

## Familial En Coupe De Sabre In Two Indian Siblings with Becker's Nevus: A Unique Rare Presentation

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### Abstract

**Observation:** En coup de sabre (frontoparietal morphea), also known as localized scleroderma, is characterized by thickening or hardening of the skin and subcutaneous tissue as a result of excess collagen deposition. Familial morphea is uncommon with only around 20 cases reported to date. To the best of our knowledge, our case reports are the only other reported case of familial en coup de sabre and the second case of horizontal transmission. There is only a single case report of coexistence of morphea and Becker's nevus so far. This report adds a unique case of concomitance of Becker's melanosis with familial en coupe de sabre to the medical literature.

### Introduction

Morphea, also known as localized scleroderma, is characterized by thickening or hardening of the skin and subcutaneous tissue as a result of excess collagen deposition. Familial morphea is uncommon with only around 20 cases reported to date [1, 2, 3]. En coup de sabre (frontoparietal morphea) has been described in a girl and her grandfather [4], and two female siblings [5]. To the best of our knowledge, ours is the only other reported case of familial en coup de sabre and the second case of horizontal transmission. There is only a single case report of coexistence of morphea and Becker's nevus so far [6]. This report adds a unique case of concomitance of Becker's melanosis with familial en coupe de sabre and idiopathic atrophoderma of Pasini and Pierini (IAPP) to the medical literature.

### Case Report

A 15 year old boy presented with single, asymptomatic, depressed, linear skin coloured lesion over forehead and scalp since 6 years which was insidious in onset, non progressive, was associated



**Figure 1.** Showing single, linear, well defined, slightly indurated plaque with dermal atrophy, and cicatricial alopecia



**Figure 2.** Showing Becker's nevus on anterolateral aspect of right thigh of the male sibling



**Figure 3.** Showing similar linear plaques in frontoparietal region in both the siblings

with decreased pinch ability of overlying skin, and hair loss over the lesion. He also developed multiple, asymptomatic, round to oval, depressed, skin coloured lesions over the back and thighs since 1 year which had been increasing gradually in size especially the lesions on the right side of the back.

His 14 year old sister, student of 9th standard also presented with similar single, asymptomatic, depressed, linear skin coloured lesion over forehead and scalp since 5 years. However, she didn't develop any skin lesions on the back, unlike her brother.

History of decrease in vision was present in both eyes in male sibling since 10 years which was more for distant vision. There was no history of headache, seizures, joint pains, any triggering factor in either of the siblings. History of autoimmunity was present in the family as their mother was a known case of hypothyroidism. No other family member had a history of similar disease.

On examination, a single, linear, well defined, slightly indurated plaque of size around 4x2cm in male sibling and 3x2cm in female sibling was present in median and right paramedian area of forehead respectively extending onto the frontal scalp, showing dermal atrophy, and cicatricial alopecia (**Figure 1**). In addition, the male sibling also had multiple, well defined, round to oval plaques of size around 1x1cm to 3x3cm on the back and thighs showing dermal atrophy, loss of hair, and prominent veins. Becker's nevus was also present on anterolateral aspect of right thigh of the male sibling (**Figure 2**). Diagnosis of en coupe de sabre was also made in his sister (**Figure 3**).

Routine investigations, radiological investigations (Chest X-ray, X-ray skull, X-ray LS spine, USG Abdomen), and thyroid function tests were normal in both the siblings. Ophthalmological evaluation of the boy revealed high myopia in right > left eye (-12

DS > -2 DS), chorioretinal atrophy (more in right eye), bilateral ptosis, and Marcus Gunn phenomenon in the left eye. His sister's ophthalmological evaluation was normal.

Histopathological examination of the linear lesions in both the siblings was consistent with the diagnosis of morphea, and Atrophoderma of Pasiini and Pierini was reported from the lesions over back in the boy.

## Discussion

En coup de sabre is an uncommon linear form of morphea that occurs on the scalp and face. Onset is usually during the first two decades of life. In addition to a morphea-like sclerosis of the skin, there can be atrophy or calcification of underlying tissues. Unlike the gradual spontaneous remissions seen in morphea, linear scleroderma tends to have a longer and occasionally progressive course [7]. Some authors suggest scleroderma en coup de sabre occurs along Blaschko lines [8], which suggests that it may arise in a mosaic clone of susceptible cells.

The etiology of localized scleroderma is unknown. Environmental exposures, immune alterations, microchimerism with autoimmunity, trauma, and Borrelia infection have been suggested as contributing to the pathogenesis of the disease [1, 7]. The vast majority of cases are sporadic. Familial morphea is uncommon and has rarely been reported in the medical literature. In 1953 Rees and Bennett documented the first familial case, which involved a father and daughter [9]. There have only been around 20 reported cases of familial morphea to date [1, 2, 3]. En coup de sabre

(frontoparietal morphea) has been described in a girl and her grandfather [4], and two female siblings [5]. To our knowledge, our patient is the only other reported case of familial en coup de sabre and the second case of horizontal transmission. It is unclear if our case represents a common household exposure, a genetic predisposition for disease, or both. Epidemiologic, genetic, familial, and twin studies suggest that scleroderma is not a heritable disease but may arise in genetically susceptible individuals when exposed to environmental triggers [1]. No clear HLA associations with localized scleroderma have been established.

Idiopathic atrophoderma of Pasini and Pierini (IAPP) and concomitant lesions of morphea have been reported commonly. Histologically the lesions are indistinguishable. IAPP appears to be a variant or an abortive form of morphea.

There have been few case reports of association of Becker's nevus with vitiligo, lichen planus, localised lipoatrophy, prurigo nodularis and more recently with osteoma cutis, and ILVEN like persistent psoriasiform dermatitis [10]. There is only a single case report of coexistence of morphea and Becker's nevus so far [6]. Our case demonstrates unique concurrence of Becker's melanosis with en coupe de sabre and IAPP, hitherto undescribed.

The chorioretinal atrophy present in the male sibling was attributed to high myopia. However, Marcus Gunn phenomenon (jaw winking syndrome) is a sporadic disorder with an autosomal dominant pattern of inheritance, but a few familial cases with an irregular autosomal dominant inheritance pattern have been reported. The wink reflex consists of a mo-

mentary upper eyelid retraction or elevation upon stimulation of the ipsilateral pterygoid muscle which may be elicited by opening the mouth, chewing, smiling, or sucking.

Further HLA typing studies and genetic investigations in familial cases are needed to substantiate the above coexistence.

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