

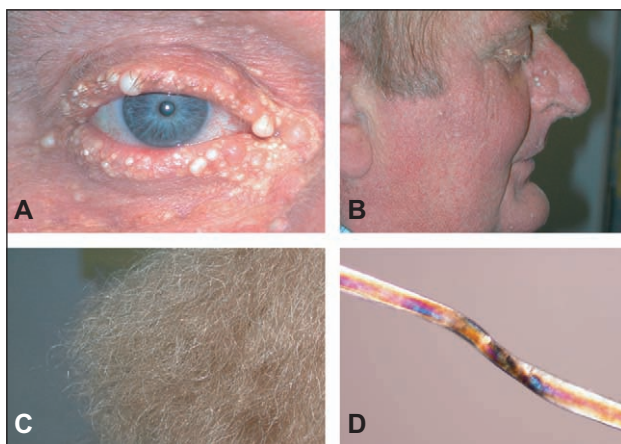


Figure 1. A) Multiple apocrine hidrocystomas of the eyelid. B) « Birdface » appearance. C) Woolly hair appearance. D) Narrowed hair shaft.

ents presented no similar manifestations and no history of consanguinity. The patient had no siblings or children. Since its initial description by Schöpf [1], 24 cases of SSP have been reported. Although the transmission seems to follow an autosomal recessive pattern, the possibility of a dominant mode remains discussed. Sporadic cases and consanguinity are reported [2]. Recently, locus homogeneity was suggested for a recessive mutation with instances of pseudo dominant inheritance [2].

The diagnosis of SSP relies on major and minor criteria [3]. The major criteria include hypodontia with a normal or aberrant temporary dentition and an absence or a severe alteration of the permanent teeth. The palmoplantar keratoderma usually appears progressively during adolescence. The bluish and/or pinkish eyelid cysts are apocrine hidrocystomas and usually appear later in life [2]. They are present in all the published cases. Calcification of these cysts may occur. Multiple eccrine hidrocystomas have been reported. The tumours can be located on the ears, the shoulders, the feet, the external genitalia, or the scalp. Their pathogenesis remains hypothetical, either deriving from adenomatous proliferations from normal Moll glands [1] or originating from ectopic residues of foetal apocrine glands. The nail appearance is highly polymorphic, including anonychia, brittle and/or furrowed ungual alterations; others may be affected by dystrophic changes. Hypotrichosis also represents a major diagnostic criterion [2].

The minor diagnostic criteria feature the following: facial dysmorphism (birdface) [1] and multiple eccrine syringofibroadenomas on the palmoplantar hyperkeratotic areas [2, 3]. As far as we know, the histological alterations of the hair in our case have never been described before. None of the patient's medications has been associated with the induction of pili torti.

SSP patients are prone to develop benign tumours such as eccrine poromas, tumours of the follicular infundibulum, precancerous keratoses, and eccrine syringofibroadenomas [4, 5]. Furthermore, basal cell and squamous cell carcinomas are not uncommon [1, 4]. Hence, long-term clinical surveillance is indicated [5]. The presence of multiple basal cell and squamous cell carcinomas in our patient may also be explained by long-term immunosuppressive therapy for cardiac transplantation. No specific treatment

is available for SSP. Long term management involves dental, dermatological, and ophthalmological follow-up and is tailored individually. Although recurrences are frequent, surgery remains the treatment of choice for the eyelid cysts, eventually with a complete excision of the anterior lamella of the eyelid. Topical trichloroacetic acid may be used, sometimes complicated by ectropion formation or corneal injury [6].

In conclusion, SSP is a rare genodermatosis of unknown pathogenesis. The pili torti appearance of the hair and the hair shaft narrowing have never been reported before and may represent a new criterion for diagnosis of SSP. ■

Acknowledgements. The authors declare no conflict of interests. No funding sources support this work.

¹Department of Dermatology,
Clinique Sainte-Elisabeth, B-5000,
Namur, Belgique

²Department of Dermatology,
CHU du Sart-Tilman, B-4000,
Liège, Belgique

³Département de Pathologie,
Clinique Sainte-Elisabeth, B-5000,
Namur, Belgique

⁴Department of Dermatology,
CHU Saint-Pierre, B-1000,
Bruxelles, Belgique
<af.nikkels@chu.ulg.ac.be>

Grégory SZEPEŦIUK^{1,2}

Olivier

VANHOOTEGHEM^{1,2}

Gebhard MULLER³

Jean Jacques STENE⁴

Arjen F. NIKKELS²

1. Schöpf E, Schulz HJ, Passarge E. Syndrome of cystic eyelids, palmo-plantar keratosis, hypodontia and hypotrichosis as a possible autosomal recessive trait. *Birth Defects* 1971; 7: 219-21.

2. Castori M, Ruggieri S, Giannetti L, Annessi G, Zambruno G. Schöpf-Schulz-Passarge syndrome: further delineation of the phenotype and genetic considerations. *Acta Derm Venereol* 2008; 88: 607-12.

3. Hampton PJ, Angus B, Carmichael AJ. A case of Schöpf-Schulz-Passarge syndrome. *Clin Exp Dermatol* 2005; 30: 528-30.

4. Starink TM. Eccrine syringofibroadenoma: multiple lesions representing a new cutaneous marker of the Schöpf syndrome, and solitary nonhereditary tumors. *J Am Acad Dermatol* 1997; 36: 569-76.

5. Alessi E, Gianotti R, Coggi A. Multiple apocrine hidrocystomas of the eyelids. *Br J Dermatol* 1997; 137: 642-5.

6. Dailey RA, Saulny SM, Tower RN. Treatment of multiple apocrine hidrocystomas with trichloroacetic acid. *Ophthalm Plast Reconstr Surg* 2005; 21: 148-50.

doi:10.1684/ejd.2009.0743

Allergic contact dermatitis induced by topical hydrocortisone butyrate propionate mimicking acute generalized exanthematous pustulosis

Herein, we report a rare case of allergic contact dermatitis induced by topical hydrocortisone 17-butyrate 21-propionate (HBP), which resembled acute generalized exanthematous pustulosis (AGEP).

A 31-year-old Japanese woman with atopic dermatitis was treated by topical application of Pandel[®] ointment, containing HBP. Soon after the start of application of the ointment, she developed edematous erythema with pustules at the sites of application of the ointment, and was referred to our department because the eruptions spread rapidly within two days to involve large parts of the body, associated with high-grade fever. At the first examination, erythematous papules and irregular erythematous

lesions with purpura were noted on the back and lower limbs (*figure 1A*). Numerous pustules were also observed on the neck, axilla and arms, however, there was no evidence of mucosal involvement. Laboratory studies revealed elevated white blood cell and neutrophil counts with a slight increase in the eosinophil count; serum tests for antistreptolysin O and viral antibodies (Epstein-Barr virus and cytomegalovirus) were negative. Bacterial and fungal cultures of blood, tonsillar swabs and material obtained from the pustules were negative.

Histopathological examination of the pustules showed that they arose in the subcorneal layer, with inflammatory cell infiltration of the dermis; there was no evidence of vasculitis (*figure 1B*). We discontinued the Pandel[®] ointment, and started her on oral betamethasone. Two weeks later, the eruptions had completely disappeared.

Positive reactions were seen to Pandel[®] ointment 0.1% pet. and HBP 0.01% pet. at 48 hr and 72hr, and 7days (*figure 1C*). In addition, patch tests conducted for hydrocortisone butyrate (HB) 0.1% pet. and prednisolone valerate acetate (PVA) 0.1% pet. also revealed positive reactions (*figure 1D*), whereas patch testing for other corticosteroids was negative.

HBP, HB and PVA, but not the other corticosteroids tested, have an ester substitution at the C17 position. Based on these results, we diagnosed the condition of the patient as pustule-forming allergic contact dermatitis induced by topical C17-substituted corticosteroid preparations.

Our patient showed an important difference from cases of systemic contact dermatitis caused by cobalt and nickel [1].

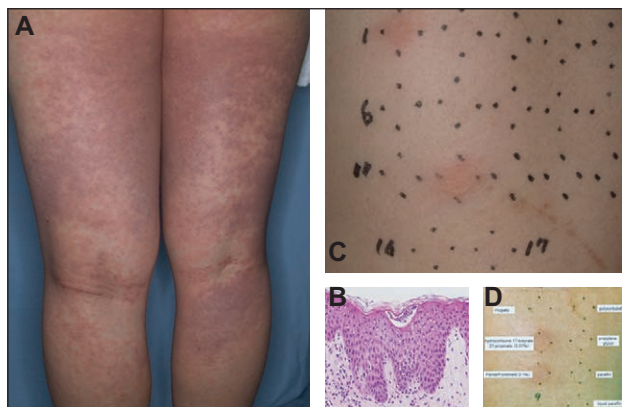


Figure 1. A) Widespread and scattered purplish erythematous lesions on the legs. B) Skin biopsy specimens showing subcorneal pustules, but no evidence of vasculitis (Hematoxylin and eosin stain, original magnification $\times 200$). C) A positive patch test reaction to Pandel[®] ointment and hydrocortisone 17-butyrate 21-propionate at 7 days; the same reactions were, however, also seen at 48 and 72 h (not shown). D) A positive patch test reaction to hydrocortisone 17-butyrate and prednisolone valerate acetate at 7 days. Cross reactivity to hydrocortisone 17-butyrate 21-propionate, hydrocortisone 17-butyrate, and prednisolone valerate acetate is evident, and all of these corticosteroids have an ester substitution at C17. The patch testing was negative for betamethasone valerate, betamethasone butyrate propionate, clobetasol propionate, clobetasone butyrate, dexamethasone propionate, diflucortolone valerate, fluocinonide, fluocinolone acetate, methylprednisolone, mometasone furoate and prednisolone.

In our case, the histopathological examination revealed subcorneal pustules and the clinical manifestations included numerous pin-sized pustules and irregular erythematous lesions with purpura. These findings are rarely observed in systemic contact dermatitis. Assessment of our patient according to the Euro SCAR group criteria revealed a score of 8 [2], however, the patch test reaction was mildly positive. Based on these findings and previous descriptions of the condition [3], we diagnosed our patient as a case of AGEF-like allergic contact dermatitis [3]. AGEF is well known to be induced by oral antibiotics and mercury, but AGEF or AGEF-like eruptions have rarely been reported to be caused by topical corticosteroids [3, 4]. A large study revealed that 1% of patients show a positive patch test reaction to HB [5]. HB, HBP and PVA have an ester substitution at the C17 position, and this C17-ester has an allergenic property, similar to C17-acetonide, such as in budesonide [6]. The mechanism of development of AGEF in our case remains unknown. However, systemic absorption of topically applied agents can cause AGEF or AGEF-like eruptions [2-4]. Our case was a rare one, but nonetheless, posed a serious problem. ■

Acknowledgements. Financial support: none. Conflict of interest: none.

Department of Dermatology,
Kitasato University School
of Medicine, 1-15-1 Kitasato,
Sagamihara, Kanagawa, 228-8555,
Japan
<hm4765@yahoo.co.jp>

Natsuko TOHGI
Hiromitsu ETO
Hideki MAEJIMA
Norimitsu SAITO
Mika NAKAMURA
Kensei KATSUOKA

1. Asano Y, Makino T, Norisugi O, Shimizu T. Occupational cobalt induced systemic contact dermatitis. *Eur J Dermatol* 2009; 19: 166-7.
2. Sidoroff A, Halevy S, Bavnick JNB, et al. Acute generalized exanthematous pustulosis (AGEP) – a clinical reaction. *J Cutan Pathol* 2001; 28: 113-9.
3. Broesby-Olsen S, Clemmensen O, Andersen KE. Allergic contact dermatitis from a topical corticosteroid mimicking acute generalized exanthematous pustulosis. *Acta Derm Venereol* 2005; 85: 444-61.
4. Mur EC, Ballesteros MGC, Fernández RS, et al. Generalized exanthematous reaction with pustulosis induced by topical corticosteroid. *Contact dermatitis* 2005; 52: 114-5.
5. English JSC. Corticosteroid-induced contact dermatitis: a pragmatic approach. *Clin Exp Dermatol* 2000; 25: 261-4.
6. Dooms-Goossens A, Anderson KE, Brandao FM, et al. Corticosteroid contact allergy: an EECDRG multicentre study. *Contact dermatitis* 2004; 51: 47-56.

doi:10.1684/ejd.2009.0744

Henoch-Schönlein purpura and aromatase inhibitors

The third-generation aromatase inhibitors (AIs) have only recently become standard care in the adjuvant treatment of postmenopausal women with endocrine-responsive breast cancer, according to internationally recognized guidelines. The Arimidex, Tamoxifen, Alone or in Combination (ATAC) trial has clearly demonstrated a statistically significant advantage of anastrozole, a reversible non-steroidal AI, over tamoxifen in disease-free survival (DFS) and time to recurrence (TTR) [1]. These results have led with time to the widespread use of anastrozole as initial therapy in the adjuvant setting of early breast cancer. Side effects more commonly encountered during AI therapy are diffuse arthralgia and myalgia, bone loss and