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ULCERATED ENTEROPATHY - WHEN TO CONSIDER HEMATOLOGICAL DISTURBANCES IN GASTROENTEROLOGY?

João Carlos Silva, Inês Rueff Rato, Rolando Pinho, Adélia Rodrigues, Ana Ponte, Margarida Badior, Xiaogang

Wen, Catarina Gomes, João Correia, Manuela Estevinho, João Carvalho

Centro Hospitalar Vila Nova de Gaia Espinho

CASE DESCRIPTION

- 56-year-old woman admitted to the ER due to abdominal pain, vomiting and fever (37.8°C).
- The patient had past medical history of HBP, type 2 diabetes and pulmonary tuberculosis.

DIAGNOSTIC WORKUP

- ✓ Complete blood count: haemoglobin 10,2g/dL, mean cell volume 103fL and 38x10E3/uL platelets
- ✓ Lactate dehydrogenase 450 U/L. C-Reactive protein 10,9mg/dL. Urinalysis was unremarkable.

Additionally, an 18 mm **hepatic nodule** was identified in the caudate lobe/right lobe, with enhancement in the arterial phase, without washing out.

Abdominal CT revealed an **ACUTE OCCLUSIVE THROMBUS OF THE PORTAL VEIN** and **PARIETAL THICKENING OF THE 3RD AND 4TH DUODENAL PORTIONS AND PROXIMAL JEJUNUM** associated with the densification of regional fat.

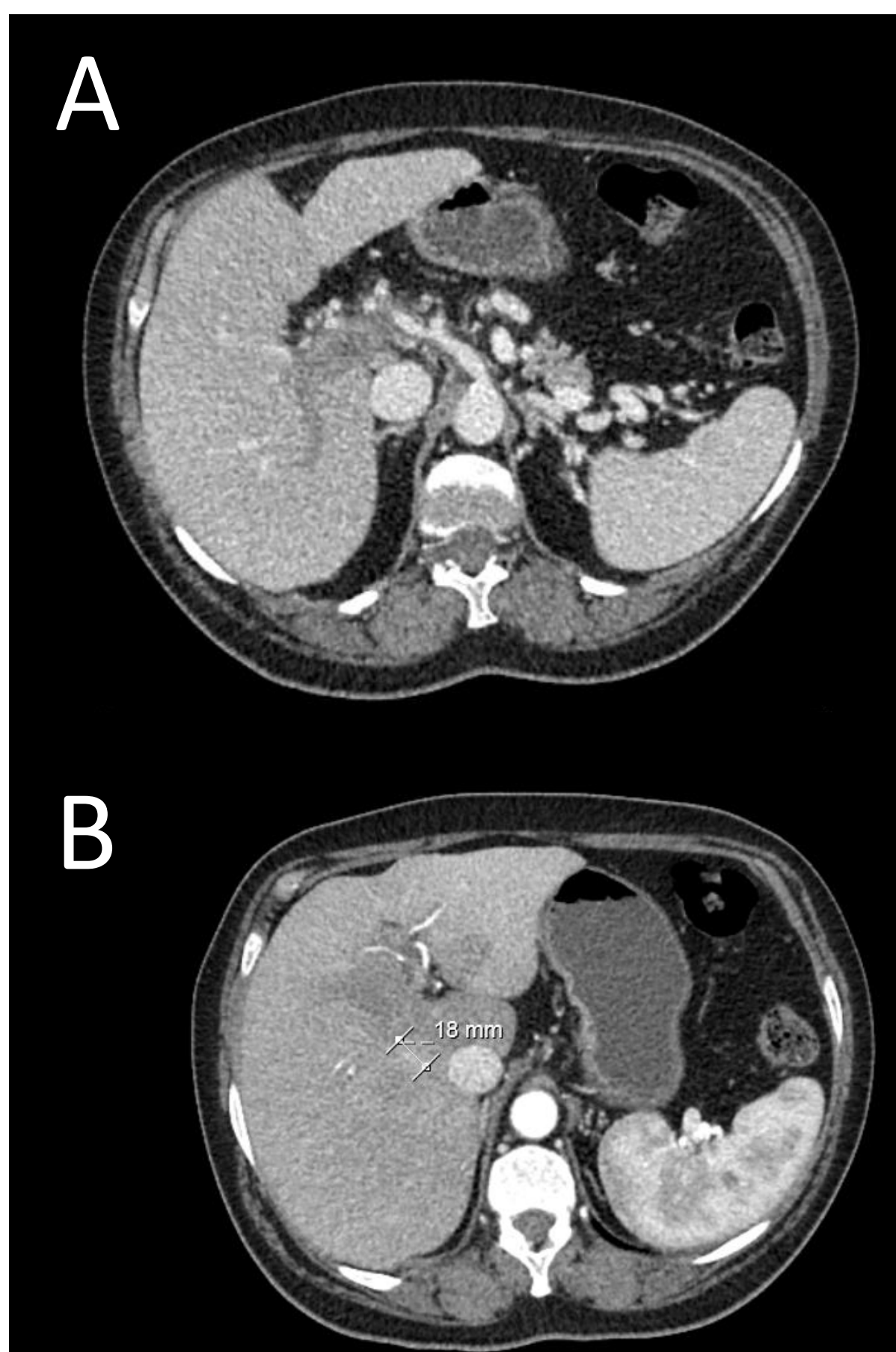


Figure 1 –Abdominal-CT findings

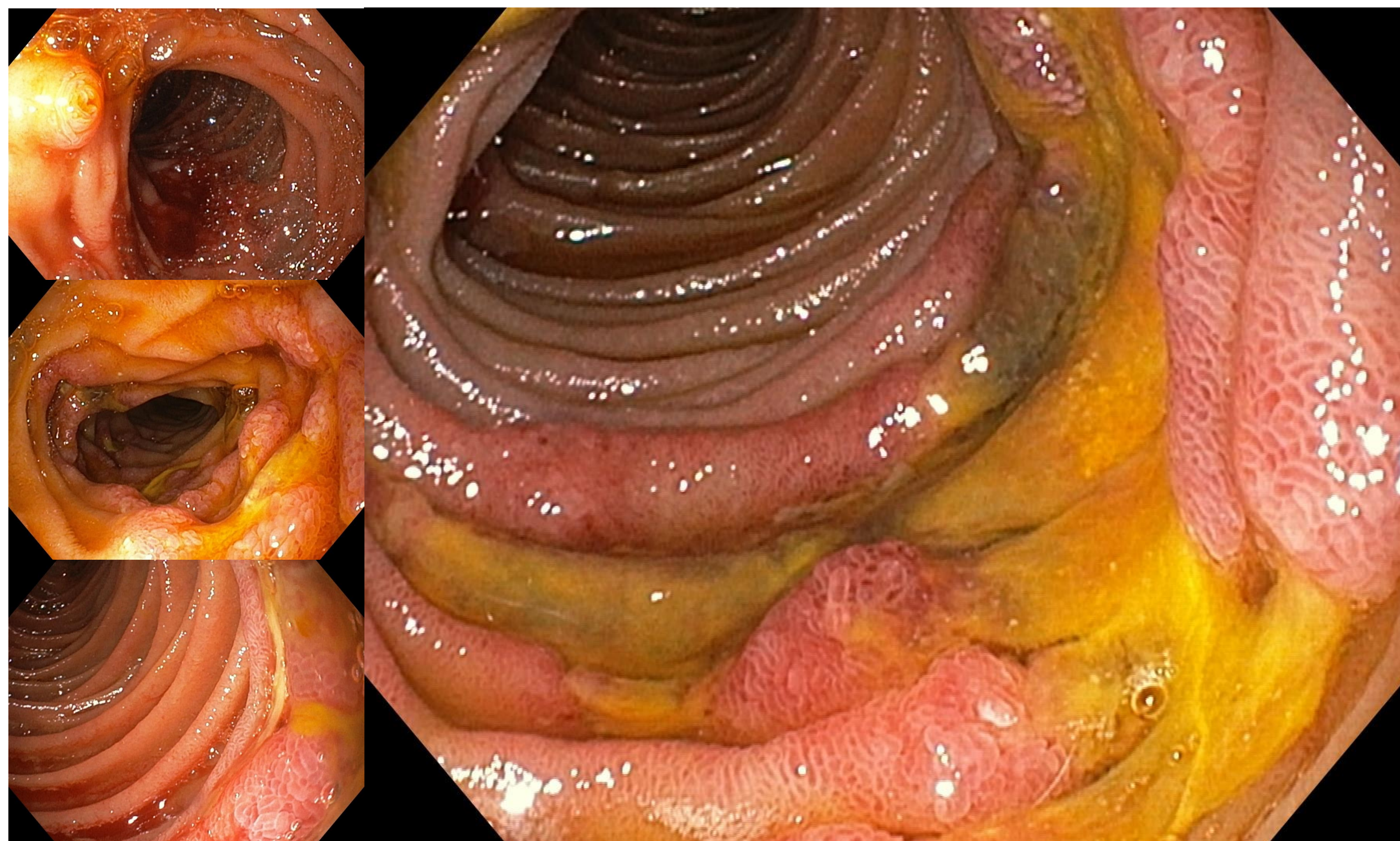


Figure 2 – Balloon assisted enteroscopy revealing 2 ulcers (8mm and 12mm) in the 4th duodenal portion and an ulcerated lesion involving ¾ of the luminal circumference in the proximal jejunum.

Differential Diagnosis:

1. Infectious enteritis
2. Intestinal tuberculosis
3. Hepatocarcinoma
4. Myelodysplastic disorder

Blood cultures, viral serologies (including CMV PCR) and direct exam for BAAR in jejunal biopsy excluded infectious etiology.

Histology revealed erosion of the superficial epithelium and the **MUCOSAL AND SUBMUCOSAL VESSEL THROMBOSIS**, mimetizing vasculitis.

For thrombocytopenia and negative coombs hemolytic anemia, flow cytometry was performed and confirmed the diagnosis of **TYPE II / III PAROXYSMAL NOCTURNAL HEMOGLOBINURIA (PNH)**

The patient presented a good clinical evolution with hypocoagulation and supportive care.

CONCLUSION

PNH is a rare hematopoietic stem cell disease characterized by intravascular hemolysis by complement as a result of clonal expansion of hematopoietic stem cells having acquired mutations in PIG-A gene. Ischemic enteritis accompanied with PNH has life-threatening complications associated with extensive intestinal infarction frequently attributed to superior mesenteric vein thrombosis. In this case, enteroscopy played a central role in the diagnosis of ischemic enteritis. The challenging diagnosis of PNH implied an integrative and multidisciplinary approach.

REFERENCES

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