Case Report / Olgu Sunumu

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A Case of Basal Cell Carcinoma Related to a Rare Condition, Bazex-Dupré-Christol Syndrome

Bazex Dupre Christol Sendromu Zemininde Gelişen Bazal Hücreli Karsinom

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Abstract / Özet

Basal cell carcinoma (BCC) is a common condition and various internal and environmental predisposing factors to BCC are described in the literature. Basex-Dupré-Christol Syndrome (BDCS) is one of the rare syndromes that predispose a patient to BCC. In these patients, BCC may be seen at a younger age, in multiple foci and with a more aggressive course. Our data show that close follow up of BDCS patients is essential. Careful observation of a simple BCC excision case can yield valuable clues for the diagnosis of a syndrome that will benefit from close follow up. We present a patient with a simple BCC lesion who was diagnosed with BDCS as a result of careful observation and investigation.

Key Words: Basal cell carcinoma, Bazex-Dupré-Christol Syndrome, familial bazex syndrome

Bazal hücreli karsinom (BCC), derinin en sık karşılaşılan malign epitelial neoplazmıdır. Literatürde BCC predispozisyonuna sebep olan çevresel ve bireysel faktörlere sıkça değinilmiştir. Bazex Dupre Christol Sendromu (BDCS), BCC predispozisyonuna sebep olan, nadir görülen sendromlardan biridir. BDCS'lu hastalarda BCC, daha erken yaşlarda, multiple odaklarda eş zamanlı olarak, ortaya çıkabilmekte ve sporadik vakalara kıyasla daha agresif seyirli olabilmektedir. Basit gibi görünen bir BCC eksizyonu vakasında bile, dikkatli bir gözlem ile karakteristik sendromik bulguları yakalamak, hastanın sıkı onkolojik takibi açısından değerli fırsatlar sunacaktır. Basit bir papüler BCC lezyonu için başvuran ve dikkatli bir inspeksiyon ve araştırma sonucu BDCS tanısı alan hastamızı sunuyoruz.

Anahtar Kelimeler: Bazal hücreli karsinom, Bazex-Dupré-Christol Sendromu, ailesel bazex sendromu

Introduction

Basal cell carcinoma (BCC), the most commonly occurring malignant epithelial neoplasm of the skin, is a slow-growing tumor that rarely metastasizes. However, if inadequately treated or left untreated, it can cause extensive local tissue destruction and slow death (1). Even in young patients, a lesion should rouse extra suspicion of BCC if the patient has a history of X-radiation exposure, arsenic ingestion, immune suppression, acquired immune deficiency syndrome, exposure to psoralens, or old scars and burns (2). Also, a clinician should be aware of nevus sebaceous of Jadassohn, porokeratosis, Gorlin's syndrome, Xeroderma pigmentosum, Rombo syndrome, albinism and linear unilateral basal cell nevus, all of which carry an increased risk for the development of BCC (3, 4).

Another condition that predisposes a patient to BCC is Bazex-Dupré-Christol Syndrome (BDCS), which is also known as Familial Bazex Syndrome and has an X-chromosome-linked dominant mode of inheritance (5). The main clinical features of this rare syndrome are follicular atrophoderma, hypotrichosis, hypohydrosis, early onset BCC and milia (6). We present a patient who had a seemingly uncomplicated BCC papule on his face who was eventually found to be affected by BDCS.

Case Report

A 35 year old man with a small non-healing papule lateral to his right lateral orbital cantus applied to our outpatient clinic (Figure 1). The papule had been growing slowly for the past 4 months. An excisional biopsy was performed because of a strong prediction of BCC. The pathological analysis yielded results that were concordant with BCC. The clinical assessment revealed follicular atrophoderma on the dorsum of his hands and elbows. The patient exhibited generalized milia. There was marked, generalized, hypotrichosis composed of sparse scalp hair, scanty eyebrows and eye lashes as well as reduced density of pubic and axillary vellus hair (Figure 2). He had been aware of generalized hypohidrosis since childhood. He stated that his two daughters had multiple milia on their faces, sparse eyebrows and hair, and follicular atrophoderma on the dorsum of their elbows. His mother, three sisters and two aunts had had milia during their childhood and both his mother and one of his sisters had undergone operations because of skin cancer (Figure 2). Genetic counseling yielded a result of an X-linked dominant mode of inheritance. The family history and the characteristic signs led us to the diagnosis of BDCS.

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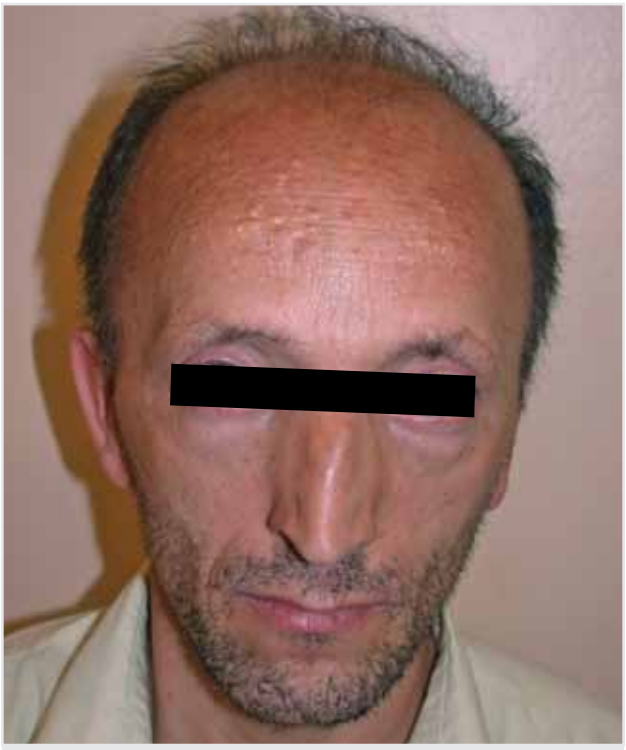

Figure 1. Prominent, multiple milia on the forehead. Hypotrichosis affecting scalp hair, eyebrows and eyelashes

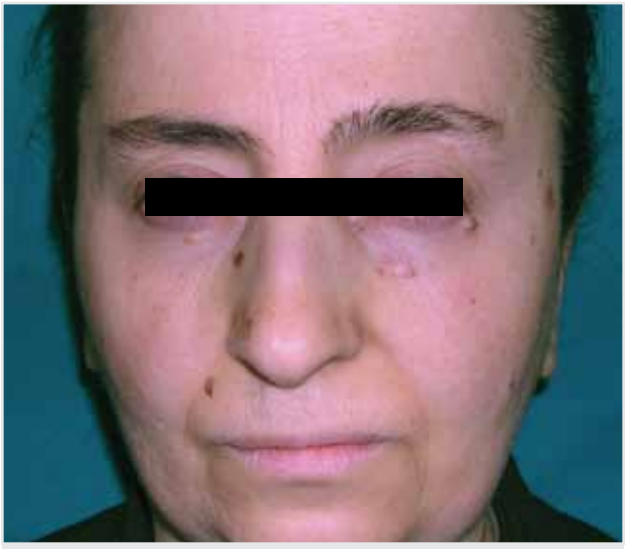

Figure 2. Patient's sister who has multiple macular and papular lesions suggestive of BCC. She already had a history of BCC excision from her left temporal area

Discussion

BDCS, first described by Bazex et al. in 1964, is a rare X-linked dominant disease and the main features of the syndrome are follicular atrophoderma, hypotrichosis, milia, hypohidrosis and BCC (5, 6). The extent and the severity of these main features vary in each individual patient. Follicular atrophoderma is encountered in about 85% of patients. It usually affects the face, the dorsum of the hands and feet, and the extensor surfaces of the elbows and knees. Hypotrichosis is another feature in about 85% of patients. It can be limited to the scalp hair, but is generalized in some patients. Milia, usually present at birth or in the infancy period, are seen in about 85% of patients and are a prominent feature in our patient and his relatives. Most often they affect only the face, but a generalized state such as milia on the face, limbs and trunk is experienced by a minority of the population. Hypohidrosis is a positive finding in about 55% of patients with BDCS (7). Early onset BCC, probably being the most noteworthy clinical feature, is seen in about 40% of patients. It is mostly localized on the face, is usually seen in the second and third decades, has an aggressive course, and is prone to relapse. All these data indicate a need for close follow up of these patients (7, 8).

We could easily differentiate our case from Rombo Syndrome and Gorlin Syndrome, which are autosomal dominant cancer syndromes. Our patient has had no skeletal abnormalities that would raise suspicion of chondroplasia punctata. Generalized basaloid follicular hamartoma syndrome (GBFHS) does not include components of BCC and atrophoderma.

Conclusion

BCC is one of the most common conditions encountered in dermatology and plastic surgery clinics. BDCS is known to cause a predisposition to BCC with a more aggressive course and multifocality. For this reason these patients deserve a close follow up with meticulous skin examinations. Our case demonstrates that deliberate assessment of incidentally recognized clinical characteristic findings, even in a case of simple BCC excision, can yield valuable results in terms of the follow up and, consequently, the prognosis of the patient.

Conflict of Interest
No conflict of interest was declared by the authors.

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Çıkar Çatışması
Yazarlar herhangi bir çıkar çatışması bildirmemişlerdir.

Hakem değerlendirmesi: Dış bağımsız.

Yazar Katkıları
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