

# Sebaceous Carcinoma

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## CASE SUMMARY

### History

A 69-year-old man presented with a one-year history of scalp nodule. It had increased in size during this period but was otherwise asymptomatic. Apart from eczema, there was no significant past medical history and drug history. There was also no family history of malignancy.

### Physical examination

There was a one-centimeter, flesh-coloured nodule on the scalp with telangiectatic blood vessels (Figure 1). There was no lymphadenopathy and general examination including abdominal examination was normal.

### Differential diagnosis

The differential diagnoses include: basal cell carcinoma, sebaceous cyst and sebaceous carcinoma.



Figure 1: A flesh-coloured nodule on scalp

### Investigations

An incisional skin biopsy of the nodule showed an infiltrating epithelial neoplasm with malignant silhouette featuring asymmetry, ulceration and necrosis (Figure 2). The neoplasm contained lobules of undifferentiated cells with focal vacuolated sebocytic differentiation and sebaceous ductal differentiation (Figure 3). High power of tumour lobules showed anaplastic undifferentiated basaloid cells and a few multi-vacuolated malignant sebocytes (Figure 4). The histological diagnosis was **sebaceous carcinoma**.

As sebaceous carcinoma may be associated with Muir-Torre syndrome, screening for underlying neoplasia was performed. Results of complete blood picture, ESR, renal and liver function tests, carcino-embryonic antigen, alpha-fetoprotein and prostate specific antigen were normal. Faecal occult blood was negative. Chest X-ray was normal.

### Management and progress

The patient was referred for colonoscopy examination to exclude underlying colonic neoplasm. He was also referred to the surgical clinic for complete excision of the remaining skin lesion. Wide excision and skin grafting was performed.

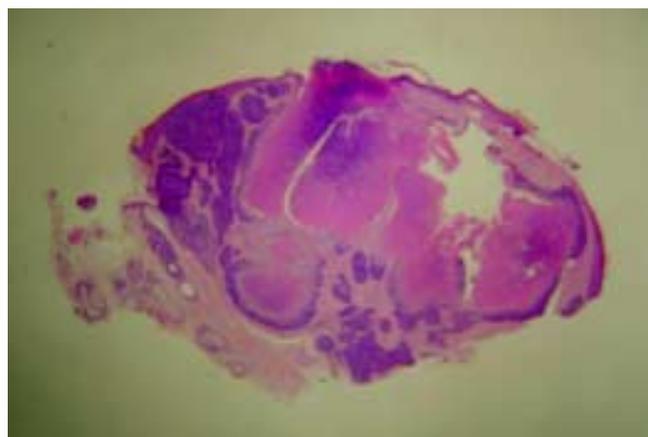
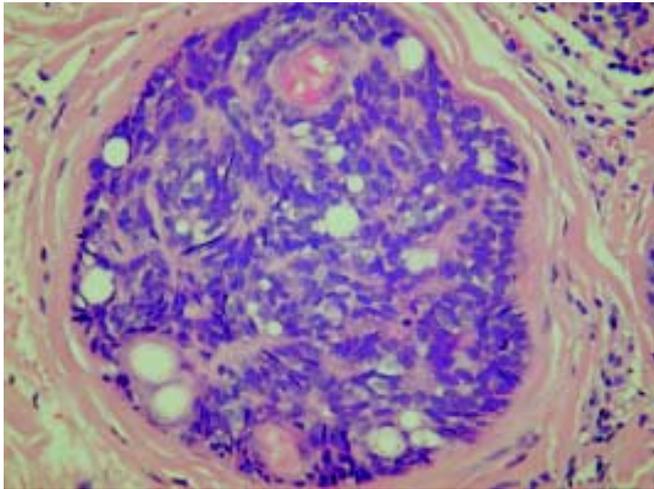
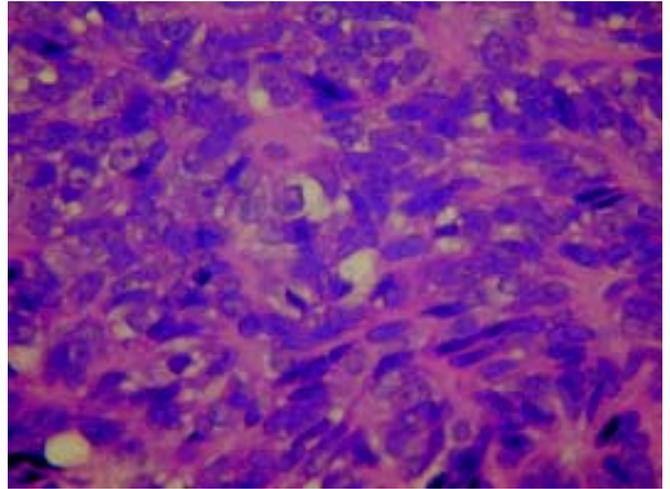


Figure 2: Picture of the entire tumour showing epithelial neoplasm with malignant silhouette featuring asymmetry, ulceration and necrosis en masse (By courtesy of Dr. W. Y. Lam, Tuen Mun Hospital)



**Figure 3: Tumour lobule showing basaloid undifferentiated cells with focal vacuolated sebocytic differentiation and sebaceous ductal differentiation (By courtesy of Dr. W. Y. Lam, Tuen Mun Hospital)**



**Figure 4: High power of tumour lobules showing anaplastic undifferentiated basaloid cells and a few multi-vacuolated malignant sebocytes. A mitosis is seen in this picture (By courtesy of Dr. W. Y. Lam, Tuen Mun Hospital)**

## REVIEW ON SEBACEOUS CARCINOMA

### Definition

Sebaceous carcinoma is a malignancy of the sebaceous glands.

### Clinical features

Sebaceous carcinoma consists of a single yellow or orange nodule. The lesion grows slowly and often appears after 40 years of age. The face (particularly the eyelids) and scalp are the most commonly affected sites. It has also been reported to arise from actinic keratoses<sup>1</sup> and Bowen's disease of the vulva.<sup>2</sup> The upper eyelid is affected two to three times more often than the lower eyelid due to the greater number of Meibomian glands. Metastases are common in sebaceous carcinomas affecting the upper eyelid, often spreading to the regional lymph nodes and viscera. Metastases are uncommon in extra-ocular lesions. Sebaceous carcinoma may be a marker for the Muir-Torre syndrome.

### Histopathology

Sebaceous differentiation is the main histological feature. The cells are arranged in lobules and are of variable size and differentiation. The cells are atypical and may show evidence of local invasion. The differentiated cells tend to be located in the center of

the lobule while the less well-differentiated cells are found near the periphery and may contain fat globules. The lesion is usually located in the deep dermis. Sebaceous carcinoma has to be distinguished from basal cell carcinoma with foci of sebaceous differentiation, sebaceous adenoma, and squamous cell carcinoma with sebaceous differentiation. This may be difficult if the cells are poorly differentiated. It has been reported that extra-ocular and ocular sebaceous carcinoma are positive for human milk fat globule subclass 1 (HMFG 1), human milk fat globule subclass 2 (HMFG 2), breast carcinoma antigen 255 (BCA 225), and epithelial membrane antigen (EMA),<sup>3-5</sup> and negative for carcinoembryonic antigen (CEA), PAS and Alcian blue staining. This enables sebaceous carcinoma to be distinguished from other tumours.

### Muir-Torre syndrome<sup>6,7</sup>

The Muir-Torre syndrome is an autosomal dominant genodermatosis consisting of sebaceous neoplasms (sebaceous adenomas, sebaceous carcinomas, and sebaceous epitheliomas) with or without keratoacanthomas and one or more low-grade visceral malignancies. Keratoacanthomas are only considered a marker for this syndrome if they are multiple and associated with two or more low-grade visceral malignancies and a positive family history of the Muir-Torre syndrome or when there is sebaceous differentiation in the keratoacanthoma. The diagnostic

criteria are shown in Table 1. In a study of 59 cases of sebaceous gland neoplasms, 42% were found to have associated malignancies.<sup>8</sup> Sebaceous carcinoma may therefore be a marker of this syndrome and may precede the onset of malignancy for more than 20 years. It is more common in males (male:female ratio 3:2) and affects patients between 31 and 89 years of age. There is a family history of malignancy in 87% of cases.

In a study of 147 patients with Muir-Torre syndrome and internal malignancy, colorectal carcinoma was the most common visceral malignancy associated with the Muir-Torre syndrome (53% of cases).<sup>9</sup> Colonic carcinomas occur one decade earlier in this syndrome than in the general population. More than one primary may be present and 58% of colonic carcinomas occur proximal to the splenic flexure, in contrast to the general population where colonic carcinoma occurs distal to the splenic flexure. Thirty-nine patients (27%) were found to have colonic polyps, and 89% of patients with colonic polyps developed colonic carcinoma. Genito-urinary neoplasms were the next most commonly associated malignancy (25%). Other associated visceral malignancies include breast, head and neck, haematological malignancies, lung and gastrointestinal tract malignancies are occasionally reported (Table 2). Visceral malignancies associated with the Muir-Torre

are often low-grade and prolonged survival (over ten years) has been reported even in the presence of metastases.

### Evaluation

Initial evaluation in the history and physical examination should be aimed at searching for a possible affected organ. Examination of the breasts, prostate, pelvis lymph nodes and rectum should be performed. Investigations including Pap smear, mammography, intravenous urogram, prostate specific antigen, chest X-ray and CT abdomen, etc. should be based on the results of the physical examination. As colonic carcinoma is commonly associated with this syndrome, carcino-embryonic antigen and stool for occult blood are often performed. Colonoscopy or barium enema is recommended at the initial evaluation and at three to five-yearly intervals.<sup>9</sup> Life-long surveillance for malignancy is required in patients with sebaceous gland tumours. Family members are also screened for sebaceous tumours and for visceral malignancies. Once detected, sebaceous lesions (sebaceous adenoma or epithelioma) should be excised. Sebaceous carcinoma should be excised with wide margins. Isotretinoin at a dose of up to 0.8 mg/kg/day has been used to prevent the cutaneous lesions of this syndrome.<sup>10</sup> As the

**Table 1. Diagnostic criteria for the Muir-Torre syndrome<sup>7</sup>**

Group A	Group B	Group C
Sebaceous adenoma	Visceral malignancy	Multiple keratoacanthomas
Sebaceous carcinoma		Multiple visceral malignancies
Sebaceous epithelioma		Family history of Muir-Torre syndrome
Keratoacanthoma with sebaceous differentiation		

Diagnosis of the Muir-Torre syndrome requires one criterion from group A and group B or all three criteria from group C without any predisposing factors for malignancy.

**Table 2. Survey of 147 patients with the Muir-Torre syndrome and internal malignancy<sup>9</sup>**

Malignancy	Percentage of patients
Colorectal	52.9
Genito-urinary	24.8
Breast	4.9
Haematological	4.9
Head and neck	3.9
Others (lung, gastro-intestinal, etc.)	8.6

malignancies in this syndrome are low-grade, surgical resection of the primary tumour may be helpful even with metastatic disease.

**Learning points:**

*Sebaceous gland tumours are uncommon but once detected, due to its association with Muir-Torre syndrome, lifelong surveillance for malignancy is required.*

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**Web sites of Dermatology & Venereology in Hong Kong**

**The homepage of the Hong Kong Society of Dermatology & Venereology**  
<http://www.medicine.org.hk/hksdv/>

**Hong Kong Dermatology & Venereology Bulletin**  
(Official Publication of the Hong Kong Society of Dermatology & Venereology)  
<http://www.medicine.org.hk/hksdv/bulletin.htm>

**Hong Kong Dermatology & Venereology Bulletin**  
(Subscription site of Blackwell Science Ltd.)  
<http://www.blacksci.co.uk/~cgilib/jnlpage.bin?Journal=hdv&File=hdv&Page=ed>

**Handbook of Dermatology & Venereology**  
(Published by Social Hygiene Service, Department of Health)  
<http://www.hkmj.org.hk/skin/>

**CME Online (Dermatology)**  
(CME Programme accredited by the Hong Kong College of Family Physicians)  
<http://www.medicine.org.hk/cme/>

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