To the Editor

Hereditary angioedema (HAE) is a rare autosomal dominant disorder characterized by a quantitative or functional deficiency of Cl esterase inhibitor (Cl-INH) [1]. Attacks of angioedema may be induced by a variety of factors, such as tissue trauma, infection, anxiety, and fatigue [1]. The condition apparently is further influenced by sexual hormones [2–4].

Case Report. A 20-year-old previously healthy woman was given Diane® (containing 0.035 mg ethinyl estradiol and 2 mg cyproterone acetate) as treatment for acne. Three weeks after institution of the therapy she experienced attacks of poorly circumscribed, nonpitting and nonpruritic cutaneous swellings, along with occasional abdominal pain. The edema involved particularly the extremities, the trunk and further pressure-exposed sites. The episodes lasted for up to 3 days and occurred weekly. After 2 months she changed to Trinordiol® (containing 0.05–0.125 mg levonorgestrel and 0.03–0.04 mg ethinyl estradiol) as oral contraceptive agent (OCA), but attacks persisted. She was therefore given Lutényl® (containing 5 mg of normegestrol acetate) and she was free of symptoms until she discontinued OCA 2 months later. Responses to repeated pressure challenges were all the time negative. Extensive laboratory investigations, including search for autoan-tibodies, did not reveal any abnormalities, except of the complement profile (table 1).

Letters to the Editor

Table 1. Complement profile 1 month after stop of OCA treatment

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<tr>
<th>Patient</th>
<th>Normal range</th>
<th>300 SIß0/ml</th>
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gests that progestational agents, such as normegestrol acetate, may be of benefit in young women with hereditary angioedema requiring an oral contraception.
Functional assay CH50 Functional Cl-INH
Formation of Cl-INH-CIr

undetectable

1

0.04

114 115 0.93 0.07
70 -135% 80 -125 %4 70 -130%5
0.11–0.26 g/l 75 -125% 67 -127% 0.75–1.40 g/l 0.14–0.35 g/l

complexes2 Exog. Cls (amidolysis3) Exog. Cls (amidolysis) Protein concentrations6 Cl-INH Clr
C2
C3/C3c C4

1 U corresponds to the amount of serum (ml) which lyses 50% of 3·106 optimally sensitized sheep erythrocytes within 45 min at 37 °C in a total volume of 0.5 ml.
2 This assay directly measures interaction of Cl-INH with its natural substrate, Clr, at physiologic concentrations of Clr and Cl-INH[according to 7].
3 Based on inhibition by Cl-INH of amidolytic activity of exogenous Cls added to the sample to be tested, using commercially available testkits.
4 Kits from Beringwerke, Marburg, FRG.
5 Kits from Immuno, Vienna, Austria.
6 Assessed by nephelometry or radial immunodiffusion using mono-specific antisera.

The father of our patient, 3 of her paternal uncles and 1 aunt had a history of repeated attacks of angioedema, occasionally in association with the intake of salicylates and insect bites.

Comment. In the present case clinical features, complement profile and family history are consistent with a diagnosis of hereditary angioedema. In this disorder, the first clinical manifestations often occur during puberty and symptoms may be further elicited or aggravated by menstruation, suggesting a direct relationship between hormonal changes and angioedema. In addition, angioedematous attacks induced by OCA have been rarely described [2–5].

In line with previous reports, our patient experienced the first symptoms shortly after institution of the OCA therapy [2–5]. The estrogen-related compounds contained in the OCA appear to precipitate the attacks [2–5]. Cyproterone acetate has not been shown to aggravate clinical manifestations in hereditary Cl-INH deficiency [6], however, it has been recently implicated in the development of angioedema in female patients with normal Cl-INH function [5]. Interestingly, the swellings disappeared while the patient was on normegestrol acetate, a progestogen derived from 17-OH progesterone. In fact, in order to avoid the frequent side effects observed in female patients with Cl-INH deficiency receiving long-term prophylactic therapy with attenuated androgens, such as danazol, clinical efficacy of synthetic progestatins with anabolic properties has been recently assessed [8].

Our observation thus confirms that OCA containing estrogens may precipitate potentially dangerous attacks of edema, and further sug-

About the Association of Lichen planus and Psoriasis
Sir,
W. We read with interest the report by Shiohara et al. about the coexistence of lichen planus and psoriasis in a single patient in a recent issue of Dermatologica [1].
Between September 1986 and February 1988 the Gruppo Italiano Studi Epidemiologici in Dermatologia (GISED) conducted a multi-