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Becker's Nevus Syndrome: A Case Report.

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Abstract

Becker's melanosis is one of the not so common nevi, which usually appears peripubertal. Hairy or not, it seldom has extra-cutaneous manifestations, however, it has been rarely reported to be associated with other developmental anomalies in skin, muscles, and bones on the same side of the nevus in the so-called "Becker's nevus syndrome".

Introduction

Becker's melanosis is a cutaneous hamartoma characterized by well circumscribed hyperpigmented macules or patches with or without hypertrichosis. The condition was first described by S W Becker in 1949 [1]. Becker's nevus syndrome is characterized by the presence of Becker's melanosis associated with other developmental anomalies in skin, muscles, and bones on the same side of the nevus [2].

Case report

A 14 years old female patient presented with unilateral left mammary hypoplasia inspite of normal development of the contralateral one, in addition to normal developmental and pubertal milestones, Closer examination revealed a well circumscribed area of macular hyperpigmentation without hypertrichosis involving part of the area of the hypoplastic breast, which was stated to be of 3 years duration according to the patient's mother. Laboratory investigations were done and revealed no

abnormalities and all sex hormones were in normal range and mammography and breast ultrasonography revealed hypoplastic left breast. At this point, the diagnosis of Becker's melanosis with ipsilateral mammary hypoplasia was concluded. Full body scan and musculoskeletal assessment were performed and revealed no abnormalities, so we reached the diagnosis of Becker's nevus syndrome.

Discussion

Becker's melanosis is a cutaneous hamartoma characterized by circumscribed hyperpigmentation with hypertrichosis on shoulder, anterior chest, and scapular region. It usually presents unilaterally [3].

The exact pathogenesis of Becker's melanosis is not known. Many authors suggested the hypothesis of androgen hyper-responsiveness evidenced by peri-pubertal onset, male predominance, acne and hypertrichosis within the lesion [4].

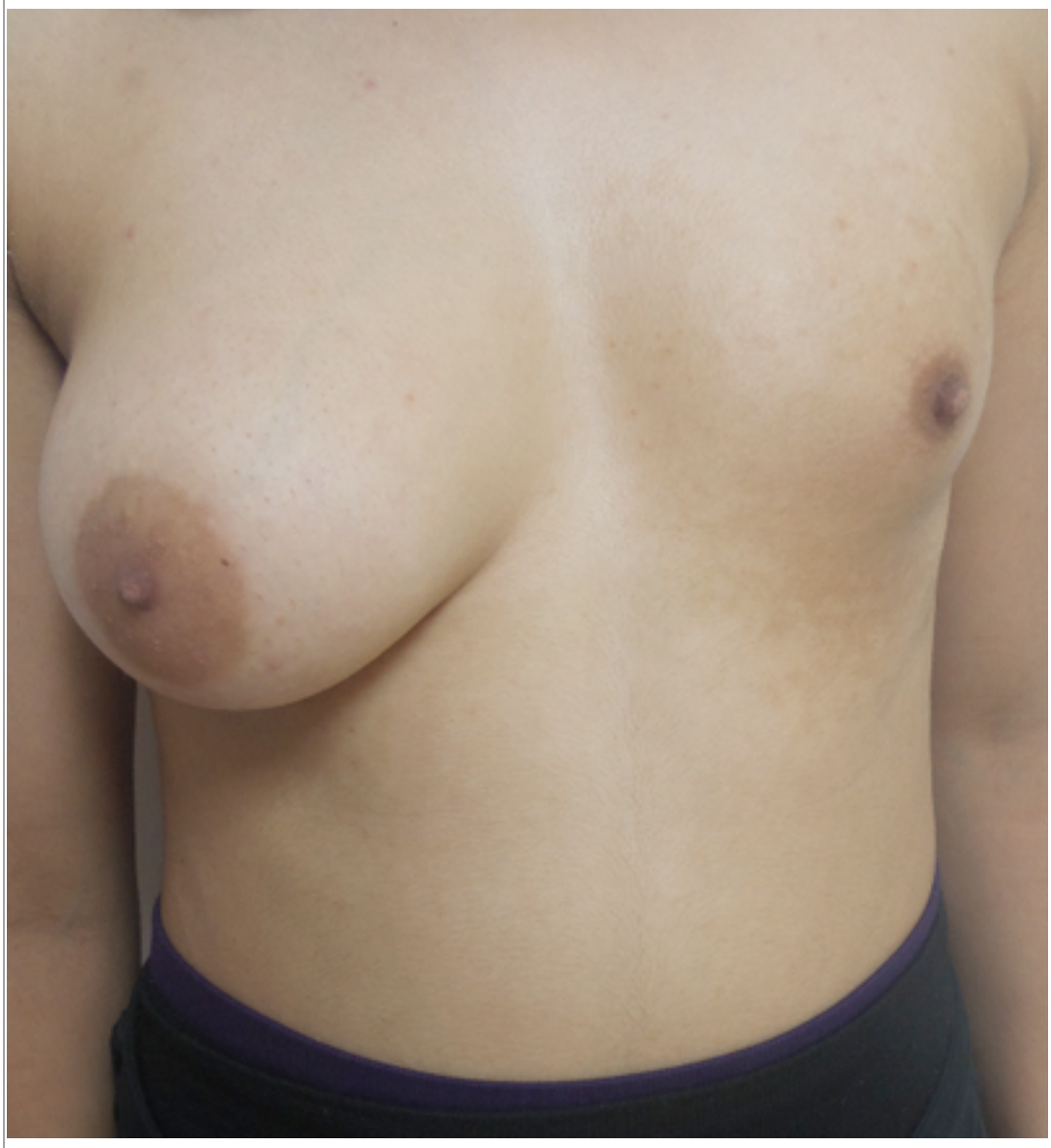
Becker's nevus syndrome is characterized by the presence of Becker's nevus with cutaneous manifestations such as; granuloma annulare, basal cell carcinoma, malignant melanoma, lymphangioma and developmental anomalies such as; osteoma cutis, hypohidrosis, spina bifida, scoliosis, pectus carinatum, congenital adrenal hyperplasia, quadriparesis, accessory scrotum[4], breast hypoplasia [5], supernumerary nipples [6], aplasia of the pectoralis major muscle [7], limb reduction [8], segmental odontomaxillary dysplasia [9], and lipoatrophy [10].

Histopathologic examination reveals acanthotic epidermis with variable degree of hyperkeratosis, elongation of rete ridges and increased amount of melanin in the basal layer [11].

Treatment modalities of the hypertrichosis component include; electrolysis, waxing, camouflage makeup and laser hair removal. Nd:YAG and Q-switched ruby laser are reported to be successful treatment modalities of the hyperpigmented part [12]. Spironolactone is a synthetic mineralocorticoid. It functions as aldosterone receptor antagonist with antiandrogen properties [13]. Spironolactone is reported to be beneficial in treatment of mammary hypoplasia as it causes down regulation of androgen receptors leading to improvement [14].

In our case, only Becker's nevus and ipsilateral mammary hypoplasia could be detected of peri-pubertal onset, and based on that the patient was set on Spironolactone 100 mg/d and is showing promising improvement in the size of the affected breast after 2 months of therapy till now,

supporting the hypothesis of the role of the androgen hyper-responsiveness in Becker's nevi.



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