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IMAGE OF THE MONTH

Primary acinic cell carcinoma of the esophagus

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A 64-year-old female suffered from retrosternal discomfort, and occasional belching and dysphagia for 6 months. The medical record are not otherwise specific, with a history of chronic atrophic gastritis. Endoscopic examination revealed a hemispherical protruding lesion (1.5×1.0 cm) locating at distal esophagus (34 cm from incisor). Endoscopic submucosal dissection (ESD) was applied for resection of the esophageal lesion. The final pathologic diagnosis was a primary esophageal acinic cell carcinoma (ACC) with a clear margin. The patient did not show relapse of the disease during the 29 months follow-up.

The tumor invaded submucosal layer under light microscopy. Normal esophageal squamous epithelium and proliferating lymphoid tissue were found at the periphery of the tumor. It was composed of major acinar structure and minor ductal structure that filled with eosinophilic secretion. The tumor cells were round or oval, with centrically or eccentrically located nuclei, visible nucleoli, and eosinophilic or vacuolated cytoplasm (Fig. 1). Pleomorphism, anaplasia, lymphovascular invasion, mitotic figures, and necrosis were not identified. The tumor cells were strongly positive for CEA, CK7, and CK19 and negative for TTF-1, p63, S-100, Mammaglobin, MUC5b, MUC5ac, Syn, CgA, CD56, and DOG1. The percentage of proliferative cells that stained by Ki-67 was 2% (Fig. 2). Periodic acid-Schiff-Diastase (PAS-D) stain showed scattered weak positivity.

Salivary-type ACC arising outside the head and neck is extremely rare. To date, approximate 20 cases of primary pulmonary ACCs have been reported [1]. They originate from tracheal or bronchial submucosal glands. The esophagus also has submucosal glands distributed, but none primary esophageal ACC has been reported. Esophageal submucosal glands have almost pure mucous acini interspersed with a minimal number of serous cells. However serous cells could show hyperplasia in certain circumstances such as gastroesophageal reflux disease, on which esophageal ACC may occur. The patient had a history of chronic atrophic gastri-

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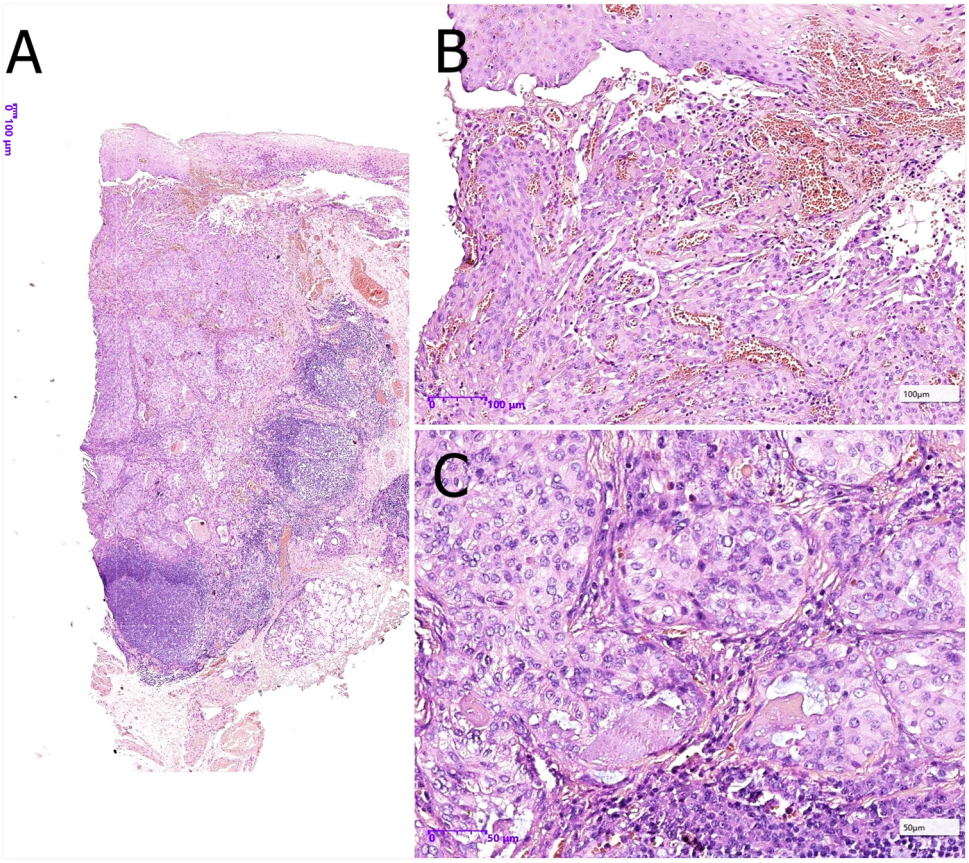

Fig. 1 Microscopical features of the esophageal ACC. A. The tumor was not well circumscribed with peripheric interstitial lymphocytic infiltration and aggregation. B. It was located under normal esophageal squamous epithelium without morphological transition. C. It was composed of dense acinar structures and minor ductal structures filled with eosinophilic secretion.

tis and a symptom of belching that implies the existence of gastroesophageal reflux disease.

The presence of intracytoplasmic PAS-D positive granules has been described as a characteristic finding of ACC, however there are reports of an absence or weak positivity in some pulmonary ACCs [2]. The tumor cells showed scattered weak positivity of PAS-D stain, which could be explained by the fact that serous cells of esophageal submucosal gland do not produce abundant digestive zymogen, instead, they secrete solute and fluid with low concentration of proteins and peptides [3]. Negative staining of DOG1 may arouse dispute, but it is not entirely positive in ACCs [4].

Differential diagnosis is important since the result decides subsequent patient management. Primary esophageal adenocarcinoma and secretory carcinomas (SC) should be considered in differential diagnosis [5]. The former usually has prominent nuclear pleomorphism, anaplasia, and mitotic figures, which are not detected in our case. In addition, the mucin markers are negative. SC has been described with similar morphology but distinct immunohistochemical and molecular features [6]. In the present case, absence of papillary-cystic growth pattern and negative staining of Mammaglobin and S100 exclude the diagnosis of SC [4,7]. ACC is a low-grade carcinoma with a low rate of recurrence and metastasis, therefore ESD is the optimal treatment to the patient, and subsequent esophagectomy is not required.

Author's contribution

Ling Nie: collection of data, manuscript preparation and writing.
Qi Wang: collection of data.
Fanqing Meng: review of the manuscript.
Final approval of the manuscript: all authors.

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Disclosure of interest

The authors declare that they have no competing interest.

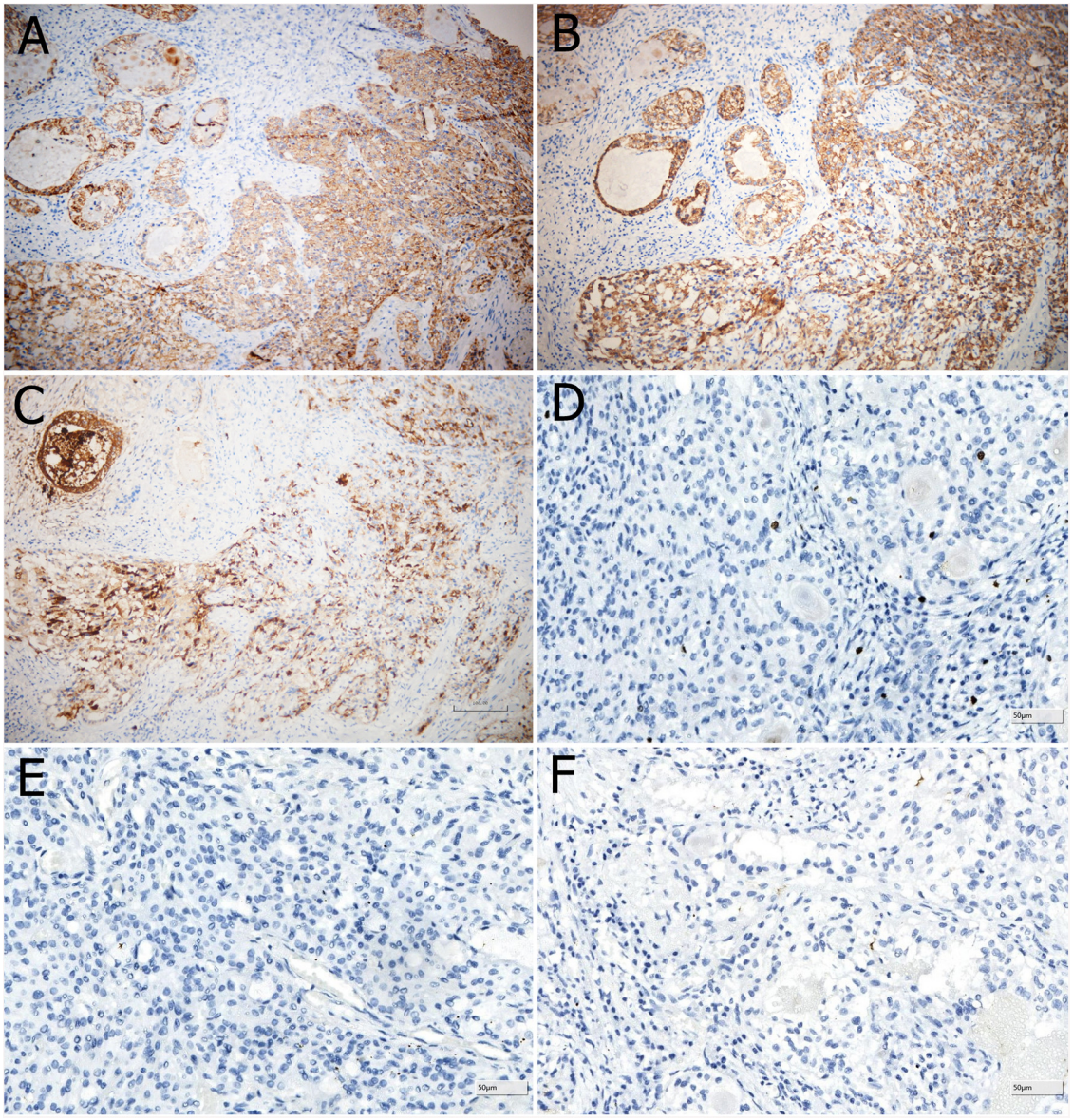

Fig. 2 Immunohistochemical results of the esophageal ACC. A, B, and C. The neoplastic cells were positive for CK7, CK19, and CEA. D. Ki-67 index was low. E, F. p63 and MUC5b were negative staining.

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