Rare disease

Synchronous primary oesophageal malignant melanoma and sigmoid adenocarcinoma

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Summary

The authors present a case of a gentleman in his 70s who was referred to the gastroenterology outpatient clinic with dysphagia. An oesophagogastroduodenoscopy was performed which showed a polypoidal black coloured mass in the oesophagus. Endoscopic biopsies confirmed malignant melanoma. Further staging investigations were organised to assess suitability for surgery which revealed a mass in the sigmoid colon. Subsequent colonoscopy and biopsy confirmed adenocarcinoma. As this was an unusual case to associate these two malignancies at the same time, there was no ideal or recognised management plan available. Different treatment options were considered and a consensus was developed regarding best surgical approach but due to the lapse in time a repeat staging CT scan was organised which unfortunately now demonstrated lymph node metastasis. Patient was managed conservatively from this point onwards and he died 12 months later.

BACKGROUND

Synchronous neoplasms of the gastrointestinal tract from different origins are rare conditions. To the authors' knowledge there is no reported case of synchronous primary oesophageal malignant melanoma (POMM) and sigmoid adenocarcinoma in the literature. POMM is a very rare and aggressive tumour and management is surgical. Sigmoid adenocarcinoma not favourable to endoscopic removal is also treated surgically. In this case as both were co-existent and there was no recognised standard approach available, there was a dilemma to which tumour should be the priority for surgical resection. This case highlights a difficult situation that can be faced in the management of synchronous gastrointestinal tract tumours and highlights the importance of multidisciplinary approach to tackle these difficult circumstances. In this case, the outcome was not favourable probably due to the aggressive nature of POMM.

CASE PRESENTATION

A gentleman in his 70s was referred by his primary physician to gastroenterology outpatient clinic with dyspepsia,

dysphagia to solids and weight loss of 2 stones over a period of 3 weeks. There were no reported bowel symptoms. Systemic examination was unremarkable.

The patient had no medical history and he was not taking any medications. His only surgical history was cholecystectomy secondary to gallstones.

INVESTIGATIONS

In view of the presenting features, a sinister cause was suspected and routine blood tests and an urgent oesophagogastroduodenoscopy (OGD) were organised for him. The blood tests including a full blood count, renal function tests, serum electrolytes and liver function tests were all in normal range. The initial OGD showed food residue in the oesophagus and a lesion at 30 cm from incisors, black in colour but not biopsied as there was a concern it might be vascular. Subsequently, an urgent CT scan of the thorax and upper abdomen was organised which showed a dilated oesophagus with food residue and thickening of the oesophageal wall just above the gastro-oesophageal junction. In view of this, a repeat OGD was organised which

Figure 1 (A,B) Primary oesophageal malignant melanoma seen on oesophagogastroduodenoscopy.

Figure 2 Melanoma cells seen on oesophageal biopsy.

showed an exophytic tumour at 30 cm from the incisors, 9 cm in length (figure 1A,B). Biopsies were taken which came back positive for malignant melanoma on histology (figure 2). As there was no primary source, a diagnosis of POMM was made. The patient was discussed in a multidisciplinary team (MDT) meeting and an endoscopic ultrasound scan (EUS) and positron emission tomography (PET) scan were organised for further staging. EUS was reported as showing a predominantly submucosal lesion but more distally there was transmural involvement as well. The PET scan confirmed the presence of mass in the distal oesophagus. However, at the same time, it demonstrated an abnormal area in the sigmoid colon highly suspicious of a sigmoid malignancy (figure 3). A colonoscopy was organised in light of these results which showed a large polypoidal stricturing lesion in the sigmoid colon and biopsies confirmed moderately differentiated infiltrating colonic adenocarcinoma. Hence, the results of the investigations concluded that the patient had two synchronous potentially operable carcinomas, one in the oesophagus and one in the sigmoid colon.

DIFFERENTIAL DIAGNOSIS

The main differential diagnosis of POMM is metastasis of skin or ocular malignant melanoma.

TREATMENT

In view of the increasing dysphagia, an oesophageal stent was inserted which led to improved swallowing.

The results of the EUS and PET scan were discussed in the upper gastrointestinal, supra upper gastrointestinal and lower gastrointestinal MDT in detail. A consensus was reached that the patient should have surgery for sigmoid adenocarcinoma first followed by urgent oesophagectomy

for the malignant melanoma. The rationale was that the recovery time after the lower gastrointestinal surgery would be less allowing for an earlier oesophagectomy.

OUTCOME AND FOLLOW-UP

After consideration of his options, a repeat CT was arranged to restage the patient. Unfortunately, this showed lymph node involvement outside the surgical resection field for his oesophageal tumour. The treatment plan then became palliative, and he died 12 months later.

DISCUSSION

Synchronous malignancies of the gastrointestinal tract are a rare occurrence. The combination of POMM and sigmoid adenocarcinoma has not previously been reported and hence poses a novel but difficult management situation.

Malignant melanoma is a predominantly cutaneous malignancy derived from melanocytes. However, melanocytes have been described in the oesophageal mucosa in up to 8% of the population.¹ POMM is rare and accounts for less than 0.1–0.5% of oesophageal malignancies² and around 0.5% of all non-cutaneous melanomas.³ It occurs predominantly in the sixth and seventh decades of life, with a male-to-female ratio of 2:1.⁴ Overall, 43% of the tumours are located in the lower third of the oesophagus as in our case, 29% in the middle third, 18% at the junction of the middle and lower thirds and 10% in the upper third.⁵ It is a biologically aggressive tumour, with high overall mortality.⁶ The prognosis is very poor; the 5-year survival is 2.2% and the median survival is approximately 10 months.² The colour of the tumour may vary from black to white (amelanotic form).⁷ In most cases, the tumour appears polypoidal, grossly pigmented and covered with a normal looking mucosa that bleeds easily on contact.^{8,9}

Figure 3 FDG PET scan showing increased uptake in the oesophagus and sigmoid colon corresponding to primary oesophageal malignant melanoma and sigmoid adenocarcinoma.

Most patients are symptomatic at diagnosis; dysphagia is the most common presenting complaint (80%), followed by weight loss (37%) and epigastric pain (33%).⁸

Endoscopic biopsy is most sensitive in making the diagnosis of malignant melanoma. The barium scan usually shows polypoid repletion defects, sometimes ulcerated, and it is difficult to distinguish the appearances from other more frequent tumour types. The CT scan can help to define the extent of the lesion, its relationship to mediastinal structures and the presence of regional lymph nodes or distant metastasis.^{8–11} Fluorodeoxyglucose PET has proved to be an excellent method for staging of metastatic melanoma. Due to its high sensitivity and ability to cover whole body in one examination, it can supplement other staging tools.

It is important to mention here that in a small percentage of cases of primary skin melanoma which subsequently metastasize, the primary melanoma lesion completely regresses for reasons that are not understood and the primary lesion is never located. This makes it difficult to be 100% sure that the lesion is a primary oesophageal melanoma.

Due to the rarity of tumour, the optimal management of primary oesophageal melanoma is still uncertain. In general, the treatment of POMM is wide surgical resection, with dissection of the lymph nodes, however, total or near-total oesophagectomy offers the best survival outcome (about 5 years, vs 9 months for local resection).⁵ Unfortunately, 30% of all tumours are unresectable at the time of diagnosis. Radiotherapy, chemotherapy or immunotherapy may play a part in management, alongside surgery or as palliative approaches.^{12–15}

Adenocarcinoma of the large bowel is one of the leading causes of cancer deaths worldwide. It usually presents as change in bowel habit, bleeding per rectum, abdominal pain, weight loss or anaemia. In our case, the patient had no symptoms secondary to the sigmoid carcinoma.

The best management plan for these tumours when present in combination is not known and poses a difficult

question to the involved professionals. In our case, a surgical approach was thought appropriate with left-sided hemicolectomy followed by oesophagectomy though one can argue that POMM being more aggressive of the two should have been tackled first. However, unfortunately before this plan could be executed, the patient developed nodal metastasis and was eventually managed conservatively.

Learning points

- ▶ Synchronous cancers of the gastrointestinal tract though rare can be difficult to manage. An MDT approach is helpful to guide management in these challenging circumstances.
- ▶ POMM is rare and endoscopic biopsy is the investigation of choice.
- ▶ POMM is a very aggressive and can metastasize very quickly and when identified should be treated as quickly as possible to prevent poor outcome.

Competing interests None.

Patient consent Not obtained.

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