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BASAL CELL CARCINOMA IN AN EYELID OF A FARMER WITH STURGE-WEBER SYNDROME

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Abstract: The paper presents the coexistence of a nevus flammeus and basal cell carcinoma affecting the left upper lid of 61-year-old farmer with Sturge-Weber syndrome. The occurrence of basal cell carcinoma in nevus flammeus is extremely rare. The etiology of the malignancy in this situation is unknown. It is difficult to diagnose neoplastic transformations that could arise in the nevus flammeus. Radiotherapy followed by complete surgical excision were used as a treatment in the described patient.

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INTRODUCTION

Basal cell carcinoma (BCC) of the skin often occurs in people exposed to sunlight which is why farmers and fishermen are especially predisposed for this cancer.

Basal cell carcinoma often appears as an ulceration, nodular or sclerotic lesion. It is very difficult to diagnose symptoms of neoplastic transformation arising in previously pathologically-changed skin. The occurrence of basal cell carcinoma within a port-wine stain or nevus flammeus is rare. Sixteen cases of basal cell carcinoma which developed in a port-wine stain or nevus flammeus have been reported to 2004 [3].

CASE DESCRIPTION

A 61-year-old farmer with Sturge-Weber syndrome presented an ulcerated thickening of the nevus flammeus situated on his left upper eyelid (Fig. 1). He had noticed for 5 months that the surface of the eyelid thickened, changed its colour to brighter, became ulcerated, and he felt discomfort when blinking. He had never had treatment of any kind for the nevus flammeus of the eyelid. However, nevus flammeus from the cheek were excised in 1968.

In the ophthalmological examination the right eye was normal. The left upper eyelid was thickened, sclerosing with pink-reddish nevus flammeus. The surface of the eyelid was nodular with ulcerated margin and loss of eyelashes. The conjunctival and episcleral vessels were tortuous and dilated. The anterior chamber angle was irregular and partially closed. The iris was atrophic with a distorted pupil. The lens showed initial opacification. The optic disc was glaucomatous and marginally excavated. There was not light perception in the left eye. The eyelid biopsy (but not excisional biopsy) was performed to determine whether or not skin cancer is present. The histological examination of the excised part of the eyelid showed a basal cell carcinoma. Many markedly dilated blood vessels engorged with erythrocytes were visible between cancer cells (Fig. 2, 3). In this specific condition, it was difficult to define the margins of carcinoma. The surrounding tissues appeared abnormal and the infiltration could be multifocal. Therefore, after the oncological consultation, we decided to treat the patient with ionizing radiation alone.

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Figure 1. Left eyelid: nevus flammeus with ulcerated basal cell carcinoma.

However, after 2 years we observed the recurrence, and complete surgical excision of affected part of the eyelid was performed. The defect was repaired with a full-thickness skin graft.

DISCUSSION

Nevus flammeus or port-wine stains are congenital lesions occasionally associated with other focal (glaucoma, epilepsy and mental retardation) or systemic (Sturge-Weber, Klippel Trehaunnay and Von Hippel-Landau syndromes) abnormalities.

The development of basal cell carcinoma in a nevus flammeus is an uncommon occurrence. The etiology of the malignancy is uncertain [3, 4].

The natural history of nevus flammeus with progression to an uneven surface and raised nodular hemangiomata, may delay the recognition of malignant change [4].

The development of basal cell carcinoma in skin damaged by prolonged exposure to sunlight, ionizing radiation, chronic ulcers, burns, or sebaceous nevi, has been well documented [3, 4]. It is possible that BCC may occur on and around the site of a thorium X treated port-wine stain (a natural isotope of radium which was the main radiotherapeutic modality for treating vascular lesions of the skin) [1, 2]. However, some reports suggest that radiotherapy is not an essential factor in the development of BCC in nevus flammeus [4]. Single and multiple basal cell carcinomas may arise within nevus flammeus in the absence of previous treatment with ionizing radiation [2].

The described patient was a farmer and therefore at risk of long-term sun damage. This was probably the main reason for development of the basal cell carcinoma (no previous radiotherapy or laserotherapy had been used as a treatment). In our opinion, those people with nevus flammeus who are

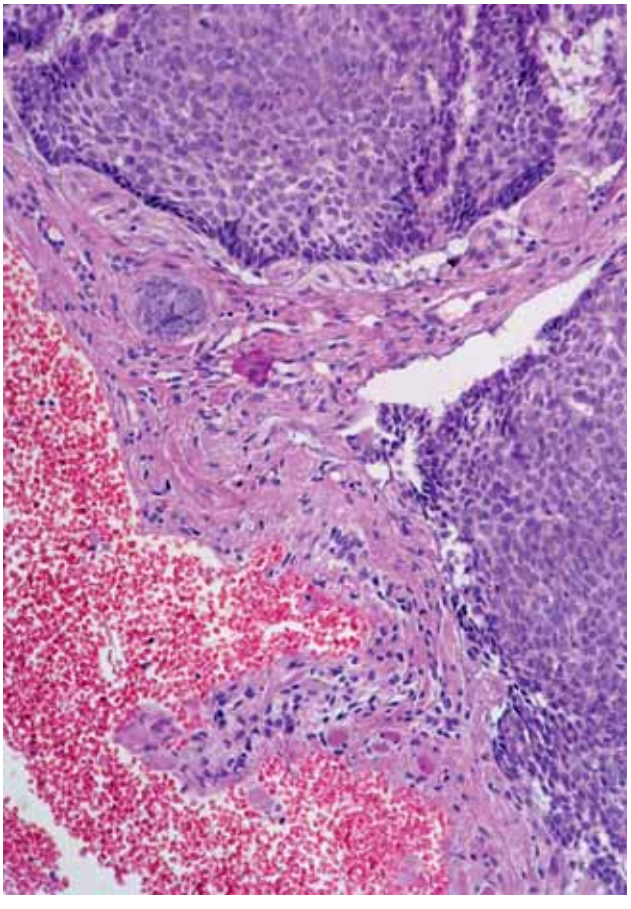

Figure 2. Histological examination of excised part of eyelid (H&E, magnification ×157).

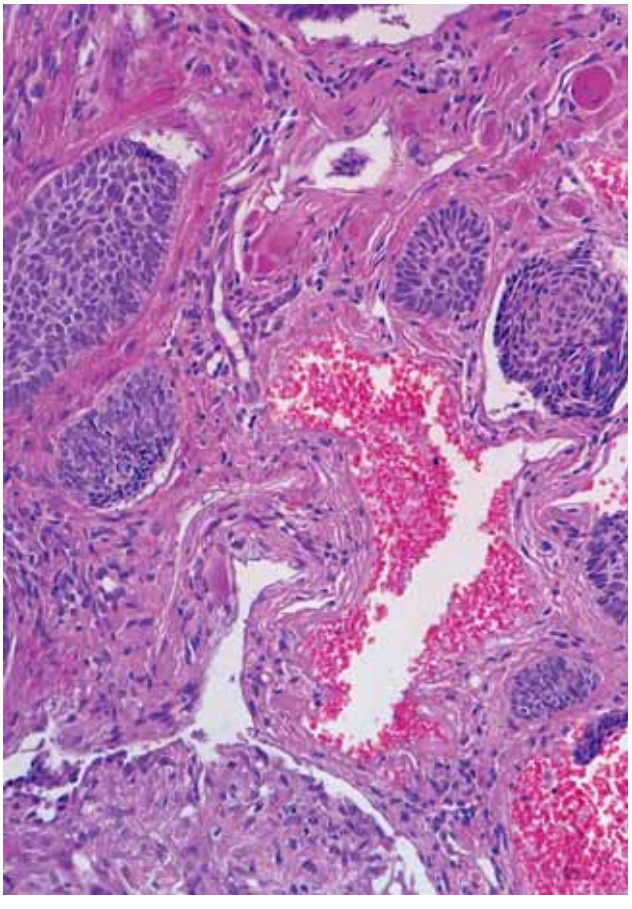

Figure 3. Histological examination of excised part of eyelid (H&E, magnification ×157).

especially exposed to the sun should pay particular attention to any changes and discoloration of the skin.

It is difficult to recognize and treat nevus flammeus because of undefined margins and possibility of massive bleeding during an operation. This is why only experienced surgeons should treat such patients.

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