Phillippe Charles Ernest Gaucher (1854-1918) described a case of the disease that bears his name in his 1882 thesis for the doctorate of medicine at Paris. His academic career continued in that city, and he rose through the ranks to professor of dermatology and syphilology, that appointment coming in 1902.

The patient was a woman, and Gaucher remarks that she had neither ascites nor leukemia, just a very large spleen. He remarks on the rarity of any type of cancer of the spleen and then presents his "conclusions" as follows:

There is a primary and idiopathic type of hypertrophy of the spleen for which I propose the designation primary epithelioma of the spleen, on the basis of its histological characters.

The clinical characters of this disease are as follows:

A marked splenic hypertrophy, progressive, although slowly so at times, accompanied by acute pain and various pressure phenomena, hemorrhages (nose-bleed, purpura and hemorrhagic gingivitis), at times icterus, due to a secondary hypertrophy of the liver, without leukemia, without intermittent fever, without ascites, and terminating in a special type of cachexia.

Its anatomic characters are:

A spleen of very great volume (4.77 kg), uniformly and regularly developed, with glossy and smooth surface, almost normal in shape and color, but hard and sclerosed.

Its histological characters are:

(1) Total substitution of the normal elements of the spleen with huge epithelial cells, irregularly rounded or polyhedral, nucleated, and confined among the normal trabeculae of the hyperplastic splenic network.
(2) Interstitial hemorrhages.
(3) Complete disappearance of the Malpighian corpuscles.
(4) Partial disappearance of the blood vessels.

Contributed by Robert Prichard, MD
Bowman Gray School of Medicine
Winston-Salem, NC