(18)FDG-PET-scan in staging of primary malignant melanoma of the oesophagus : a case report

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Abstract

Primary malignant melanoma of the oesophagus is a rare disease, only 262 cases being reported up to June 2005. In general, the prognosis is dismal because of its tendency to present as an advanced neoplasm with aggressive biological behaviour. (18)FDG-PET-scan is a useful tool for evaluation of metastatic disease and locoregional lymph node metastasis. We present herein a case of a young adult with primary malignant melanoma of the oesophagus, followed by a literature review on the subject. (Acta gastroenterol. belg., **2006**, 69, **12-14**).

Key words : malignant melanoma, oesophagus, PET-scan.

Case Report

A 35-year old man presented with a 2 month history of dysphagia, postprandial pain and 12 kg weight loss over that same period. Hematemesis and melena were denied. Medical history was negative but he consumed large quantities of alcohol up to 3 months before entry and there was heavy tobacco use. Physical examination revealed hepatomegaly with an irregular margin and a palpable epigastric mass. No pigmented skin, eye or anal lesions were evident. Laboratory analysis revealed a normocytemic normochromic anaemia and moderate elevation of lactate dehydrogenase, alkaline phosphatase and GGT, but normal liver transaminases.

A barium swallow revealed a large bulky tumour in the distal oesophagus (Fig. 1). Upper GI-endoscopy confirmed these findings with the tumour located from 30 to 40 cm of the incisors. The lesion was concentric and substenotic, with a heterogeneous surface, showing zones of moderate pigmentation (Fig. 2). There was no spontaneous bleeding but a remarkable mucosal friability was noted, readily bleeding after biopsy taking. Microscopic examination revealed a solid proliferation of poorly differentiated neoplastic cells, with vesicular nuclei and prominent nucleoli. There was pronounced anisonucleosis and some neoplastic cells showed melanin accumulation in the cytoplasm. Immunohistochemical stains for S-100-protein and HMB-45 were positive, consistent with malignant melanoma. (18)-FDG-PET-scan showed increased captation in the mediastinum, abdomen, left thigh and a brain localisation in the postero-parietal region (Fig. 3). A few days later, the patient was seen on the emergency department after a drop attack. Computed tomography confirmed multiple brain metastases. Computed tomography of thorax and abdomen further revealed massive adenopathies in the supraclavicular, mediastinal and retroperitoneal regions. More-over multiple liver metastases were present. High dose corticoid treatment was started together with pancranial radiotherapy (5×4 Gy). Two weeks later systemic chemotherapy with Dacarbazine (DTIC) in a dose of 800 mg/m^2 was started but the patient's disease was rapidly progressive and his general condition did not allow retreatment. His condition was further complicated due to bronchopneumonia and dilated cardiomyopathy, leading to pulmonary oedema. He died within 2 months of the initial diagnosis.

Discussion

Primary malignant melanoma of the oesophagus is a very rare condition accounting for 0,1 to 0,2% of all malignant tumours of the oesophagus. Volpin et al reported 238 cases in 2002 (1). Since then 24 additional case are reported to our knowledge (2-16). Our case represents most of the typical characteristics of malignant melanoma of the oesophagus, except for the age of presentation. Generally malignant melanoma is a disease of the elderly, most patients being in the 6th or 7th decade. Only rarely the disease is described in children (17) or young adults (18,19). Symptomatology is non specific, never suggesting the diagnosis on clinical grounds. Most patients complain of dysphagia, vague retrosternal pain and weight loss, as with other oesophageal tumours (20). The tendency of the tumours as to present as a large, bulky, non-ulcerating, polipoid or irregular mass, renders them easy to delineate with a barium swallow (21). Even with huge tumours, bleeding (hematemesis and/or melena) is a rare condition (4), although friability and bleeding during biopsy taking is generally excessive. Pigmentation is not necessarily present, making the interpretation of endoscopic biopsies sometimes difficult. Positive immunohistochemical stains for HMB-45 and S-100-protein support a diagnosis of malignant

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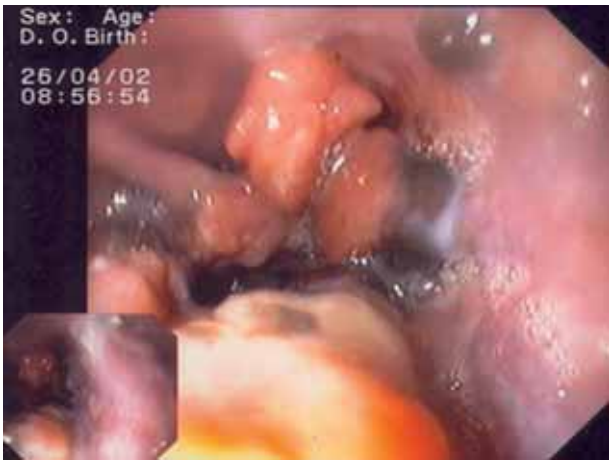

Fig. 1. — A barium swallow shows a large bulky tumour in the distal part of the esophagus.

FIGURE 2 MANQUE !

Fig. 2. — Endoscopic view of the upper part of the tumour with irregular protuberances and zones of moderate pigmentation.

melanoma. Since metastatic deposits to the oesophagus are more common than primary lesions, it is important to distinguish between primary and secondary lesions. To be acceptable as a primary melanoma, the tumour should be seen to arise from, or be surrounded by squamous epithelium, showing junctional change (22). Demonstration of an area of junctional change containing atypical melanocytes and pagetoid radial growth is not always possible on endoscopic biopsies, rendering the distinction between primary and secondary disease challenging. Ninety per cent of tumours are located in the distal two thirds of the oesophagus. The treatment of choice is radical surgery, requiring wide resection margins. Even then, long term survival is less than 5% (10). This type of tumour in general has a very aggressive biological behaviour, with 50% of patients having already metastases at the time of diagnosis, excluding them from curative treatment. In a series of 45 postmortem examinations, the most common sites of metastases were the liver (31%), the mediastinum and mediastinal lymph nodes (29%), the lung (17%) and the brain (13,2%) (23). The prognosis in these cases is very cumbersome because, in general, the tumour is resistant to radiotherapy and chemotherapy. Evaluation for metastatic disease and locoregional lymph node metastasis should be done in all cases with the appropriate radionuclide or computed tomographic scans. In primary malignant melanoma of the skin, (18)FDG-PET-scan is the functional imaging of choice for evaluation of suspected metastatic melanoma (24,25). This technique allows greater patient convenience, has lower radiation dosimetry and provides incremental and clinically important information in around 10% of patients as compared to 67-Ga-SPECT-scan. Immunoscintigraphy with indium-111-labeled monoclonal antibody and with technetium-99m-labeled melanoma monoclonal antigens are also able to identify most of metastatic deposits larger than 1cm in size (26,27). Due to the rarity of the tumour type, no comparative studies exist for primary malignant melanoma of the oesophagus. Kinuya et al reported good demonstration of metastatic lesions with Ga-67-SPECT-scan (28). In our case (18) FDG-PET-scan accurately predicted metastatic lesions in the abdomen, in the liver, in the mediastinum, in the skeleton and even in the brain.

FIGURE 3 MANQUE !

Fig. 3. — The (18)FDG-PET-scan shows increased captation in the mediastinum, in the abdomen, in the liver, in the left thigh and in the left postero-parietal region of the brain.

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