Verrucous squamous cell carcinoma of the oesophagus: a rare tumour with a typical presentation

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Abstract

Introduction

Verrucous squamous cell carcinoma is a rare tumour of the oesophagus with a few reported cases in the literature. While it is an uncommon malignancy, the natural history is substantially more favourable compared to adenocarcinoma and squamous cell carcinoma. A pre-operative tumour biopsy with unyielding results is not uncommon for this type of tumour. In the following report, we present a patient with this malignancy to outline the salient aspects of the clinical management of patients with verrucous squamous cell carcinoma.

Case Report

This is a report of a 65-year-old male who presented with dysphagia to solids and 20 pound weight lost. Multiple esophagogastroduodenoscopies (EGD) showed a large fungating partially obstructing mass in his distal oesophagus with multiple biopsies which were negative for malignancy. Due to the symptomatic nature of the mass and the concern for malignancy, he underwent minimally invasive transhiatal esophagectomy. Final pathology revealed a rare type of oesophageal cancer called verrucous squamous cell carcinoma.

Conclusion

Verrucous squamous cell carcinoma is rare, but it has a typical presentation.

More than 75\% of adenocarcinomas occur in the distal third of the oesophagus, whereas SCC is evenly distributed in the distal two-thirds\textsuperscript{1}. However, other malignancies of the oesophagus have been reported.

Verrucous squamous cell carcinoma (VSC) is a rare tumour of the...
Case report

A 65-year-old male with no past medical history presented with four months of dysphagia and 20 lb weight lost. Symptoms were at first shown only to solid food, however, recently it became difficult for the patient to even tolerate liquids. He was an active smoker and occasional alcohol user, however, denied any personal or family history of malignancy. Physical examination was unremarkable.

An EGD was performed showing a large fungating mass starting at 35 cm and extending to 42 cm (Figure 1). Biopsies showed ulcerated and inflamed squamous epithelium and pseudohyphae and budding yeast consistent with Candida infection. He was started on Fluconazole. There was no evidence of dysplasia or malignancy. Following treatment for candidiasis, EGD was repeated twice more with no new findings except severe esophagitis, no evidence of yeast or pseudohyphae. Multiple biopsies were repeated and were all negative for malignancy. An endoscopic ultrasound was performed showing a bulky tumour at 35 to 40 cm with some spread into gastric cardia. It was circumferential and extended through the muscularis propria and up to the adventitia (Figure 2). Biopsies showed squamous cell proliferation with ulceration, erosion, and inflammation, but without dysplasia or malignancy. A PET scan was performed and showed FDG avidity in the distal oesophageal mass and peri-oesophageal lymph nodes.

A minimally invasive trans-hiatal oesophagectomy was performed. Pathologic specimen revealed a circumferential polypoid lesion 4.5 × 5.5 cm in size above and involving the gastro-oesophageal junction. Non-involved oesophagus and stomach were normal in appearance. All surgical margins were negative for malignancy. Histologically, the mass showed well-differentiated squamous epithelium with minimal cytologic atypia. No malignancy was noted in the ten lymph nodes. Histologic findings are consistent with a pT2N0M0 verrucous squamous cell carcinoma of the oesophagus.

Discussion

Originally described in 1966, (VS) is a malignant papillary tumour composed of well-differentiated squamous epithelium with minimal cytologic atypia and blunt, pushing margins. Pre-operative diagnosis of VS is difficult. Findings of non-diagnostic biopsies showing inflammation, ulceration, and reactive changes to the squamous mucosa are typical and the diagnosis if often made after final pathological examination following surgical extirpation of the tumour.

Similar verrucous carcinomas have been found in the oral and nasal cavity, larynx, genitalia, bladder, and anus. VSC of the oesophagus is a rare entity with only 26 previously reported cases in the literature. These lesions are most commonly located in the lower oesophagus, but reports throughout the organ have been described. Dysphagia is the most common symptom.

Because of its well-differentiated nature, biopsies of VSC are typically unyielding even when multiple attempts are made. VSC is twice more common in men than women. The aetiology for VSC remains unclear, but has a predictable presentation and natural history. With only a couple of dozen cases reported in the English literature, VSC remains a highly unlikely tumour to entertain in a differential diagnosis without a high degree of suspicion. Fortunately, this tumour is a slow growing variant of squamous cell carcinoma with low metastatic potential. Pre-operative diagnosis is difficult due to the well-differentiated nature of the tumour which makes pathologic diagnosis difficult. The following report describes a case of a patient with VSC and outlines the diagnostic dilemma seen with this disease.

Figure 2: Endoscopic ultrasound showing extension through the muscularis propria and up to the adventitia (arrow).
but an inflammatory and/or infectious process might play a role. For instance, human papilloma virus (HPV) was identified as a possible causative agent\(^{2,6,7}\). HPV has been associated with VSC of the genitalia, and in some cases of oesophageal VSC. Similarly, smoking, chronic bile reflux, esophagitis, achalasia, lye injuries, and alcohol use have been associated with VC\(^4\). In the present case, the patient tested negative for HPV.

Endoscopic ultrasound has been used in only a few cases\(^2,6,7\). In these cases, EUS have shown a hyperechoic mass with nonspecific wall thickening involving some of the layers of the oesophagus, which mimics an inflammatory process more than a neoplastic process.

Regional and metastatic spread of the tumour is rare. Although rare, this tumour is associated with low recurrence rates and better than average survival\(^2,5–7\). Follow-up mirrors typical SCC tumours with surveillance EGD and imaging.

**Conclusion**

VSC is rare, but it has a typical presentation. Only a few cases exist in the literature. Pre-operative diagnosis is challenging. The aetiology remains unknown. Resection with follow-up similar to SCC tumours is typically undertaken for VSC.

**Consent**

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

**Abbreviations list**

HPV, human papilloma virus; SCC, squamous-cell carcinoma; VSC, verrucous squamous cell carcinoma.

**References**