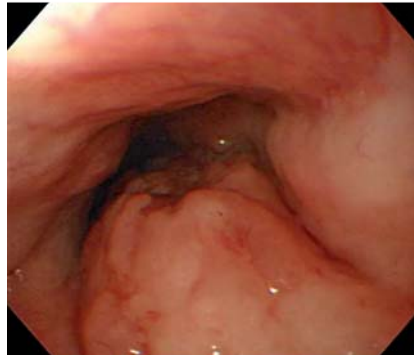


## Primary pure signet ring cell adenocarcinoma of non-Barrett's esophagus

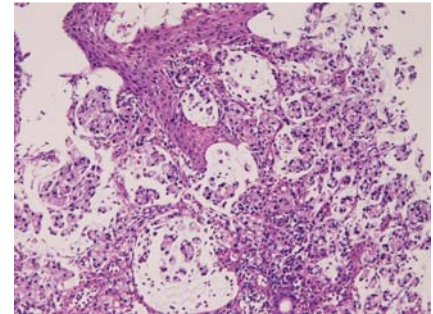
Primary signet ring cell adenocarcinoma (SRCA) of the esophagus is very rare, with only two case reports [1,2] and one case series [3] in the published literature. Most primary SRCAs arise in Barrett's esophagus [2,3], with occurrence in non-Barrett's esophagus being extremely rare [1]. Moreover, primary pure SRCA (i.e., composed only of SRCA cells) has not been reported in the literature.

A 73-year-old man attended our hospital because of dysphagia. Upper gastrointestinal endoscopy showed a tumor in the mid and lower esophagus (● Fig. 1). Histological examination of the biopsy specimen showed SRCA (● Fig. 2). No Barrett's esophagus was found and there was no evidence of tumor in the stomach and duodenum. Esophagectomy was carried out, and gross examination of the resected esophagus revealed a tumor measuring 5 × 5 × 1 cm in the mid and lower esophagus (● Fig. 3). No Barrett's esophagus was seen. Microscopically, the tumor was pure SRCA, with mature squamous epithelium of the esophagus seen in some areas (● Fig. 4). Histochemically, the tumor cells were positive for mucins. An immunohistochemical study carried out using Dako's EnVision method as previously described [4,5] revealed the SRCA cells were positive for cytokeratin (CK) AE1/3, CK CAM5.2, CK18, CK19, CK20 (● Fig. 5a), CEA, CA19-9, CDX-2, MUC2 (● Fig. 5b), and MUC5AC. They were negative for CK34βE12, CK5/6, CK7, CK8, CK14, vimentin, MUC1, and MUC6. There was distant metastasis and the prognosis was considered poor. The patient died of carcinomatosis 15 months after the operation.

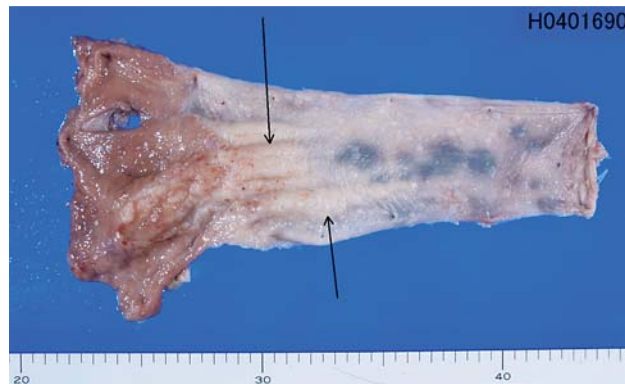
The pathogenesis of the present primary SRCA remains unclear. Clearly, the SRCA did not arise from the squamous epithelium because no squamous cell carcinoma was seen. In the esophagus, esophageal glands may give rise to mucoepidermoid carcinoma and adenoid cystic carcinoma. We speculate that in the present case, the carcinoma was the result of malignant transformation of the cells of the esophageal glands. There have been no immunohistochemical studies of primary SRCA of the esophagus and the immunohistochemical profile reported here may



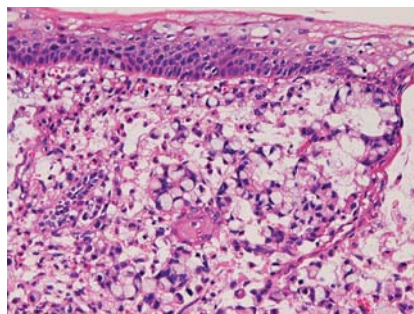
**Fig. 1** Esophageal endoscopy showing a tumor in a 73-year-old man with dysphagia.



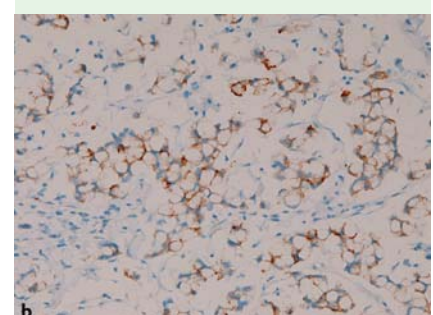
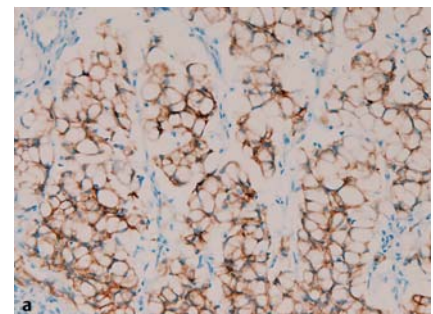
**Fig. 2** Biopsy findings: signet ring cell carcinoma cells are seen along with non-neoplastic squamous epithelium of the esophagus (hematoxylin and eosin, magnification × 40).



**Fig. 3** The resected esophagus with tumor measuring 5 × 5 × 1 cm (arrows) is seen in the mid and low esophagus.



**Fig. 4** Histological section of the resected esophagus. Signet ring carcinoma cells are arranged in a medullary pattern with non-neoplastic squamous epithelium near the surface (hematoxylin and eosin, magnification × 100).



**Fig. 5** Immunohistochemical findings: the signet ring adenocarcinoma cells were positive for cytokeratin 20 (a) and MUC2 (b) (immunostaining, magnification × 200).

contribute to initial knowledge of pure SRCA of the esophagus. Survival in primary SRCA is poor [3], as was also in the case reported here.

Endoscopy\_UCTN\_Code\_CCL\_1AB\_2AC\_3AB

**Competing interests:** None

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