Autopsy Case Reports

Image in Focus

Gamna-Gandy bodies of the spleen in sickle cell disease

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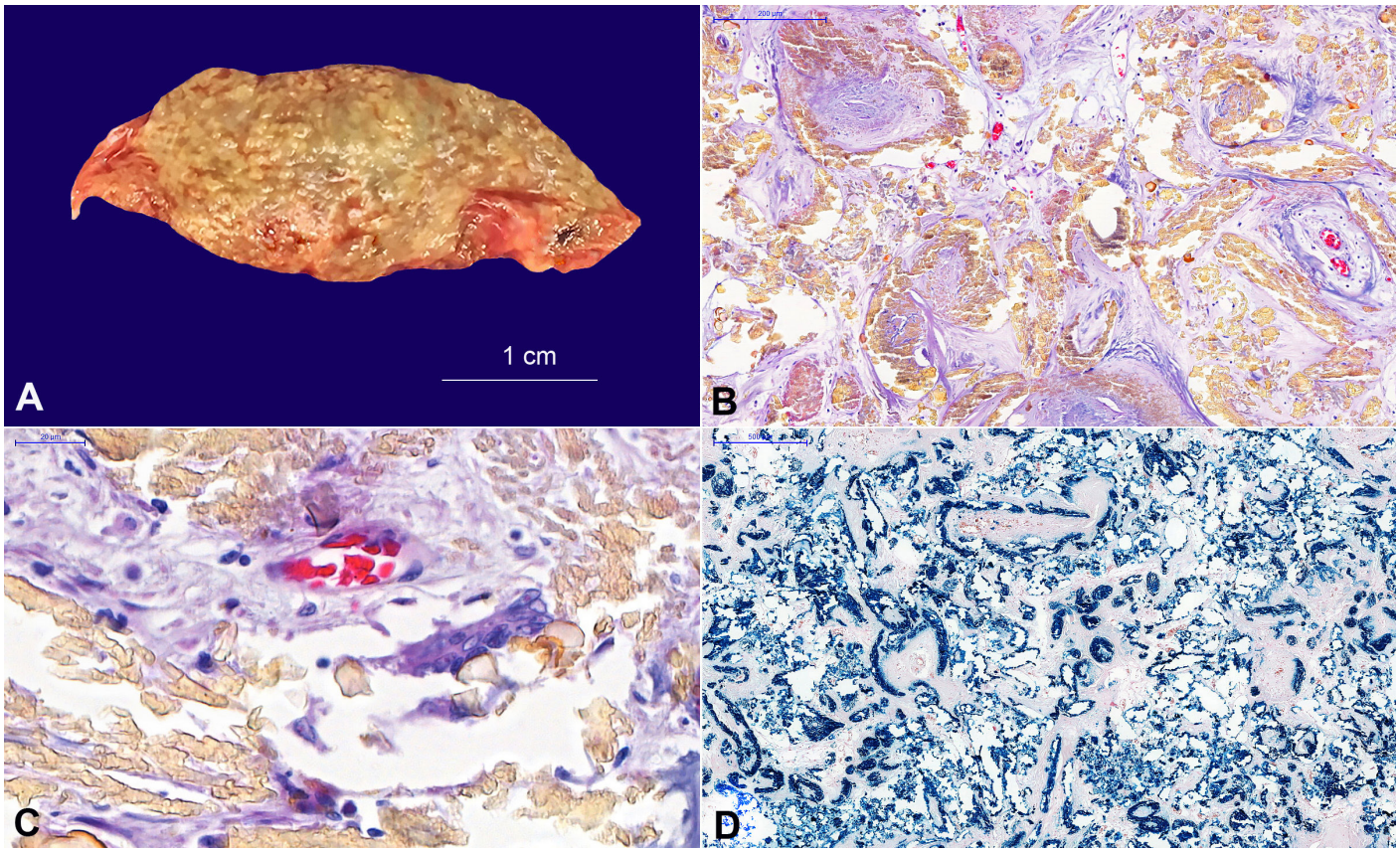

Figure 1. A – Gross examination of the atrophic spleen with a light brownish external surface; **B, C**, and **D –** Photomicrographs of the spleen; **B –** shows the foci of interstitial fibrosis with iron deposits (H&E, 200X); **C –** shows the granulomatous reaction with multinucleated foreign-body cells surrounding the iron deposits (H&E, 400X); **D –** shows iron pigmentation associated with fibrous tissue (Pearls, 200X). Scale bars: B, C, D = µm.

Gamna-Gandy bodies (GGBs), also called tobacco flecks or siderotic nodules, appear as yellow-brownish and spheroidal foci within the splenic parenchyma, are composed of deposits of iron pigments and calcium salts, and are associated with granulomatous inflammatory reactions with multinucleated foreign-body giant cells and fibrous tissues (Figure 1).[1,2] GGBs can vary in size, ranging from 10 to 49 microns in the largest dimension.[3]

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The first description of GGBs dates back to 1902 when Marini described the siderotic nodules in the spleen. Three years later, the French physician Charles Gandy found these structures in the spleen of a patient with biliary cirrhosis, but the significance of these structures was not elucidated.[1,4,5] In 1910, the same finding was described in the lung during the autopsy of a patient who died of endocarditis. At that time, GGBs were associated with a fungal etiology since they microscopically resembled spores. In 1921, the etiology of this pathological finding was better described when Carlos Gamna, an Italian pathologist, found GGBs in the spleen of a patient who died of chronic hemolytic anemia. Gamna observed that the amorphous material was composed of iron and calcium sulfate deposits and was therefore named *Splenogranulomatosi siderotica*. In 1963, the name "Gamna-Gandy bodies" became established and has been widely used ever since.[2]

Portal hypertension is one of the pathophysiological mechanisms that explains the formation of these nodules. Patients with increased blood pressure in the splenic circulatory system are likely to have episodes of minimal bleeding in the splenic parenchyma, which is organized as hemosiderin (iron), calcium salts and fibrotic tissue.[1,2,6,7] GGBs in the spleen can occur with three main patterns: i) fine granular deposits in the red pulp; ii) iron deposits associated with subcapsular infarcts; and iii) periarterial deposits associated with fibrosis.[2] Other histological characteristics of GGBs include blood vessels with proliferation of the tunica media, macrophagic reactions and spheroid fibers.

Sickle cell disease (SCD) is another major entity in which GGBs can be found.[8] The pathophysiological mechanism of GGBs in SCD is due to chronic episodes of vaso-occlusion and hemolysis in the central arteriole of the white pulp with periarteriolar hemorrhages. Then, mineral elements of the blood will deposit to form GGBs. Piccin and colleagues analyzed the spleen of 17 patients (all African American and mostly male) with diagnoses of SCD and found GGBs in 65% of the patients. The authors determined that the chemical composition of GGBs was carbon (47.1%); oxygen (29.7%); phosphorus (9.0%); iron (7.4%); calcium (6.4%); and potassium (0.4%).[2] The presence of GGBs is not pathognomonic of SCD since GGBs can occur in other diseases and in several other organs (Table 1).[1-7,9]

Figure 1 refers to a study of a spleen sampled from the autopsy of a 46-year-old male African American patient with a history of sickle cell anemia, smoking, and alcoholism. The organ weighed 4 g (reference range; 150-250 g) and measured 3.0 cm on the largest dimension. Externally, the capsule was fibrinous. The cut surface of the spleen parenchyma was rugged and had a sandy texture with structural distortion. The patient was admitted to the emergency room with sudden onset dorsalgia and chest pain and died a few minutes after admission. An autopsy was performed, which confirmed death due to a massive pulmonary thromboembolism.

The authors retain an autopsy informed consent form.

Keywords
Anemia, Sickle Cell; Autopsy; Spleen; Splenic Diseases.

Table 1. Conditions associated with Gamna-Gandy bodies

Neoplastic Diseases	Thymoma
	Thyroid Follicular Adenoma
	Pituitary Adenoma
	CNS Neoplasms
	Hairy Cell Leukemia
	Hodgkin and non-Hodgkin Lymphoma
	Cardiac Myxomas
	Angiosarcoma
	Ovary Carcinoma
	Liver Carcinoma
	Renal Cell Carcinoma
	Breast Cancer
	Gastric Neurinoma
Non-neoplastic Diseases	Portal Hypertension
	Thrombotic Thrombocytopenic Purpura
	Idiopathic Thrombocytopenic Purpura
	Hemolytic Anemia
	Paroxysmal Nocturnal Hemoglobinuria
	Acquired Hemochromatosis
	Congestive Splenomegaly
	Retroperitoneal Lymph Nodes

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