

MALIGNANT MELANOMA OF THE OESOPHAGUS – A CASE REPORT

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ABSTRACT

Primary malignant melanoma of the oesophagus is a rare presentation for melanoma and is also an unusual and aggressive form of oesophageal neoplasm. We present a 57-year-old Chinese gentleman who underwent successful resection of a primary malignant melanoma of the oesophagus.

Keywords: malignant melanoma, oesophagus

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INTRODUCTION

Primary malignant melanoma of the oesophagus is exceedingly rare and accounts for 0.1% of all oesophageal neoplasms. Since Baur reported the first case in 1906⁽¹⁾, only 139 cases have been reported in the English literature until 1989⁽²⁾. Most of these cases present at a late stage when they are symptomatic with advanced disease. Hence the resectability rate is usually low and the prognosis grave.

We report a case of primary malignant melanoma in a male patient who underwent successful surgical resection.

CASE REPORT

A 57-year-old Chinese man presented with progressive dysphagia to solid food for 6 months prior to consultation. He was able to take only soft diet and complained of excessive secretion of saliva in the mouth that was occasionally blood stained. This was associated with significant loss of weight. He had never undergone surgery for any skin lesions.

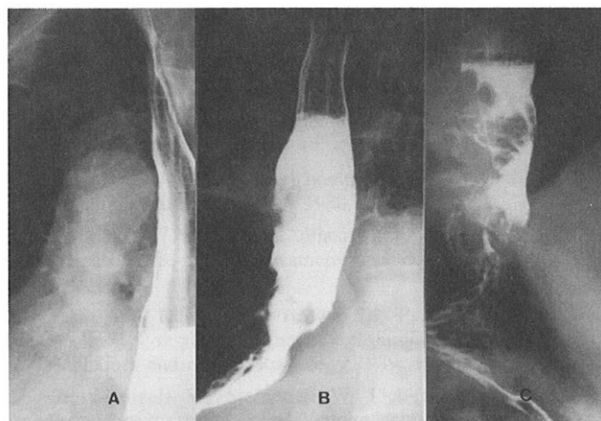
Physical examination showed a well-nourished gentleman. There was no pallor or jaundice. There were no enlarged lymph nodes in the neck, axilla or groin and no palpable liver or spleen. There was no pigmented or depigmented lesion on the skin or mucosa of the oral and anal region.

An upper gastrointestinal endoscopy revealed a polypoidal lesion in the distal third of the oesophagus, 5 cm from the cardia. The stomach and duodenum were normal. Biopsy of the lesion revealed it to be a malignant melanoma.

Barium swallow showed a large polypoidal and irregular lesion at the lower end of the oesophagus that protruded into the fundus of the stomach (Fig 1a, b, c). The total length of the irregularity on barium swallow was 10 cm with the largest cross sectional diameter being 7 cm.

The patient underwent an Ivor Lewis type of oesophagogastrectomy. There were no secondaries noted in the liver and no enlarged nodes in the abdomen but a few pigmented lymph nodes were present in the mediastinum. The tumour in the oesophagus extended from 2 cm below the

Fig 1 - Barium swallow of the patient. (A) Normal upper 1/3 of the oesophagus. (B) Lower 1/3 of the oesophagus where there is dilatation of the lumen with mucosal irregularity. (C) Polypoidal nature of the lesion can be seen at the cardio-oesophageal junction.



azygos vein to the cardia of the stomach. The oesophagus together with the cardia of the stomach was resected with the proximal resection margin 1 cm away from the tumour and the distal resection margin 3 cm beyond the tumour (Fig 2.) The operation was completed with an intrathoracic anastomosis of the remnant stomach to the proximal oesophagus at the level of the azygos vein.

The patient made an uneventful recovery and was discharged on the 13th postoperative day.

He was readmitted 4 months later for complaints of breathlessness and loss of 10 kg of weight. He did not have any dysphagia but had severe loss of appetite. A chest X-ray revealed a large left pleural effusion. Following aspiration of 700 cc of blood stained fluid, a repeat chest X-ray showed that there were multiple cannonball lesions in the left lung fields. An ultrasound of the abdomen revealed metastatic disease in the liver. An upper gastrointestinal endoscopy revealed a pigmented lesion in the first part of the duodenum and biopsy proved it to be malignant melanoma.

DISCUSSION

The existence of primary malignant melanoma was initially disputed, until De la Pava et al⁽³⁾ found the presence of melanocytes in the stratum basale of the oesophageal mucosal epithelium. Others have subsequently confirmed that primary malignant melanomas arise from this cell. Although benign melanocytosis occurs in 4% to 8% of normal oesophagus⁽⁴⁾, malignant melanoma of the oesophagus is extremely rare and

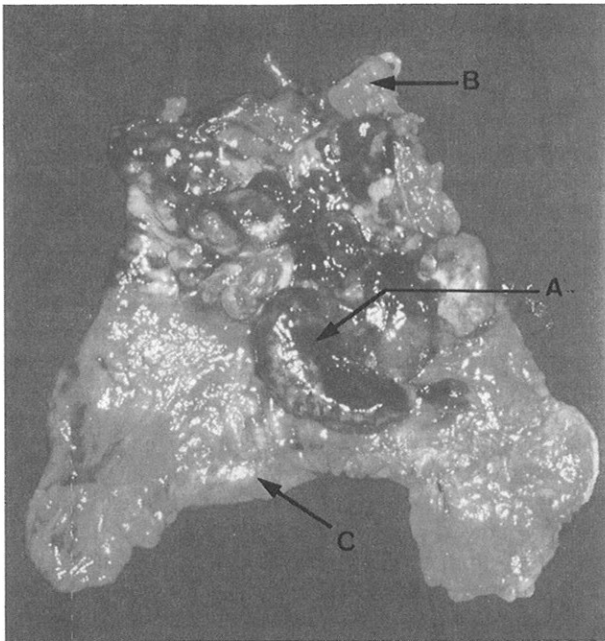
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Fig 2 - Photograph of the resected specimen.
(A) Pigmented polypoidal lesion. (B) 1 cm clear proximal margin. (C) 3 cm clear distal margin.



accounts for only 0.1% of all primary oesophageal malignancies⁽⁵⁾.

Like squamous carcinoma of the oesophagus, most cases of primary malignant melanoma are diagnosed in the 6th or 7th decade with a slight male preponderance and a male-to-female ratio of 2:1⁽²⁾.

The diagnosis of this condition is difficult to make clinically. The presenting symptom is no different from other oesophageal cancers, with progressive dysphagia to solids being the most common symptom. None of the cases reported in the literature had the diagnosis made clinically.

Barium contrast studies and endoscopy are both useful investigations but not specific. The radiological features suggestive of a malignant melanoma include a large, polypoidal tumour often causing obstruction, usually at the middle or lower third of the oesophagus (Fig 1). Endoscopy may be more diagnostic due to pigmentation of the tumour which may be present in most cases (85%)⁽²⁾ or melanosis of the oesophageal mucosa (23%)⁽²⁾. Unfortunately, these features were not identified at endoscopy in our patient.

The diagnosis is usually made by histological examination of endoscopic biopsies, as in our case. The accepted diagnostic criteria include the typical melanoma structure and the presence of melanin, origin from squamous epithelium with junctional activity, and junctional activity with melanotic cells in the adjacent epithelium⁽⁶⁾. Some authors have found junctional activity in less than half the cases and believe that the rapidly growing and expanding tumour may have destroyed any adjacent junctional activity. The growth pattern of this tumour is similar to that of malignant lentigo⁽⁷⁾, with a pushing rather than an infiltrative deep radial margin below normal epithelium. Therefore, false negative biopsies may occur because of the submucosal nature of the tumour. Malignant melanoma of the oesophagus also has a propensity for vertical growth, and in our patient the tumour had invaded the muscular mucosae but not the muscularis propia. In fact

no reported resected specimens contained malignant melanoma confined to the epithelium.

Microscopically, these tumours have a pleomorphic appearance mostly with oval and polygonal cells and pigmentation in 90% of the cases.

Both haematogenous and lymphatic metastases are common and may occur in nearly 50% of cases at first presentation. In a large postmortem study, 40% had liver secondaries, 34% to the mediastinal lymph nodes, 24% to the lungs, 20% to the pleura and 20% the supraclavicular lymph nodes⁽²⁾.

Surgery remains the mainstay of management of these patients. Radical resections are often required because of the widespread intramucosal component of these tumours. But this is not often possible because of the extensive local disease and nodal metastasis. In our case, the tumour involved the lower third of the oesophagus to the cardio-oesophageal junction, making necessary a cardio-oesophagectomy. Even though a clear margin was achieved during operation, detailed histological examination revealed that the proximal resection margin was involved by extensive submucosal spread. In reported series, the survival following local excision of the tumour was less than 9 months with no survivors at the end of one year. Even after radical resection the 5-year survival was only 4.2%, although 2 cases of long term survivors treated by surgery alone are on record, one for 6.5 years⁽⁸⁾ and the other for 10 years⁽⁹⁾. No other form of adjuvant therapy either in the form of radiotherapy or chemotherapy has shown any survival benefit so far.

The aggressive nature of malignant melanoma is well illustrated in this case. In a period of 4 months the patient developed multiple secondaries in both the lung and liver. But the occurrence of the metastatic lesion in the duodenum is of some interest. Although transmural deposits of malignant melanoma have never been reported before, we believe that tumour cells must have deposited at the pyloroplasty site during mobilisation of the tumour at the time of operation. The prognosis in this patient is dismal and the treatment has provided relief of dysphagia for only a little over three months.

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