A Case of Muire-Torre Syndrome Presenting with Sebaceous Carcinoma Arising from Bowen's Disease of the Penis and Colonic Adenocarcinoma

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Muir-Torre syndrome is characterized by the presence of sebaceous tumors and at least one internal malignancy. Sebaceous carcinoma arising from Bowen's disease is a very rare phenomenon. We describe a case of Muir-Torre syndrome initially presenting with a sebaceous carcinoma arising from Bowen's disease on the penis, which led to subsequent discovery of a colonic adenocarcinoma. We report this case to emphasize that the presence of sebaceous neoplasms should alert physicians to the possible coexistence of internal malignancy. (Dermatol Sinica 21: 255-258, 2003)

Key words: Muir-Torre syndrome, Bowen's disease, Sebaceous carcinoma

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INTRODUCTION

Muir-Torre syndrome is an autosomal dominant genodermatosis which is characterized by the presence of sebaceous tumors and at least one internal malignancy. The sebaceous tumors required to establish the diagnosis of Muir-Torre syndrome include sebaceous adenoma, sebaceous epithelioma, or sebaceous carcinoma. Sebaceous carcinoma arising from Bowen's disease is a very rare phenomenon. Only one case of Muir-Torre syndrome with sebaceous carcinoma arising from Bowen's disease on the vulva has been reported in the English language literature. We describe a patient with a sebaceous carcinoma arising from Bowen's disease of the penis leading to the subsequent discovery of colon cancer.

CASE REPORT

A 63-year-old man was seen in November 1999 with a slowly growing tumor (Fig. 1) on the dorsal aspect of his penis. The tumor had been present for six months without symptoms. Physical examination revealed a firm, brown, warty tumor measuring 2 x 1.5 cm on the proximal portion of the penis. There was no tenderness or regional lymphadenopathy. His family history was unremarkable.

Fig. 1
A 2x1.5 cm firm, brown and warty tumor on the proximal portion of the penis.

Fig. 2
Bowen's disease with atypical keratinocytes and mitosis. (H & E stain, x200)

Fig. 3
Lobules of sebaceous carcinoma with foamy cytoplasm arising from Bowen's disease. (H & E stain, x200)

Fig. 4
Adenocarcinoma of the colon. (H & E stain, x40)
A biopsy of the specimen of the tumor showed Bowen's disease and invasive lobules of tumor cells showing indicating sebaceous differentiation (Fig. 2 & 3).

We suspected our patient was a case of Muir-Torre syndrome. An adenocarcinoma of the colon was found after a thorough examination. The patient underwent hemicolecction. The tumor measured 6.5 X 4 cm in size. Histologically, it was moderately-differentiated adenocarcinoma (Fig. 4). No lymph node metastasis was found. The penile lesion was completely excised. Two years later, the patient had no clinical evidence of recurrence or metastasis.

**DISCUSSION**

Muir-Torre syndrome is an autosomal dominant syndrome with the presence of at least one sebaceous tumor (sebaceous adenoma, sebaceous epithelioma, or sebaceous carcinoma) or keratoacanthoma with sebaceous differentiation and a visceral malignancy.¹ In a recent review² of 205 cases of the Muir-Torre syndrome reported in the literature, sebaceous neoplasms were diagnosed before the internal malignancy in 45 patients (22%), simultaneously in 12 (6%), and after the internal malignancy in 114 (56%). Sebaceous carcinoma was found in 44 cases of Muir-Torre syndrome; 17 of 44 were neoplasms of the Meibomian gland. Gastrointestinal cancers were the most common internal malignancies, followed by genitourinary cancers. Our patient presented with a sebaceous carcinoma arising from Bowen's disease on the penis and concurrent adenocarcinoma of the colon. Sebaceous carcinoma arising from Bowen's disease has rarely been reported. There was only one previous case report of a sebaceous carcinoma arising from Bowen's disease on the vulva with Muir-Torre syndrome.³

Sebaceous neoplasms are found mostly on the eyelid, scalp and face. Sebaceous carcinoma occurring on the genital area is very rare. Only four previous reports were found in the literature; two sebaceous carcinomas on the penis⁴,⁵ and two on the vulva.³,⁶ Our patient was the first reported case of Muir-Torre syndrome with sebaceous carcinoma arising from Bowen's disease of the penis.

It has been estimated that 3% to 5% of patients with Bowen's disease may develop invasive carcinoma.⁷ The characteristic histologic picture of Bowen's disease with invasive carcinoma is the presence of Bowen's disease in the overlying epidermis and an invasive dermal tumor. Squamous differentiation is the most common pattern of differentiation of the invasive dermal tumor, followed by basaloid differentiation, pilosebaceous differentiation and glandular (adenoid) differentiation.⁷ The association of atypical keratinocytes and malignant sebaceous cells in our patient raises the problem of histogenesis. It has been suggested that Bowen's disease might originate from the pluripotential epidermal cells. There were some previous reports in which sebaceous differentiation within a keratinocytic lesion were described.⁶,¹⁰ A case of sebaceous carcinoma arising from Bowen's disease of the vulva was reported by Jacobs et al., in which they mentioned the pluripotential nature of the cells giving rise to invasive squamous and sebaceous neoplasms. In conjunction with previous reports, our patient seemed to show a picture of adnexal carcinoma with the differentiation of squamous cells and sebaceous cells, as described by Kao,⁷ rather than showing that of a sebaceous carcinoma arising directly from Bowen's disease.

To our knowledge, the only reported case of a sebaceous carcinoma arising from Bowen's disease was found on the vulva of an 89-year-old woman with a history of colonic cancer, who was regarded as a patient with Muir-Torre syndrome.³ Our patient had the unusual presence of sebaceous carcinoma arising from Bowen's disease on the penis, which led to a thorough investigation of internal malignancy and subsequent discovery of the adenocarcinoma of the colon. We should have a high index of suspicion in patients with sebaceous neoplasms for the presence of internal malignancies.
REFERENCES