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

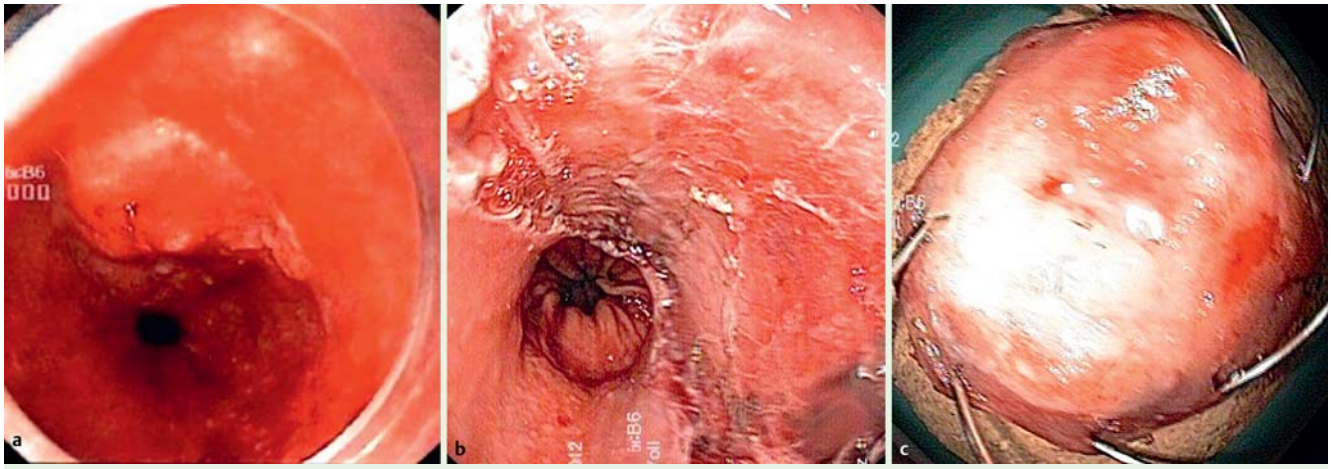

Fig. 1 Endoscopic view in a 68-year-old man presented with a rapidly growing mass at the esophagogastric junction. **a** Overview. **b** The tumor bed after resection. **c** Macroscopic view of the resected specimen after removal (courtesy of H. Schulz).

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 carcinoma. 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.

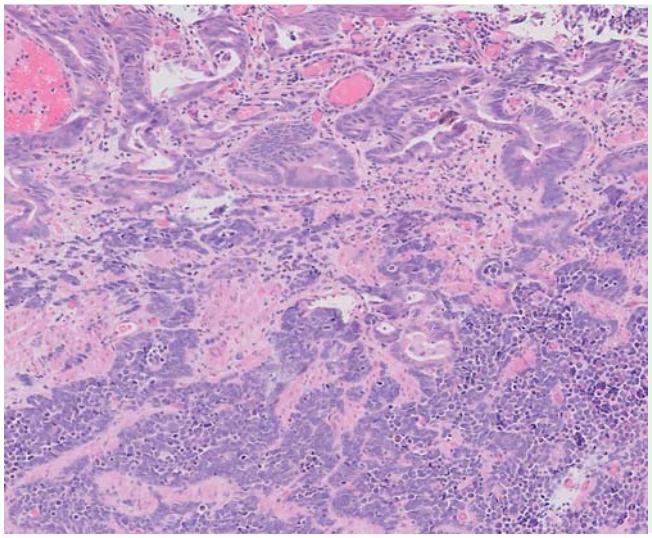

Fig. 2 Abrupt transition between neuroendocrine carcinoma and adenocarcinoma (hematoxylin and eosin, magnification × 100).

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

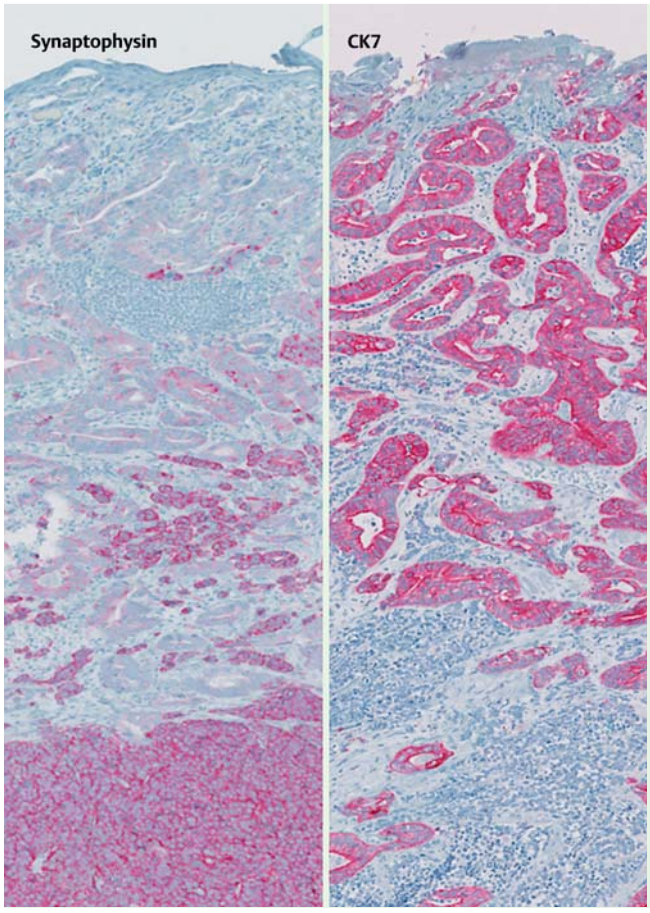

Fig. 3 Immunohistochemical staining with synaptophysin (left panel) and CK7 (right panel) (magnification ×50).

L. Veits[1], C. Lang-Schwarz[1], H. Volkholz[1], C. Falkeis[1], M. Vieth[1], H. Schulz[2]

[1] Institute of Pathology, Klinikum Bayreuth, Bayreuth, Germany
[2] Gastroenterologist, Private Practice, Bremen, Germany

References

1 *Bosman FT, Carneiro F, Hruban RH* et al. (eds). WHO Classification of Tumours of the Digestive System. Lyon: IARC; 2010
2 *Cary NR, Barron DJ, McGoldrick JP* et al. Combined oesophageal adenocarcinoma and carcinoid in Barrett's oesophagitis: potential role of enterochromaffin-like cells in oesophageal malignancy. Thorax 1993; 48: 404 – 405
3 *Maru DM, Khurana H, Rashid A* et al. Retrospective study of clinicopathologic features and prognosis of high-grade neuroendocrine carcinoma oft he esophagus. Am J Surg Pathol 2008; 32: 1404 – 1411
4 *Law SY, Fok M, Lam KJ* et al. Small cell carcinoma of the esophagus. Cancer 1994; 73: 2894 – 2899

Bibliography

DOI http://dx.doi.org/10.1055/s-0032-1326113
Endoscopy 2013; 45: E16–E17

Stuttgart · New York
ISSN 0013-726X

Corresponding author

L. Veits
Institute of Pathology
Klinikum Bayreuth
Preuschwitzerstraße 101
95445 Bayreuth
Germany
Fax: +49-921-4005609
lothar.veits@klinikum-bayreuth.de