Secondary Cutis Verticis Gyrata Due to Pilar Smooth Muscle Proliferation: Multiple Leiomyomas or Pilar Muscle Hyperplasia?

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We report a 66-year-old man with two closely neighbored, asymptomatic, markedly elevated nodu-loplaques over each eyebrow symmetrically for about 1 to 2 years. The convoluted folds and deepening furrows formed from thickened skin of and between the eyebrows resembled the pattern of cutis verticis gyrata which typically affected the scalp. Histologically, it presented as smooth muscle proliferation. However, it is unlikely for an acquired tumor, whether leiomyoma or smooth muscle hamartoma, developed on both eyebrows simultaneously. We prefer it is pilar muscle hyperplasia. Unlike previously reported cases of secondary cutis verticis gyrata, it may be the first case caused by pilar muscle hyperplasia. (Dermatol Sinica 26: 22-27, 2008)

Key words: Cutis verticis gyrata, Smooth muscle hyperplasia, Pilar muscle hyperplasia, Pilar leiomyoma, Smooth muscle hamartoma

INTRODUCTION

Cutis verticis gyrata (CVG) is a skin affection characterized by hypertrophy and folding of the skin. The lesions are mostly located on the parietal scalp but can also manifest on other parts of the body. One rare cause of secondary CVG is the skin tumor in which a dermal or subcutaneous component causes hypertrophy of the skin. The histological findings reflect the underlying condition.

Smooth muscle tumors arising in the skin have been classified traditionally into hamartomas, leiomyomas, and leiomyosarcomas. Smooth muscle hamartoma (SMH) usually occurs in association with a Becker nevus and is uncommon on the head and neck region. Multiple pilar leiomyomas, the most common type of leiomyoma, may rarely distribute in a group or in a linear pattern on the face.1,2

We presented a rare case of pilar muscle hyperplasia occurring symmetrically on the eyebrows. The differential diagnosis included SMH and pilar leiomyoma. The clinical pattern of the convoluted skin folds and deepening furrows of and between the eyebrows might be categorized in the group of secondary CVG due to pilar smooth muscle proliferation.
CASE REPORT

A 66-year-old man had two noduloplaques over each eyebrow with the development of convoluted skin folds and deepening furrows of and between the eyebrows for about 1 to 2 years. The lesions were asymptomatic. They had been first noted by the patient when he was 64 turning 65 years old and gradually became more prominent. He was certain that these lesions had not been presented during childhood.

Examination revealed two closely neighbored, markedly elevated noduloplaques over each eyebrow symmetrically (Fig. 1, 2). There was no hyperpigmentation. The hair over the noduloplaques was coarse and long but normal in density as the common eyebrows. There were 5 convoluted folds and 4 furrows formed from the thickened skin of and between the eyebrows resembling the cutis verticis gyrata pattern which typically affected the vertex and occipital region of the scalp. The folds were soft and spongy and could not be corrected by pressure or traction. The direction of the folds was longitudinal. No vermiform movement of the lesions was noticed and a pseudo-Darier’s sign could not be elicited on rubbing. The clinical impression included lipoma, leiomyoma and adnexal tumor.

A skin biopsy specimen was obtained from the most elevated area of the left medial noduloplaque. Hematoxylin and eosin staining revealed proliferative smooth muscle bundles and evenly scattered individual smooth muscle cells distributed randomly within the dermis but were still closely associated with hair follicles (Fig. 3). The discrete bundles were composed of spindle cells with eosinophilic cytoplasm and elongated, cigar-shaped nuclei. The epidermis was normal, and no basal layer hyperpigmentation was seen. The histological findings were impressed as a pilar leiomyoma or pilar muscle hyperplasia.

Immunohistochemically, the tumor masses stained positive with smooth muscle actin (Fig. 4). There was no proliferation of CD34+ dermal dendritic cells interspersing among the smooth muscle cell population; such cases of proliferation of CD34+ dermal dendritic cells had been recently described as an integral part of smooth muscle hamartoma.

Since the patient had no remarkable symptoms and he did not suffer from the cosmetic problems, no therapy was prescribed because of the benign clinical and histological nature of the lesion.
DISCUSSION

There are three types of cutaneous smooth muscle lesions: hamartoma, leiomyoma, and leiomyosarcoma. Hamartoma is a tumor-like malformation composed of an abnormal well-differentiated mixture of non-neoplastic tissue elements normally indigenous to a given tissue and therefore includes smooth muscle proliferation. Cutaneous SMH usually arises on the trunk or an extremity and presents as flesh-colored or slightly hyperpigmented plaques that often contain long vellus hairs. It is usually congenital but can also emerge later in life. In 1989, a congenital SMH on right eyebrow and eyelid was reported in an 11-year-old girl. The lesion was noted at birth. It was a 1.0 x 2.0 cm in size, pink-tan plaque with granular surface and short, dark and thick hair. The clinical manifestation of that patient was similar to our case except the age of onset and the single side.

Acquired SMH (ASMH) is rare and has been reported most frequently in association with a Becker nevus, but there are also reports of ASMH alone. The lesions distributed diffusely on genital area and neck but located unilaterally on other sites of the body.

Leiomyomas of the skin are generally divided into three categories: pilar leiomyoma, angioleiomyoma, and dartoi leiomyoma. Most are acquired. Multiple pilar leiomyomas, by far the most common type of leiomyoma, are small, firm, red or brown intradermal nodules arranged in a group or in a linear pattern. The tumors can coalesce to form plaques. The extensor surfaces of limbs, trunk, and sides of face and neck are most commonly involved. Unlike SMH, pilar leiomyoma usually grossly appears as a solitary discrete nodule. The smooth muscle bundles in pilar leiomyoma are more tightly interlacing around follicular structures, while the scattering pattern of smooth muscle bundles throughout the dermis is typical of SMH. Leiomyosarcoma displays some pleomorphism, mitotic activity, and necrosis. The tumor may extend widely into the subcutis.

Nevertheless, all these three types of cutaneous smooth muscle tumors are alleged to arise from the smooth muscle cells in the arrectores pilorum muscles, the walls of dermal blood vessels and lymphatics, the dartos muscle of the scrotum, vulva, nipple, and areola, or the cutaneous adnexa. Even though high-molecular-weight caldesmon may differentiate true smooth-muscle cell tumors from tumors of myofibroblastic or fibroblastic origin, the histologic origin of these smooth-muscle tumors cannot be assessed because the smooth-muscle cells of the various structures cannot be distinguished reliably.
In this case, we would rather call it as a pilar muscle hyperplasia than a SMH or a pilar leiomyoma because of some reasons. First, most SMH occur in association with a Becker nevus. Moreover, it is unusual for an ASMH occurring on the head and neck region. Finally, this patient had similar lesions on bilateral eyebrows. It is unlikely for an acquired tumor, whether hamartoma or leiomyoma, developed on both eyebrows simultaneously. Therefore, we prefer it is pilar muscle hyperplasia.

Cutis verticis gyrata (CVG) is a descriptive term for a condition manifesting as convoluted folds and furrows formed from thickened skin resembling cerebriform pattern. It predominantly affects the scalp and rarely the face, trunk, palms or soles. One can distinguish primary, or essential CVG, from the secondary form. In secondary forms, hypertrophy of the skin is caused by an underlying disease such as inflammation and neoplasm. The histological findings reflect the underlying condition. Solid tumors such as intradermal nevi, neuromas and neurofibromas have been reported as tumorous causes of secondary CVG. Cerebriform intradermal nevus, which shows abnormalities of epithelial and mesenchymal structures leading to inclusion in the spectrum of hamartoma, is the most common cause of secondary CVG. The case presented in this report developed several convoluted folds and deepening furrows between the eyebrows due to the underlying pilar muscle hyperplasia. We think that, our patient might be categorized in the group of secondary CVG due to pilar smooth muscle proliferation.

The treatment of secondary CVG depends on the underlying condition. Excision of the lesion may be indicated for the complications of the chronic bacterial and mycotic superinfection in the furrows. If the patient suffers from serious cosmetic impairment, partial or complete surgical removal of the lesion is indicated as well. In this case, since he had no remarkable symptoms and he did not suffer from the cosmetic problems, no therapy was prescribed because of the benign clinical and histological picture of pilar muscle hyperplasia.

In conclusion, this is a very rare of pilar muscle hyperplasia occurring symmetrically on the eyebrows, complicating by secondary CVG.

REFERENCES
我們報告一位六十六歲的男性，於兩側眉毛對稱出現各兩塊緊鄰、無症狀且明顯突起之結節性斑塊，出現時間約一到兩年。在眉毛和眉間因皮膚增厚而形成隆起的皺褶和深溝，形似典型出現在頭皮之迴狀頭皮樣表現。病理下顯示為平滑肌增生，然而後天性同時發生於兩側眉毛之腫瘤較不似平滑肌瘤或平滑肌錯構瘤的表現，因此較宜稱之為毛囊肌增生。和之前所報告過的續發性迴狀頭皮不同，它可能為首例起因於毛囊肌增生之個案。(中華皮誌 26: 22-27, 2008)