Rare disease

Cutaneous relapse of an ampullary carcinoma: an unusual presentation

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Summary

This is the first ever reported case about a cutaneous relapse of small bowel adenocarcinoma. The ampulla of Vater is the area of the small bowel that presents more frequently malignant transformation, nevertheless ampullary adenocarcinomas are rare but aggressive diseases. However, distant metastases are infrequent. Cutaneous metastases constitute the 5.3% of skin tumours, and are usually found in the 12% of malignancies. Their management usually includes local treatment if they are unique and systemic treatment when they are multiple. Chemotherapy schemes used in ampullary adenocarcinoma are those used in cholangiocarcinomas, pancreas and tumours of the gallbladder; nevertheless, given the intestinal origin of these tumours combinations of capecitabine and oxaliplatin are commonly used.

BACKGROUND

This is the first reported case of cutaneous relapse of an ampulla of Vater adenocarcinoma: a rare tumor with an unusual way of relapse.

We present the case of a man with multiple skin metastasis of an ampullary carcinoma after 20 months of disease-free interval. Cutaneous metastases are common in tumours such as breast cancer, especially in advanced stages of the disease. In tumours of the biliary tract, skin involvement is rare. It has been described cases related to cholangiocarcinoma, tumours of the gallbladder and there is a case related to a periampullary leiomyosarcoma.¹ It has not been documented any case of cutaneous metastases secondary to ampullar or small bowel tumours; in fact, this is the first case report of an ampulloma with skin involvement.

CASE PRESENTATION

We report a case of a 72-year male with a history of chronic obstructive pulmonary disease, dyslipidaemia and diabetes. The patient was admitted in October 2009 with upper gastrointestinal bleeding. Gastroscopy and endoscopic retrograde cholangio-pancreatography was performed and the patient was diagnosed with a localised signet ring cells adenocarcinoma of ampulla of Vater (T2N1M0). Whipple procedure was used as radical surgery. Pathology analysis of the piece revealed a localised stage of the tumour without involvement of surgical margins. As it had lymph node invasion (metastasis in 6 of 11 loco-regional lymph nodes was analysed), adjuvant chemo-radiotherapy (gemcitabine-based scheme) was performed. He completed the treatment in March 2010, when he started the surveillance. In February 2011, progressive elevation of tumour markers (CEA 12), without signs of local or distant recurrence on imaging techniques (CT) was found. Looking for relapsed disease, he completed the study with positron emission tomography (PET)-TC (despite low sensitivity for tumours of signet ring cells) which was negative except of a pathological uptake in cavum and paracervical region. While performing these additional tests, the patient reports multiple,

painless and superficial small (about 1 cm in diameter) nodular lesions on cervical and thoracic region. Suspecting lymph node involvement and a head and neck new primary tumour, a fibroscopy with multiple blind biopsies was done with no patologic results. Finally, skin lesions were analysed by fine needle aspiration (figure 1). Pathology showed metastasis of a signet ring adenocarcinoma.

So, this is the first case of cutaneous relapse of an ampullary adenocarcinoma. At present the patient is on systemic treatment with cisplatin-gemcitabine scheme,² with good tolerance. Principio del formulario

DISCUSSION

Tumours of the biliary tract are classified in tumours of the gallbladder, cholangiocarcinoma and ampullary adenocarcinomas. They are low incidence tumours (1–3 cases per 100000 population in USA). These tumours are more prevalent in patients between 50 and 70 years, with higher incidence in males and more than 90% are adenocarcinoma.³

Figure 1 Skin lesions.

The ampulla of Vater is the area of the small intestine that presents more often malignant transformation; nevertheless ampullary adenocarcinomas are rare malignancies usually associated with hereditary polyposis syndromes. They generally have a better prognosis than cancers of the pancreas with overall survival after five years of 84% in stage I, 70% stage II, stage III 27% and 0% stage IV.

The aggressive course of these neoplasms is due to the delay on diagnosis, as they have no symptoms until very advanced stages of the disease. Their most usual presentation is a biliary obstruction: obstructive jaundice, right upper quadrant pain and weight loss. Ampullary adenocarcinomas are commonly associated to steatorrhea due to malabsorption. Distant metastatic disease most frequently involves the regional lymph nodes, liver, peritoneum, lungs and bone. It has not documented any case of cutaneous metastases until now, as far as we know.

Cutaneous metastases constitute 5.3% of skin tumours, and are usually found in 12% of malignancies, predominantly in breast, lung, colorectal, ovarian, renal and gastrointestinal cancers.⁴ They are more common after the sixth decade and are considered an advanced disease stage, with poor survival (no more than 12 months) especially if it is a multiple presentation.⁵ In most cases, they occur as the distant involvement of a known neoplastic disease, but in 22% of cases it is the only presentation of the tumour and sometimes the relapse of the disease (most cases after a 36 months of disease-free survival). Predominantly skin metastases presents as nodular lesions damaging the dermis and subcutaneous fat (epidermal involvement is found in a minority of cases), no bigger than 3 cm diameter and most often asymptomatic (although they may be accompanied by pain, ulceration and bleeding). Nevertheless, in more than 46% of cases, they have atypical presentation. Most times they are very similar to benign lesions.

Cutaneous spread can be locoregional (in context of punctures or biopsies), in-transit or distant in which case the most common sites are the chest (30.3%), abdomen (20.2%), scalp (12.6%) and extremities (7.4%).⁶ Periumbilical lesions, typical of gastric tumours, are called Sister Mary Joseph nodules. Iatrogenic subcutaneous spread of malignant diseases after diagnostic procedures is seen in the clinical practice.

Diagnosis is based on clinical suspicion and biopsy, accompanied by image techniques (TC and PET).

Treatment is based on local and systemic approaches. Although local treatment will not change the prognosis of the disease, surgery, electrocoagulation, radiotherapy or intralesional injection of miltefosina, interferon or interleukin-2⁷ may be considered for aesthetic problems when skin involvement is limited. Anyway, the patient must receive also the chemotherapy based systemic treatment for the systemic disease. Comparing the size of lesions during chemotherapy treatment can help monitoring the response to it, especially in Caucasians where it is considered that the skin involvement is directly proportional to disease spread.

Learning points

- This is the first reported case of a cutaneous relapse of an ampulla of Vater adenocarcinoma: a rare tumour with a unique form of presentation of relapsed disease.

Competing interests None.

Patient consent Obtained.

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