

Littoral-cell angioma*Angioma de células litorâneas*Vitorino Modesto Santos ¹

To Editor

Recently, I read the illustrative article by Du J et al. about littoral cell angioma (LCA) of the spleen, in which the authors highlighted the exceeding rarity of this usually benign tumour, its origin in the red-pulp of sinuses, the expression of histiocytic and endothelial antigen, and the challenging diagnosis.¹ Three male patients were related with splenomegaly, and mean age was 53.7 ± 11.1 years; the main manifestations were abdominal pain and distention, and the treatment of choice was total splenectomy.¹ The weight of the spleens ranged between 350 and 420 g, and the cut surface appeared greyish-red. Imaging studies involved ultrasound scanning, magnetic resonance (MRI), and computed tomography, and diagnostic criteria based on histopathology and immunohistochemical classical features, including splenic littoral cells positive for CD31, CD68, FVIII and lysozyme, and negative for CD34 staining.¹ Worthy of note was the characterization of three cases of this very rare condition in the same hospital, fact that might give origin to concerns about possible under

diagnosis or underreporting elsewhere. Notwithstanding, I strongly believe that previous case study can have enhanced the suspicion index.

Due to its rarity, etiological factors and biological behaviour of LCA are not well understood.²⁻⁴ Therefore, additional comments might be useful in this setting, with base on case studies in which associations between LCA and other entities suggest the possibility of etiopathological relationships.²⁻⁴ Cordersmeyer *et al.* reported a 43-year-old man with LCA, and previous diagnosis of lung sarcoidosis; the patient had abdominal pain and splenomegaly, and was successfully treated by total splenectomy.² These authors hypothesized the role of TNF- α released by macrophages in the pathogenesis of LCA, based on its effect in mediating inflammation and cellular immune response, in addition to promoting tumour development and angiogenesis, as well as migration and differentiation of endothelial cells.² Johansson *et al.* described an LCA in a 20-year-old woman with abdominal pain, splenomegaly, and pyrexia, and diagnosis of

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Crohn's disease. The clinical suspicion was based on MRI abdominal images indicative of LCA. The patient was submitted to a laparoscopic splenectomy, but evolved presenting with episodes of pyrexia, which were not associated with manifestations of active Crohn's disease.³ The authors also commented the possibility of TNF- α playing a role in the pathogenesis of LCA.³ Furtado *et al.* for the first time described LAC and sea-blue histiocytosis (SBH), in a 46-year-old woman with abdominal pain and splenomegaly. She became asymptomatic after splenectomy by laparotomy; histopathology and immunohistochemistry studies characterized the uncommon entities.⁴ SBH is a very infrequent benign condition with unclear pathogenesis, characterized by splenomegaly and low platelets, in addition to cells presenting with cytoplasmic accumulation of lipid granules, which appear with a typical sea-blue discoloration by Giemsa staining; hyperesplenism may occur too.⁴ The main disease was probably LCA, and SBH would be a secondary phenomenon to cell turnover.⁴

Current data about etiopathological factors of these rare conditions are scarce, and

did not support the hypothesis of eventual shared mechanisms of pathogenesis; notwithstanding, more good case studies could help remedy this situation.⁵

References

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