Congenital Triangular Alopecia in Phakomatosis Pigmentovascularis: Report of 3 Cases

Sir,

Congenital triangular alopecia (CTA) was first described by Sabouraud (1) in 1905, but since then has rarely been reported in the literature. The alopecia is circumscribed, non-cicatrical and non-inflammatory, and is usually located in the frontotemporal region. Associated systemic abnormalities are colonic polyposis, glaucoma, iris hamartoma, anaemia, alopecia and mental retardation.

We report here 3 cases of CTA that were seen in patients with phakomatosis pigmentovascularis (PPV).

CASE REPORTS

Patient 1 was a 3-year-old girl with naevus flammeus on the trunk and upper extremities, and mongolian spot on the trunk and predominantly on the lower extremities (Fig. 1A). Bluish irregular patches consistent with naevus of Ota were noticed on her both cheeks and right temporal scalp area. Patient 2 was a 7-year-old boy with naevus flammeus on the trunk, both extremities and left hemifacial area. Patient 3 was a 13-year-old girl with naevus flammeus on the face, trunk and upper extremities, and naevus of Ota on the peri-orbital area and naevus of Ito on both shoulders. These patients visited our clinic for treatment of their vascular and pigmented skin lesions. None of the patients had a history of other concomitant illnesses, such as seizure, glaucoma, bony abnormalities or mental retardation.

During physical examination, alopecic patches on the temporal area of the scalp were noticed in all patients. Their parents told that the patches had been there since birth. They all denied any physical trauma to their hair. A relatively well-defined triangular-shaped alopecic patch with some vellus hairs was seen on the right temporal area of patient 1 (Fig. 1B). Patient 2 also had an alopecic patch with scanty hairs on the right temporal area, and patient 3 had it on the left side. All the patients stated that the patch of triangular alopecia had remained unchanged in shape and size since it had first been noticed.

Biopsy specimens were taken from the alopecic patches and perilesional normal skins in all 3 patients to compare hair patterns between 2 areas. Biopsy of the lesional skin in patient 2 showed tiny follicles with immature hairs compared with normal scalp (Figs 2A and 2B). Only sparse inflammatory cells were seen around hair follicles. Biopsies from the other 2 patients showed similar pathology.

DISCUSSION

PPV is characterized by a distinctive association of extensive naevus flammeus with a pigmented naevus and, in some cases, with a naevus anemicus (2–5). The pathogenesis of this rare disorder remains to be clarified, although several hypotheses have been made to explain this disorder (4, 6, 7). Various other associated abnormalities have been described. The most frequent are psychomotor retardation, epilepsy, intracranial calcifications and cerebral atrophy (8). PPV associated with alopecia is rarely reported.

CTA is a circumscribed, non-cicatrical, non-inflammatory hypotrichosis that is usually located in the frontotemporal region. It is manifested at 3–5 years of age by unilateral or, less frequently, bilateral patches of alopecia and seems not to have a sex predilection (9–11). There appears to be no obvious phylogenetic or embryological explanation for these lesions. CTA is not known to be of any medical importance, except that it should not be misdiagnosed as alopecia areata.

When it is misdiagnosed as alopecia areata, adverse side-effects of long-term treatment, such as skin atrophy due to steroids, may develop in the treated area (10). CTA specimens show no inflammatory or cicatrical changes and the absence of mature hairs, whereas a specimen of alopecia areata reveals perifollicular infiltrates and the presence of dystrophic hairs and melanin incontinence in fibrous tracts (11, 12). Histo-
logical examination of temporal alopecic lesions in our patients showed decreased hair follicle sizes without distinctive perifollicular inflammatory cell infiltration, which were compatible with the findings in the lesion of CTA. The lesion remains unchanged throughout the patient’s life and there is no agreement about the treatment of CTA, although hair grafts or excision of the lesion may be useful (11–13).

Both PPC and CTA are uncommon disorders, but there have been a few cases in which both PPV and CTA were found in the same patients (14–16). Because of the rarity of this association, we cannot elucidate any possible explanation as to whether their association is coincidental or significant. Further observations will be necessary.

REFERENCES


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