Paris Portal Vein thrombosis meeting

VALDIG
VASCULAR LIVER DISEASE GROUP

Porto-Sinusoidal Vascular Diseases & Portal Vein Thrombosis

2 separate entities ?

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Inserm

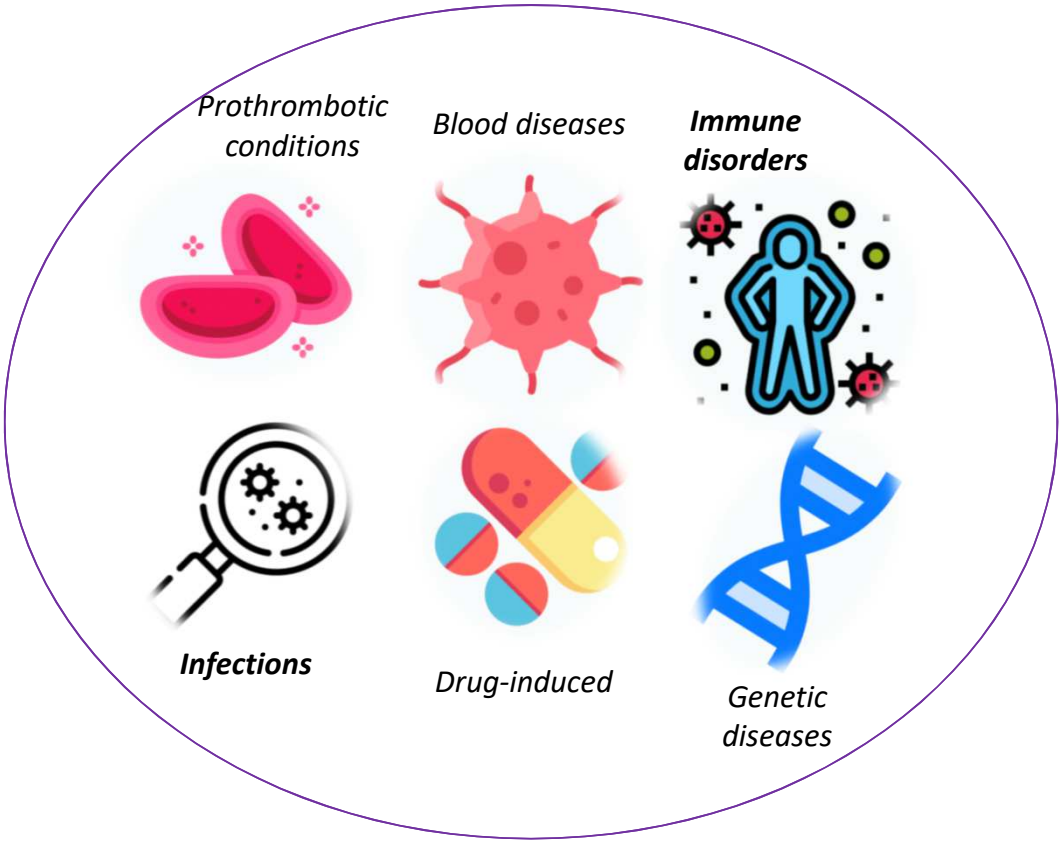
FHU MOSAIC

Université Paris Cité

PSVD & PVT: A single entity, BUT ...

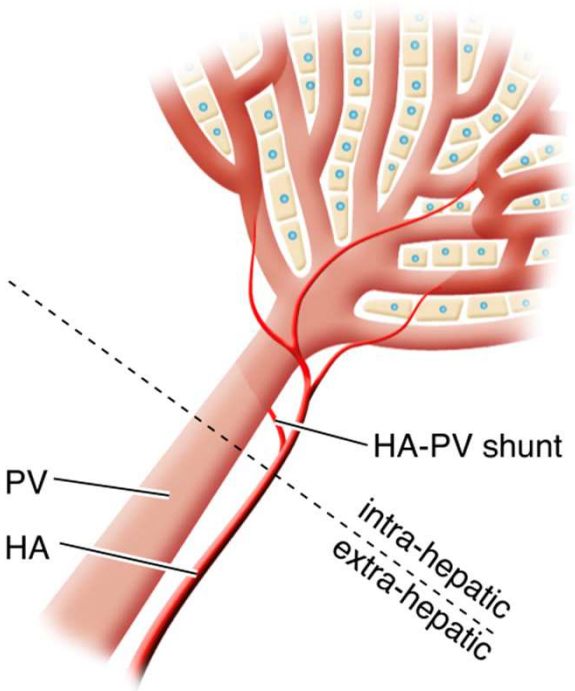
Share clinical conditions

BUT with ≠ frequency



Affect portal vascularisation

BUT @ ≠ level



From Vollmar B Physiol Rev 2009

PORTO-SINUSOIDAL VASCULAR DISEASES

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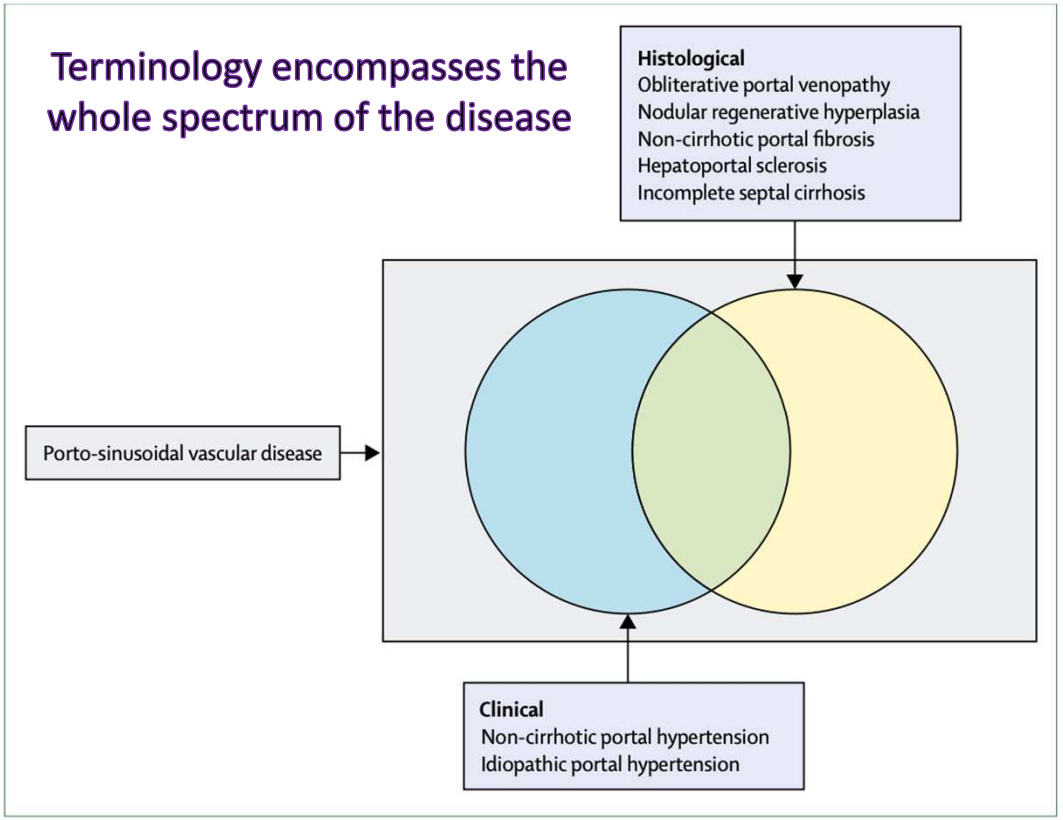
Porto-sinusoidal vascular disease: proposal and description of a novel entity

Multidisciplinary symposium → topics related to idiopathic non-cirrhotic portal hypertension

February 2017



De Gottardi A Lancet Gastroentrol Hepatol 2019

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Porto-sinusoidal vascular disease: proposal and description of a novel entity

Definition of PSVD

A multidisciplinary diagnosis

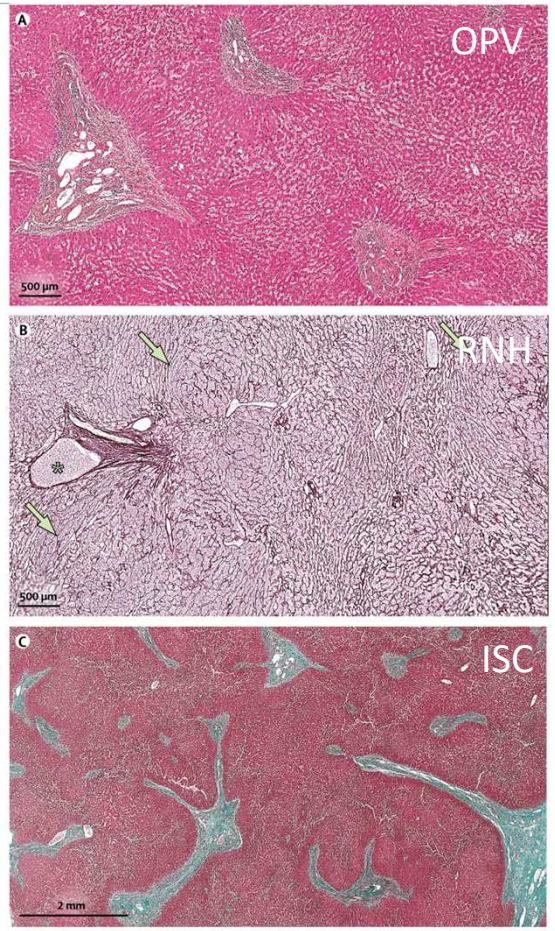
Liver biopsy ≥20 mm without cirrhosis	+	1 sign specific for portal hypertension or 1 histological lesion specific for PSVD

OR

Liver biopsy ≥20 mm without cirrhosis	+	1 sign not specific for portal hypertension and 1 histological lesion not specific for PSVD

Criteria in the definition of PSVD

	Signs of portal hypertension	Signs of portal hypertension
Specific	Gastric oesophagael, or ectopic varices Portal hypertensive bleeding Porto-systemic collaterals at imaging	Obliterative portal venopathy (thickening of vessel wall, occlusion of the lumen, and vanishing of portal veins) Nodular regenerative hyperplasmia Incomplete septal fibrosis or cirrhosis
Not specific	Ascites Platelet count <150 000 per µL Spleen size ≥13 cm in the largest axis	Portal tract abnormalities (multiplication, dilation of arteries, periportal vascular channels, and aberrant vessels) Architectural disturbance: irregular distribution of the portal tracts and central veins Non-zonal sinusoidal dilation Mild perisinusoidal fibrosis



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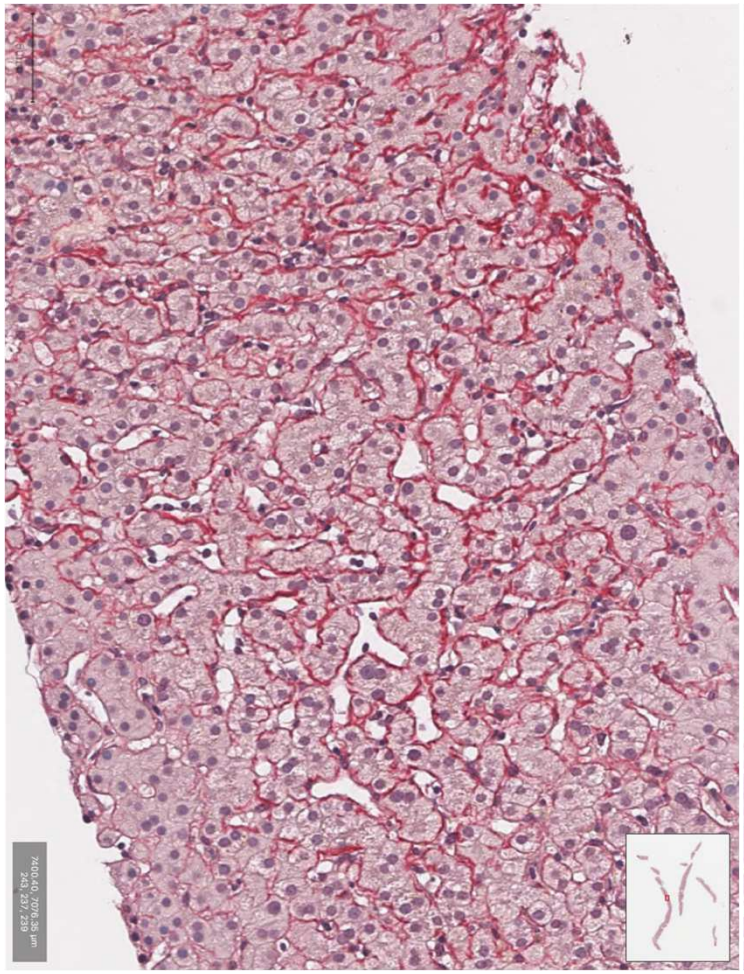
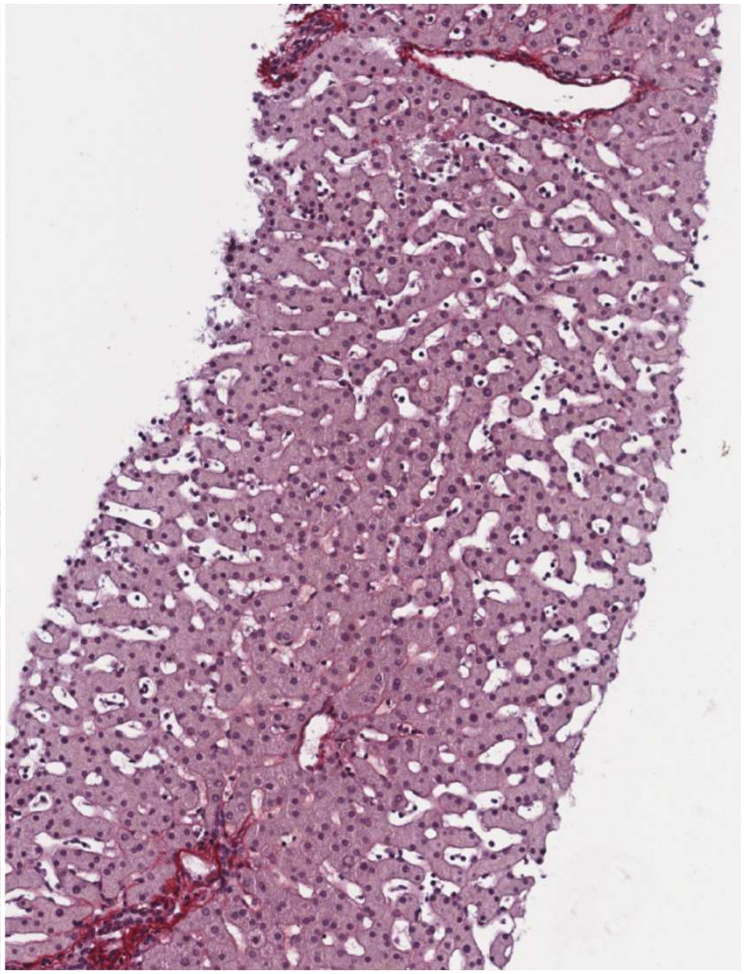
Not specific morphological lesions

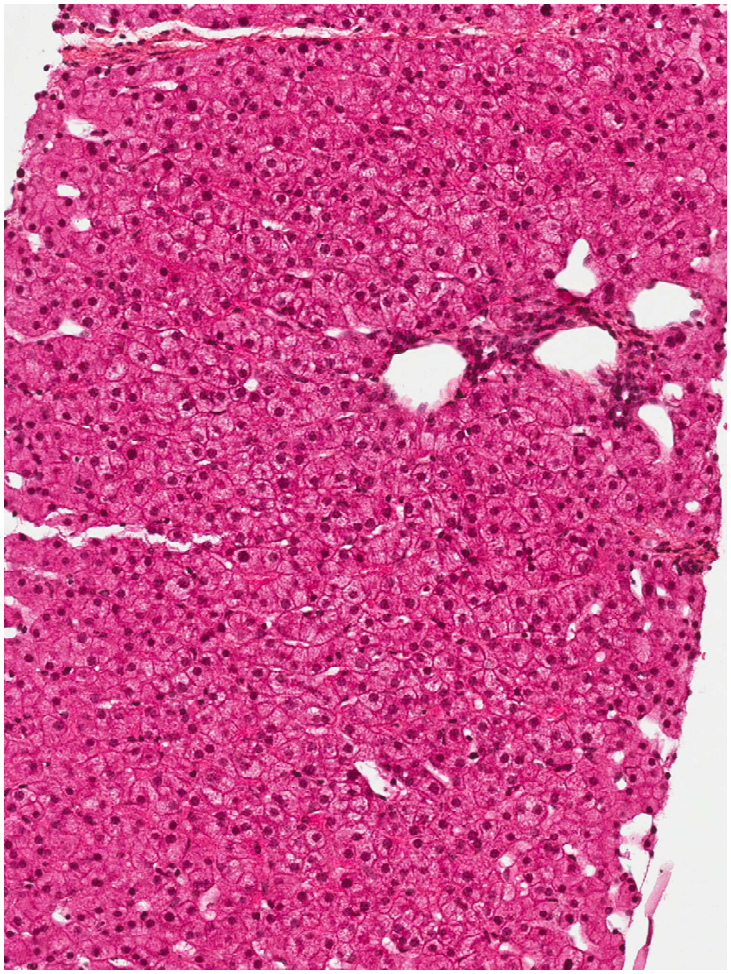
Sinusoidal fibrosis



Sinusoidal dilation

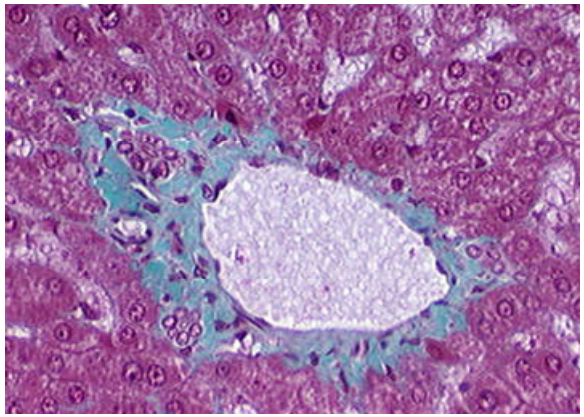


Aberrant vessels



Histology of portal vascular changes associated with idiopathic non-cirrhotic portal hypertension: nomenclature and definition

Maria Guido,[1] Venancio A F Alves,[2] Charles Balabaud,[3] Prithi S Bathal,[4] Paulette Bioulac-Sage,[5] Romano Colombari,[6] James M Crawford,[7] Amar P Dhillon,[8] Linda D Ferrell,[9] Ryan M Gill,[9] Prodromos Hytiroglou,[10] Yasuni Nakanuma,[11] Valerie Paradis,[12] Alberto Quaglia,[13] Pierre E Rautou,[14] Neil D Theise,[15] Swan Thung,[16] Wilson M S Tsui,[17] Christine Sempoux,[18] Dale Snover,[19] Dirk J van Leeuwen[20]
The International Liver Pathology Study Group



Portal vein present in > 70% of PT

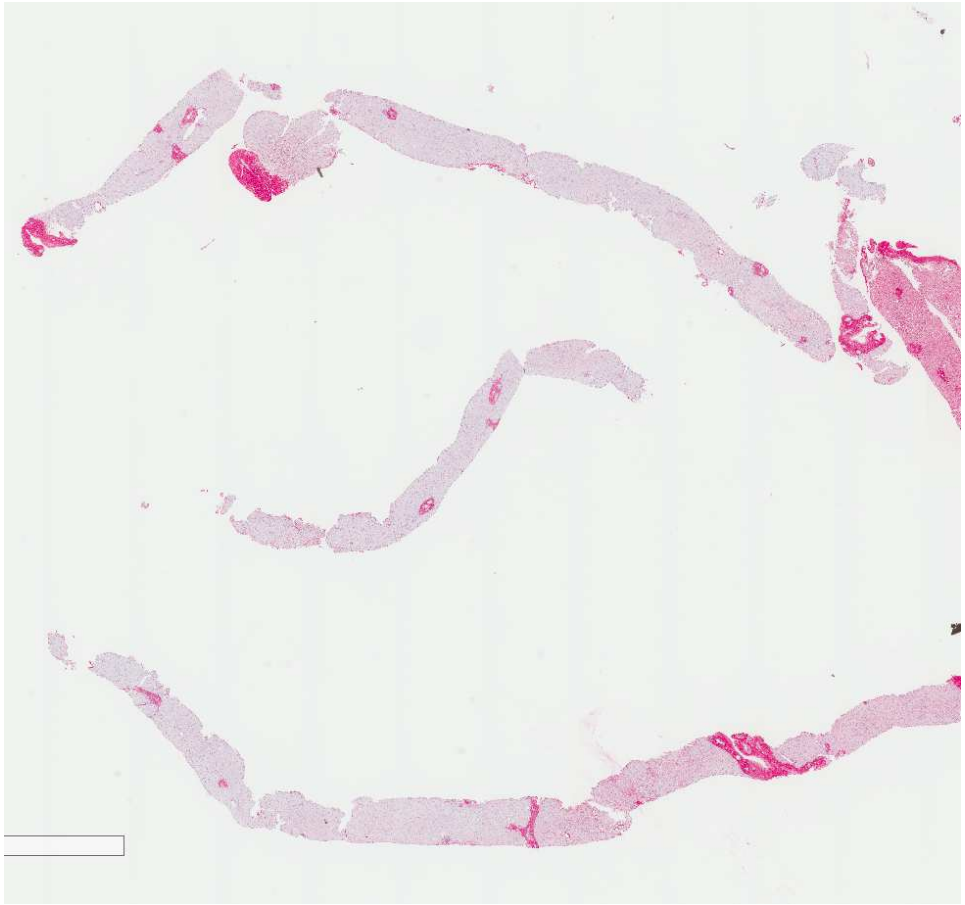
Table 1. Summary of proposed nomenclature and definition of portal/periportal idiopathic non-cirrhotic portal hypertension lesions

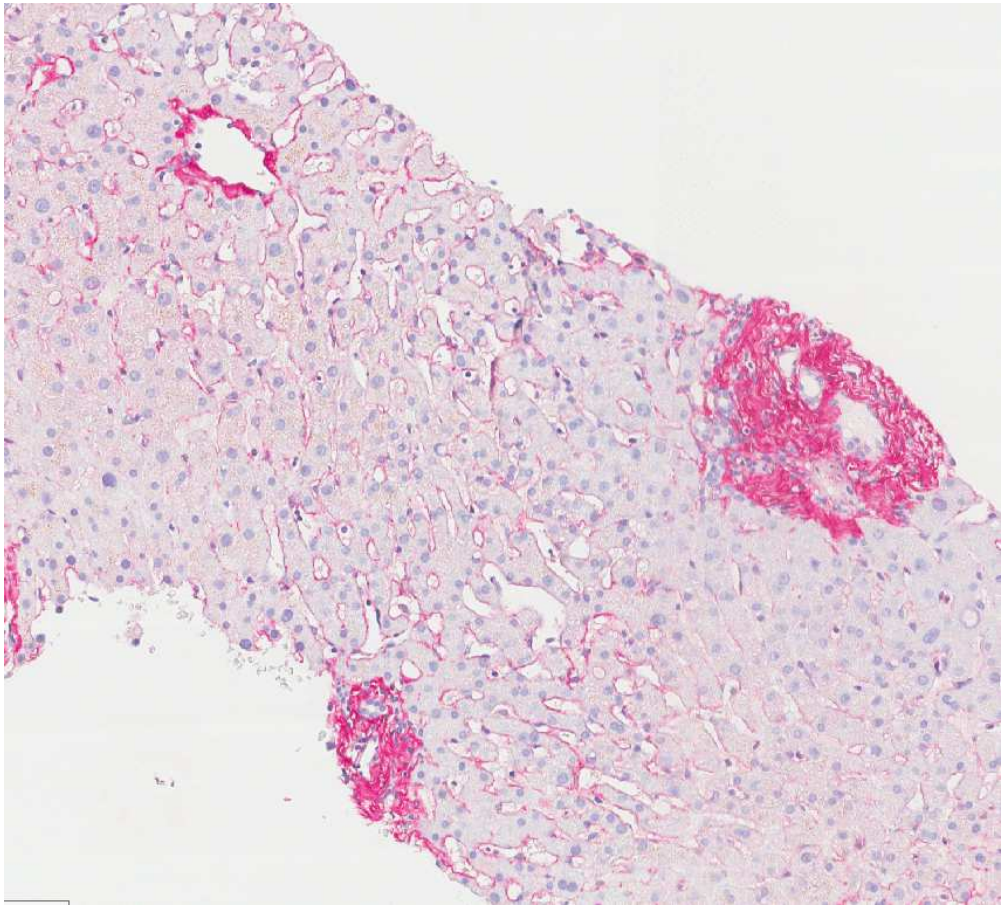
Proposed term	Definition	Terms to be discarded
Portal vein stenosis	Incomplete (Figure 1A) or complete (Figure 1B) obliteration of portal vein branches ± thickening of the wall	Phlebosclerosis; obliterative portal venopathy; portal vein obliteration; hepatoportal sclerosis
Herniated portal vein	A portal vein from the portal tract directly abutting periportal parenchyma (Figure 2A,B)	Aberrant vessels; hepatoportal sclerosis; shunt vessels
Hypervascularised portal tract	Multiple thin-walled vascular spaces in the portal tract (Figure 3A,B)	Fragmentation of portal vein radicles; portal tract angiomatosis; angiomatoid lesion; angiomatous transformation of the portal tract
Periportal abnormal vessels	Single or multiple thin-walled vascular spaces of different calibre outside, but in close contact with, the portal tract (Figure 4A,B)	Aberrant vessels; cavernous transformation; paraportal shunting vessels; megasinusoids

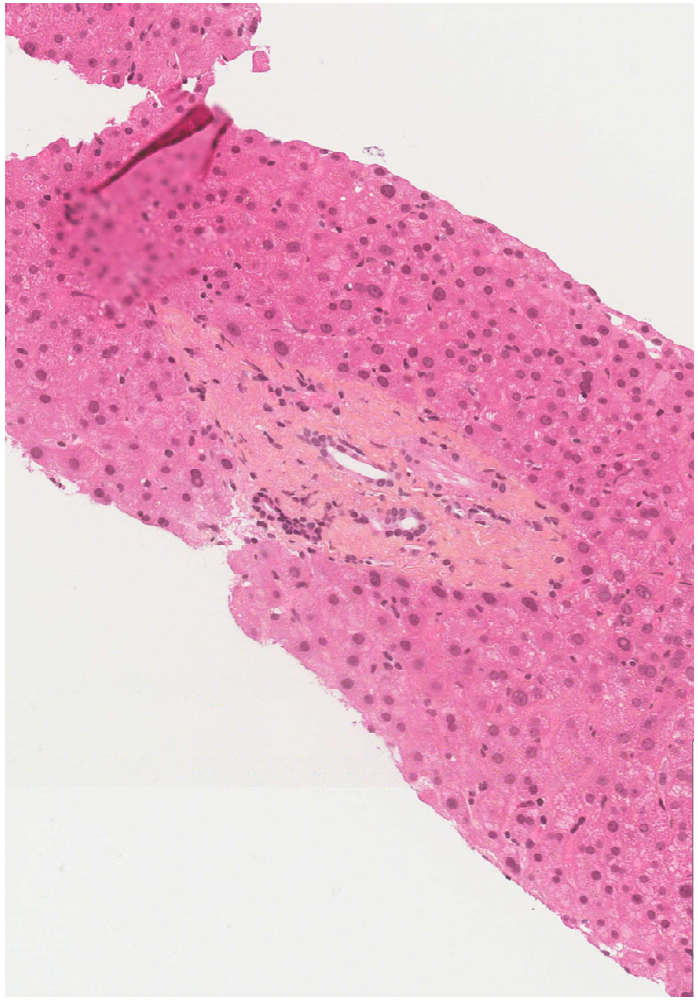
- Portal/ periportal lesions may be variably combined in a single biopsy
- Different lesions may coexist in a same portal/periportal area

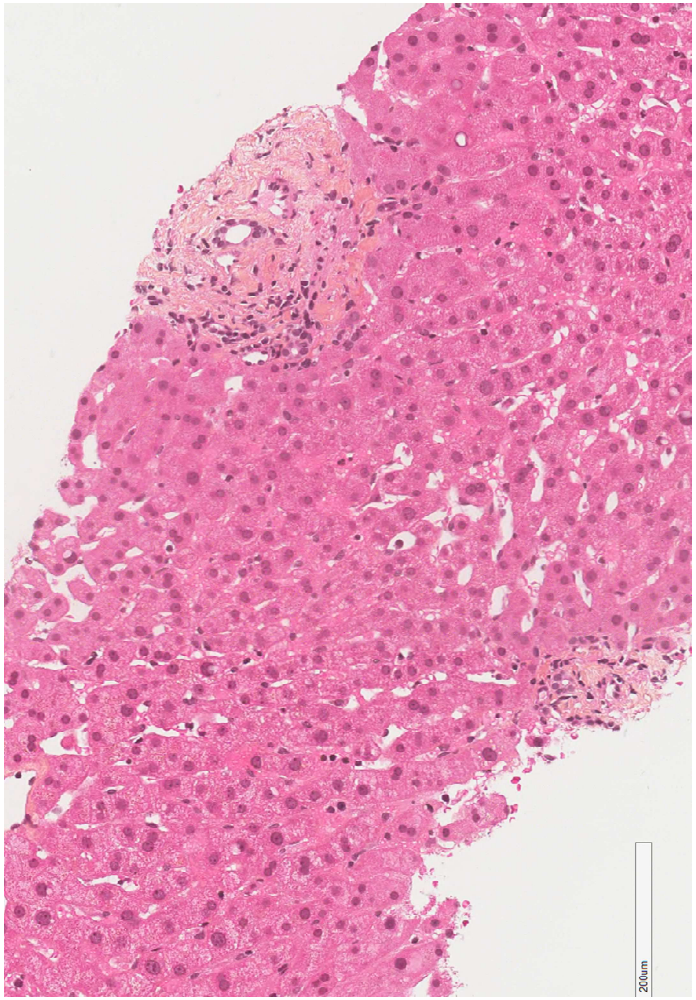
Histopathology 2019

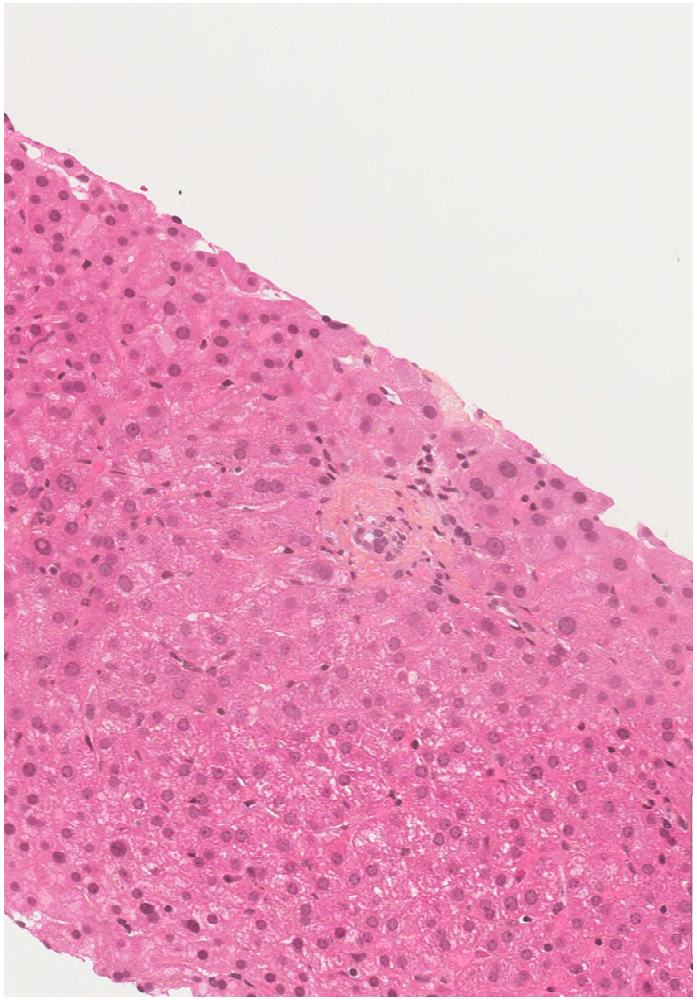
73-year old man, Oesophageal varices, dysmorphic liver, Elastometry 11 kPa, HVPG 17 mm Hg





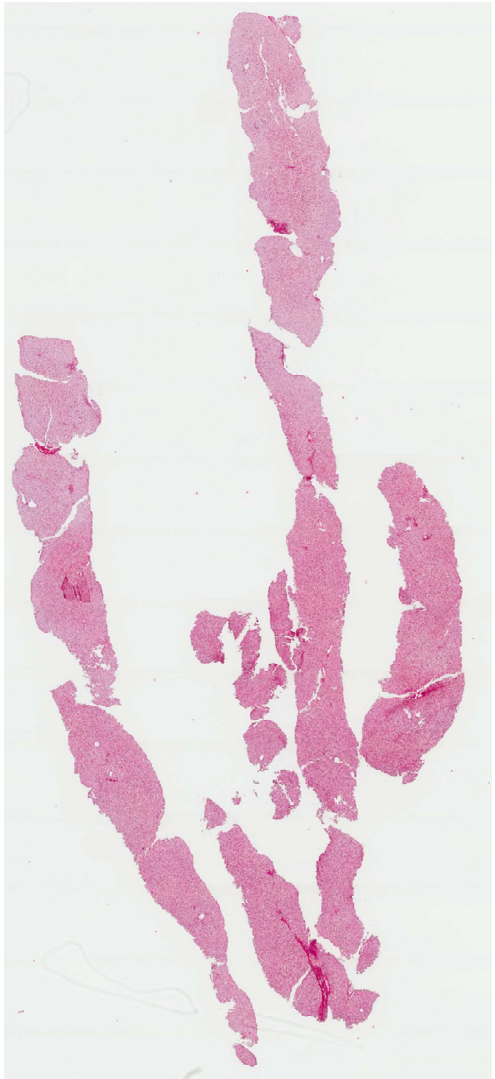


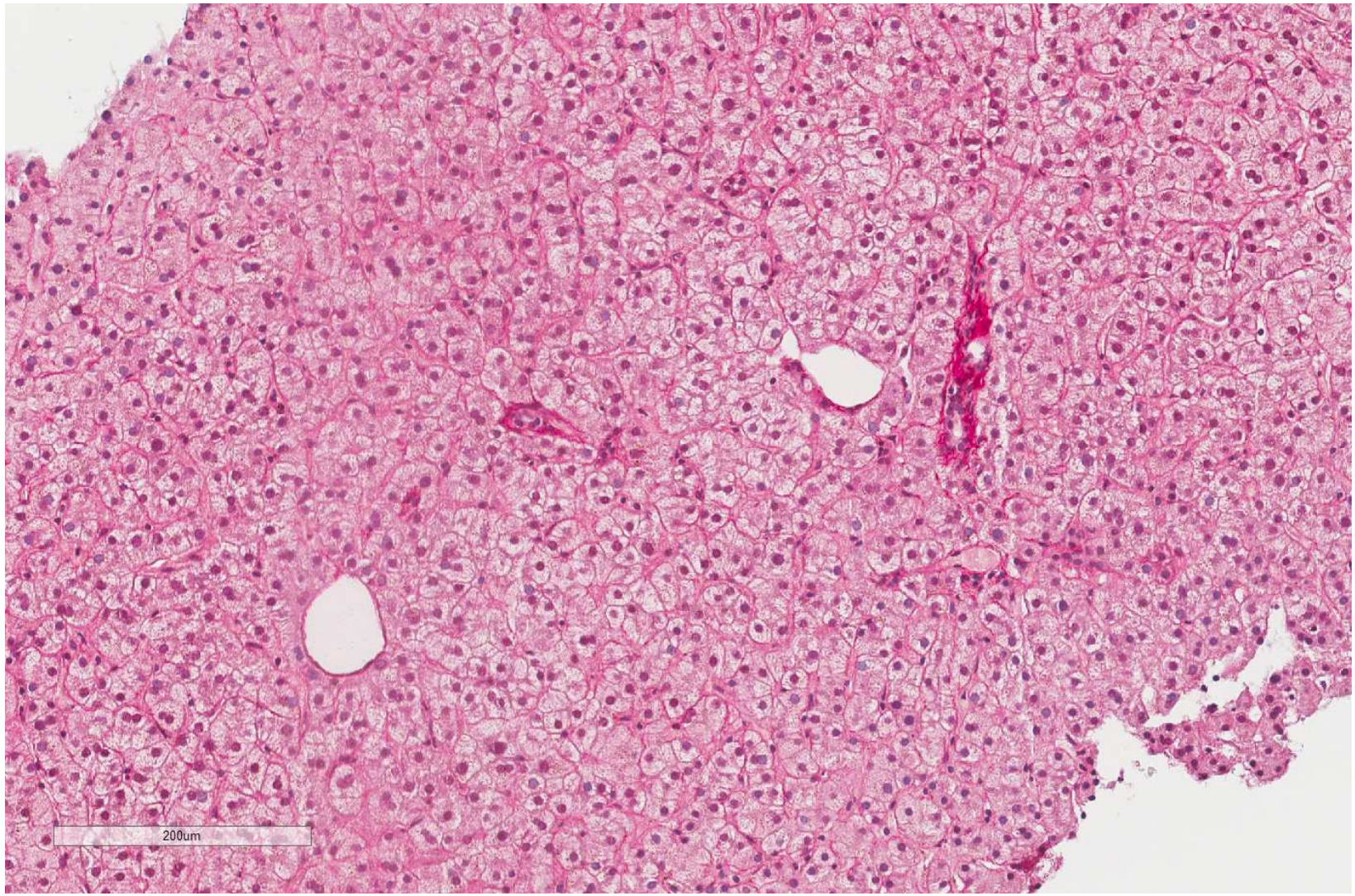


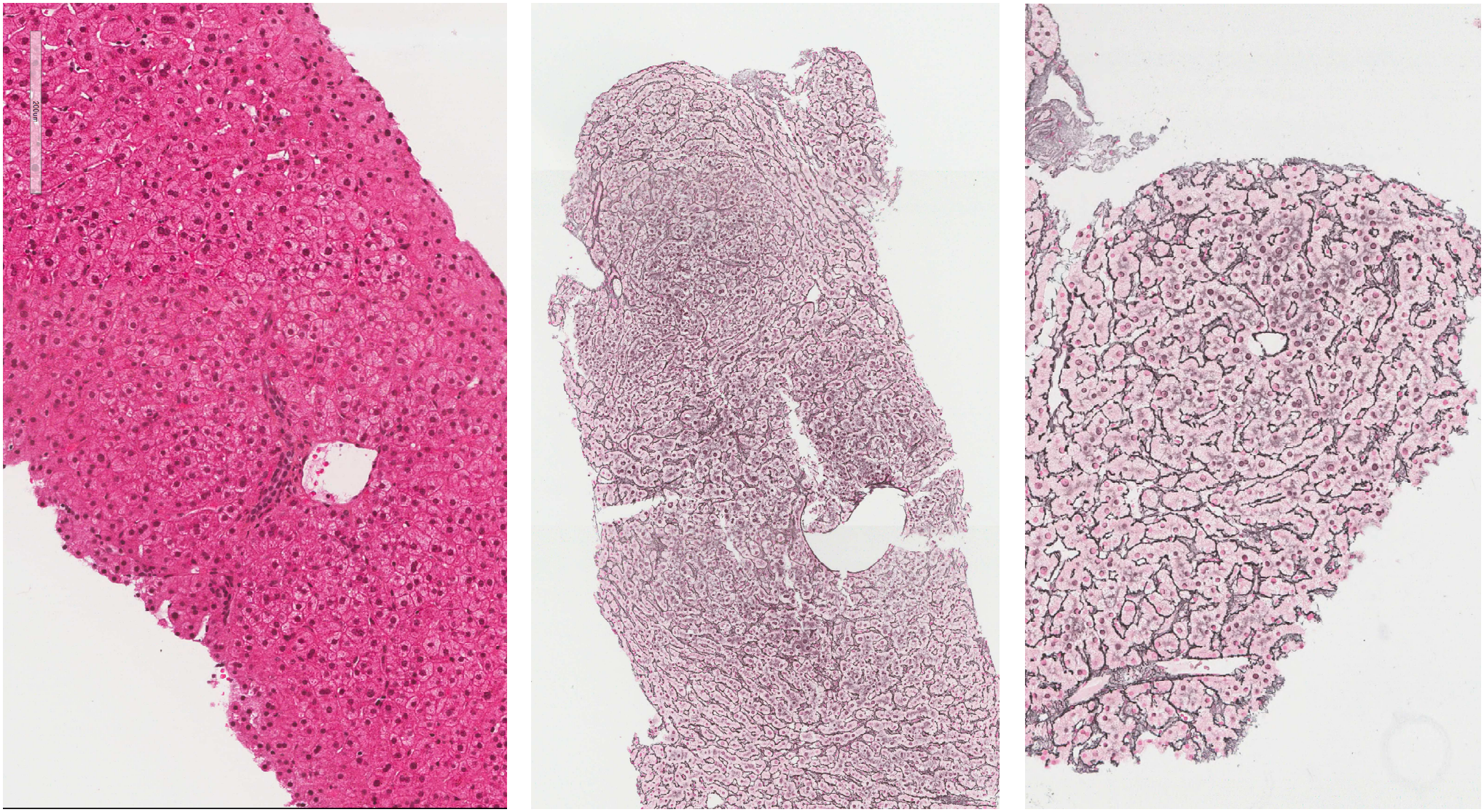


PSVD (Portal vein stenosis / Obliterative portal venopathy)

58-year old man, HIV +, dysmorphic liver, HVPG 10 mm Hg







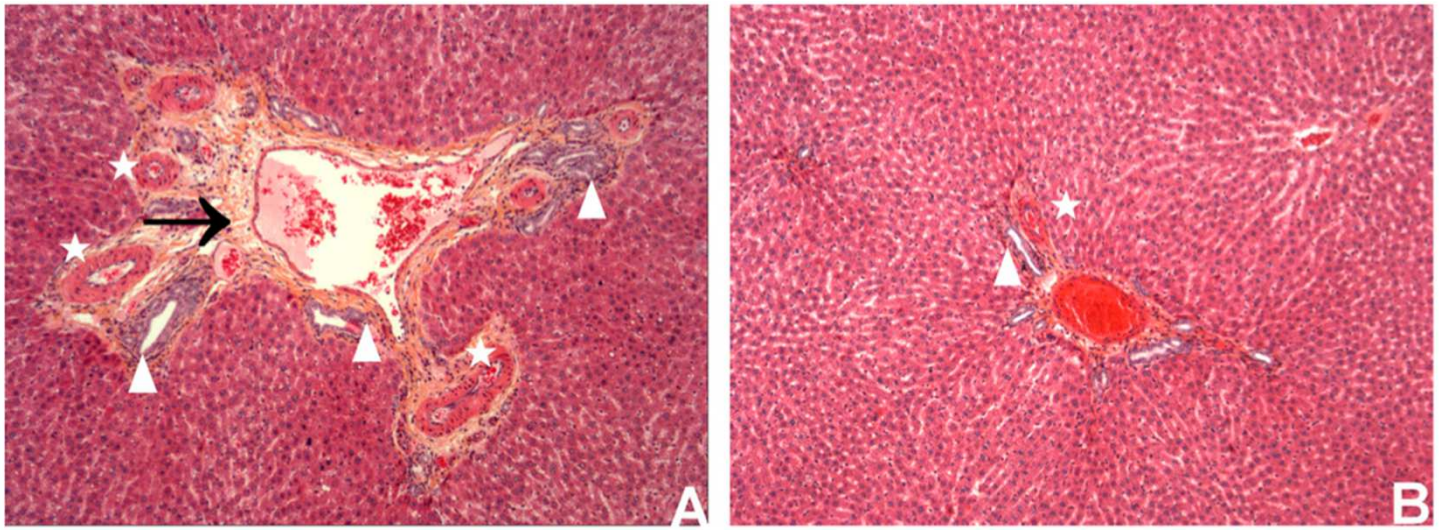
PSVD (Portal vein stenosis / Obliterative portal venopathy + RNH)

PORTAL VEIN THROMBOSIS

Hepatic Proliferation and Angiogenesis Markers Are Increased after Portal Deprivation in Rats: A Study of Molecular, Histological and Radiological Changes

Portocaval shunt & Portal vein ligation

Histological lesions @ 6 months: Periportal fibrosis, Ductular reaction & vascular multiplication



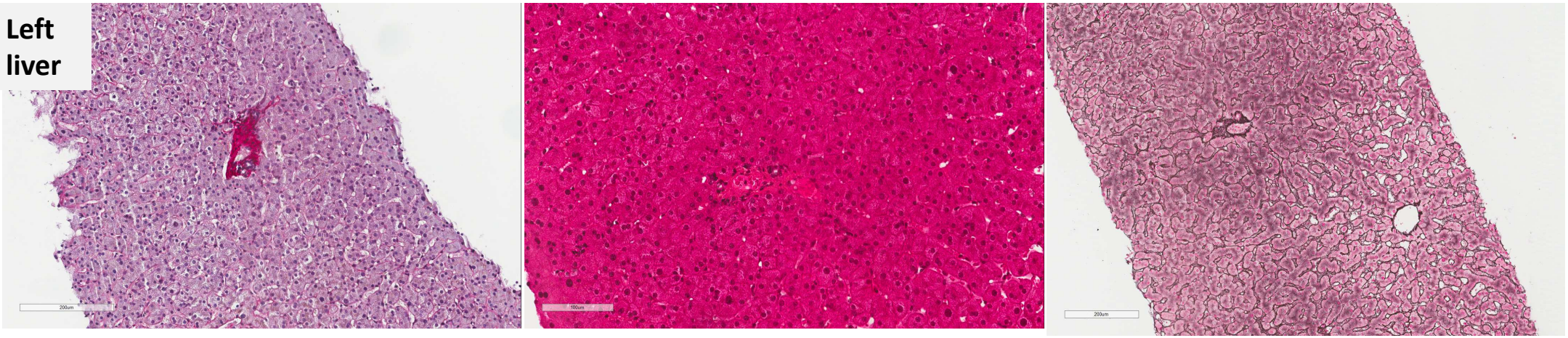
Guérin F Plos One 2015

44-year old woman, Protein S deficiency

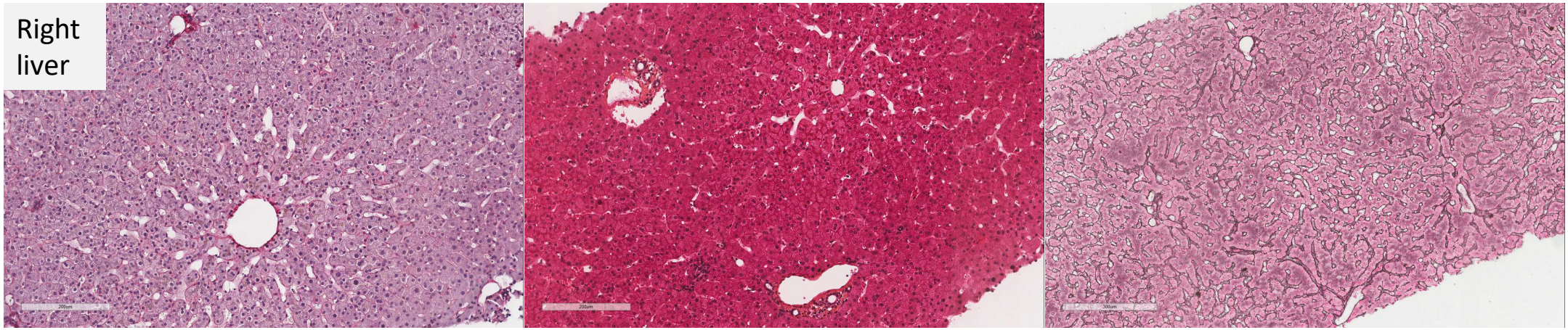
Left portal thrombosis (treated with anticoagulation)

Liver biopsy @ 5 y

Left liver

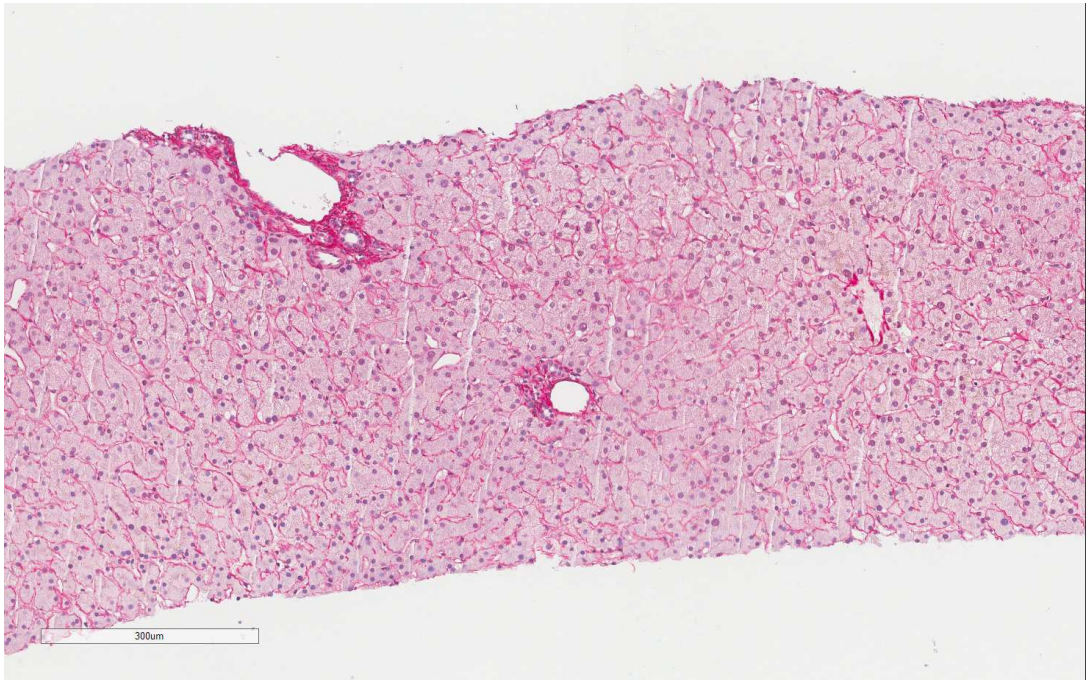


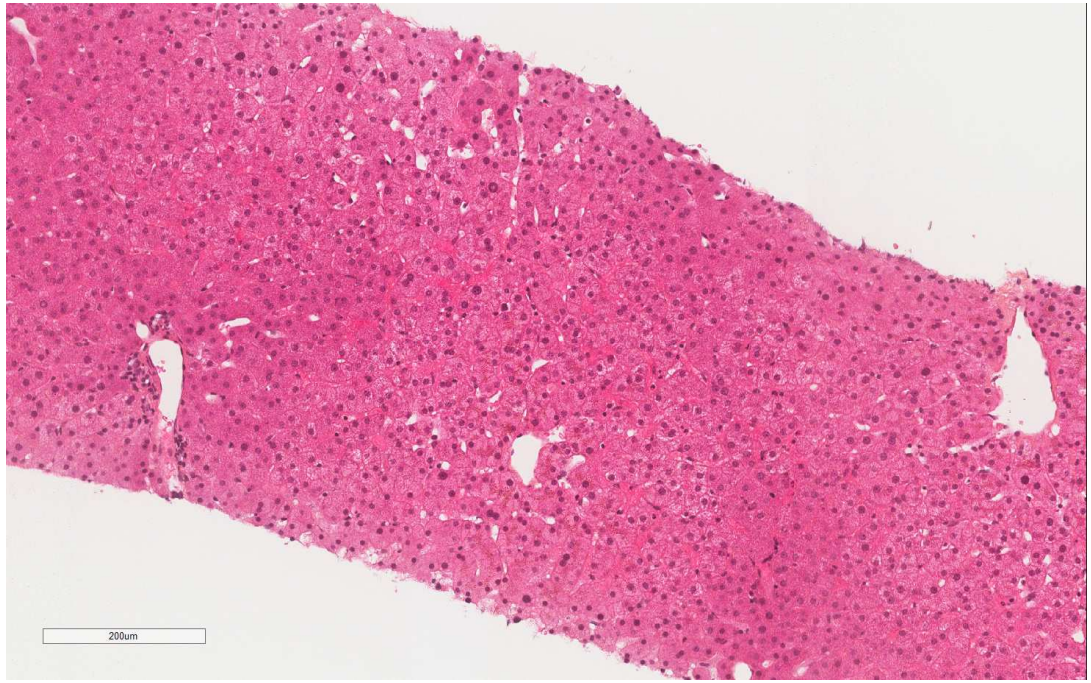
Right liver



46 year-old man

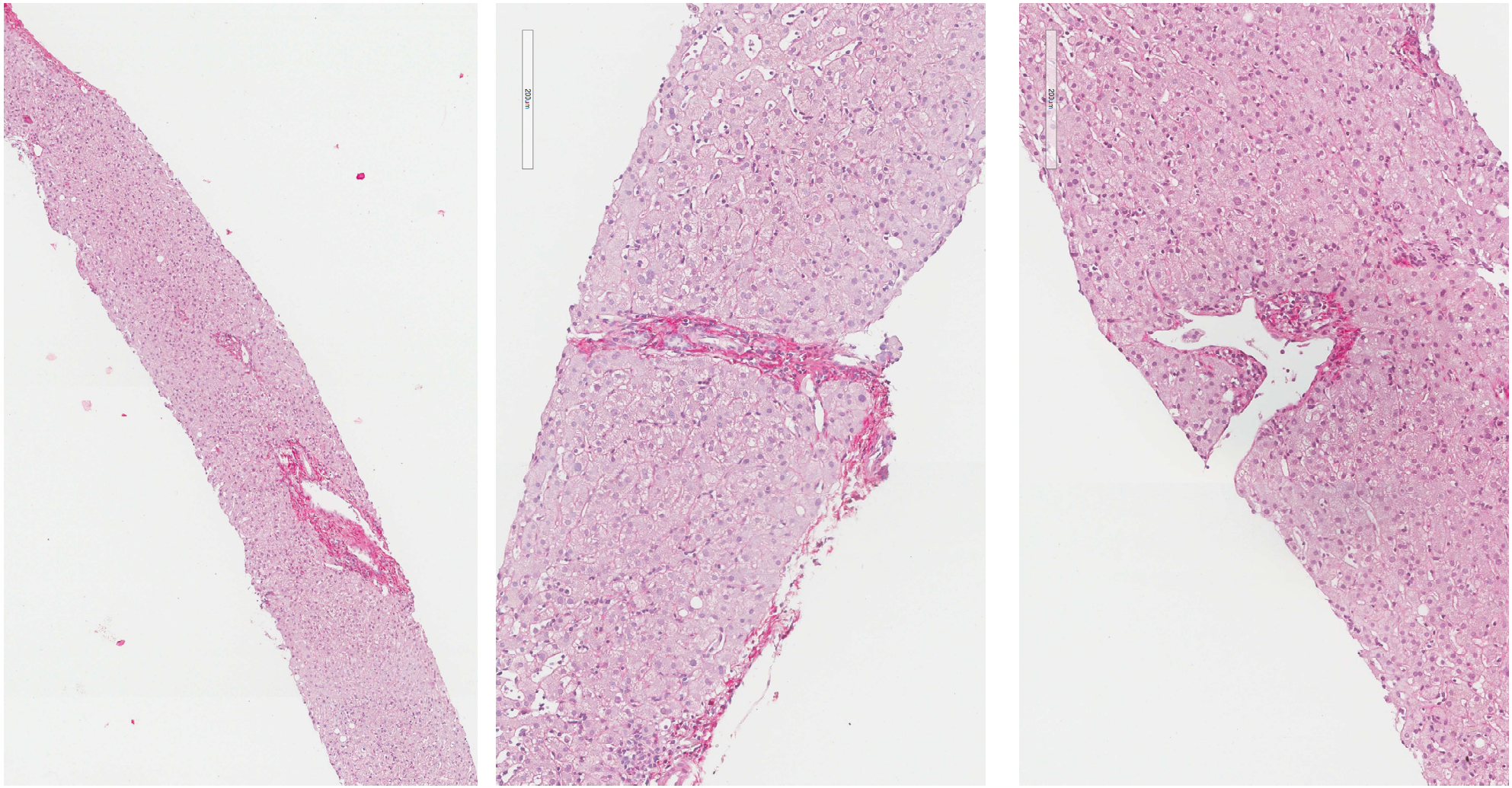
Portal Thrombosis (appendicitis), portal cavernoma

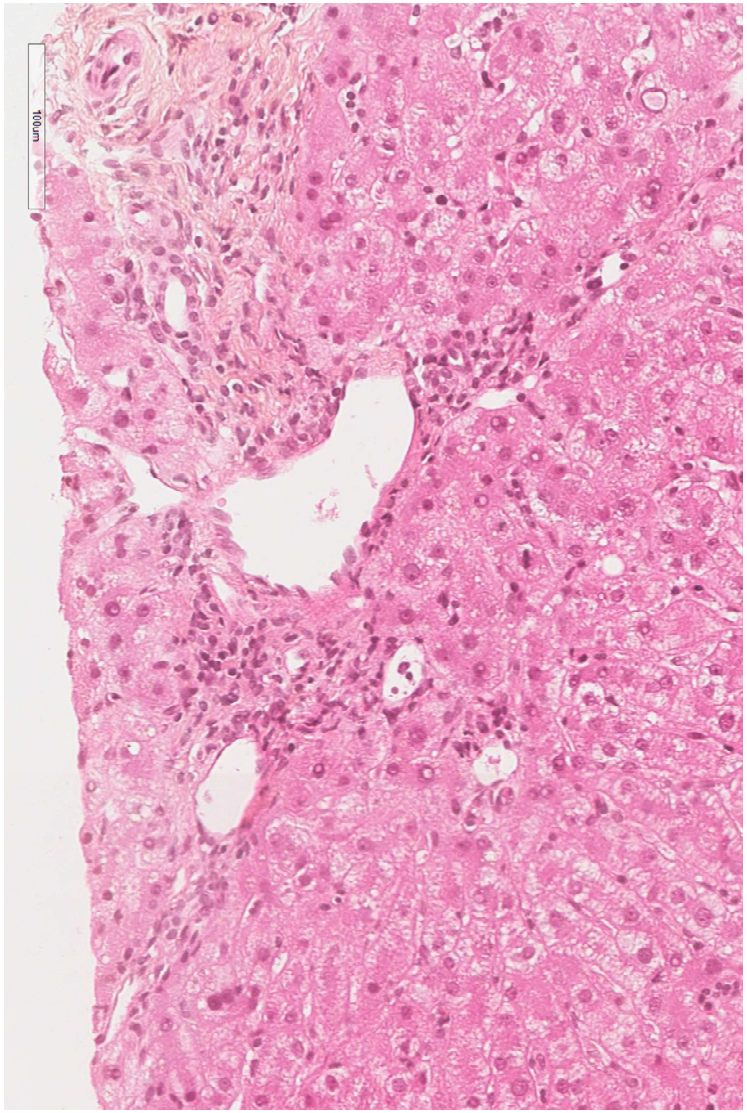


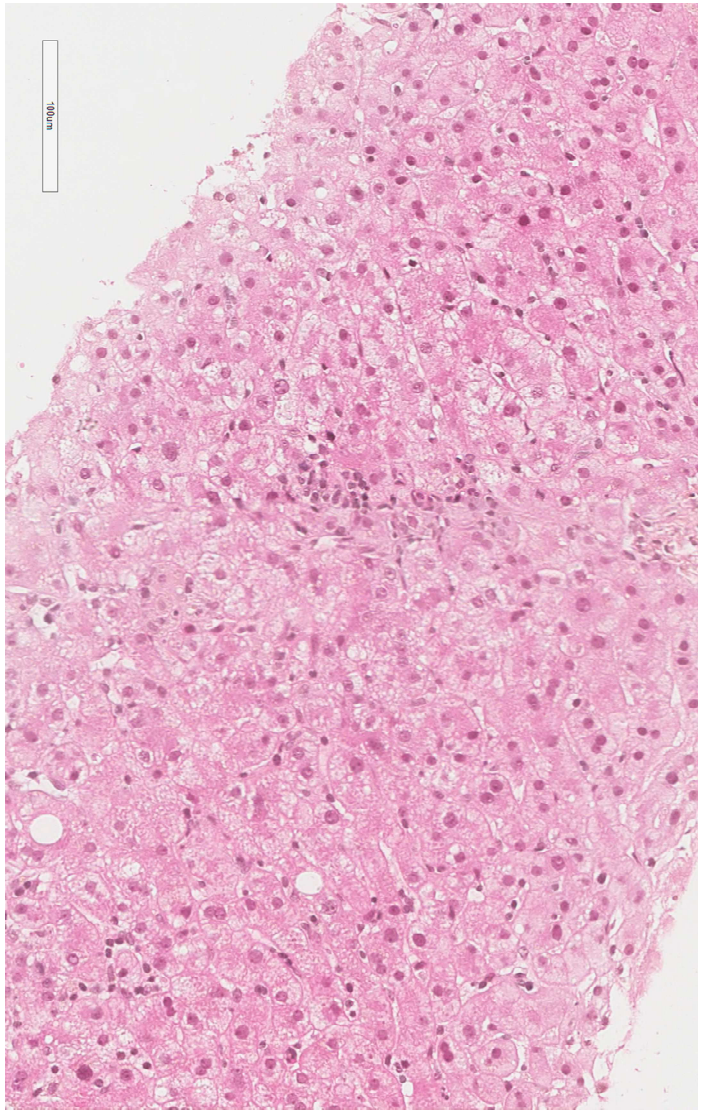


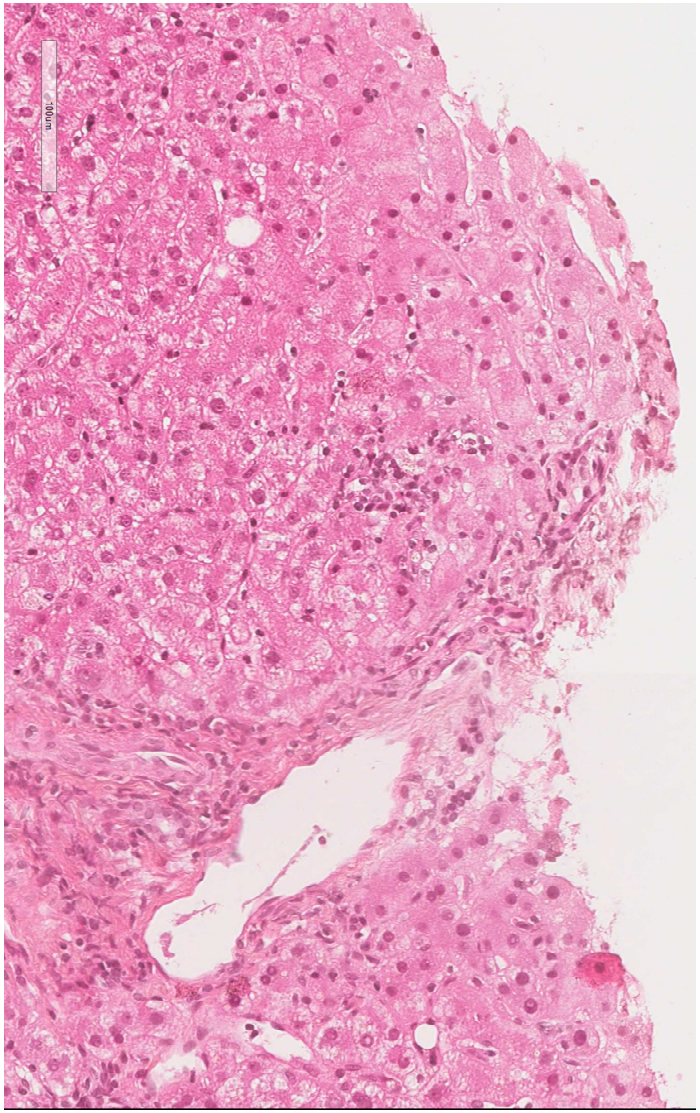
No significant portal and parenchymal lesions

39 year-old man, familial history of vein thrombosis (Protein C deficiency & Factor II mutation)
Superior mesenteric venous thrombosis + intestinal necrosis





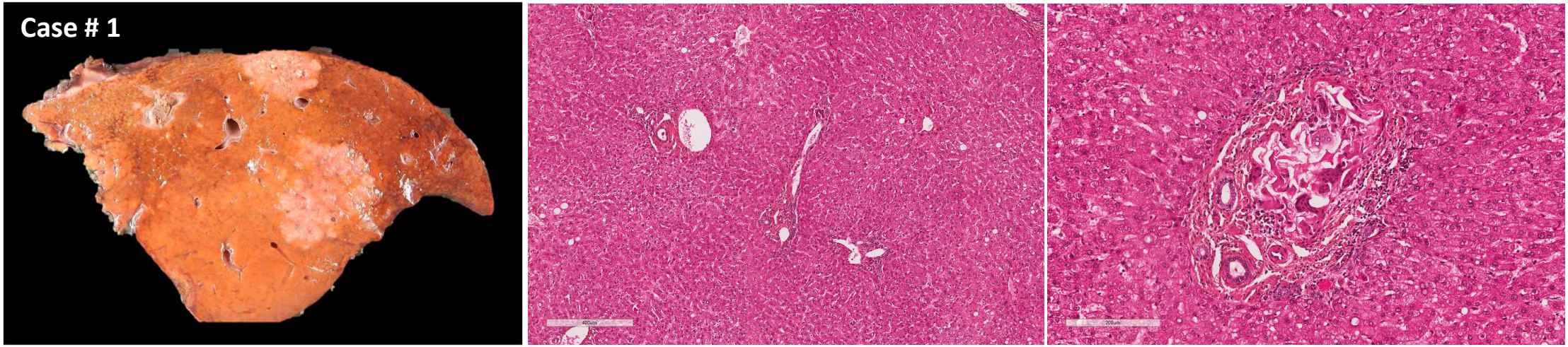




CMV-associated PVT (likely-PSVD)

De Broucker et al, J Hepatol 2022

Liver metastasis →Neoadjuvant chemotherapy → **Right portal embolisation** → Right hepatectomy **(+ 2 mo)**



Obliterative portal venopathy: A comparative study of 184 cases of extrahepatic portal obstruction and 469 cases of idiopathic portal hypertension

Table 7 Liver pathology. A comparison of EHPO and IPH

	Percentage of cases	
	EHPO ($n = 184$)	IPH ($n = 469$)
Gross findings		
Normal	69.0	41.2
Fibrosis	10.9	33.9
Irregular surface	5.4	11.3
Nodule formation	3.3	4.7
Other findings	3.3	2.1
Not described	8.2	6.8
Portal fibrosis	**28**	**72**

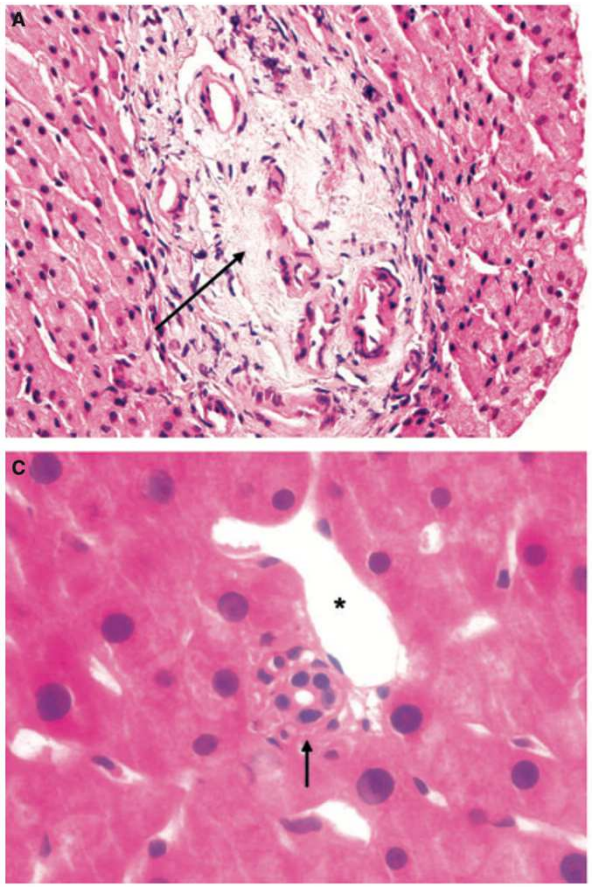
Vascular portal lesions not specifically detailed

Kameda H J Gastroenterol & Hepatology 1986

Histological features in western patients with idiopathic non-cirrhotic portal hypertension

Table 3. Comparison of histological characteristics of patients with idiopathic noncirrhotic portal hypertension (INCPH) (n = 70) and noncirrhotic portal vein thrombosis (PVT)

Histological lesions	INCPH (n = 70)	PVT (n = 23)	P
Phlebosclerosis	67 (95)	15 (65)	<0.001
Paraportal shunt vessels	62 (89)	22 (96)	0.57
Portal tract remnants	32 (46)	0 (0)	<0.001
Portal vein arterialization	11 (16)	0 (0)	0.06
Portal vein dilatation	24 (34)	18 (78)	<0.001
Thin incomplete septa	7 (10)	0 (0)	0.18
Increased vascular channels in portal tracts	50 (71)	18 (78)	0.60
Nodular regeneration	39 (56)	5 (22)	0.003
Increased parenchymal draining vessels	29 (41)	6 (26)	0.22
Sinusoidal dilatation	66 (94)	23 (100)	0.61
Perisinusoidal fibrosis	64* (97)	23 (100)	0.95
Perivenular fibrosis	43* (65)	7 (30)	0.026
Steatosis (10–50%)	4 (6)	0	0.6



Verheij J Histopathology 2013