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Case Report

Signet Cell Adenocarcinoma with Colorectal and Facial Metastasis - A Case Report

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Received: November 12, 2018; **Published:** December 21, 2018

Abstract

Cutaneous metastatic signet ring cell carcinoma originating from the appendix is rare and represents widespread disease with poor patient outcomes. We report a case of a 59-year-old male who presented primarily with facial cutaneous lesions and resultant positive biopsy for signet cell adenocarcinoma. Colonoscopy identified synchronous metastatic lesions in the cecum, transverse colon, and rectum. Negative work-up prompted re-examination of previous pathology reports identifying a small focus of signet ring cell carcinoma from an earlier appendectomy. Our case represents the only documented presentation of metachronous colorectal signet ring cell carcinoma with facial cutaneous lesions.

Keywords: *Signet Cell Adenocarcinoma; Colorectal; Facial Metastasis*

Introduction

Cutaneous metastasis are rare, occurring within an approximate range between 0.7 - 10.4% of all malignancies [1-3]. The highest rate of cutaneous metastasis occurs in breast cancer with a 24% incidence rate [4]. Specifically, cutaneous metastasis of rectal carcinoma occurs in less than 4% of all patients with rectal cancer [3,5-7].

The development of cutaneous metastasis may indicate widespread disease; with poor prognosis [7]. Most skin metastasis from colorectal cancer present within two years of the diagnosis and treatment of the primary cancer [8]. Further, they often present with metastasis to other organs, including the liver and lungs [9]. Although cutaneous metastases are rarely present at the initial diagnosis of the primary cancer (1%), primary diagnoses are not made in 19% of cutaneous metastatic cases [1,12,13].

Facial cutaneous metastasis are also quite rare, occurring in less than 0.5% of patients with metastatic disease [1].

We present a case of mucinous adenocarcinoma of the appendix with metachronous metastasis to the colon, rectum and facial skin.

Case Report

A 59 year- old Caucasian male with a past medical history of chronic lymphocytic leukemia (CLL), diagnosed over 8 years ago, no treatment has been required. He had experienced episodes of diverticulitis in the past and had a previous appendectomy secondary to a clinical picture of acute appendicitis. The patient has a smoking history of 25 pack years' duration and occupational history of chemical exposure. Currently he is on disability after a work related physical injury.

He presented initially, with redness and swelling under his left lower eyelid. His primary care provider treated him as periorbital cellulitis (Figure 1). Due to complete lack of improvement with antibiotics the patient was referred to a plastic surgeon. A punch biopsy of the lesion was performed; this revealed poorly differentiated mucinous adenocarcinoma with signet cell features in the dermis and subcutaneous tissue. The tumour was positive for cytokeratin 20 and CDX2, suggesting possible metastatic disease from a lower gastrointestinal origin.

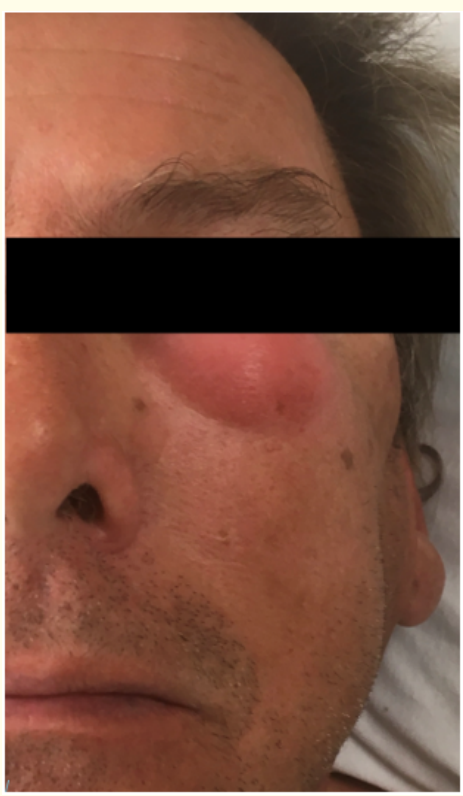

Figure 1

Consideration was immediately given for an endoscopic assessment as well as extensive investigations. Upper and lowers scopes were performed, a gastroscopy was unremarkable however the colonoscopy identified synchronous lesions in the cecum, transverse colon,

and rectum. Macroscopically those lesions had an atypical appearance, looking more like an inflammatory process rather than fungating masses, they were friable and hard in consistency (Figure 2). Biopsies were taken; the pathologist concluded that all the three areas were positive for signet ring cell carcinoma, predominantly in the submucosa with areas of lamina propria infiltration, thus favoring metastatic deposits rather than a primary carcinoma. No mutations for BRAF, KRAS or NRAS were identified.

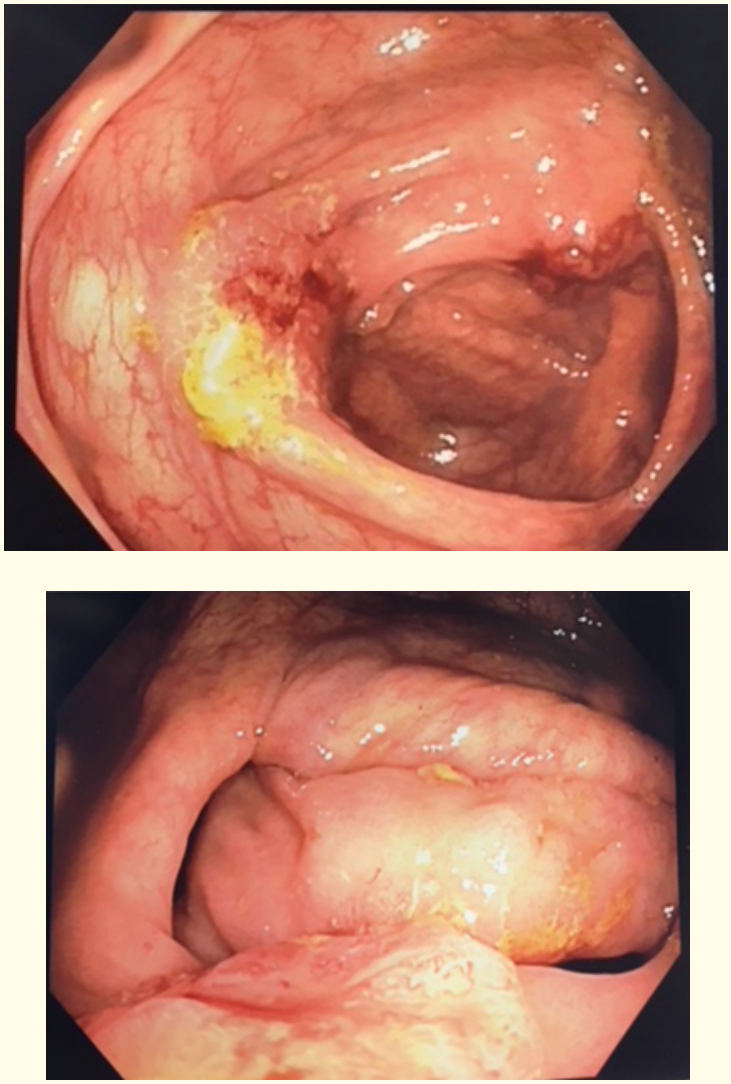

Figure 2

The rest of the work up, included: a bone scan, which was negative for metastasis. CT scan of the head displayed left peri-orbital soft tissue swelling. CT of the chest, abdomen and pelvis demonstrated adenopathy, previously attributed to the patients CLL. Further, lumbar MRI did not demonstrate metastatic marrow changes or retroperitoneal adenopathy. Specific CEA and PSA blood tests were measured 14 ug/L and 1.7 ug/L, respectively.

No evidence of a primary tumor was present at this time, the case was discussed at the multidisciplinary cancer conference, any considerations for surgery were dismissed once the biopsy results suggested metastatic disease, with no yet identified primary.

The patient was initiated on chemotherapy with the FOLFOX-6 modified regimen. Consideration for PET scan was suggested but precluded due to its poor sensitivity toward identifying mucinous metastasis [25]. A consultation with the head and neck surgery service, also considered there was no value for surgical excision. Radiation was then contemplated as an option, and commenced aiming to achieve local control of the swelling and slow progression of the disease.

Unfortunately, 4 weeks into chemotherapy treatment, the patient was diagnosed with perforated diverticulitis and hospitalized. He was managed medically with antibiotics and his chemotherapy treatment was held. This admission triggered a re-examination of previous pathology reports which identified a small focus of signet ring cell carcinoma in the sub-mucosal tissue from an appendectomy in 2015. This was a very subtle finding and different pathologists had to review the slides in order to conclude it was a positive finding.

Once the diverticulitis episode was under control, as the patient seemed to be responding locally to the radiation and remained asymptomatic, a decision was made to complete his radiation, hold systemic therapy and abandon any consideration for surgery

Discussion

Signet cell carcinoma is a rare malignant adenocarcinoma occurring in 0.1 - 2.6% of colorectal cancers [25]. Most cases are diagnosed at late stage with poor prognosis [9,10]. In our patient signet ring cell carcinoma was then confirmed in biopsies from the transverse colon, cecum, and rectum. The mucinous component was only identified on the punch biopsies, however all the sites demonstrated signet cells.

Most case reports focused on signet cell adenocarcinoma with facial metastasis present with a single lesion [7,9,13,14]. Hashimi and Dholakia discuss a case of facial cutaneous metastasis with a primary presentation of bowel habit changes [18]. Following a literature search, we present the only case of synchronous colorectal signet ring cell carcinoma with facial cutaneous metastasis.

Kilickap., *et al.* reported a case on a patient who was found to have signet cell carcinoma of the rectum. Subsequent resection, radiotherapy and adjuvant chemotherapy were performed. Posteriorly the patient developed cutaneous nodules on the chest wall, positive for signet ring cell features [8].

Most cases present with the cutaneous metastasis months following resection and treatment of the primary tumour, Cherif., *et al.* present a case where the metastatic lesion in the orbit triggered the investigations for aetiology. Their case presented with visual impairment and periorbital facial pain, with soft tissue mass discovered on CT of the orbit. CT of the chest, abdomen and pelvis demonstrated thickened walls, leading to endoscopy and confirming adenocarcinoma of the rectum [14]. In most situations a primary colon cancer is followed by cutaneous metastasis months after resection and chemoradiation treatment [3,7,9,12,13,15]. Our case presented primarily with a skin lesion, most likely a metastasis from an appendiceal adenocarcinoma from two years prior, and no gastrointestinal symptoms.

Primary adenocarcinoma of the appendix is rare, with incidence of less than 0.5% [16-21]. Only 4% of all appendiceal tumours are signet ring cell carcinoma [22]. In the absence of any other primary focus, our patient was then diagnosed and treated as having metastatic disease from the appendiceal tumor. Although cutaneous metastases are rarely present at the initial diagnosis of the primary cancer (1%), primary diagnoses are not made in 19% of cutaneous metastatic cases [1,11,12].

Prognosis for appendiceal adenocarcinoma has been reported as an overall 5 year survival rate of 18 - 27% [22-24]. Further, the median survival of cutaneous metastasis of colorectal cancer is approximately 3 - 34 months following the appearance of the lesions [1,7,25].

Conclusion

In conclusion, cutaneous metastases are rare and represent widespread disease with poor patient outcomes. Following literature review, our case represents the only report of adenocarcinoma with signet ring cell features of the appendix with metachronous metastasis to the colon, rectum and facial skin. Literature on appendiceal adenocarcinoma with facial cutaneous lesions as primary presentation is not well documented.

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Volume 6 Issue 1 January 2019