

Primary malignant melanoma of the oesophagus

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An 80-year-old woman presented with a history of dysphagia and a 13 kg weight loss in the last 2 months. She had no particular medical history. The physical examination was unremarkable. Blood test showed an iron deficiency anaemia. A gastroscopy was performed and showed a voluminous protruding polypoid purple-black lesion in the lower third of the oesophagus, extending towards the gastric cardia (Figures 1 and 2). Biopsies were performed during initial endoscopy and histology revealed a malignant melanoma with expression of S100 and SOX10 on immunohistochemistry with no mutations detected (BRAF, NRAS, c-KIT). A complete clinical work-

up, including endoscopic ultrasound, total body computed tomography (CT)-scan, PET-CT-scan, cerebral magnetic resonance imaging, dermatologic and ophthalmologic examination, did not show any other suspect lesions. Thus, the patient was addressed to surgery and the surgical team decided to treat as a Siewert type 3 tumour (total gastrectomy and distal oesophagectomy with an eso-jejunal

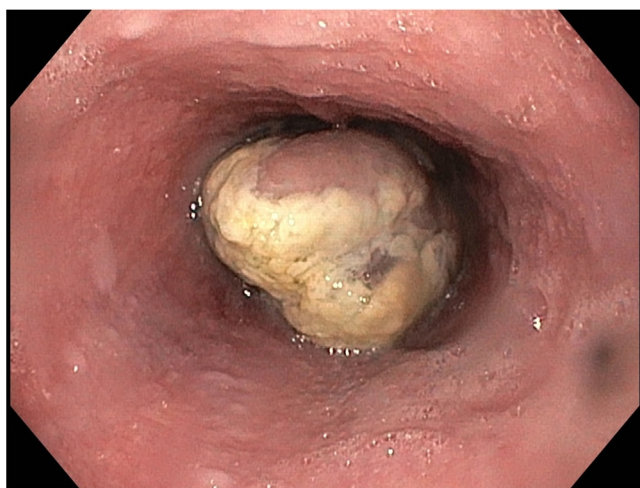


FIGURE 1 Visualisation of the oesophageal mass, upper endoscopic approach.

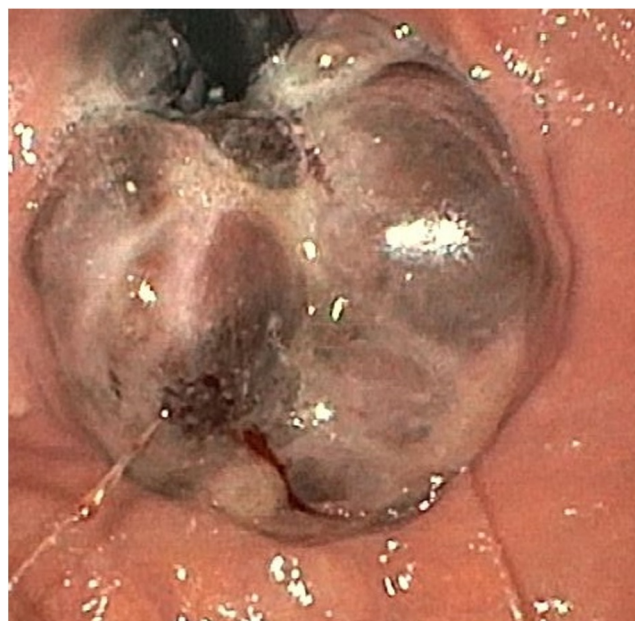


FIGURE 2 Macroscopic resected gastrectomy specimen.

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reconstruction) by robotic surgery according to their experience and recommendations 1 month after diagnosis.

The pathological examination was consistent with a primary malignant melanoma of oesophagus (Figure 3–6). Follow-up after 3 months including PET-CT and endoscopy did not show any recurrent lesion. Adjuvant immunotherapy was proposed but refused by the patient.

Primary malignant melanoma of oesophagus is exceedingly rare and has a reported prevalence of 0.1%–0.3% of all oesophageal malignancies.¹ These tumours usually appear as lobulated and darkly

coloured masses with intact mucosa or occasional ulceration, mostly located in the middle or distal third² of the oesophagus probably because of the greater concentration of melanocytes in these regions.³ The diagnostic criteria require the presence of melanin granules within the tumour cells as well as melanocytes in the overlying epithelial layer.⁴ The diagnosis is confirmed by immunohistochemistry revealing a positive antibody-specific cytoplasmic reactivity to S-100 proteins. The diagnosis of primary oesophageal melanoma can be accepted only in patients with no history of melanoma and no evidence of melanoma involving the skin, eye, anus, or vagina.^{4,5}

The endoscopic presentation is highly specific and should immediately suggest the diagnosis of oesophageal melanoma, enabling the pathologist to direct his immunohistochemical examination and thus save time in confirming the diagnosis, helping to reduce the risk of dissemination.



FIGURE 3 Visualisation of the oesophageal mass—retroflexed endoscopic approach.

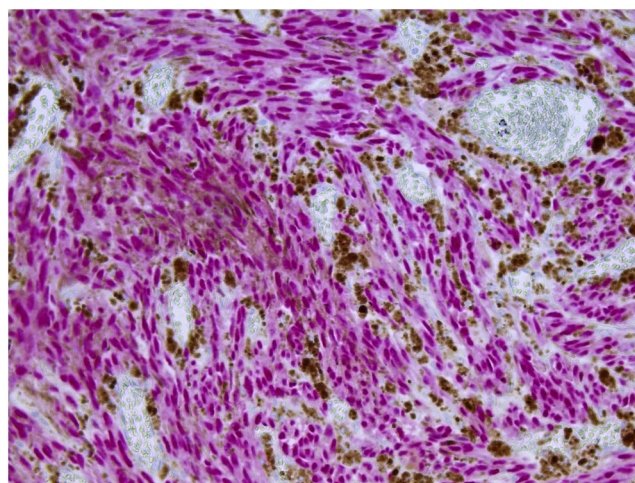


FIGURE 5 Microscopic histopathologic figure—SOX 10 colouration.

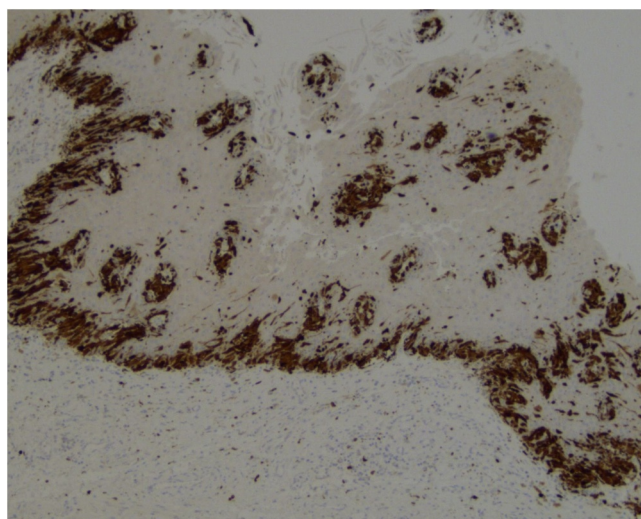


FIGURE 4 Microscopic histopathologic figure—S100 colouration.

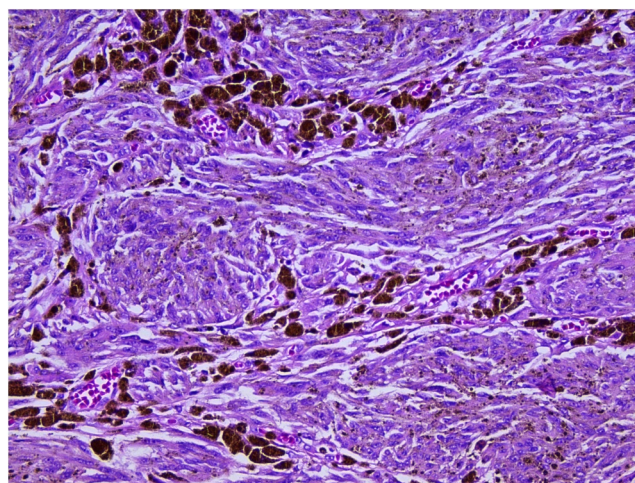


FIGURE 6 Microscopic histopathologic figure—HES IV colouration-X200. HES, Hématoxyline - Eosine - Safran coloration.

CONFLICT OF INTEREST STATEMENT

The authors declare no conflicts of interest.

DATA AVAILABILITY STATEMENT

The data that supports the findings of this study are available in the supplementary material of this article.

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