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## Scientific letters

### Verrucous Carcinoma of the Esophagus: A Rare Entity With a Difficult Diagnosis<sup>☆</sup>

### Carcinoma verrucoso esofágico: una rara entidad de difícil diagnóstico

Verrucous carcinoma (VC) is a rare variant of squamous carcinoma that is difficult to diagnose and whose therapeutic management should be individualized. Its location in the esophagus, first described by Minielly in 1967,<sup>1</sup> is rare, and only 50 cases have been reported in the literature.

Its exact etiology is unknown, but VC is strongly associated with prolonged inflammatory factors, such as achalasia, gastroesophageal reflux disease, alcohol and tobacco use. The most recently published cases suggest a possible relationship with papillomavirus (HPV) infection, which is why this cause should be considered in all patients with this suspected diagnosis.<sup>2</sup>

VC predominantly affects males (ratio 2:1) with a mean age of 60 (range 36–79 years). The main clinical manifestation is dysphagia, which is usually accompanied by weight loss. Other symptoms may include hematemesis, cough and odynophagia.

The suspected diagnosis can be established by endoscopy, with a typical macroscopic appearance consisting of a whitish wart-like mass. The exophytic nature of these lesions makes it very difficult to obtain biopsies in which the invasive component of the tumor is visualized. Frequently, only nonspecific inflammatory changes with the absence of malignancy are observed. Therefore, complete tumor resection is usually necessary to reach a definitive diagnosis.<sup>3</sup>

The pattern of superficial growth, limited lymph node and distant involvement, and the response to chemoradiotherapy differentiate VC from squamous cell carcinoma, making endoscopic resection and neoadjuvant treatment valid alternatives to esophagectomy.

The objective of this article is to describe a case of esophageal VC that demonstrates the difficulty to diagnose and stage this entity, and, therefore, to choose its treatment.

The patient was a 50-year-old man with progressive weight loss and dysphagia in previous months. He was a heavy

drinker, smoker, and had a history of gastroesophageal reflux disease and achalasia, which had been treated with a Heller myotomy 12 years earlier.

Three esophagogastrosopies were performed with biopsies, none of which contributed to the definitive diagnosis. All reported a mucosa that was completely covered by whitish, non-cottony exudates between 20 and 32 cm from the dental arch that could not be removed with washing; there was also adhered food debris and a papilliform polypoid nodular mass 26 cm from the arch (Fig. 1). The pathology studies showed evidence of a papillomatous and verruciform hyperplastic keratinizing epithelium, with notable superficial acute and deep chronic inflammatory infiltrate, no signs of malignancy and with fungal structures.

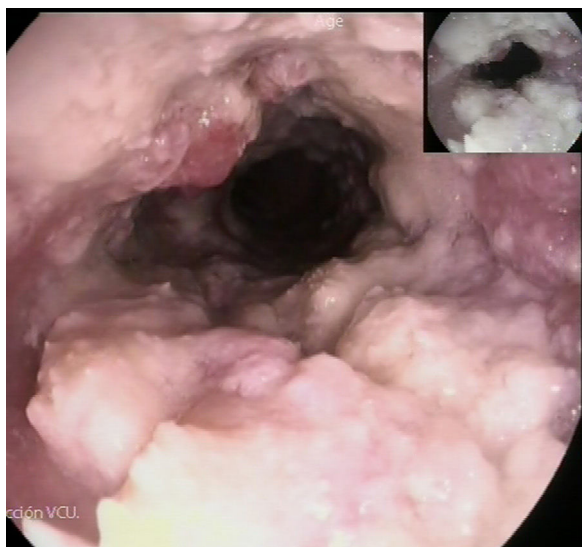
The study was completed with a computed tomography scan, hybridizations for HPV and lab work-up, including tumor markers, which were negative.

Given the high suspicion of VC that was not confirmed, the patient was referred to another hospital for an endoscopic ultrasound, which reported the same endoscopic findings as well as loss of structure of the esophageal wall and loss of layered pattern, hypoechogenic appearance, indentations in the esophageal adventitia (suggestive of T3 if the tumor nature is confirmed), and no invasion of neighboring structures or lymphadenopathies associated with the tumor or in the area of the celiac trunk.

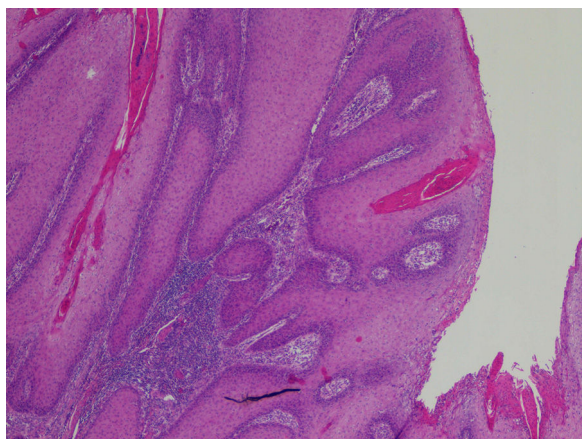
Despite not having histological confirmation, after assessment of the case in the Digestive Tumors Committee, we decided to perform a transhiatal esophagectomy with sleeve gastropasty, ruling out endoscopic resection due to the endoscopic ultrasound findings.

The postoperative period ran its course with dehiscence of the cervical anastomosis and secondary respiratory distress, associated with symptoms compatible with alcohol withdrawal syndrome. The patient was managed conservatively,

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**Figure 1 – Esophagogastroscopy: papilla-like formation with candidiasis.**



**Figure 2 – Non-invasive squamous epithelium (hematoxylin-eosin stain 40×).**

developing dysphagia secondary to stenosis of the esophago-gastric anastomosis, which was resolved after endoscopic dilatations.

In the histological study of the surgical specimen, a well-differentiated malignant epithelial neoplasm was observed with acanthotic and hyperkeratotic superficial papillomatous growth with minimal basal atypia and chronic deep inflammation. The neoplasm showed extensive superficial growth, predominantly intramucosal, although the *muscularis propria* was free of neoplastic infiltration. The definitive pathology diagnosis was well-differentiated verrucous squamous cell carcinoma, with free surgical margins, absence of lymph node metastasis and associated areas of Barrett's esophagus (TNM stage: pT1N0) (Fig. 2).

As in our case, it is not uncommon to find disagreement between the endoscopic ultrasound and the definitive

pathology study, due to the high level of chronic deep inflammation that may lead to overstaging by endoscopic ultrasound.<sup>4</sup>

The differential diagnosis of VC should include benign lesions associated with stenosis or dysphagia, such as esophageal leiomyoma, papilloma or esophageal tuberculosis, and with malignant lesions like adenocarcinoma of the esophagus.<sup>5</sup>

As this pathology is rare, there are no precise recommendations for its treatment. Endoscopic mucosal and submucosal resections have been described for early stages, with good results.<sup>6</sup> Other authors, including Brandalise et al.,<sup>7</sup> have described a positive response to neoadjuvant treatment in patients with involvement of the entire esophagus, allowing for more conservative resections.

However, despite its slow growth and high degree of differentiation, due to the frequent diagnostic delay in locally advanced stages, esophagectomy continues to be the most common therapeutic option.<sup>8</sup>

Morbidity and mortality are associated with local invasion (pulmonary, bronchial, pleural and pericardial involvement) and surgical complications (fistulas). No distant metastasis has been reported in the literature.<sup>9</sup>

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## Transanal Endoscopic Microsurgery Treatment of Twice Recurred Tail-gut<sup>☆</sup>

### Tratamiento mediante microcirugía endoscópica transanal de un hamartoma quístico retrorrectal birrecidivado



Retrorectal cystic hamartoma, or tail-gut, is a rare malformation derived from embryonic remnants of the intestine that can manifest as pain, obstruction or superinfection. Several surgical approaches have been described, including transperineal, transabdominal, laparoscopic and, recently, TEM access. Complete removal of the epithelium of the cyst is an essential condition for healing, as its persistence would lead to recurrence. We present the case of a patient who had been treated surgically five times: thrice by perineal access, one laparoscopy-assisted, and finally the transrectal access using TEM.

The patient is a 37-year-old woman whose only history of interest was perianal suppuration since childhood. Initially, she had been diagnosed with a perianal abscess that debrided spontaneously. Subsequently, she underwent elective surgery for perianal fistula flattening at the ages of 16 and 20. Given the recurring perianal disease, a rectal MRI was performed in 2008, which demonstrated the existence of a retrorectal hamartoma (tail-gut cyst).

Given this diagnostic evidence, elective surgery was performed in October 2008, and, using a left sacrococcygeal perineal approach with resection of the coccyx, the cystic lesion was resected and the external sphincter was repaired. The pathology study showed a cystic lesion covered with non-keratinized squamous epithelium compatible with retrorectal cystic hamartoma with extension to the periosteum of the coccyx. After an asymptomatic period, in 2012 the patient presented a new episode of anal suppuration while pregnant, which was self-limiting with antibiotic treatment. After childbirth, a new MRI demonstrated recurrence of the retrorectal cystic lesion. In February 2013, the patient was operated on again, involving laparoscopic excision of the cystic lesion. The patient remained asymptomatic until September 2017.

In a follow-up study due to the reappearance of discomfort in the perineal area, MRI (Fig. 1) and 3-D CT reconstruction (Fig. 2) showed a cystic lesion measuring 26 × 30 × 10 mm in

diameter in the right ischioanal fossa above the sphincter complex, compatible with recurrence/persistence of a retrorectal cystic hamartoma. Given the anatomical location of the lesion in the lower-third of the rectum and the probable existence of perilesional fibrosis, we decided to perform a transanal approach using TEM, and the cystic lesion was able to be completely removed. The patient was discharged without incident 48 h later. The pathology study confirmed the recurrence of a retrorectal cystic hamartoma with disease-free margins.

Tail-gut or cystic hamartomas are congenital in origin, derived from embryonic epithelial cells of the intestine. According to the retrorectal tumor (RT) classification by



Figure 1 – Pelvic nuclear magnetic resonance image.

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