Primary malignant melanoma of the oesophagus with a left atrial metastasis

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Abstract

A 75 year old man with an eight month history of dysphagia and weight loss underwent pericardiocentesis for a massive pericardial effusion. The echocardiogram showed a mass in the left atrium, and computed tomography also showed distal oesophageal narrowing, which was found to be due to a malignant melanoma. The left atrial mass, investigated by immunoscintigraphy with technetium-99m labelled monoclonal antibody, was diagnosed as metastatic melanoma. This represents a rare case of primary oesophageal melanoma with left atrial metastasis.

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Primary oesophageal melanoma is a rare form of neoplasm that accounts for 0.1% of all oesophageal tumours.^{1,2} Since its first description by Baur in 1906 almost 100 cases have been reported, in only 10% of which was the melanoma primary.³

Malignant melanoma has a high incidence of myocardial and combined pericardial and myocardial metastases,⁴ the frequency of cardiac metastasis increasing with the number of distant metastases.

In this report we present the case of a man with a metastatic malignant melanoma in the left atrium whose primary tumour was in the oesophagus.

Case report

A 74 year old man was admitted to hospital with clinical signs of cardiac tamponade. Physical examination showed orthopnoea and enlarged, pulsatile neck veins; the liver was palpable 4 cm below the right costal margin and was also pulsatile. The heart rate was 150 beats/min and the blood pressure 80/50 mm Hg. An electrocardiogram showed atrial fibrillation and small voltage complexes. A chest radiograph showed a considerably enlarged cardiac silhouette and small opacities were noted in the right lower lobe. A two dimensional echocardiogram showed a large pericardial effusion and a large echogenic mass in the left atrium. The results of liver function tests were severely abnormal.

The patient gave an eight month history of mild dysphagia with solid foods, with weight loss of almost 10 kg.

Under echographic guidance an 8F pigtail catheter was introduced in the pericardial space through the left costoxiphoid angle, and 2650 ml of bloody fluid was drained in one hour. A sample was sent for chemical and cytological analysis.

Afterwards an echocardiogram showed a residual pericardial effusion of about 400 ml, lying mainly in the posterior pericardial space, and improved myocardial contractility. The pigtail catheter was left in place and a further 450 ml of fluid was drained over the next 48 hours. The pericardial fluid did not collect again and the catheter was removed 72 hours later.

Computed tomography identified a 3-4 cm narrowing of the distal oesophagus, two masses in the lower lobe of the right lung, and a large, non-thrombotic mass in the left atrium.

Endoscopy showed a 4 cm irregular mass in the distal oesophagus, and a biopsy specimen of this showed malignant melanoma cells. No skin lesions were found at physical examination but atypical epithelial cells were discovered in the pericardial fluid.

The patient was then referred for more detailed assessment and entered an experimental protocol that used ^{99m}Tc labelled antimelanoma monoclonal antibody (a fab fragment). Whole body and selected regional images were obtained with a gamma camera seven to eight hours after administration of the labelled antibody. There were two areas of increased uptake in the right lung corresponding to the abnormalities on the chest radiograph and the computed tomogram, and a focus of increased radioactivity in the heart as well as in the distal oesophagus (figure).

No surgical treatment was contemplated owing to the advanced stage of the disease, and the patient was referred for medical treatment.

Discussion

Primary melanoma of the oesophagus accounts for 0.1% of all primary oesophageal malignancies. There are no clinical differences from other forms of oesophageal cancer, and the histological diagnosis is often difficult, the submucosal site of the tumour being responsible for false negative histological and cytological appearances.⁵

The diagnostic criteria of a primary form of melanoma were clearly defined by Allen and Spitz⁶ as: (1) typical melanoma structure and presence of melanin; (2) origin from squamous epithelium with junctional activity; and (3) junctional activity with melanotic cells in the adjacent epithelium.

The prognosis is extremely poor with a four year survival of 4%. Malignant melanoma and oesophageal and breast cancer have the highest rate of metastasis affecting the heart,^{7,8} cardiac metastases occurring in up to 60% of patients with known melanoma.⁹ The

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Areas of increased uptake of technetium-99m labelled antimelanoma monoclonal antibodies in the right lung, the distal oesophagus, and the heart shown by whole body immunoscintigraphy.

diagnosis of cardiac metastasis is often difficult during life⁸ and in most cases it is discovered at necropsy.

As suggested by Waller,¹⁰ in patients with melanoma and without previously known heart disease one or more of the following signs suggest cardiac metastasis: (1) acute pericarditis; (2) pericardial effusion; (3) cardiac tamponade or constriction; (4) second or third degree atrioventricular block; (5) congestive heart failure.

The antibody imaging procedure represents a sensitive and relatively simple method for detecting the presence and extent of spread of malignant disease.^{11,12} The basis of this technique is the presence of a highly specific tumour associated antigen, the oncofetal antigen P97, which is strongly expressed on most melanoma cells, but only weakly on normal cells of human adult tissue. Consequently, by using a specific monoclonal antibody labelled with technetium-99m, metastatic foci can be detected by simple immunoscintigraphy. As with most diagnostic imaging procedures, the size of metastatic lesions is an important variable that influences imaging efficiency.¹³ When analysed by size, lesions of over 2 cm are seen more frequently than lesions of 2 cm or less. Nevertheless, this antibody imaging procedure represents a useful addition to routine chest radiographs and computed tomograms.

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