Histopathological Atlas of Vascular Liver Disorders
Atlas Histopatológico de Distúrbios Vasculares do Fígado

Fátima Carneiro[1], Christine Sempoux[2]

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Palavras-chave: Doenças do Fígado; Doenças Vasculares; Hepatopatia Veno-Oclusiva; Síndrome de Budd-Chiari.

Vascular liver disorders (VLD) are less frequent than other liver diseases and include a variety of clinico-pathological entities commonly classified according to the affected vascular compartment.[1] The liver's dual blood supply and the interconnection between all vascular structures within the liver lead to some morphological overlap between the entities. Patients are often young and carry a risk of decreasing life expectancy if not correctly managed. The diagnosis is made clinically, and a biopsy is not always indicated or necessary.[2]

On the histopathological side, it is important to be aware of the range of variations in the structure of the normal portal tracts (Fig. 1) and the existence of dyads, as well as to remember that the lesions can be subtle, patchy, and found also outside of the VLD setting in other chronic liver diseases. In this context, even more than in other medical liver diseases, correlation with clinical data and imaging is essential.

When the hepatic artery is concerned, ischaemic hepatitis with centrilobular necrosis can develop in case of systemic hypotension or shock, whereas ischaemic cholangiopathy will be the consequence of hepatic artery thrombosis or obstruction because bile ducts depend exclusively on arterial supply.[1]

Sinusoidal dilatation with congestion is a common pattern of different VLD, observed in sinusoidal obstruction syndrome (SOS) and venous outflow obstruction of various aetiologies, mainly Budd-Chiari syndrome (BCS) and the so-called "cardiac liver". SOS, formerly known as veino-occlusive disease, results from an initial toxic damage of the sinusoidal endothelial cells in different clinical conditions. It is characterized primarily by endothelial cell detachment, subendothelial extravasation of red blood cells, sloughing towards central zones and subsequent occlusion of central veins (Fig. 2). It can evolve towards

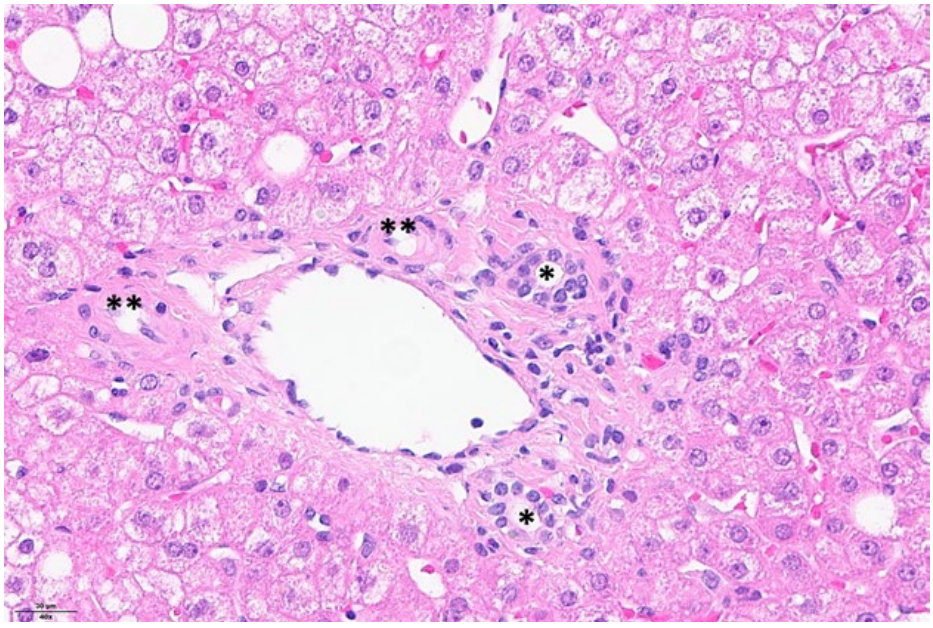

Figure 1: **Normal portal tract** - A normal portal tract contains one portal venule which is the largest structure, located centrally and surrounded by collagen, together with one or two bile ducts (*) with their corresponding arterial branch (**). They have the same calibre. (H&E stain).
H&E: haematoxylin-eosin.

[1]Ipatimup/i3S, Faculdade de Medicina da Universidade do Porto & Unidade Local de Saúde São João, Porto, Portugal.
[2]Institute of Pathology, Lausanne University Hospital and University of Lausanne, Lausanne, Switzerland.

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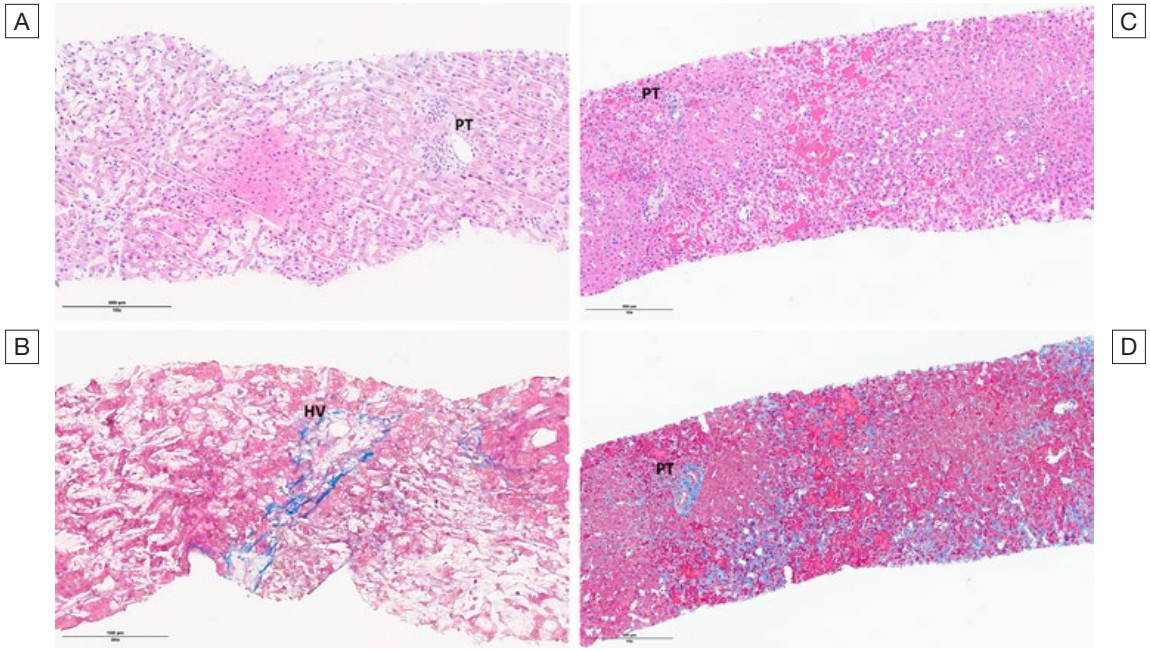

Figure 2: **Sinusoidal obstruction syndrome** - [**A-B**] Initial lesions with perivenular sinusoidal dilatation and congestion (A-H&E stain), sloughing towards central zones and partial occlusion of the central hepatic vein (B-MT stain). [**C-D**] Evolving lesions with sinusoidal congestion, atrophy of the liver cell plates (C-H&E stain) and sinusoidal fibrosis, all in the perivenular area (D-MT stain).
H&E: haematoxylin-eosin; HV: hepatic vein; MT: Masson's trichrome; PT portal tract.

fibrosis.[3] By contrast, peliosis that has various causes shows randomly distributed cystic dilated spaces filled with red blood cells, which results from the rupture of sinusoidal walls. BCS is clinically associated with different prothrombotic conditions and characterized by obstruction of the large hepatic veins, diagnosed by imaging procedures. Biopsies are performed in case of uncertain diagnosis and will show severe perivenular sinusoidal congestion with or without thrombi, progressing to fibrosis and stenosis, hence the development of new thromboses (Fig. 3).[4] Constrictive pericarditis, chronic right-sided heart failure or Fontan palliation are responsible for chronic passive congestion within the liver, starting in the perivenular area and

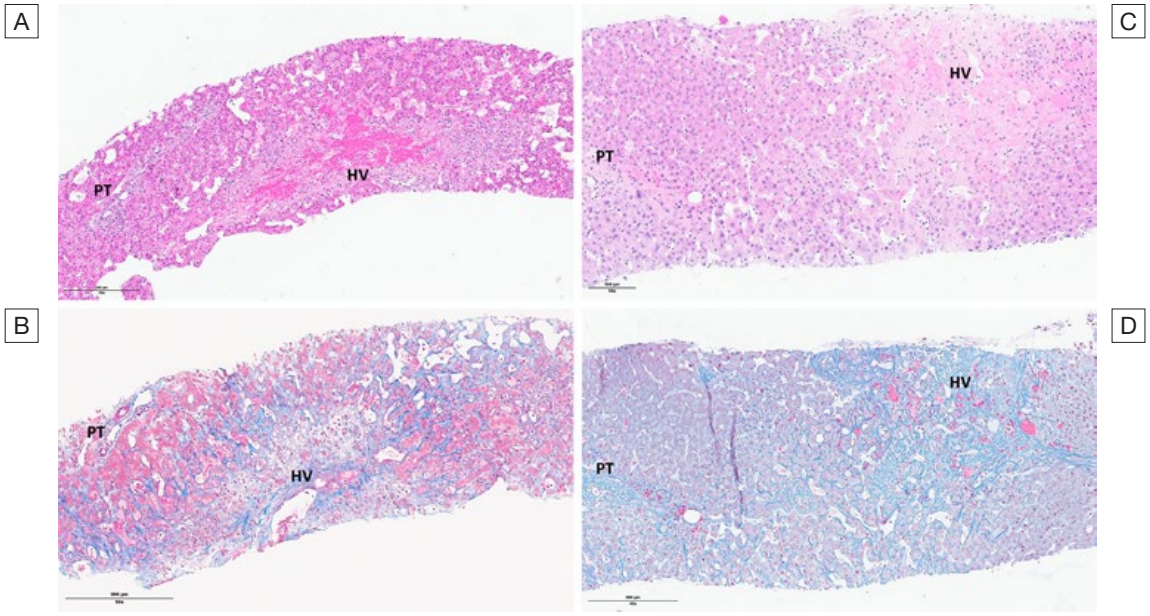

Figure 3: **Budd-Chiari syndrome** - [**A-B**] Initial lesions with perivenular sinusoidal dilatation and organized thrombus in the central hepatic vein (A-H&E stain), better seen on the Masson's trichrome stain that also underlines the atrophic hepatocytes plates (B-MT stain). [**C-D**] Evolving lesions with complete obliteration of the central hepatic vein (C-H&E stain) and fibrosis extending towards the portal tracts (D-MT stain).
H&E: haematoxylin-eosin; HV: hepatic vein; MT: Masson's trichrome; PT portal tract.

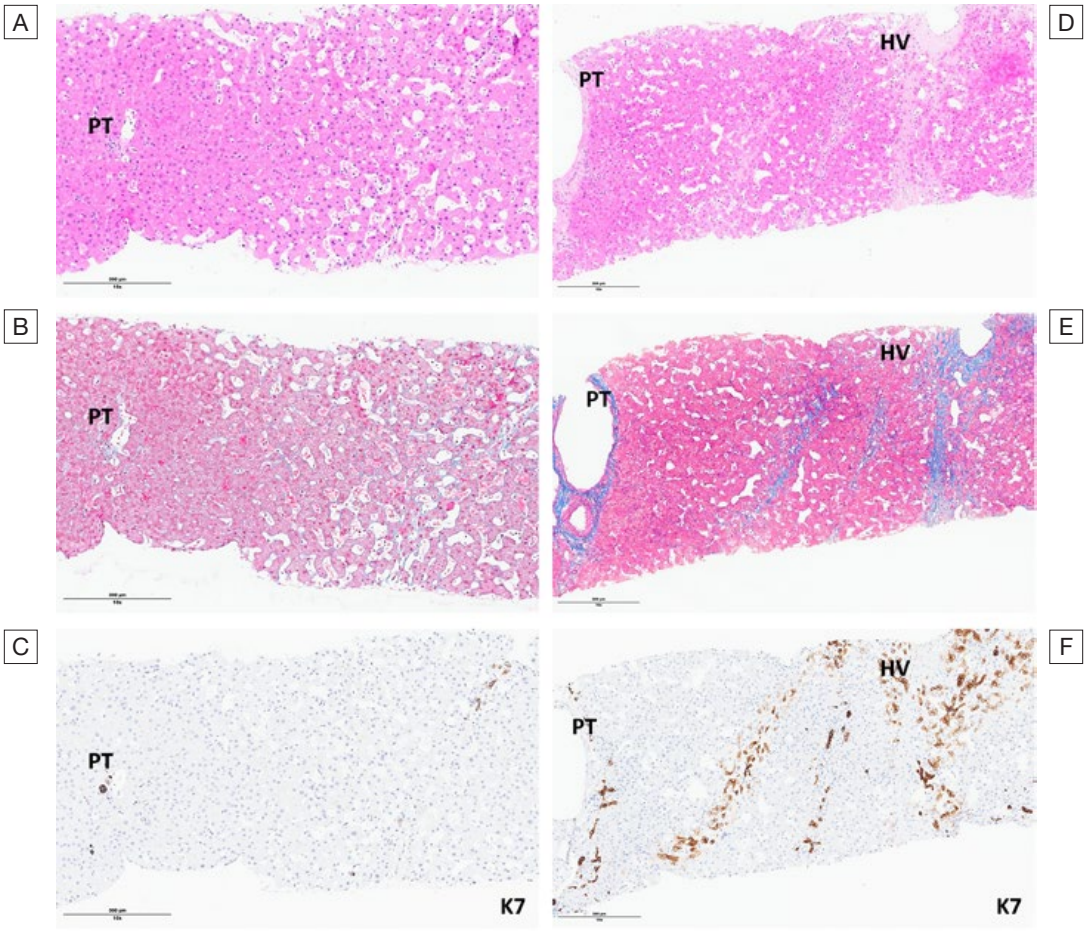

Figure 4: **Congestive hepatopathy** - [**A-B-C**] Initial lesions with perivenular sinusoidal dilatation and congestion (A-H&E stain) without fibrosis (B-MT stain) and almost no keratin 7 (K7) expression in centrilobular hepatocytes (C-K7 immunohistochemistry). [**D-E-F**] Evolving lesions with sinusoidal dilatation (D-H&E stain), fibrosis (E-MT stain) and atrophy of the liver cell plates in the perivenular area underlined by the expression of keratin 7 by hypoxic hepatocytes (F- K7 immunohistochemistry).
H&E: haematoxylin-eosin; HV: hepatic vein; MT: Masson's trichrome; PT portal tract.

being progressively accompanied by a linear sinusoidal fibrosis then by the development of fibrous septa radiating out of the central zone with reversed lobulation (Fig. 4). A congestive hepatic fibrosis score has been developed and validated in these clinical situations to assess the stage.[5]

In case of portal vein thrombosis and congenital portosystemic shunts, the modifications are predominating in the portal tracts. The calibre of the portal venules is reduced, and arterial hyperplasia of the arterial branches develop through a compensatory mechanism (Fig. 5).[6] These portal

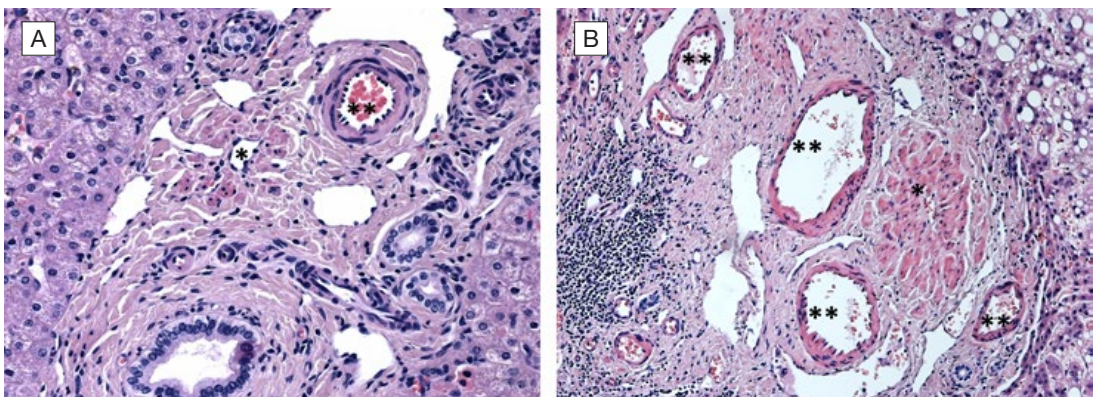

Figure 5: **Congenital portosystemic shunt** - [**A-B**] Two examples of portal tracts with portal venule stenosis and muscularized wall (*) associated with hyperplasia of arterial branches (**). Dilated irregular lymphatic channels are also observed (H&E stain).
H&E: haematoxylin-eosin.

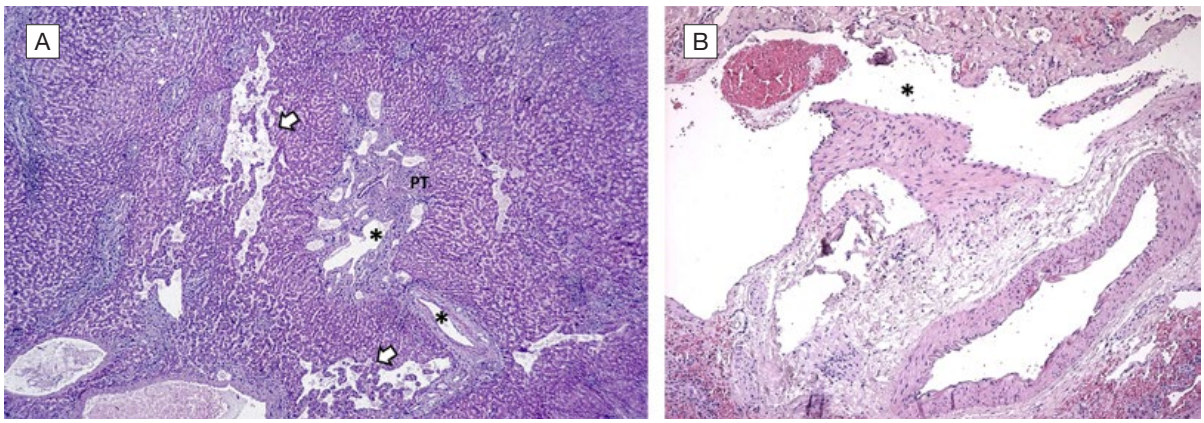

Figure 6: **Hereditary haemorrhagic telangiectasia** - Arteriovenous shunts (*) are seen in portal tracts [**A and B**] together with lobular telangiectasias (A, arrows) (H&E stain).
H&E: haematoxylin-eosin.

abnormalities are associated or not with sinusoidal congestion. In hereditary haemorrhagic telangiectasia, biopsy should be avoided because of the risk of bleeding. In surgical specimens, portal arterio-venous shunts, telangiectasia and sinusoidal congestion are seen (Fig. 6). In addition, patients are at risk of developing parenchymal perfusion disorder, with numerous regenerative nodules, or ischemic cholangiopathies with biliary necrosis requiring liver transplantation.[7]

The entity known under both the names non-cirrhotic portal fibrosis and porto-sinusoidal vascular disorder is currently under the process of a worldwide multisocietal initiative to address existing controversies and to develop an international consensus on definition and diagnostic criteria. It is agreed that the diagnosis requires a liver biopsy to exclude cirrhosis.[8,9] At the microscope, the major histopathological features are mainly the presence of portal venules stenosis or of nodular regenerative hyperplasia (Fig. 7).[10,11] When these subtle histological lesions are found in a liver biopsy in the absence of portal hypertension the clinical management is still a matter of debate.

Finally, in case of a decrease in portal venous flow, a compensatory arterial inflow develops, leading to chronic perfusion abnormalities. This results in secondary morphological

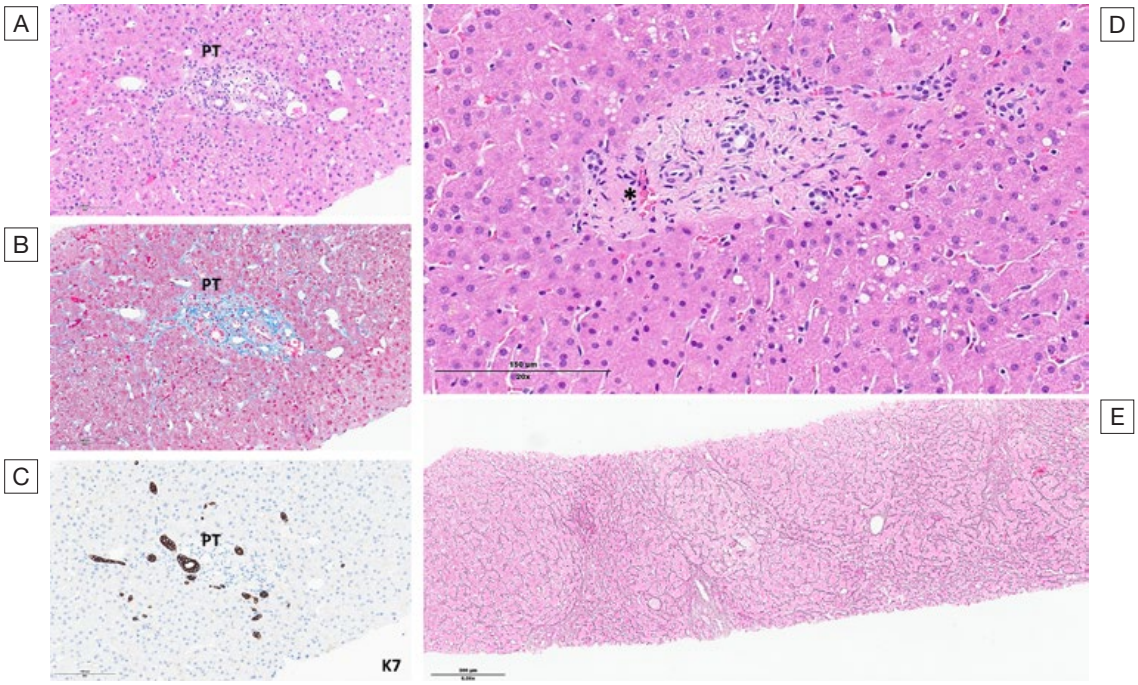

Figure 7: **Porto-sinusoidal vascular disorder** - [**A-B-C**] The same portal tract with complete portal venule stenosis (A-H&E stain; B-MT stain: C-K7 immunohistochemistry). [**D**] Incomplete portal venule stenosis (*) (H&E stain). [**E**] Nodular regenerative hyperplasia (Reticulin stain).
H&E: haematoxylin-eosin; MT: Masson's trichrome; PT portal tract.

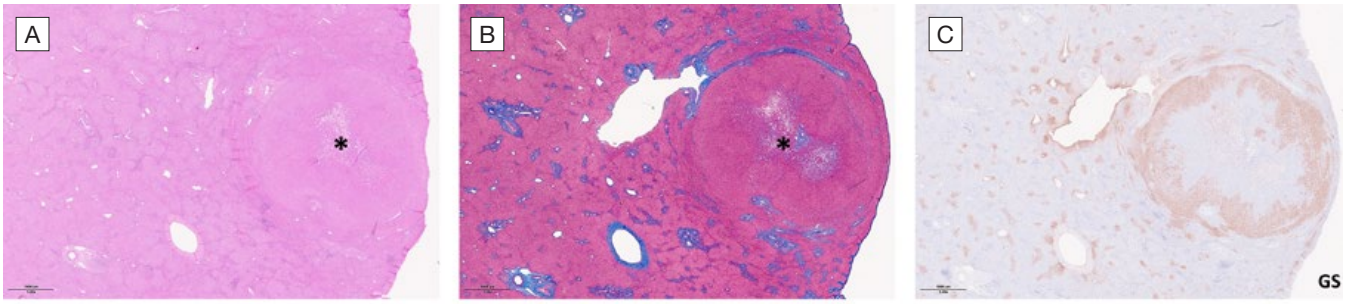

Figure 8: **FNH-like nodule in VLD** - **[A-B-C]** Focal nodular hyperplasia-like nodule with central scar (*), sharp delineation and map-like pattern of Glutamine synthetase (GS) staining by immunohistochemistry (A-H&E stain; B-MT stain; C-GS immunohistochemistry).
H&E: haematoxylin-eosin; MT: Masson's trichrome; PT portal tract.

alterations including the possible development of nodules that are mostly reactive and show FNH-like features (Fig. 8).[12] Targeted biopsies can be performed to exclude hepatocellular carcinoma also reported in some entities.[1]

Contributorship Statement

FC and CS - Study design and manuscript writing, image collection, critical revision and final approval.
All authors approved the final version to be published.

Declaração de Contribuição

FC e CS - Desenho e escrita do artigo, colheita de imagens, revisão crítica e aprovação final.
Todos os autores aprovaram a versão final a ser publicada.

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Corresponding Author / Autor Correspondente

Christine Sempoux - Christine.Sempoux@chuv.ch
Institute of Pathology, Lausanne University Hospital and University of Lausanne, Lausanne, Switzerland
Rue du Bugnon 25. CH-1011 Lausanne, Vaud, Suisse

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