PRIMARY MUCOEPIDERMOID CARCINOMA OF THE ESOPHAGUS - A CASE REPORT

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Abstract
Primary mucoepidermoid carcinoma (MEC) of the esophagus is an uncommon neoplasm account for 0.05 to 2.2 percent of all cases of primary oesophageal cancer. Diagnosis depends on precise microscopic criteria characterized by a diffuse mixture of squamous and mucus-secreting glandular carcinoma cells. It is believed to arise from the submucosal glands or ducts or from the stratified squamous epithelium of the esophagus. Mucoepidermoid carcinoma of the esophagus is extremely aggressive and has a poor prognosis. The poor prognosis of esophageal MEC may be caused by high proliferative and metastatic potential. Treatment for this tumor has primarily been surgical resection. One such rare tumor was reported at the Institute of pathology madras medical college is being published for its rarity.

Keyword: Mucoepidermoid carcinoma, esophagus, primary

INTRODUCTION:
Mucoepidermoid carcinomas occur commonly in the salivary, lacrimal and tra...
Figure 1: Gross appearance
Gross features: Transhiatal esophagectomy specimen of 9.5 cm esophagus length with 5 cm length stomach was received. On opening the esophagus, esophagogastric junction showed a friable ulcerative circumferential growth measuring 3x2x3 cm. Cut section appeared solid gray white, extending up to the muscle coat. Adjacent gastric mucosa appeared flattened [Figure 1]. Microscopic features: Section showed esophageal mucosa with a neoplasm underneath composed of sheets, islands and trabeculae of malignant squamous epithelial cells exhibiting moderate to marked pleomorphism with areas of necrosis intimately mixed with mucin filled cystic spaces lined by cuboidal epithelium and intermediate type of cells in cords and trabeculae. The neoplasm extended from the mucosa to the subserosa and into the adjacent gastric mucosa. Proximal and distal resected margins are free of tumor infiltration [Figures 2 to 5]. Two lymph nodes dissected are negative for tumor deposits. Special stains: Alcian Blue - PAS and Mucicarmine stains were done to demonstrate the presence of sialomucins in the mucus secreting elements of the tumor [Figure 6]. A Diagnosis of Primary Mucoepidermoid carcinoma of esophagus was made.
DISCUSSION

Mucoepidermoid carcinomas occur commonly in the salivary, lacrimal and tracheobronchial glands. However, they have been reported in the oesophagus also very rarely. Clinical features including age, sex symptoms at presentation, site distribution, length of tumor, and appearance of tumor did not differ from those of squamous cell and adenocarcinoma. The mean age was 64 years and the male to female ratio was 4.5:1. The mean diameter of these tumours was 4 cm. 27% of these tumours were in the upper, 46% in the middle and 27% in the lower portion of the oesophagus. World Health Organization classification defines mucoepidermoid carcinoma as a tumor composed of an "intimate" mixture of squamous and mucin-secretting elements, and an adenosquamous carcinoma as a tumor that shows the two cellularelements separate from, but "intermingled with," each other, clearly some tumors show both of these features.

Mixed squamous and glandular differentiation is noted in some esophageal carcinomas. Usually the mixture is morphologically and diagnostically insignificant: a squamous cell carcinoma may contain a negligible glandular or mucus-secreting element; or, conversely, an adenocarcinoma can incorporate small, bland squamous foci. Areas showing glandular or mucus-secreting differentiation were in greater part located in the submucosa and the lamina propria mucosae, thereby suggesting an origin from submucosal esophageal glands or their ducts. Because of intraepithelial spread of the tumor and the close relationship between foci of invading carcinoma and regions of dysplastic epithelium, it is also suggested neoplastic transformation of a totipotent primitive stem cell in the basal region of the squamous epithelium that undergoes heterogeneous differentiation within a single tumor. The glandular component of this group of tumors histochemically differentiated in the direction of oesophageal glands: examination of the mucin secretory component in squamous cell carcinoma in resected specimens is therefore required for recording the true incidence of this type of tumor. The mucin produced by these tumors was mixed and included a variable content of enzyme labile sialomucin (positive for mucicarmine, periodic acid Schiff, and alcian blue). Electron microscopy shows features of both adenocarcinoma (The intracellular microcysts an ultrastructural marker of adenocarcinoma) and squamous cell carcinoma (bundles of keratin filaments). Some tumor cells in mucoepidermoid...
carcinomas may be positive with immunohistochemical stains for CEA [Carcino Embryonic Antigen], SC[Secretory Component], or lactoferrin. Differential diagnosis: 1. Adenosquamous carcinomas: These are rare neoplasms confused histologically with mucoepidermoid carcinoma in which, the adenocarcinomatous component is usually tubular. The squamous and adenocarcinomatous components are more clear-cut with some areas of purely glandular epithelium and mucin with increased nuclear pleomorphism. Occasional keratin pearls enable a differentiation to be made. These tumors have commonly been reported in association with Barrett esophagus, but can also occur de novo. Mucoepidermoid carcinoma is also composed of both squamous and glandular elements, but displays a more intimate admixture and no clear distinction between the squamous and adenocarcinoma components in the tumor nests compared to adenosquamous carcinoma, with islands of squamous carcinoma with mucus-secreting cells and occasional ductlike structures. 2. Collision tumor: These tumors are represented by a merging of two originally separate squamous cell carcinoma and adenocarcinoma. CONCLUSION: Primary esophageal mucoepidermoid carcinoma is a rare disease and prone to be misdiagnosed. Lymph node metastasis and operation are independent prognostic factors. Surgical resection is the primary treatment, but the prognosis poor 7.

REFERENCES: