

Mixed adenoneuroendocrine carcinoma (MANEC) of the esophagogastric junction predominantly consisting of poorly differentiated neuroendocrine carcinoma

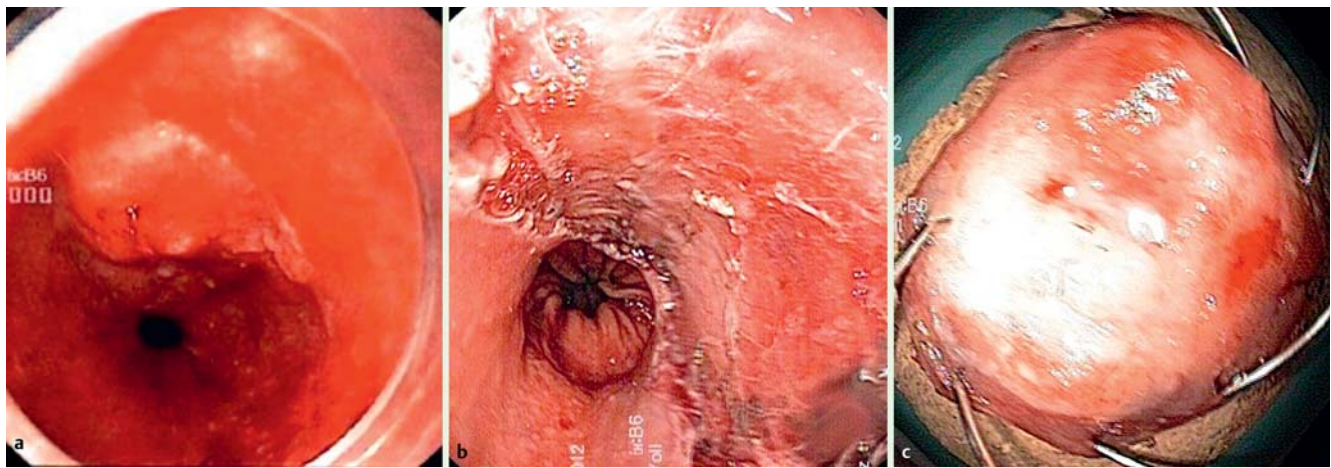


Fig. 1 Endoscopic view in a 68-year-old man presented with a rapidly growing mass at the esophagogastric junction. **a** Overview. **b** The tumor bed after resection. **c** Macroscopic view of the resected specimen after removal (courtesy of H. Schulz).

A 68-year-old man presented with a rapidly growing mass at the esophagogastric junction. The tumor was initially biopsied and diagnosed as well-differentiated Barrett's carcinoma. Subsequently the tumor was resected with endoscopic submucosal dissection (Fig. 1). On histopathological examination, the tumor showed two distinct components with abrupt transition (Fig. 2). The part of the tumor in the superficial mucosal layer was a well-differentiated adenocarcinoma, resembling common Barrett's carci-

noma. The underlying component consisted of a poorly differentiated small round blue cell tumor that made up about 80% of the entire tumor mass. On immunohistochemistry this component was strikingly positive for synaptophysin and showed a Ki67 proliferation rate of >95%, which confirmed it as poorly differentiated neuroendocrine carcinoma. Within the adenocarcinoma, only a scattering of synaptophysin-positive cells were visible. The results for CK7 were exactly the opposite as the adenocarcinoma was strongly

positive for it and the neuroendocrine carcinoma almost negative (Fig. 3), with the neuroendocrine carcinoma infiltrating at least the lower third of the submucosal layer (sm3) and reaching the basal resection margin. Therefore, according to the current World Health Organization (WHO) criteria the final pathological diagnosis was poorly differentiated mixed adenoneuroendocrine carcinoma (MANEC) of the esophagogastric junction.

MANECs are exceedingly rare, because of which there are no recommended therapeutic strategies; however, according to WHO, surgery is the treatment of choice [1]. These tumors often occur, as in our case, in association with Barrett's carcinoma, but combinations with squamous cell carcinoma have also been reported [2, 3]. It is believed that MANECs have a slightly better prognosis than pure neuroendocrine carcinomas in the same location, but which unfortunately is very poor (<6 months overall survival rate [4]). In conclusion, clinicians should have a high index of suspicion for MANEC in cases with a diagnosis of Barrett's carcinoma when a rapidly growing tumor is reported.

Endoscopy_UCTN_Code_CCL_1AB_2AC_3AB

Competing interests: None

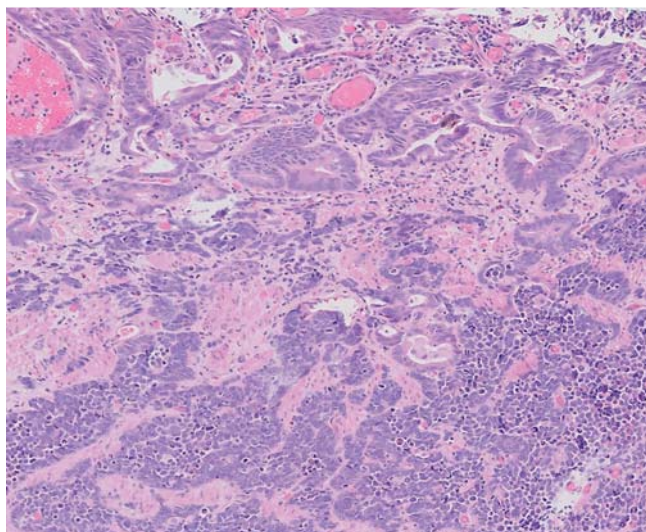


Fig. 2 Abrupt transition between neuroendocrine carcinoma and adenocarcinoma (hematoxylin and eosin, magnification $\times 100$).

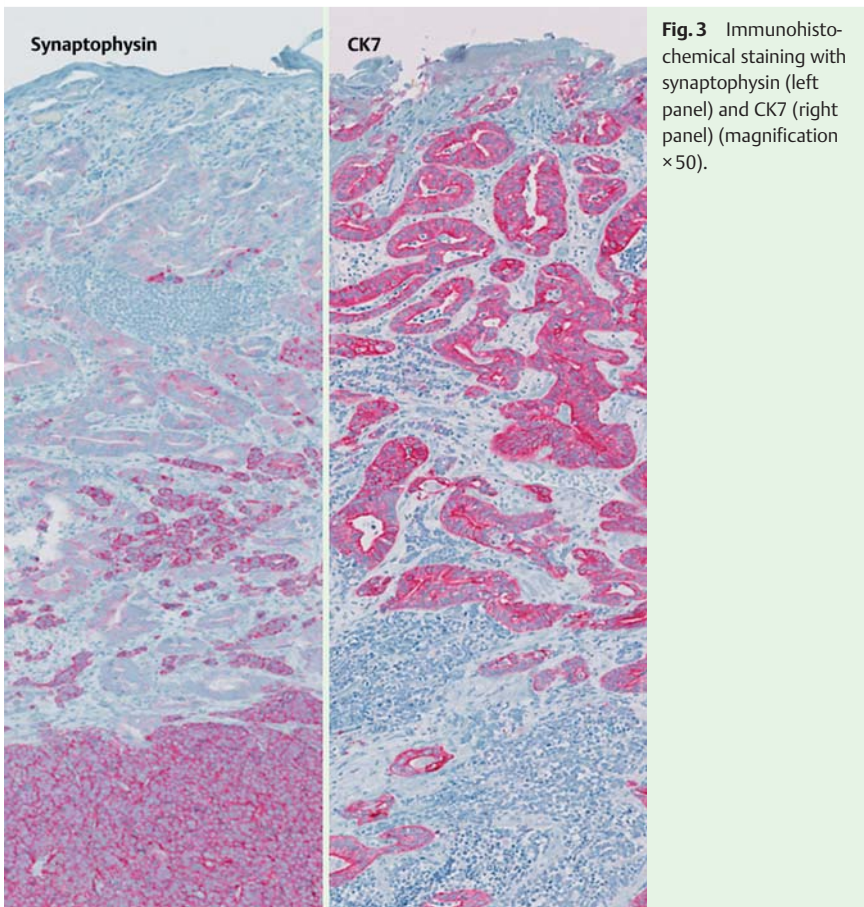


Fig. 3 Immunohistochemical staining with synaptophysin (left panel) and CK7 (right panel) (magnification $\times 50$).

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