

Views and Practice

Truncal nodule in a patient with known colorectal cancer

WZ Loh, HY Chia, MT Chio

Introduction

Sebaceous differentiation is common in cutaneous neoplasms, in lesions with predominantly sebaceous differentiation (sebaceous adenoma, sebaceoma and sebaceous carcinoma) or in neoplasms with other primary lines of differentiation with focal sebaceous areas. However, sebaceous carcinomas are rare, with an estimated incidence rate of approximately 1 to 2 per 1,000,000 per year.¹ The majority (98%) of cases are older than 40 years of age with a peak incidence of 60 and 79 years.² Of these, most sebaceous carcinomas are ocular, with extra-ocular lesions only accounting for approximately 25% of sebaceous carcinomas.

Case report

A 73-year old Chinese male was referred from a nursing home for a rapidly growing and bleeding nodule on his left anterior chest wall of one month duration. He had a history of stage II

hepatic flexure adenocarcinoma of the colon diagnosed four years previously that was treated with right hemicolectomy and follow-up interval imaging had not shown any evidence of recurrence. Cutaneous examination showed a 1.5 cm well defined flesh-coloured ulcerated nodule with areas of bleeding, which was firm to the touch and fixed to the surface skin (Figure 1). There was no known family history of cancer. A shave excision for histopathology showed multiple irregularly sized tumour lobules seen arising from the epidermis, composed of a disordered mixture of basophilic germinative sebaceous cells with oval nuclei and nuclear pleomorphism, and areas with mature sebaceous cells with eosinophilic bubbly cytoplasm (Figure 2). This was suggestive of well-differentiated sebaceous carcinoma. In view of the history of right-sided colorectal cancer and sebaceous carcinoma, a diagnosis of Muir-Torre syndrome (MTS) was strongly considered. However in view of the patient's poor pre-morbid function and because he was unable to consent or agree to further investigations, he was expectantly managed.

Discussion

Most extra-ocular sebaceous carcinomas are located around the head and neck.¹ Diagnosis is confirmed on histology which reveals neoplastic cells with various degrees of differentiation arranged in lobules as in this case or at other times sheets of cells separated by a fibrovascular stroma. In well-differentiated tumours, sebocyte-like cells with vacuolated, foamy cytoplasm

National University Hospital, Singapore

WZ Loh, MD, MRCP(UK)

National Skin Centre, Singapore

HY Chia, MD

MT Chio, MD

Correspondence to: Dr. WZ Loh
National University Hospital, 1E Kent Ridge Road, Singapore
119228

are present. Differential diagnosis of bleeding truncal nodules with sebaceous cells includes other malignancies such as basal cell carcinoma, squamous cell carcinoma and adnexal neoplasms, which can have secondary sebaceous differentiation or present with cytoplasmic clearing appearing similar to sebaceous differentiation.³ Other characteristic features of the various cutaneous neoplasms, as well as immunohistochemistry such as adipophilin to identify intracytoplasmic lipids suggestive of true sebaceous differentiation can help to confirm the diagnosis.⁴ Other differentials



Figure 1. Clinical photo of the ulcerated nodule on left central sternum 1.5 cm by 1.7 cm.

include benign lesions like sebaceous adenoma and sebaceoma, which is usually distinguished by a lack of cytological atypia and infiltrative growth. Ulceration, as seen in this patient, is also less typical of benign lesions.⁵ Sebaceous carcinomas, though rare, appears to be more common in Asian populations,^{2,6} and hence a high index of suspicion must be maintained in such patients.

Sebaceous carcinomas that occur on the trunk are rare. When identified, they are less likely to be sporadic and should prompt evaluation for underlying MTS,⁷ a phenotypic variant of Lynch syndrome characterised by at least one sebaceous skin tumour and at least one visceral malignancy. In patients diagnosed with sebaceous carcinoma, approximately 2% are associated with MTS. Unlike Lynch syndrome, MTS can be diagnosed without a family history of malignancy. The most common visceral malignancy is colorectal cancer especially in the proximal colon. Other associated malignancies include urogenital tract, breast, pancreatic, gastric, lung, and haematological cancers.⁸ Sebaceous neoplasms can precede or occur simultaneously with visceral cancer, although in more than half of the reported cases they are diagnosed afterwards.⁹ Additional clinical features of MTS include a personal and family history of

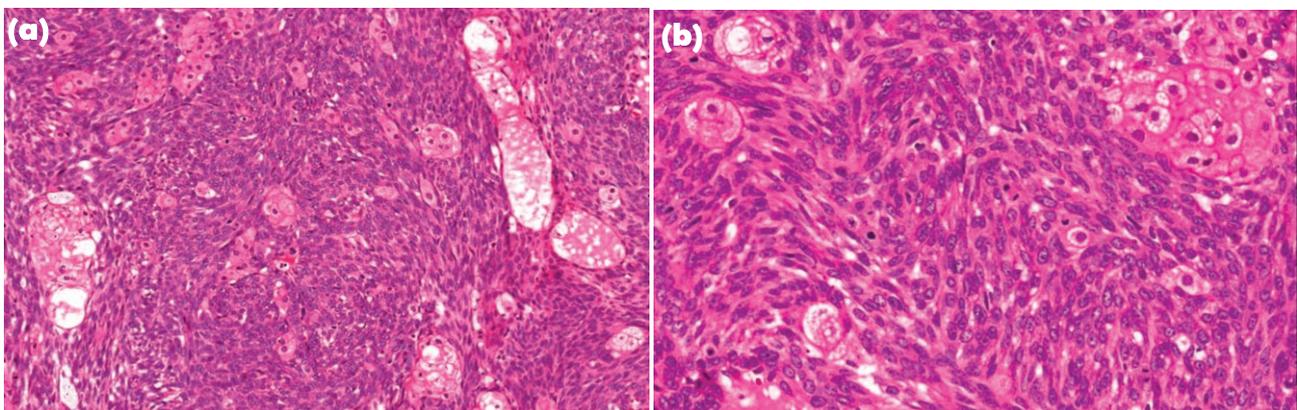


Figure 2. H&E stain of skin nodule. The epidermis is ulcerated. There are multiple irregularly sized tumour lobules showing connection with the overlying epidermis. The tumour lobules consist of a disorderly mixture of basophilic germinative sebaceous cells which show nuclear pleomorphism, and areas of mature sebaceous cells. Conspicuous mitotic figures, some of which are atypical, are seen within the tumour. (a) 200 x magnification. (b) 400 x magnification.

associated cancers and Fordyce spots in the oral mucosa which are also more common in patients with germline mismatch repair mutations.¹⁰ Before proceeding with costly microsatellite instability and germline mutation analysis, microsatellite instability (MSI) immunohistochemical staining for MSH2, MLH1, MSH6 and PMS2 protein can serve as an easy screening test for MTS, with approximately 90% sensitivity.¹¹ Comprehensive management includes complete surgical excision with wide margins, aggressive screening for associated cancers and genetic counselling for family members if a germline mutation is identified. The overall 5 and 10 year survival rates of patients with sebaceous carcinoma are approximately 92% and 79%, respectively, with most of extraocular sebaceous carcinomas curable with excision.¹ MSI-associated and extraocular sebaceous are associated with a lower rate of metastatic disease,¹² although the 5-year survival is similar.^{1,2}

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