substitutional treatment and differential diagnosis. Due to persisting abdominal pain with negative X-ray of abdomen, contrast-enhanced CT examination was performed. CT showed a hemoperitoneum, hematoma in the spleen with active arterial bleeding (Figures No.1 and 2). The patient has repeatedly denied recent injury or trauma. Subsequently, she was moved to the surgical department and referred to acute surgical exploration. Laparotomy verified a hemoperitoneum with approximately 500ml of blood and coagulum, with a 5cm hematoma of the spleen with superficial rupture and leakage of blood. A splenectomy was performed. During awakening from anaesthesia, the patient had an episode of hemoptysis. In the context of this information, we began to search the pathological conditions. She made a good recovery without any other complications. We performed a complex differential diagnosis, examination of blood, urine (with the finding of erythrocyturia), and autoantibodies. Hemato-oncological malignancies and inflectional diseases like EBV, CMV were excluded. Definitive histology has shown the picture of necrotising eosinophilic granulomatosis arteritis, which may correspond with a diagnosis of Churg-Strauss syndrome (Figures No.3 and 4). Patient was immediately referred to rheumatological examination.

Discussion

Atraumatic rupture of the spleen is an extremely rare condition, with an estimated prevalence of 0,1-0,5 %, twice more common in men (3). Risk factors include splenomegaly, hemato-oncological diseases, and infections, such as malaria or infectious mononucleosis (4). Splenic ruptures are extremely rarely described in autoimmune diseases, such as Wegener granulomatosis, lupus erythematosus, and polyarteritis nodosa (5). There has been no reported case of spontaneous splenic rupture as a first manifestation of Churg- Strauss syndrome so far. Of course, the possibility of an injury that the patient is not aware of must also be taken into account.

Churg - Strauss syndrome (also known as eosinophilic granulomatosis with polyangiitis) is a very sporadic condition presented as disseminated necrotizing vasculitis and extravascular granulomas (6). This syndrome typically occurs in patients with preexisting asthma and eosinophilia

Fig. 1, 2. *Active arterial bleeding in spleen (contrast enhanced CT, arterial phase)*

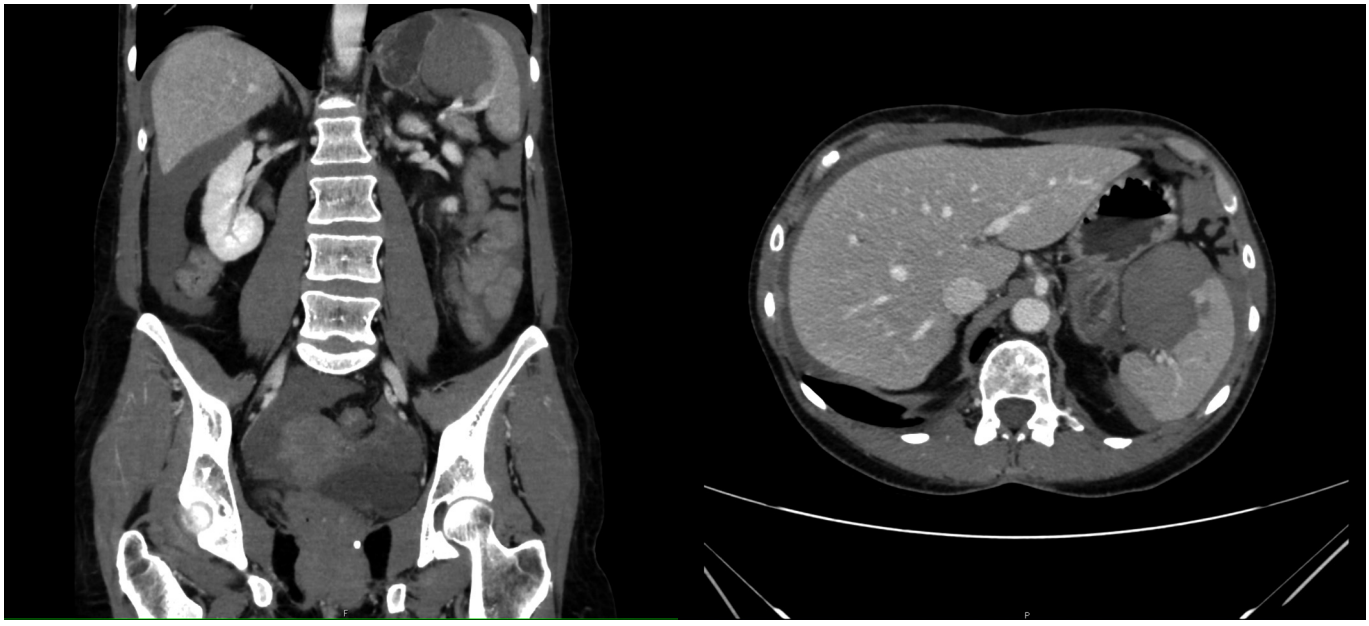
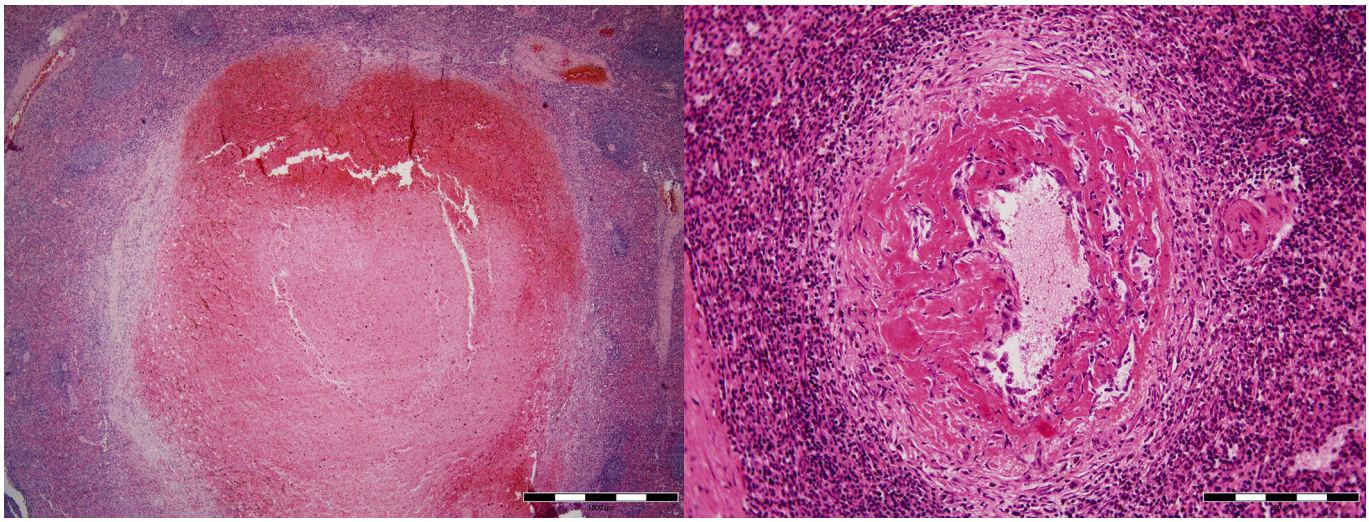


Fig. 3, 4. *Fibroid necrosis of medium large vessel (left, HE 200X) and hemoragic infarction of spleen due to necrotizing vasculitis (right, HE 40x)*



and affects small or medium-size vessels of any organs (7). Together with Wegener's granulomatosis and microscopic polyangiitis belong to vasculitis associated with the presence of antineutrophil cytoplasmatic autoantibodies (ANCA) (8). According to available papers, traces of ANCA antibodies have been found in 40-75% of patients. Estimated incidence is approximately 2,5 cases per 100 000 adults per year. The average age of patient is 50 years (9). Diagnosis can be difficult. The American College of Rheumatology suggested six criteria for diagnosis Churg-Strauss syndrome: presence of asthma, eosinophilia of more than 10 % in peripheral blood, paranasal polyps or sinusitis, pulmonary infiltrates, histological proof of vasculitis with extravascular eosinophils and mononeuritis multiplex (or polyneuropathy) (10). The prognosis depends on the start of the treatment. Without treatment, 5-year survival rate of patients is about 25% (11). The disease is obscure, with minimal symptoms and a variable clinical picture. Spleen involvement is rare and usually presents as splenic infarction. There is no reported case of asymptomatic splenic rupture associated with Churg-Strauss disease.

Conclusion

Spontaneous spleen rupture is an extremely rare condition and usually is not considered in the differential diagnosis of acute abdominal pain. In this report, we presented a clinical case of spontaneous spleen rupture in a patient with unknown Churg-Strauss disease, as a first manifestation of disease.

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REFERENCES

1. Tunçyürek Ö, Tunçyürek P, Ertekin E et al. Pathological rupture of the normal spleen: Review with the literature. Int J Surg Case Rep. 2016;26:163-5. doi: 10.1016/j.ijscr.2016.07.029. Epub 2016 Jul 28. PMID: 27497938; PMCID: PMC4975709.

2. Wehbe E, Raffi S, Osborne D. Spontaneous splenic rupture precipitated by cough: a case report and a review of the literature. Scand J Gastroenterol. 2008;43(5):634-7. doi: 10.1080/00365520701763472. PMID: 18415760.

3. Abbadi SE, Rhouni FZ, Jroundi L. Rupture spontané de la rate: à propos d'un cas et revue de la littérature [Spontaneous splenic rupture: about a case and review of the literature]. Pan Afr Med J. 2017;27:62. Published 2017 May 29. doi:10.11604/pamj.2017.27.62.12451.

4. Baibars M, Ohrum P, Alraiyes AH et al. Asymptomatic spleen rupture in patient with endocarditis, QJM: An International Journal of Medicine, Volume 106, Issue 11, November 2013, Pages 1047–1048, https://doi.org/10.1093/qjmed/hcs190.

5. Greco A, Rizzo MI, De Virgilio A et al. Churg-Strauss syndrome. Autoimmun Rev. 2015 Apr;14(4):341-8. doi: 10.1016/j.autrev.2014.12.004. Epub 2014 Dec 11. PMID: 25500434.

6. Nguyen Y, Guillevin L. Eosinophilic Granulomatosis with Polyangiitis (Churg-Strauss). Semin Respir Crit Care Med. 2018 Aug;39(4):471-481. doi: 10.1055/s-0038-1669454. Epub 2018 Nov 7. PMID: 30404114.

7. Keogh KA, Specks U. Churg-Strauss syndrome. Semin Respir Crit Care Med. 2006 Apr;27(2):148-57. doi: 10.1055/s-2006-939518. PMID: 16612766.

8. Boyer D, Vargas SO, Slattery D et al. Churg-Strauss syndrome in children: a clinical and pathologic review. Pediatrics. 2006 Sep. 118(3):e914-20.

9. Masi AT, Hunder GG, Lie JT, et al., The American College of Rheumatology 1990 criteria for the classification of Churg-Strauss syndrome (allergic granulomatosis and angiitis). Arthritis Rheum. 1990 Aug. 33(8):1094-100.

10. Keogh KA, Specks U. Churg-Strauss syndrome: update on clinical, laboratory, and therapeutic aspects. Sarcoidosis Vasc Diffuse Lung Dis. 2006 Mar. 23(1):3-12.

11. Hattori N, Ichimura M, Nagamatsu M et al. Clinicopathological features of Churg–Strauss syndrome-associated neuropathy, Brain, Volume 122, Issue 3, March 1999, Pages 427–439, https://doi.org/10.1093/brain/122.3.427.