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Vanek jejunal tumor, a rare cause of anemic syndrome. Review of the literature

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Summary

fibroid polyps (IFP), commonly referred to as Vanek Tumor, are rare, benign lesions of mesenchymal origin. They can affect both sexes with a majority incidence between 50-70 years of age^{1 2}

We present the case of a 53-year-old woman referred to outpatient clinic for severe iron deficiency anemia, gastroscopy and colonoscopy without relevant findings. A capsule endoscopy study was completed, identify an ulcerated polypoid mass, which caused a slight stenosis of the lumen allowing the passage of the capsule endoscopy compatible with Vanek tumor.

Keywords: Vanek. Fibroid polyp. Anemia. Capsule.

Dear Editor,

fibroid polyps (IFP), also called Vanek tumors, are rare, benign lesions of mesenchymal origin. They can affect both sexes with a majority incidence between 50-70 years of age¹,²

We present the case of a 53-year-old woman who was referred to an outpatient clinic for severe iron deficiency anemia with clinical tests that highlighted Hb 8 g/dL, iron 25 μ g/dL, ferritin 5.1 μ g/dL, STI 6 and positive SOH. The patient has reported asthenia



without weight loss and without gastrointestinal bleeding. She has had negative celiac antibodies and negative Helicobacter pylori. An endoscopic exploration with gastroscopy and ileocolonoscopy was performed with no relevant findings.

Despite iron supplementation, Hb controls persist around 10 g/ dl. Consequently, the study has been extended with an endoscopic capsule that has identified a ulcerated polypoid mass in the distal jejunum-proximal ileum (at 3:20 h from the pylorus and 45 min from the cecum). The polypoid mass partially obstructs the intestinal lumen but allow passage of the endoscopic capsule to reach the cecum.

We suspected a intestinal tumor, for this reason, A CT thoracoabdominal- pelvic has been performed, which showed the lesion referred to in the hypogastrium, with a endoluminal origin, polypoid and pedunculated, measuring 5 x 3 cm with peripheral uptake and a necrotic centre causing a short invagination of the loop without obstructive signs. There were not distant metastasis.

A laparoscopic procedure was chosen to treat this 5cm lesion due to the potential complication to invagination described in TC and the difficulty endoscopic access, according to the surgery service too. The laparoscopic approach was with excision of the jejunal tumor with section of the meso of the small intestine and preparation of a side-to-side aniso peristaltic.

The jejune's surgical resection piece measures 12 cm in length and 4.5 cm in diameter. Upon opening it, approximately 1.4 cm from the nearest surgical edge of the piece, an elongated polypoid lesion of 6 x 2.5 cm is recognized, an apparently submucosal growth bulging the mucosa with flattening of folds. Histologically of submucosal origin that bulges the mucosa, identifying areas of erosion and ulceration. The lesion grows at the expense of a submucosal proliferation of stromal cells with elongated cytoplasm, with slightly stellate nuclei without remarkable cytological atypia, identifying areas with looser stroma and slight focal myxoid degeneration and with vessels of various sizes, some with somewhat thicker and hyalinized walls than others and in other areas vessels around which the stromal cells are arranged concentrically giving images in "onion layers". An immunohistochemical study is performed, observing in said stromal proliferation: CD34 positive and S100, Ckit, DOG1 and STAT6 negative. All of this is compatible with Vanek tumor.



The patient had an excellent clinical evolution, with no post-surgical complications and subsequent analytical controls showing no signs of anemia or iron deficiency.

Vanek tumors or inflammatory fibroid polyps (IFP) are benign and rare lesions, and their origin is unknown. They were first reported by Vanek in 1949. They generally present as single, polypoid lesions of variable size that can be located throughout the gastrointestinal tract, with the gastric antrum and ileum being the most common locations. Jejunal location is rare^{1 2}

The clinical course is variable, from being asymptomatic, occurring within the context of anemic syndrome or even debuting through a complication such as gastrointestinal bleeding, intestinal obstruction or intussusception as in the case recently described in this journal by Hueso et al. that required a surgical approach as in our patient. Due to the characteristics of its clinical presentation, Vanek 's tumor should be included in the differential diagnosis of other small bowel neoplasms such as gastrointestinal stromal tumor (GIST), leiomyoma, leiomyosarcoma, schwannoma and inflammatory myofibroblastic tumors^{3 4}

The definitive diagnosis is anatomopathological with positivity for CD34 and histologically it presents a submucosal mesenchymal origin with an ulcerated surface characterized by spindle cells arranged in onion peels with an eosinophilic component and vascular infiltration².

Within its therapeutic approach, it can be considered as other benign gastrointestinal polyps in addition to the use of endoscopic ultrasound to better identify the lesion if applicable. The choice of treatment is based on both the patient's clinical picture and the location and size of the polypoid lesion.

The treatment of choice is endoscopic with endoscopic mucosal or submucosal resection if its location and size allow it. Although the small intestine is a less frequent location, surgical intervention is reserved especially for large lesions that are difficult to reach by endoscopy or with obstructive manifestations as in our case. Finally, after surgical resection there is no data on recurrence^{3 5}

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Figure



Inflammatory fibroid polyp or Vanek tumor. On the left, capsule endoscopy images showing a stenosing polypoid lesion. In the center, sagittal section image of an abdominal CT scan showing this lesion with a short loop invagination. On the right, post-surgical macroscopic piece. Capsule endoscopy images. Pathological anatomy of fibroid polyp 4x" and " Fibroid polyp 4x" vessels: stromal cells are arranged around them forming "onion layers".