

Primary Mucoepidermoid Carcinoma of Esophagus: A Rare Case Report

Dr. Anchana Gulati¹, Dr. Shikha Sharma², Dr. Brij Sharma³, Dr. Gauri Nakra⁴

¹Professor, Department of Pathology, IGMC Shimla

²Senior Resident, Department of Pathology, IGMC Shimla

³Professor & Head, Department of Gastroenterology, IGMC, Shimla

⁴Junior Resident, Department of Pathology, IGMC Shimla.

Corresponding Author: Dr. Gauri Nakra

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ABSTRACT

Primary mucoepidermoid carcinoma (MEC) of the esophagus is an uncommon malignant esophageal neoplasm characterized by a diffuse mixture of squamous and mucus secreting glandular carcinoma cells. The diagnostic challenges are associated with the distinction between adenosquamous carcinoma and MEC. Here we report a case of 60 year old male with dysphagia. Due to its rarity, the prognosis of this disease has not been well established.

Keywords: Mucoepidermoid Carcinoma, esophagus, diagnostic challenge.

INTRODUCTION

Mucoepidermoid carcinoma (MEC) is the most common malignant neoplasm in the major and minor salivary glands [1]. It may also arise in other organs, including the bronchi, lacrimal sac, thyroid gland and, rarely, the esophagus. Mucoepidermoid carcinoma of the esophagus (EMEC) is an unusual variant of esophageal cancer that comprises a mixture of mucus-secreting adenocarcinoma with occasional duct-like structures and squamous cell carcinoma [2]. It is a very rare malignancy, with an incidence of less than 1% of all esophageal

carcinomas (3). It is characterized by poor clinical recognition, pre-operative diagnostic challenges and a lack of standardized therapeutic guidelines.

CASE REPORT

60 year old male presented with dysphagia. On upper GI endoscopy a nodule with ulceration around 1 cm in diameter was seen in the esophagus 30 cms from the incisors. Endoscopic biopsy was received as seven grey white to grey brown soft tissue pieces approximately measuring 0.5 cm³. On microscopy, there were separately lying fragments of variably acanthotic stratified squamous epithelium along with fragments of tumor comprised of closely packed nests of intermediate epithelial cells having uniform to mildly pleomorphic vesicular nuclei, fine chromatin, inconspicuous nucleoli and moderate amount of eosinophilic finely vacuolated cytoplasm with interspersed mucous cells. Few glands with intraluminal mucous and occasional nests of squamoid/epidermoid cells were also seen and hence, the histopathological diagnosis of Esophageal Mucoepidermoid Carcinoma of intermediate grade was given

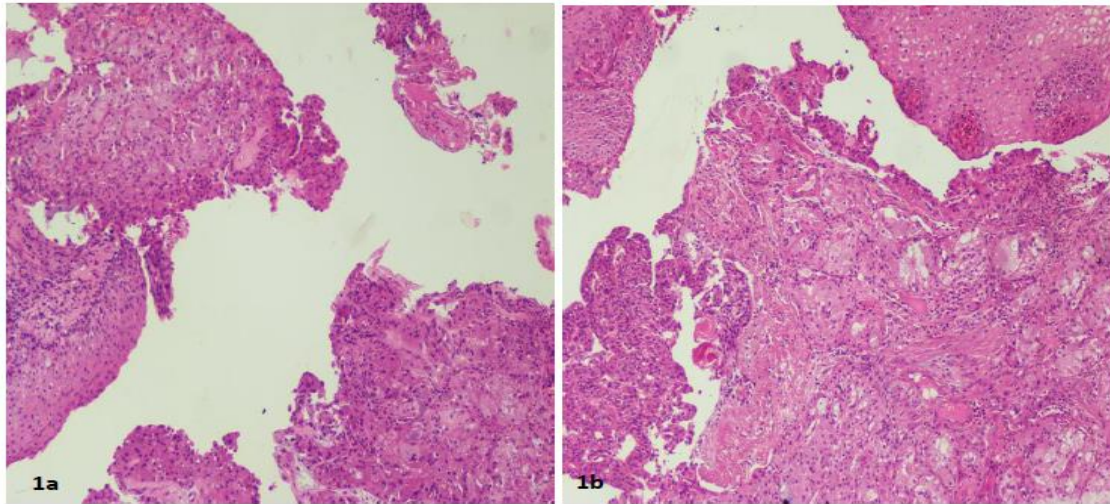


Figure 1a & b: Low power view of esophageal biopsy revealing a separately lying acanthotic stratified squamous epithelium along with tumor tissue fragments comprised of nests of intermediate epithelial cells with interspersed mucus cells (H&E, 100X)

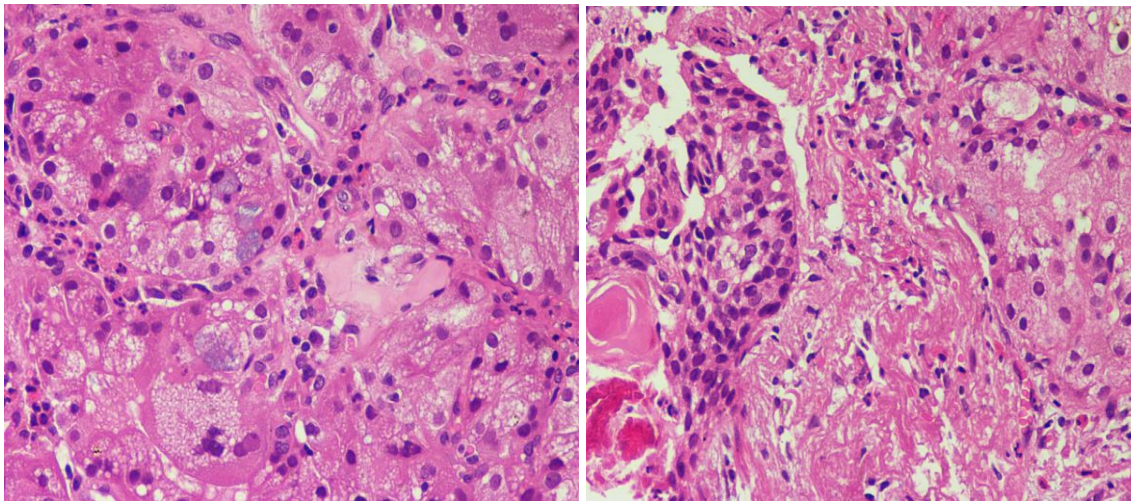


Figure 2a & b: High power view of esophageal biopsy revealing nests of intermediate epithelial cells, squamoid cells and mucous cells (H&E, 400X)

DISCUSSION

Primary mucoepidermoid carcinoma (MEC) of the esophagus is an uncommon malignant esophageal neoplasm characterized by a diffuse mixture of squamous and mucus secreting glandular carcinoma cells [3]. EMEC occurs mainly in men in the sixth decade. MEC predominantly involves the middle and lower thirds of the esophagus[4], while the exact micro-anatomic origins of these tumors are still debatable.

The hypothesis that these tumors arise from the esophageal glandular or ductal epithelium [5] is supported by their submucosal location, microscopic findings of normal stratified squamous epithelium overlying the EMEC [6] and the common

embryological derivation of esophageal and salivary glands [6]. Some authors also believes that EMEC arises through dysplasia in metaplastic surface of squamous epithelium [7].

Most common clinical presentation is dysphagia and weight loss.

Radiologically, tumor shows constricting or stenotic appearance with or without ulceration [8]. It is difficult EMEC from squamous or adenocarcinoma on endoscopy. In the review article by Kumagai et.al.[9], only 14.3% of the patients were diagnosed with EMEC by endoscopic biopsy, and most of them (77.1%) were misdiagnosed with Esophageal Squamous cell carcinoma (ESCC). Chen et al [10]in a review of 20

patients with EMEC diagnosed over a 20 year period demonstrated a 100% false pre-operative diagnoses; 18 were misdiagnosed as squamous cell carcinoma, and the remaining 2, as adenosquamous carcinoma. Diagnostic difficulties are attributed to poor attention to the exact anatomic location of the tumor, surface epithelial dysplastic alterations and the range of tumors that contain squamous and glandular elements [11]. On microscopy there are mucous, squamous, and intermediate cells in variable amounts in solid sheets or lining spaces filled with mucinous secretions. Mucous cells are cubical, columnar, or goblet shaped with intracytoplasmic mucin collections. Squamous cells show keratinization and intercellular bridges with occasional pearl formation. Intermediate cells are smaller, forming stratified lamina lining mucin spaces.

The diagnostic challenges are associated with the distinction between adenosquamous carcinoma and MEC. Recognition of the squamoid and glandular components on endoscopic esophageal biopsies is critical. The identification of intermediate cells may be an unrealistic expectation in small biopsies, and histochemical mucin stains are not helpful in differentiating between MEC and adenosquamous cell carcinoma.

In the extrasalivary MECs, advanced squamous differentiation and pleomorphism of squamous, glandular and solid cell growth have been documented [3-5,7]. The morphological features of MEC that may help in their diagnostic distinction from adenosquamous carcinoma includes the presence of mucin pools, extravasation of mucin, intermediate cells and the deep location of the tumor. While the absence of surface epithelial dysplasia is a helpful diagnostic feature, its identification does not exclude MEC [11]. Because of the glandular and squamous co-differentiation of MEC IHC markers of glandular or squamous origins are not helpful in the distinction. There is variable S100 protein staining in salivary MEC [12]. However, S100

immunopositivity in an esophageal tumor with glandular and squamous elements may favor a mucous glandular origin and a diagnosis of MEC. Some authors report it to have an aggressive course while some state it to have favourable prognosis.

CONCLUSION

Primary MEC of esophagus is a rare esophageal carcinoma and it is often misdiagnosed as adenosquamous carcinoma. Due to its rarity, the prognosis of this disease has not been well established. Therefore, further studies are needed to ascertain the management and prognosis of esophageal MEC.

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