

(3) Secondary to the cytokine storm itself (high levels of TNF-alpha and others).³ Theory number 3 is the most likely hypothesis in our patient.

Although haemophagocytosis and activated macrophage proliferation in the liver biopsy are common, in almost half of all cases only haematogenous pigments are found, as was the case in our sample. Other common findings include sinusoidal dilatation and hepatocyte necrosis. In our patient, haemophagocytosis was not detected in the liver biopsy but rather in the bone marrow aspiration.^{3,5}

The HPS mortality rate exceeds 70%, primarily due to sepsis and intracerebral haemorrhage. Apart from in cases of fulminant hepatic failure, liver disease does not usually represent the cause of death, although it does constitute a severity marker together with ferritin and thrombocytopenia.² The administration of steroids resolved our patient's hepatitis, with the gradual normalisation of bilirubin and transaminase levels as well as the gradual resolution of kidney failure, whilst maintaining high ferritin levels. Given progressive cytopenias, treatment with dexamethasone and etoposide was initiated in accordance with protocol HLH. The patient died due to sepsis and intracerebral haemorrhage.

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McKittrick-Wheelock syndrome: A rare cause of metabolic coma[☆]



Síndrome de McKittrick-Wheelock: una causa infrecuente de coma metabólico

Colorectal adenomas are benign tumours with the potential to become malignant. Their incidence ranges from 2.9% to 11.5% depending on the series.¹ Villous adenomas represent 10% of all colorectal adenomas and are normally asymptomatic, although 3% are hypersecretory.² In 1954, McKittrick and Wheelock³ were the first to report hydroelectrolyte depletion secondary to severe diarrhoea caused by this type of tumour.

We present the case of a 72-year-old female patient with a history of hypertension, type 2 diabetes mellitus and hospitalisation two years prior due to pneumonia and hyponatraemia. She attend A&E following three days of epigastric pain together with melaena in the last 24 h. The patient's relatives reported occasional vomiting and severe

diarrhoea in the previous days. Although the patient presented bradylalia and bradyphrenia, she was awake and oriented during the physical examination. She was found to be haemodynamically stable with vital signs within normal range. The abdomen was rounded but soft and depressible with signs of peritoneal irritation. There were signs of skin and mucosa dehydration. During her stay in A&E, the patient presented a reduced level of consciousness, suffered tonic-clonic seizures and went into a coma. The complete blood count conducted in A&E revealed severe hyponatraemia with sodium levels of 101 mEq/l; potassium 4.6 mEq/l; plasma hypoosmolarity without metabolic acidosis and normal kidney function.

The patient was admitted to the intensive care unit after being intubated and put on mechanical ventilation. A cranial computed tomography was performed that found no acute lesions. Volume replacement with hypertonic saline was implemented, and hyponatraemia was gradually corrected until levels of 131 mEq/l were reached. The patient regained consciousness as sodium levels were being corrected, coherently responding to verbal commands but with bradyphrenia.

Given the patient's profuse diarrhoea, a colonoscopy was performed that revealed a villous polyp (0-IIa on the Paris classification) at the hepatic flexure measuring more than 3 cm and endoscopically unresectable owing to its size, with no endoscopic signs of malignancy. The lesion showed abundant surface mucus and signs of recent bleeding (Fig. 1). The

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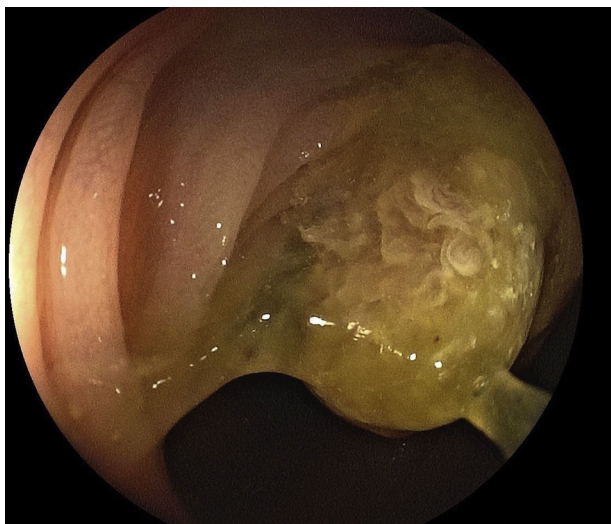


Figure 1 Villous polyp at the hepatic flexure of the colon. A polyp larger than 3 cm covered with abundant mucus (0-IIa on the Paris classification) and endoscopically unresectable owing to its size can be seen.

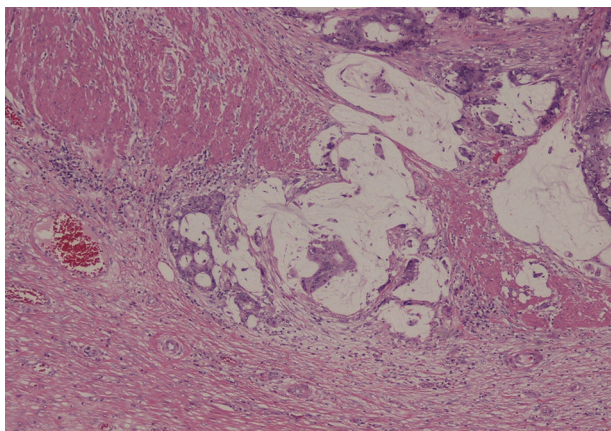


Figure 2 Histological section of the lesion after surgical excision. Tumour invasion of the colon wall's muscular layer as well as cellular atypia and abundant mucin can be seen.

endoscopic biopsy revealed a fragment of villous adenoma in the large intestine with low grade dysplasia. Given that the polyp was secreting mucus, a laparoscopic right hemicolectomy was performed, subsequent to which the patient's condition improved and all symptoms abated. Sodium plasma levels after the surgery were 134 mEq/l. The anatomopathological study of the surgical specimen revealed a villous adenoma measuring 4.5 cm × 3.5 cm × 1.2 cm, in which a moderately-differentiated mucinous adenocarcinoma was found (low grade dysplasia) that measured 1.5 cm microscopically (Fig. 2). The tumour invaded the muscle itself and just reached the subserosal fat, isolating 16 lymph nodes, all without histological features of malignancy (pT3N0).

McKittrick and Wheelock³ reported a syndrome characterised by mucous diarrhoea associated with hyponatraemia, hypokalaemia, severe dehydration and pre-renal azotaemia. Although our patient presented severe hypona-

traemia and physical signs of dehydration, serum potassium and creatinine levels were normal at admission.

Colorectal villous adenomas are benign tumours that can histologically be divided into secretors and non-secretors. Secreting tumours produce a large amount of mucous and are composed of a greater number of mucus-secreting cells, resulting in higher levels of cyclic AMP and adenyl cyclase. The mediator involved in hypersecretory diarrhoea is prostaglandin E2 (PGE2), levels of which can be up to three times higher in secretory adenomas than in non-secretory adenomas.⁴ This causes profuse diarrhoea resulting in severe hydroelectrolyte depletion, which is definitively treated by polyp resection. To alleviate symptoms while awaiting resection, 400 mg/day of indomethacin has been proposed due to its inhibitory action on PGE2,⁵ which could reduce adenoma secretion levels.

This type of tumour is most commonly found in the sigmoid and the rectum. Tumour size, location and how villous the tumour is have been proposed as factors that could influence symptoms.¹ Diarrhoea and mucosal secretion seem to be more common in large lesions and those located in the rectum, probably because the distal location inhibits the colon's ability to absorb liquids. Nevertheless, small lesions in the right colon, such as in our patient, may manifest with very acute symptoms. Although the known side effects of secretory diarrhoea include central nervous system dysfunction due to hyponatraemia, such extreme dysfunction as metabolic coma and seizures have not been reported in the literature.

Surgical resection of the lesion is considered to be the definitive treatment of choice.⁶ Depending on the location of the lesion, the most appropriate surgical resection technique is chosen based on the principles of surgical oncology.

Whenever the patient has a history of chronic or hypersecretory diarrhoea, McKittrick-Wheelock syndrome should be considered in the differential diagnosis of severe metabolic disorders associated with central nervous system dysfunction. For early diagnosis, a diagnostic colonoscopy should be performed in all patients with these symptoms. Although polyp size has been correlated to symptom severity, small villous adenomas located in the proximal colon may trigger extremely acute symptoms, as with our patient.

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Rectal neuroendocrine tumor with an exceptional behaviour[☆]



Tumor neuroendocrino de recto de comportamiento excepcional

Neuroendocrine tumors are malignancies that derive from the diffuse endocrine system of the intestine, which secrete

up to 40 different cytokines and hormones.^{1,2} 75% manifest in the digestive system, 34% of which in the rectum, where they tend to be asymptomatic and present in the form of sessile masses or thinning of the rectal wall.³ Prognosis will depend on the extent of the vascular or lymphatic invasion. Tumors measuring less than 1 cm present metastasis in fewer than 3% of cases, while the rate of metastasis for tumors larger than 2 cm increases to up to 70%. Tumor size

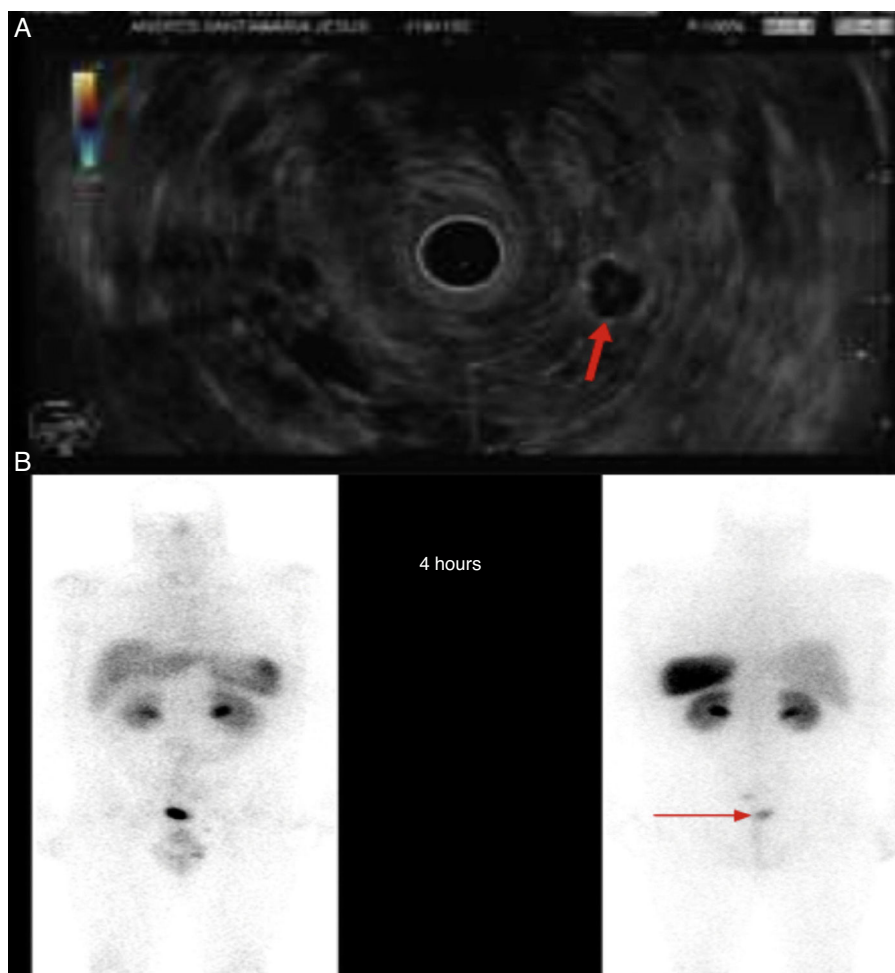


Figure 1 (A) Lower echo-endoscopy revealing perirectal lymphadenopathy. The resulting biopsy was positive for carcinoid tumor. (B) Octreoscan with pathological uptake in the left perirectal area.

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