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Review

Sclerosing angiomatoid nodular transformation of spleen: A report about a new clinical observation and literature review

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Sclerosing angiomatoid nodular transformation of spleen is a benign primitive vascular tumor of unknown etiology first described in 2004 by Martel et al. This condition can occur at any age with a female predominance but has been rarely described in children. The clinical signs found in our study are mainly splenomegaly and weight loss occurring in a context of chronic inflammatory syndrome and mixed microcytic anemia in a 39-year-old patient. CT and abdominal MRI used preoperatively helped the diagnosis of sclerosing angiomatoid nodular transformation of spleen (SANT) even though the radiological characteristics of splenic lesions observed were little specific. Immunohistochemistry after biopsy of the spleen helped make the diagnosis of certainty of SANT. Pathological examination of SANT reveals a splenic parenchyma replaced by well circumscribed angiomatoid nodules separated by an inflammatory fibro sclerotic stroma. Splenectomy by laparoscopic surgery with curative purpose ended all chronic inflammatory process maintained by this benign vascular lesion of spleen.

Keywords: Sclerosing angiomatoid nodular transformation of spleen, vascular injury, splenectomy, immunohistochemistry.

INTRODUCTION

Sclerosing angiomatoid nodular transformation of spleen (SANT) is a benign primitive vascular tumor of spleen. It was first described in 2004 by Martel et al. in a series of 25 cases (Martel al., 2004). From 2004 to date approximately less than 100 cases have been reported in the literature worldwide (Bagul et al., 2015) Sclerosing angiomatoid nodular transformation of spleen (SANT) is a rare tumor whose exact etiology remains unknown.
SANT is usually asymptomatic, and about 50% of cases are discovered fortuitously at imaging (Kakisaka T and al., 2014). The abdominal pain is a common symptom, followed by splenomegaly in symptomatic patients (Falk et al., 2012). SANT can simulate a neoplastic condition. Radiological and immunohistochemical examinations help make the diagnosis of SANT.
We report the case of a recent description of a pathological tumor, SANT, treated by laparoscopic splenectomy.

OBSERVATION

The 39-year-old female patient, a single person with no particular history was followed from 2007 to 2014 in

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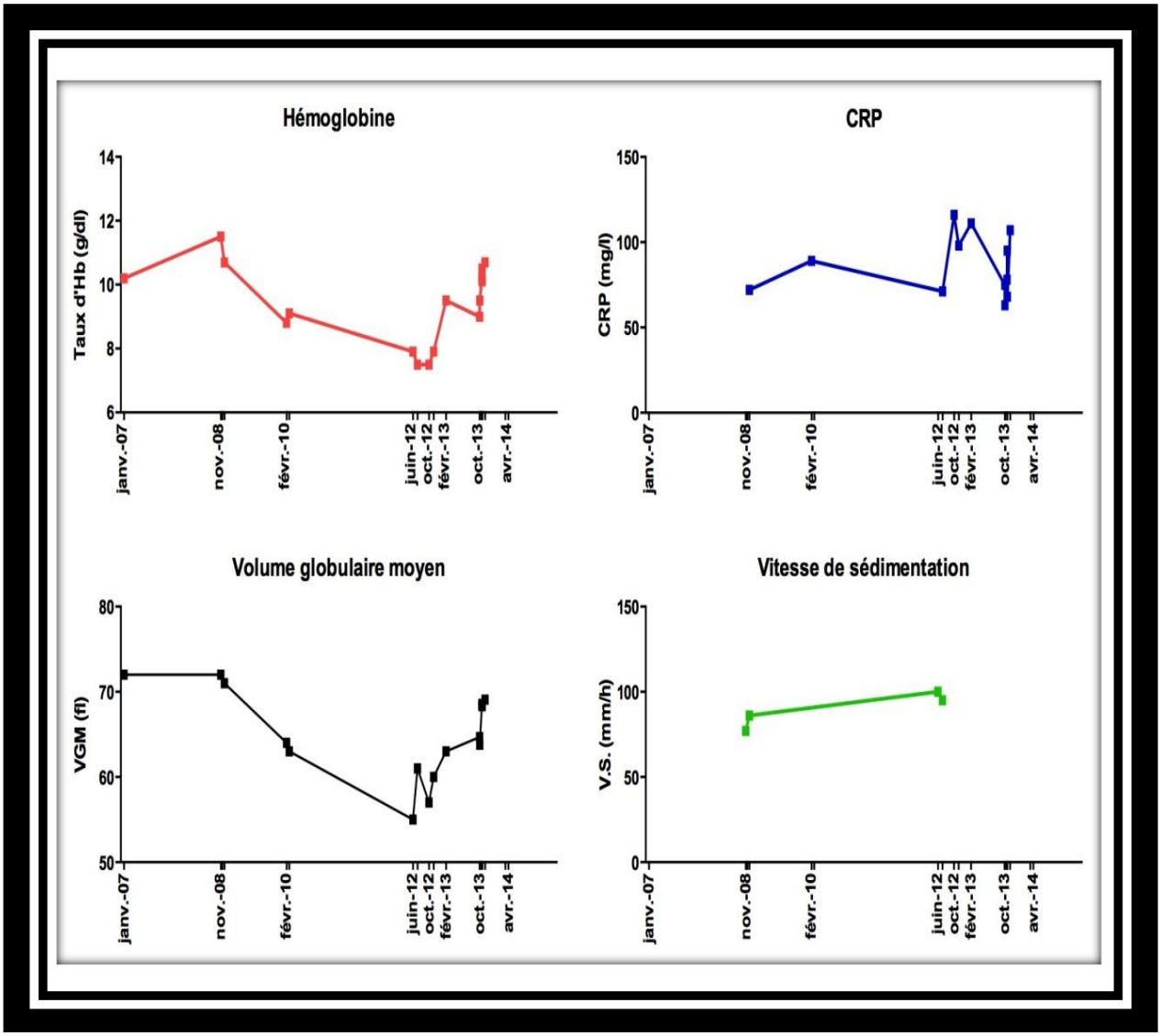
Fig.1. Graph summarizing the evolution of anemia and chronic inflammatorysyndrome from 2007 to 2014 before splenectomy.



internal medicine for chronic inflammatory syndrome, microcytic anemia and weight loss (fig 1).

She was hospitalized in internal medicine from 30/09//13 to 03/10/13. On clinical examination and on admission the weight = 55 kg. Size = 166 cm. BMI = 19.96. BP = 108/70 mmHg. Pulse = 79/min. the oxygen saturation = 99%. Temperature = 36.30 c. The patient describes discomfort in the left hypochondrium, which is accentuated on palpation. In addition, the splenomegaly is palpable, of type II according to Hackette's classification. There is no left scapular radiation. The rest of the examination is unremarkable. The review carried out revealed a microcytic iron deficiency anemia (Hb = 10.2 g/dl. VGM = 68.4 fl. Serum iron = 5.3 micromol/l. Transferrin = 1.6 g/l. Total capacity of iron binding = 41 micromol/l. Ferritin = 752 microg/l), a persistent inflammation (Fibrinogen = 7.01 g/, CRP = 78 mg /l albuminemia= 29 and polyclonal hypergamaglobulinemia) and a splenic nodule of 7 cm in diameter associated to hepatic hemangioma in abdominal CT (fig 2).

An endoscopy and a colonoscopy performed are normal as well as histology. Abdominal ultrasound revealed increased splenic nodule (8.3cm VS. 6.5cm in 2009) very poorly vascularized making us suggest hamartoma.

Aregenerative iron deficiency anemia persistence with normal lymphocytic under populations.

Bone marrow biopsy found a reaction rich marrow without lymphoid tumor infiltration. The performance of a guided biopsy of the splenic nodule will bring out sclerosing angiomatoid nodular transformation of spleen. Laparoscopic splenectomy is performed in March 2014 due to the persistence of inflammation and weight loss of the patient (Figure 3).

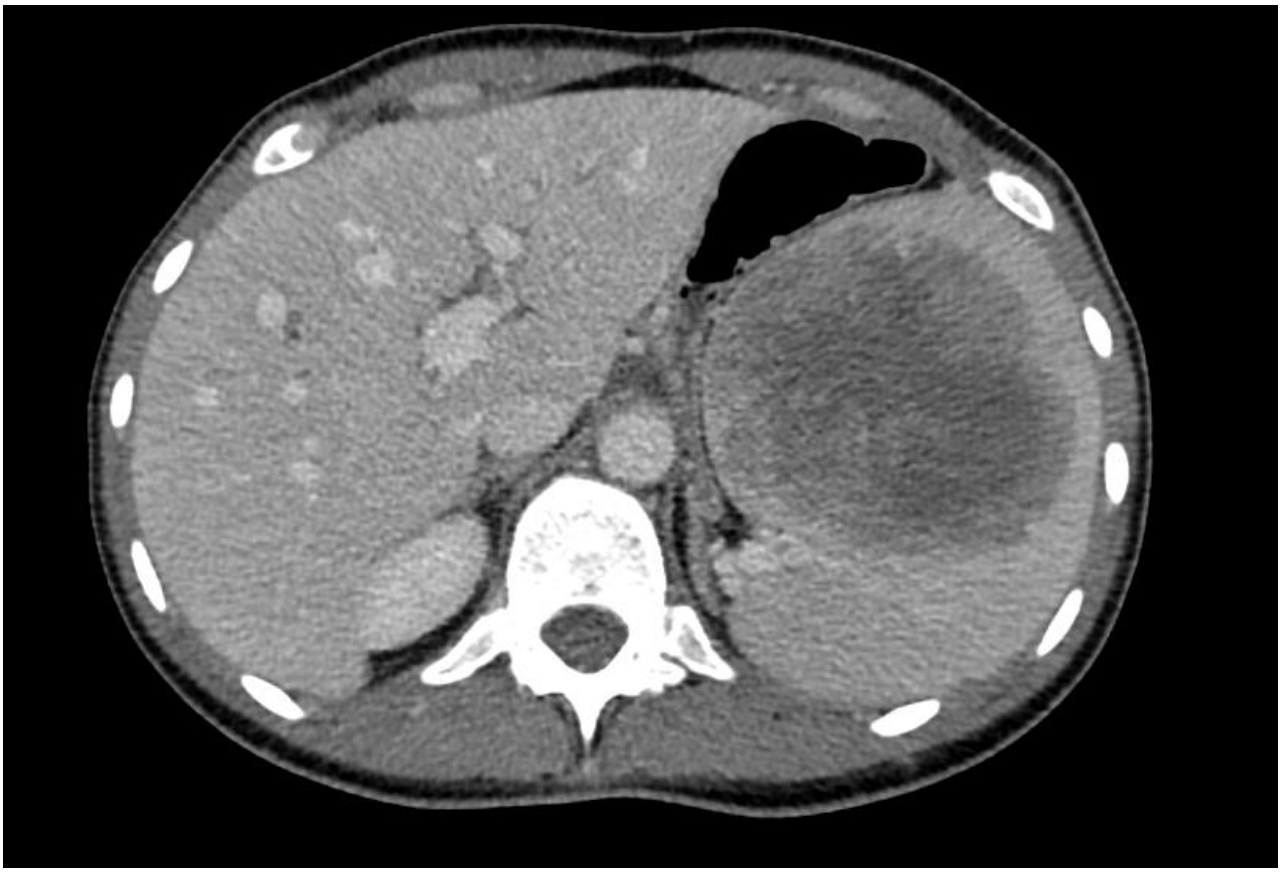


Fig 2. CT by improved contrast shows an iso-dense mass in the spleen with liver angiomas.

The macroscopic appearance reveals a mass of 3 to 17cm, non-encapsulated, well demarcated from the rest of the spleen. The splenic parenchyma is the site of angiomatousred-brown nodules separated by a c (fig. 4).
The microscopic appearance helps discover nodules of varying size, composed of vascular structures whose immunohistochemical analysis finds different normal vascular components of red pulp that are sinusoidal lining cells, capillaries and small veins. Immunohistochemical staining of the small vessels in the lesion were positive for CD34, CD31, CD68 and CD163, and negative for CD8. Histological sections and immunohistochemical staining performed on the lesion of the spleen confirmed the diagnosis of SANT.
The outcome to a month after splenectomy compared to previous results is characterized by the disappearance of inflammation, correction of anemia (fig.5).

DISCUSSION

Splenic tumors can be classified into tumors of lymphoid or vascular origin (Kakisaka et al., 2014). The vast majority of primary splenic lesions are vascular (Thacker et al., 2010). SANT is a new entity of primitive benign vascular tumor of the spleen newly diagnosed then long ignored.
SANT usually affects middle-aged adults with a slight female predominance; the sex ratio is 2/1. In the series of Martel et al. the average age was 53.7 years, with extremes ranging from 22 to 74 years (Martel et al., 2004). Cases of SANT have been described in pediatrics (Kuybulu et al., 2009). SANT is usually asymptomatic but patients may experience intermittent abdominal pain, splenomegaly, growth retardation in the child, weight loss, fever and elevated ESR (Bamboat et al., 2010). Indeed symptoms are non-specific and are variable from one person to another.
Although preoperative imaging can be used to help diagnose SANT, radiologically, the characteristics of splenic lesions are often little specific **(**Abbott et al., 2004). However the literature suggests that the result at the scanner evocative of SANT is a hypo-vascularized lesion, centered with a peripheral enhancement extending to the center of the lesion through partitions giving an aspect "of spokes"(Bamboat et al., 2010).This radiological aspect is observed in harmatoma, which as

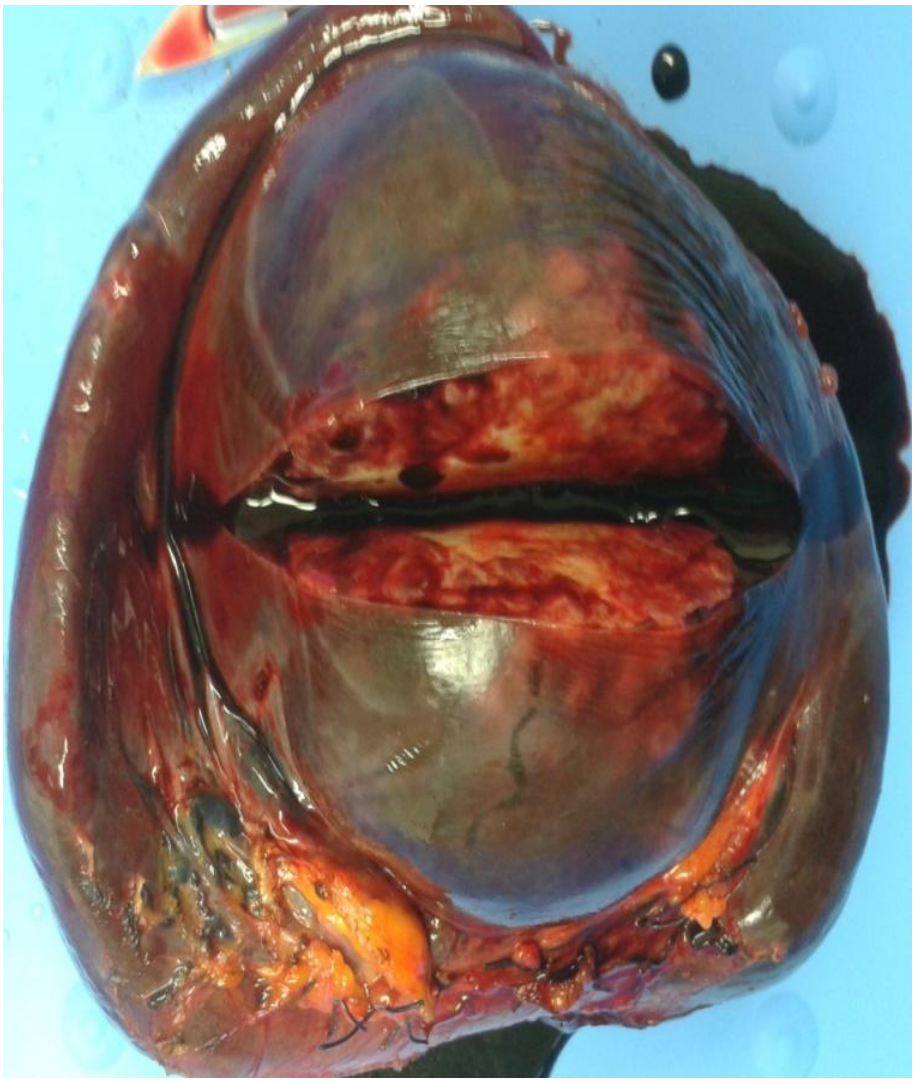


Fig. 3 hemorrhagic cross section of SANT after splenectomy

even been suspected on the ultrasound scan in our patient. SANT sometimes takes the aspect of a solitary mass, as in our case; defined, it measures between 3 and 17 cm in large diameter with a bumpy surface separated by a dense fibrotic stroma, gray-white with hemorrhagic sites that may evoke an inflammatory pseudo tumor of spleen **(**Abbott et al., 2004). As for MRI, it brings out the dual component in the form of areas in T2 hyper-signal predominantly peripheral, performing radial spans, with enhancement after injection gradually and centripetal corresponding to the intra-lesion vascular phase that contrasts with the fibro-sclerotic component that is less vascularized (Bamboat et al., 2010). In fact the suspicion of the diagnosis of SANT is established from radiological images and confirmation is made by histological and immunohistochemical examinations (BurneoEsteves et al., 2012).

Histologically, SANT is a non-encapsulated tumor lesion, although well limited by the spleen tissue. Typical angiomatoid nodules of SANT are composed of different endothelial cells from the red pulp. They are separated by a fibro-sclerotic stroma.

At the microscopic level, the fibro-sclerotic stroma contains macrophages loaded with hemosiderin, lymphocytes and plasma cells. The intercellular stroma may show large areas of hyalinization.

Immunohistochemical study of SANT, as in our patient, showed that in angiomatoid nodules, endothelial cells of vessels had various immunophenotypic expressions defining three distinct types of vessels intertwined: capillary type of endothelial cells that express for CD34 (+) / CD 31 (+) / CD8, the lining cells of the sinusoids expressing for CD8 (+) / CD 31 (+) / CD34 and finally the type venous of endothelial cells that are positive for CD31 and negative for CD8 and CD34.

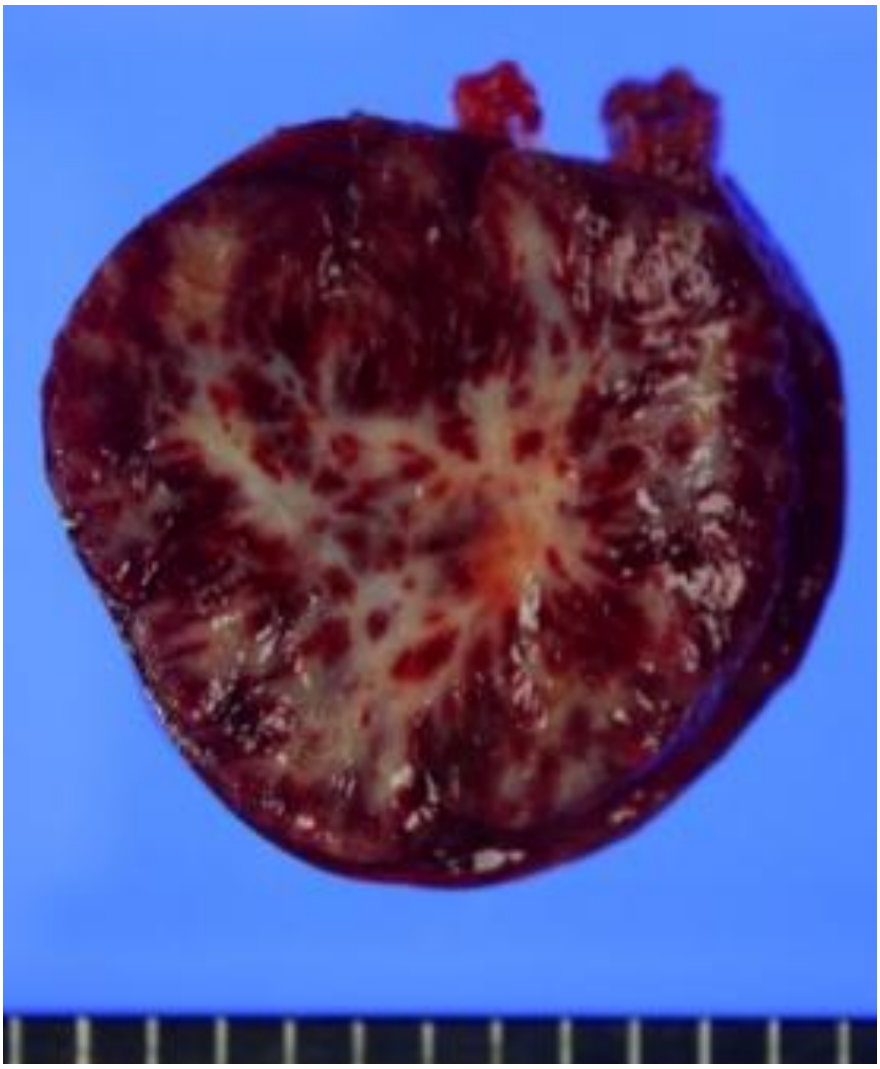


Fig 4. Mass of 3 to 17cm, non-encapsulated, well demarcated from the rest of the spleen, site of angiomatous red-brown nodules separated by a fibrosclerotic stroma.

Post-splenectomy immunohistochemistry is the only method to confirm the diagnosis of SANT (BurneoEsteves et al., 2012). However because of preoperative diagnostic difficulties, percutaneous biopsy of the spleen was advocated by (Gutzeit et al 2009). Wang et al., (2012). This needle splenic biopsy was used as in our case by Weinreb et al. that suggests that this method helps distinguish SANT from other lesions (Gutzeit et al., 2009). Although SANT is a distinct histological entity, several vascular lesions both benign and malignant, such as inflammatory pseudo-tumor of spleen, splenic hemangioma, splenic hamartoma, littoral-cell angioma, endothelialhemangioma, angiosarcoma and primary splenic lymphoma can be discussed. However, there is concern about the risk of intra-peritoneal dissemination if the biopsied lesion proves to be angiosarcoma. Other complications such as rupture of spleen and splenic post puncture biopsy bleeding may occur (Ilan et al., 2007]. Thee indication of splenectomy by laparoscopy with therapeutic purpose has been recommended in this patient. Recently, laparoscopic splenectomy has become the standard technique for the surgical treatment of tumors and hematological diseases and splenic tumors (Kim et al., 2011).

The theory on the active part of inflammatory components in the pathophysiology of SANT could be supported by the complete disappearance of biochemical parameters of inflammation after surgical removal of the tumor.

CONCLUSION

Sclerosing angiomatoid nodular transformation of spleen is a rare primitive entity to be evoked in the presence of the fortuitous discovery of a solid splenic lesion at

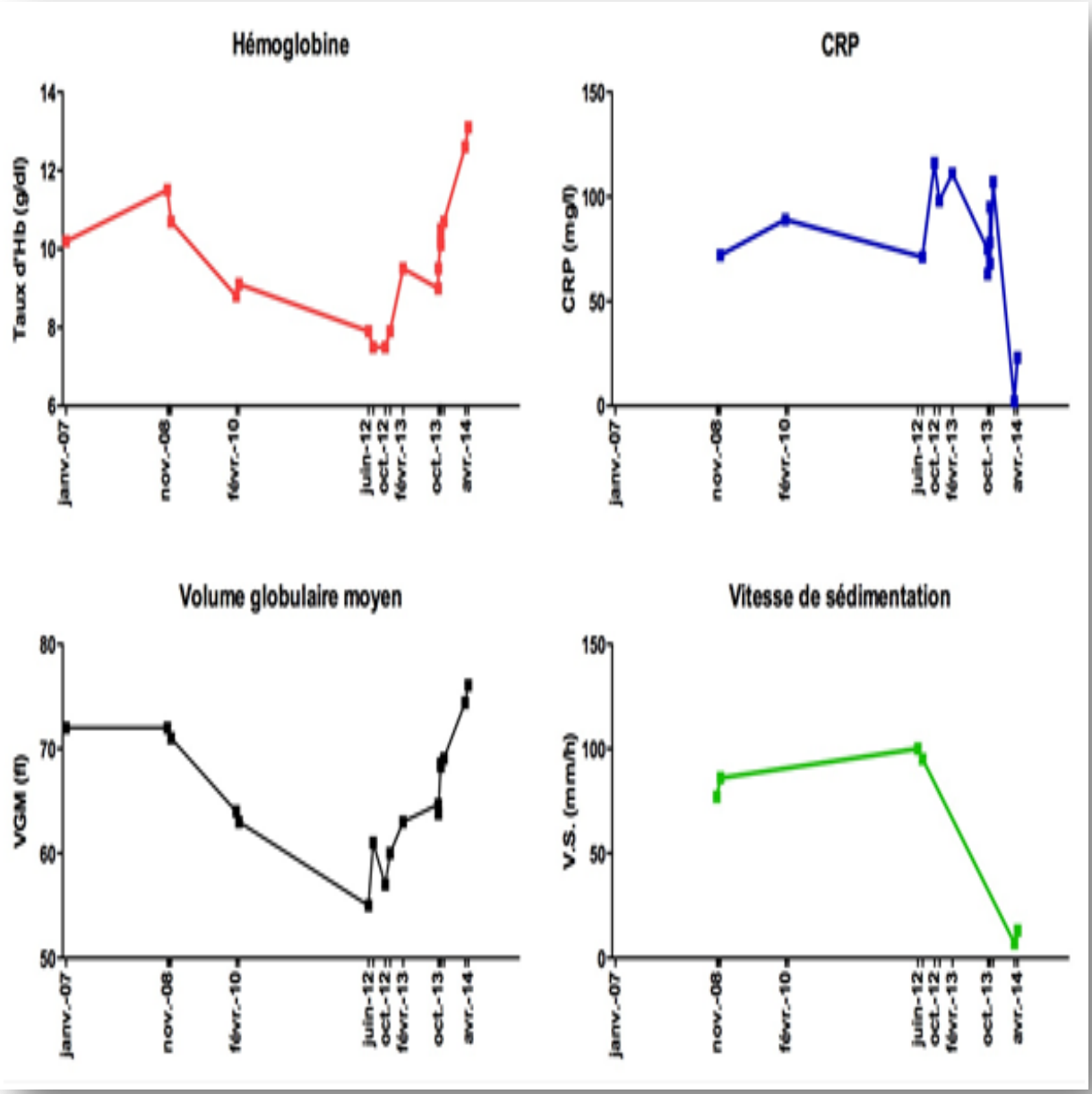


Fig.5: graphic describing the disappearance of chronic inflammatory syndrome and anemia correction at one month post splenectomy

imaging. Immunohistochemistry is the only diagnostic means of SANT. It has a good prognosis because no recurrence or secondary unfavorable outcome is currently reported with SANT after splenectomy.

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