

Primary malignant melanoma of the cervical oesophagus – a rare tumour in a very rare location

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Summary

We report a case of primary malignant melanoma arising most unusually in the cervical oesophagus. A 78-year-old woman presented with an eight-month history of progressive shortness of breath and stridor, and an enlarging neck lump associated with dysphagia. Apart from a multinodular goitre and a parathyroid adenoma, investigations revealed a polypoid mass in the cervical oesophagus close to the cricopharynx.

Biopsy of the lesion showed a primary malignant melanoma. No pigmented lesions of the skin or eyes were found.

Endoscopic debulking of the tumour was performed and the patient received palliative percutaneous radiotherapy.

Although 238 cases of primary oesophageal malignant melanoma have been reported in literature, none of these malignancies was localised in the cervical oesophagus.

Key words: melanoma, oesophagus

Streszczenie

Przedstawiono przypadek pierwotnego czerniaka złośliwego, niezwykle rzadko wyrastającego z szyjnego odcinka przełyku. Choroba wystąpiła u 78-letniej pacjentki z 8-miesięcznym wywiadem postępującej zadyszki, świstu oddechowego i rosnącego guza szyi, z towarzyszącymi zaburzeniami połykania. Oprócz woli wieloguzkowych tarczycy i gruczolaka przytarczyc u chorej stwierdzono polipowatą masę w szyjnym odcinku przełyku niedaleko krtani. Biopsja zmiany chorobowej wykazała obecność pierwotnego czerniaka złośliwego. Nie stwierdzono żadnych zmian barwnikowych w obrębie skóry i oczu. Metodą endoskopową uzyskano zmniejszenie masy guza, a następnie chora została poddana paliatywnej, przezsłkórnej radioterapii.

Mimo że w literaturze 238 przypadków czerniaka złośliwego w obrębie przełyku, żaden z nich nie był zlokalizowany w jego szyjnym odcinku.

Słowa kluczowe: czerniak złośliwy, przełyk

Introduction

Primary malignant melanoma of the oesophagus (PMME) is an extremely rare tumour comprising 0.1-0.2% of all primary neoplasms of the oesophagus [1, 2]. It is more common in men and occurs in the sixth and seventh decades of life. PMME is regarded as a tumour of high grade malignancy because of its tendency to present as an advanced neoplasm with aggressive biological behaviour. The most favourable outcome is achieved with surgical resection which is rarely feasible for its progressed tumour stage at presentation [3]; however, radiotherapy, chemotherapy and immunotherapy might be considered palliative treatment options [3].

Case Report

A 78-year-old woman presented with an eight-month history of progressive shortness of breath, enlarging neck lump and associated mild dysphagia. Physical examination showed only evidence of a small goitre. On ultrasound scan, the goitre turned out to be multinodular and suspicion of a parathyroid adenoma was raised. A Sestamibi scan showed tracer retention in the upper pole of the right lobe of the thyroid, again suggestive of a parathyroid adenoma. This was confirmed by raised serum PTH and calcium levels.

A contrast swallow revealed an intraluminal cervical oesophageal lesion. Computed tomography gave evidence of a four-centimetre upper oesophageal mass obliterating the

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Fig. 1. CT scan demonstrating the oesophageal lesion compressing the trachea

lumen and at places compressing the trachea and ruled out metastatic spread (figure 1). On oesophagoscopy a five-centimetre polypoidal mass of varying consistency was found at the level of the cricopharynx, with several more distal mucosal skip lesions. Endoscopic biopsies confirmed a malignant melanoma. A bronchoscopy showed merely extrinsic compression of the trachea. A detailed examination of the eyes and skin did not reveal a primary melanoma site elsewhere.

The close proximity of the tumour to the cricopharynx, confirmed skip lesions and advanced age rendered surgical resection inappropriate. The patient underwent endoscopic debulking of the tumour to improve her swallowing and palliative radiotherapy thereafter.

Discussion

The first case of PMME was reported by Baur in 1906 [4]. PMME affects mostly men over 60 years of age. Ninety percent of PMMEs occur in the middle or distal oesophagus [5]. It accounts for 0.1 to 0.2% of all oesophageal tumours [1, 2] and to date only 238 cases have been reported in literature [3]. PMMEs are associated with dysphagia of short duration (73%), weight loss (72%), retrosternal or back pain (44%) or melaena (10%) [6]. Physical signs are uncommon and non-specific. Melanoma of the skin, eye, rectum and vagina should be excluded. The colour of the tumour varies from black to white in the amelanotic form, which accounts for 10-25% of all PMMEs. Satellite lesions are present in about 12% of the patients, sometimes distant to the primary tumour [2].

The diagnostic criteria for a PMME were defined by Allen and Spitz [7]:

- (1) A typical histological pattern of melanoma, the presence of melanin granules within the tumour cells

- (2) origin from squamous epithelium with junctional activity
- (3) junctional activity with melanotic cells in the adjacent epithelium.

A more accurate diagnosis is achieved by immunohistochemical staining, as PMME stains with HMB-45 antibody, S 100 protein and neuron-specific enolase, but not with cytokeratin or CEA [5].

Contrast studies or endoscopy show a lobular, polypoid, ulcerated predominantly intraluminal mass. Chest and upper abdominal computed tomography usually reveal an oesophageal mass. On endoscopic ultrasound (EUS), we conventionally find a hypoechoic or mixed echogenic mass. FDG-PET scan has a sensitivity in detecting mediastinal lymph node metastases varying from 83% for 6- 10 mm lesions to 100% for lesions greater than 10 mm [8].

Despite its poor prognosis, total or near total oesophagectomy is the preferred method of treatment in operable patients, yielding a longer survival than other treatment modalities [2]. Several cases with survival between five to twelve years have been described in patients who underwent total or near total oesophagectomy [9], whereas after local resection mean survival has been reported to be only 9 months [2]. The resection margin should be as long as possible because of the tendency of this tumour to spread longitudinally along the submucosa and the likelihood of satellite lesions [6].

Chemotherapy and immunotherapy do not have a major role in the management of PMME. In several cases, radiotherapy has been used in conjunction with surgery or alone as palliation. Radiotherapy does not seem to significantly affect survival; it is likely to be effective as adjuvant therapy in reducing the possibility of relapse in a subset of patients [2].

Approximately 85% of the patients die with disseminated disease; the common sites involved are the liver, lung, mediastinum, pleura, peritoneum and brain [2].

In our patient, surgical resection was considered inappropriate due to the close proximity of the tumour to the cricopharynx and the associated satellite lesions.

In conclusion, PMME is a very aggressive tumour regarded as a potentially systemic disease at the time of diagnosis, regardless of local extension. The longest survival is achieved by radical resection, although numbers in the literature are small. Radiotherapy, chemotherapy and immunotherapy may be considered for palliative treatment.

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