

Perineurioma: an uncommon lesion in the gastrointestinal tract

A 45-year-old man was referred to our hospital with a gastric nodule. Gastroscopy revealed a flat, pale-colored lesion, approximately 15 mm in diameter, in the gastric body. The lesion showed some nodularity but seemed to be covered with a slightly inflamed mucosa (● Fig. 1). Endoscopic ultrasound (EUS) showed a thickened mucosal layer, as well as scattered, small hypoechoic areas in the submucosal layer (● Fig. 2).

A biopsy was taken but the final diagnosis was not definitive because of the small size of the samples. Endoscopic submucosal dissection (ESD) was performed to obtain a precise diagnosis, as well as for removal of the lesion. Histopathologically, the lesion consisted of a focal proliferation of spindle cells without atypia that were present from the lamina propria to the

submucosa (● Fig. 3a). Immunohistochemistry revealed that the spindle cells were positive for EMA and claudin-1 (● Fig. 3b,c). Therefore, this lesion was eventually diagnosed as a gastric perineurioma with no evidence of malignancy.

Benign peripheral nerve sheath tumors, which occur uncommonly in the gastrointestinal tract, include ganglioneuromas, neurofibromas, and schwannomas. Perineuriomas are rare, benign peripheral nerve sheath tumors that include soft tissue, intraneural, and sclerosing variants [1]. The first soft tissue perineurioma was described by Lazarus and Trombetta in 1978 as an intramuscular neurofibroma-like tumor of the calf with ultrastructural features characteristic of perineurial cells [2]. To date, there are only two case reports of gastric perineuriomas [3,4].

It is important to emphasize that distinguishing perineuriomas from other spindle cell neoplasms of the gastrointestinal tract can be facilitated by immunostaining for EMA and claudin-1. Agaimy and Wuensch suggested that this exceedingly rare mesenchymal neoplasm might have been under-recognized or misinterpreted as a gastrointestinal stromal tumor (GIST) before the era of KIT immunostaining. Perineurioma should therefore be included in the differential diagnosis of spindle cell GISTs with unusual features, especially the so-called KIT-negative GISTs. Gastroenterologists, as well as pathologists, should be aware of this type of submucosal lesion.

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Fig. 1 Endoscopic view under white light showing a 15-mm diameter, flat, pale-colored, nodular lesion covered by mildly inflamed mucosa within the gastric body.

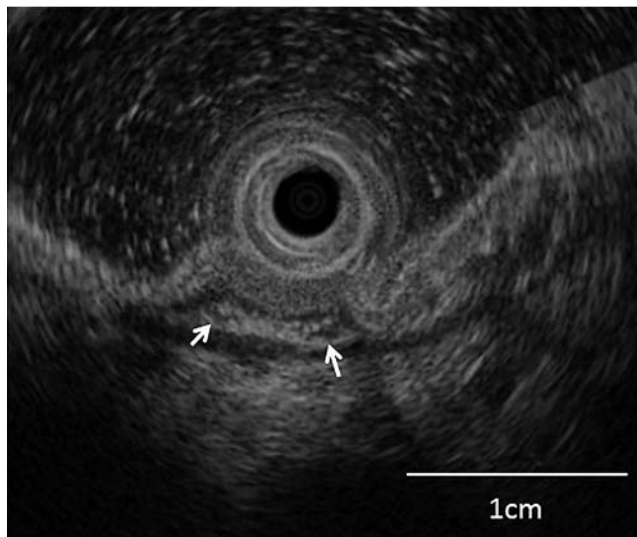


Fig. 2 Endoscopic ultrasound (EUS) image of the gastric lesion showing a thickened mucosal layer with small hypoechoic areas (arrows) in the submucosa.

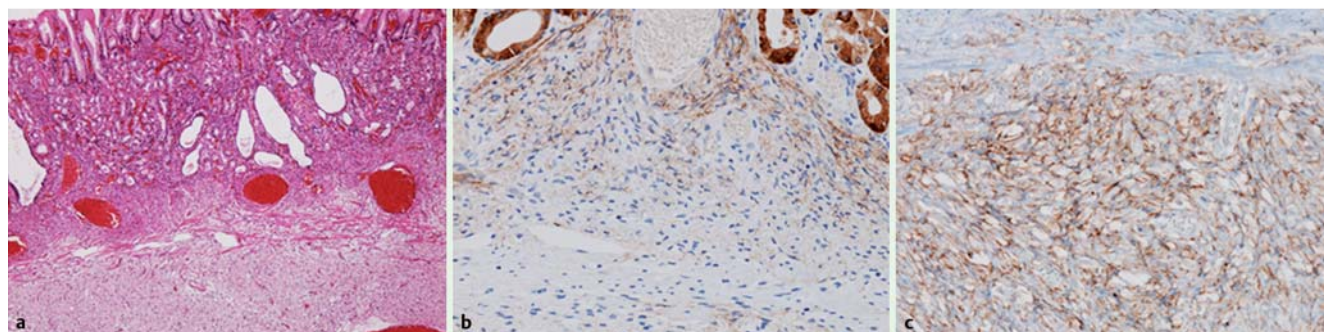


Fig. 3 Histopathological examination of the samples obtained by endoscopic submucosal dissection (ESD) of the lesion showing: **a** focal proliferation of spindle cells without atypia from the lamina propria to the submucosa (low power view); **b** weak positivity for EMA by immunohistochemical staining; **c** strong positivity for claudin-1 by immunohistochemical staining, consistent with the diagnosis of a perineurioma.

Competing interests: None

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