CLINICAL CASE

Diffuse primary malignant melanoma of the upper gastrointestinal tract

Mélanome primitif et diffus du tractus œsogastroduodénal


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Summary  Primary malignant melanomas of the GI tract are very rare. Their symptomatology is not specific. We report a 78-year-old Tunisian woman hospitalised with a 6-month history of recurrent abdominal pain, loss of appetite, weakness and weight loss. She had no personal history of cutaneous or ocular melanoma. Upper gastrointestinal endoscopy revealed multiple small, raised darkly pigmented tumours. Theses lesions were found in the oesophagus, the stomach, the bulb and the duodenum. Biopsy specimens were taken and histology showed the presence of melanocytic cells with abundant melanin pigment. Immunohistochemically, tumour cells were positive for HMB-45. Morphological examinations revealed hepatomegaly with multiple nodules with small lymph nodes at the celiac axis. All available diagnostic procedures failed to identify any other site of ocular or cutaneous melanoma, the present case was considered as primary GI melanoma. Palliative chemotherapy was not possible because patient was extremely cachectic and she died one month later.


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de tumeur primitive oculaire ou cutanée étaient normales, ainsi le diagnostic de mélanome digestif primitif a été retenu. Une chimiothérapie palliative a été proposée mais récusée du fait d’une altération profonde de l’état général de la patiente qui décédait un mois après le diagnostic.

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Introduction

In clinical practice and the literature, malignant melanoma usually appears in typical sites where melanocytes can be found (skin, eyes, meninges and anal region) [1]. Malignant melanomas of the gastrointestinal (GI) tract are usually metastatic [1]. Primary and diffuse upper GI tract melanoma is rare and only a few descriptions of this presentation have been found in the literature [2,3]. The present report describes a new case of diffuse and primary malignant melanoma of the GI tract.

Case presentation

A 78-year-old Tunisian woman was admitted to the Department of gastroenterology because of a 6-month history of recurrent abdominal pain, loss of appetite, weakness, weight loss and anaemia. She had no personal history of medical disease especially cutaneous or ocular melanoma, but she had family history of malignant brain and prostate tumours in first degree relatives.

On admission the patient was pale, in bad general condition and abdominal examination revealed painful multinodular hepatomegaly. The rest of physical examination was normal and did not reveal any cutaneous melanoma.

Laboratory investigations confirmed noniron deficiency anaemia with 10 g/dL haemoglobin’s rate and C reactive protein rate was 32 mg/L (< 5 mg/L). Liver tests revealed cholestasis with γ glutamyl transpeptidase at 501 IU/L (9—36 IU/L) and alcaline phosphatase at 33 IU/L (40—150 IU/L) and aminotransferases (aspartate aminotransferase at 87 IU/L (5—34 IU/L), alanine aminotransferase at 53 IU/L (5—55 IU/L). Total bilirubin rate was 18 μmol/L (3.4—20.5 μmol/L) and prothrombin rate was 92%. Upper gastrointestinal endoscopy revealed twelve small, darkly pigmented tumours (Fig. 1). Some of these lesions were raised and volcanoid whereas the other were flat. Theses lesions were found in the oesophagus, the stomach, the bulb and the first and second parts of duodenum. Biopsy specimens were taken and histology showed the presence of melanocytic cells with abundant melanin pigment in oesogastroduodenal mucosa (Fig. 2). Immunohistochemically tumour cells were positive for HMB-45 and negative for cytokeratin and melan A (Fig. 3).

Abdominal ultrasonography and computed tomography (CT) contrast scan of the abdomen showed hepatomegaly with multiple high density metastatic lesions in the liver, the largest measuring 5.4 cm and 5.8 cm, respectively in segments VI and VII (Fig. 4). In addition, CT contrast scan showed a neoplastic partial portal thrombus with small lymph nodes at the celiac axis. The colonoscopy did not identify any neoplastic process up to the small bowel. Dermatological and ophthalmologic examinations were normal. Brain and spinal cord CT scan did not show any metastatics lesions.

The diagnosis of primary advanced melanoma was made and because of the extremely poor condition of the patient, pal-

Figure 1 Endoscopic aspect of multiple and small raised darkly pigmented tumours in the stomach.

Figure 2 Melanocytic cells with abundant melanin pigment, HE × 400.

Figure 3 Tumour cells positive for HMB-45. Immunohistochemical study × 200.
Diffuse primary malignant melanoma of the upper gastrointestinal tract

Figure 4  Multiple liver high density metastatic lesions on computed tomography (CT).

Discussion

While malignant melanoma is a common skin tumour, primary growth of this tumour in another organ is exceedingly rare [1].

Primary malignant melanomas of the GI tract are very rare and occur frequently in the anorectum and oesophagus [3]. They can occur in the tongue, stomach, small bowel and colon. The endoscopic appearance of GI lesions are described as volcanic like dark ulcers, polyps or masses or small black spots [4]. The present case was a diffuse primary melanoma involving the oesophagus, stomach, bulb and duodenum. Criteria for primary melanoma include lack of concurrent or previous removal of skin melanoma and lack of involvement of other organs [3].

The presence of melanoma in the GI tract has been discussed in many publications [2–4], and most are metastases of skin tumours. For this reason, the diagnostic methods of the present report may not have been enough to identify discrete and primary sites of melanomas.

The poorer outcome of primary GI melanoma is related to earlier dissemination because of the rich vascular supply of GI mucosa and detection of disease in the advanced clinical stage. The liver is the most common initial site of metastatic involvement and morphological studies generally show multiples nodules in the liver [5]. Percutaneous liver biopsy is not necessary except in hepatic metastatic miliaria spread. In these cases, morphological studies are normal and only biological abnormalities such as icteric cholestasis and hepatocellular insufficiency may suggest the diagnosis [6].

In the present case, a family history of malignant tumour suggests the existence of family syndrome. In fact, recent studies have suggested that there may be an increased risk of melanoma in relatives of patients with brain, pancreatic and colorectal cancer [7,8].

Despite the many advances in cancer treatment in the past few decades, the prognosis for patients with advanced melanoma remains poor and the 5-year survival rate in patients with distant metastases is less than 10% [9].

Radiation therapy is primarily to relieve symptoms and although newer chemotherapy agents and biotherapy have been developed, they does not significantly increase overall survival of patients of metastatic melanoma [9]. Surgery significantly improves survival especially when resection is considered complete following microscopic examination. It is also recommended for palliative treatment when GI melanoma is symptomatic [9].

Conclusion

Although malignant melanoma is usually described as a metastatic lesion, in this case because another primary site of melanoma was not identified the present case can be considered a primary upper GI tract melanoma. This is a rare but aggressive disease, often associated with metastatic spread at presentation.

References