Verrucous carcinoma of the esophagus

Dear Editor,

Verrucous carcinoma of the esophagus is a rare variant of squamous cell carcinoma and only few cases have so far been reported in the international literature.

Case report

A 74-year-old male presented with 1 year history of progressive dysphagia for solids and liquids. Additionally, he had early satiety, anorexia and 30 lb loss of weight. He was a heavy smoker. There was no history of ingestion of caustic substances, esophageal trauma or alcohol use. Initial evaluation elsewhere included two upper endoscopies which showed a constricting white, villous lesion in the distal esophagus. The stomach and duodenum were normal. Endoscopic biopsies showed nonspecific acanthosis with chronic inflammation.

When seen at our institution he appeared cachetic, but the physical examination was otherwise unremarkable. It was decided to repeat upper endoscopy which showed a diffusely thickened, white, friable, exophytic, lumen-constricting polypoid mass in the lower esophagus from 32 to 41 cm (Fig. 1) and to make deeper full-thickness biopsies, which revealed foci of hyperkeratosis and parakeratosis with moderate to severe cytologic atypia. This was suspicious for verrucous carcinoma (VC). An endoscopic ultrasound (EUS) performed using a radial echoendoscope showed thick circumferential hypoechoic lesion arising from the mucosa and extending to the muscularis propria (Fig. 2). There were several enlarged paraesophageal lymph nodes. A thoracic and abdominal computerized tomography (CT) scan was also performed which excluded metastasis. Since this lesion demonstrates T2N1M0 and the biopsies were suspicious for VC, the patient underwent esophagectomy. The resected specimen showed invasive well differentiated squamous cell carcinoma with prominent acanthosis and hyperkeratosis. In situ hybridization for human papillomavirus (HPV) subtypes was negative. The post operative was complicated with hypovolemic shock and the patient died four days after surgery.

Discussion

VC was first described by Ackerman in 1948 (1) and is a very rare esophageal cancer. Esophageal VC is a rare distinct, well
differentiated squamous cell carcinoma that is typically associated with chronic mucosal irritation or a long-term local disease process (2). There are less than 25 cases reported in the international literature. VC is strongly associated with the chronic use of tobacco (3) and according to recent studies, human papillomavirus (HPV) could have a potential role in the tumoral development and progression, although this topic is still under discussion (4). In our case, the patient was a heavy smoker but HPV was not found. This cancer has a highly unfavorable outcome, despite its high degree of differentiation and slow growth, because symptom onset is insidious and there is usually a long delay between development of dysphagia and the detection of the lesion (5). Usually this neoplasm invades the surrounding organs and has a very low potential for metastasis (6). Morbidity and mortality are usually due to complications from local invasion.

Endoscopically, esophageal VC appears as exophytic warty, papillary, spiked, or cauliflower-like masses (1). A conclusive diagnosis of esophageal VC is difficult to make because superficial biopsies tend to show only nonspecific acanthosis, parakeratosis, or hyperkeratosis, with associated acute or chronic inflammation. Deep biopsies with additional diagnosis modalities such as EUS are helpful (7). Surgery is the option of choice for early stage lesions and advanced VC does not seem to benefit from chemotherapeutic regimes (8).

In our case the diagnosis of esophageal VC was challenging, because superficial endoscopic biopsies fail to confirm this diagnosis. Only deeper biopsies and EUS made the suspicious for esophageal VC and the resected specimen made the definitive diagnosis.

Esophageal VC resection can be curative however it has high mortality and morbidity. The reported outcome in patients with esophageal VC suggests that this is a deadly disease, despite the bland histology. Approximately 2/3 of the reported subjects died of disease or direct complications of therapy (6). This interesting condition requires a high index of clinical suspicion and persistence in obtaining a definitive diagnosis.

Fig. 2. EU S: circumferencial hypoechoeic lesion arising from the mucosa and extending to the muscularis propria.