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Case Report

# Splenic Infarction by Wegener's Disease a Practically Non-Existing Pathology

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## Abstract

**Introduction:** Granulomatous Polyangiitis or Wegener's Disease is an extremely rare disease; it is defined as a systemic autoimmune vasculitis. On the other hand, splenic infarction is equally rare and consequent on another pre-established disease.

**Objective:** Case report and literature review.

**Methods and Results:** It is the presentation of a 30-year-old male patient with a nonspecific clinical picture but with data of a systemic inflammatory response, with emergency medical attention, where in laboratory and cabinet studies; he undergoes an emergency surgical procedure and the histopathological report confirms the diagnosis as the genesis of splenic infarction.

**Discussion:** Splenic infarction has been linked to various etiologies, such as cardioembolic, aortic, vascular, anatomic (errant spleen), embolism, septic, autoimmune, neoplastic, hematological, iatrogenic, traumatic, and idiopathic embolisms. There are few published cases, being practically anecdotal. It must be clarified that they are two pathological entities that must be identified and treated differently, arguing their therapy due to their etiology Wegener's granulomatosis and its splenic infarction consequence.

**Conclusions:** In our country (Mexico), splenic infarction due to granulomatous polyangiitis is practically unknown and/or has not been documented.

Primary treatment should focus on the etiology and solve without delay the consequence of splenic infarction with surgical management as the only option and in an energetic manner.

***Keywords:*** *Polyangiitis; Granulomatosis Vasculitis Autoimmune; Heart Attack; Splenic*

## Introduction

Granulomatosis with Polyangiitis or Wegener's disease (GP) is an extremely rare disease; in 1931 Klinger identified the first case, but it was described by Friedrich Wegener in 1936, who managed to associate the clinical and pathological features of this disease [1,2]. It is defined as a systemic autoimmune vasculitis caused by the action of cytoplasmic antibodies to anti neutrophils (ANCAs), proteinase-3 (PR3) produced by neutrophils. Typically, its condition is in the upper and lower respiratory tract, as well as in the kidney area [3].

On the other hand, splenic infarction (SID) is equally rare; not very clearly defined as an isolated pathological entity, but as in a great majority of cases, it is of a secondary consequence to an already established condition or disease [4].

## Objective of the Study

Case report and literature review.

## Case Report and Result

A 30-year-old male patient with no significant history who came to the emergency department with chronic, colicky abdominal pain of mild intensity and occasional presentation in the left upper quadrant over 3 months of evolution. 72 hours prior to his admission, he becomes an oppressive pain, of sudden onset, of high intensity, continuous, fixed in the left hypochondrium; accompanied by abdominal distension, nausea without vomiting, hyporexia and unquantified and intermittent fever of 12 hours of evolution. Physical examination with tachycardia of 100 X´ FR: 26x´, T/A: 110/70 mm Hg. Temperature: 37.8°C. With abdominal facies, abdomen with diminished peristalsis, on palpation of the abdomen with minimal splenomegaly, pain on superficial and deep palpation with data of peritonism and abdominal rebound; rest with no data to add. Laboratories with leukocytosis of 17,800, neutrophilia, lymphopenia, thrombocytosis of 598,000. Rest of laboratories in normal parameters. Report of the abdominal tomography study with intravenous contrast highlights that the spleen presents homogeneously decreased densities, with minimal heterogeneous nodular enhancement, both kidneys with multiple hypo-densities in their parenchyma. Figure 1 emergency surgery is performed with laparoscopic splenectomy. Figure 2 and 3 shows the histological report describes a splenic infarction with necrosis associated with vasculitis with granulomatosis and eosinophils: Granulomatosis with Polyangiitis or Wegener's disease (GP). He has a satisfactory evolution, he graduates after management and control and for the specialty of Rheumatology.

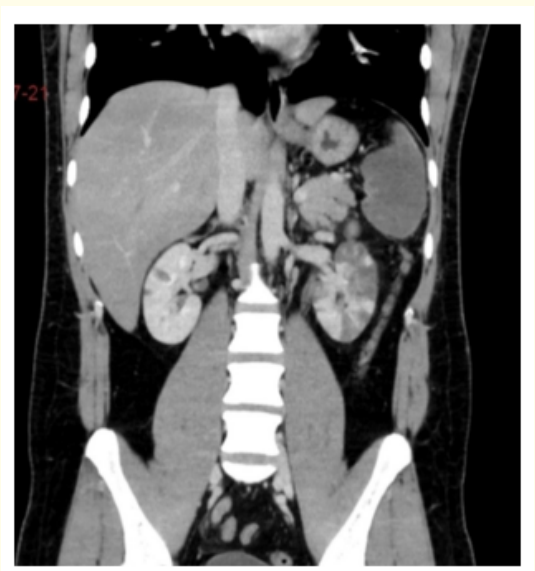

***Figure 1:*** *Tomographic reconstruction showing splenic infarction.*

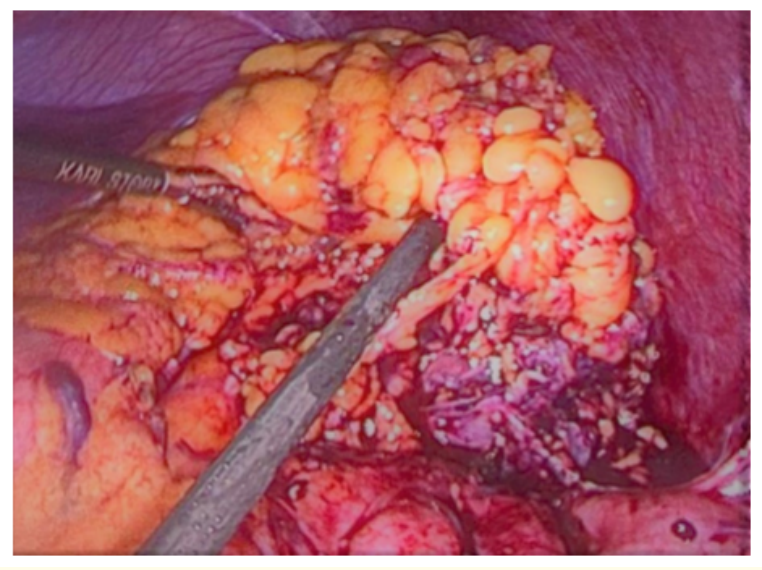

***Figure 2:*** *Laparoscopic splenectomy.*

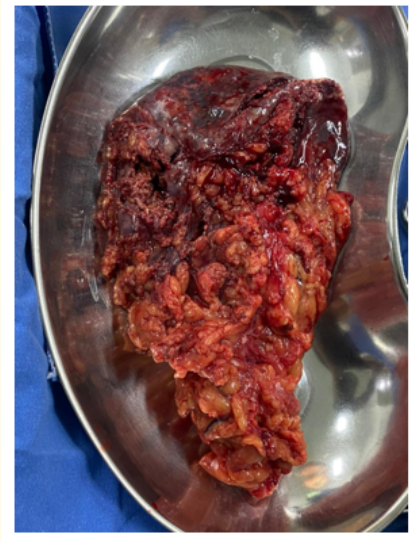

***Figure 3:*** *Surgical specimen: Spleen.*

## Discussion

Describing a patient with an EI associated with an etiology of GP is extremely rare indeed; with a nonspecific clinical picture, so the diagnosis is extremely difficult [5,6]. IE has been linked to various etiologies, such as cardioembolic, aortic embolism, vascular, anatomic

(wandering spleen), embolism, septic, autoimmune, neoplastic, hematologic, iatrogenic, traumatic, and idiopathic [5,7-10]. There are few published cases in which it is concluded that patients with IE have an etiology due to GP; That is why there is no real incidence or prevalence of the disease, being practically anecdotal [11-13].

Regarding the use of diagnostic support with extension studies by office, the tomographic study can confirm the previous clinical diagnostic suspicion of an IE, if it were to occur; where, therefore, the viability of the organ in question can also be evaluated [5,11-13]. No other desk study is documented in the literature that allows greater diagnostic sensitivity and specificity, such as the hepatosplenic scintigraphy or selective arteriography.

Regarding the treatment, this is very diverse, not concluding in any way a behavior or criteria that delimit or not, the conservative medical management of the surgical management in relation to IE. It must be clarified that they are two pathological entities that must be identified and treated differently, arguing their therapy based on their etiology (GP) and their consequence (IE). That is why they should be treated differently and the behavior clarified, both as for the primary treatment (cause) and as for the secondary treatment (consequence). Primary treatment is highly variable and will depend on its origin, however secondary treatment (or consequence) of EI is fearful according to the few references; [14-16]. it is mentioned to carry out with great importance a follow-up of the patient due to the risk of abscess, rupture and hemorrhage [17]. That is why the IE leaves many questions regarding its therapeutic behavior; This is with regard to what has been found in the references in world literature and regarding its evolution and prognosis; that following these behaviors, puts the patient's life at risk.

## Conclusions

- IE is a sequela to the presentation of another already existing disease, which is actually extremely rare, and which occurs due to very diverse etiologies; in our country (Mexico) EI due to GP is practically unknown and/or has not been documented.
- GP is a practically exclusionary diagnosis, which in most cases requires histopathological confirmation and thus concludes the genesis of IE.
- The primary treatment should focus on the etiology and solve without delay the consequence that is IE, with a surgical conduct; this is justified by determining that the permanent sequelae (necrotic tissue) are not recoverable, of the organ involvement. All of the above leads the authors to conclude that surgical management is considered as the only option and energetically.

## Conflict of Interests

The authors declare that they have no conflict of interest.

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