7). In our patient there was no history of any injury to the head and neck or parotid disease, physical examination revealed no neurological signs, no associated Horner's syndrome or other autonomic dysfunction was observed and magnetic resonance imaging of the brain and brainstem were normal.

There are few reports demonstrating the presence of enlarged sweat glands in the affected skin of patients with localized hyperhidrosis and the lesions have been considered as variants of the pure anatomical eccrine naevi or as functional naevi which showed secondary hypertrophy of the glandular elements (1, 9, 10). But this finding has not been reported in most of the patients with localized hyperhidrosis, and skin biopsies in our patient failed to reveal such findings.

According to some authors (4), localized hyperhidrosis may be caused by a non-demonstrable disturbance of autonomic nervous system, but the reason for this rare condition remains to be elucitated.

Because of the limited number of cases reported, there is no standard therapy for idiopathic unilateral localized hyperhidrosis and the condition often proves resistant to therapy. Sweating may partially be controlled by treatments with anticholinergic drugs, sedatives, tranquillizers, calcium-channel blockers, astringents, topical antiperspirants, iontophoresis and surgical sympathectomy. However, these treatments have various side-effects and topical therapies tend to cause contact dermatitis (1, 5, 10). In our patient treatment with amitryptiline in a dosage of 20 mg daily combined with topical aluminium chloride cream 20% was not found to be effective.

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Pityriasis Rubra Pilaris: A Retrospective Analysis of 43 Patients

Sir,

Pityriasis rubra pilaris (PRP) is a rare skin disease of unknown aetiology. Since 1980 we have seen 43 patients with this diagnosis. This study describes these patients and the course of their disease, based on a retrospective analysis.

MATERIALS AND METHODS

Patients

The study comprised 43 patients (12 females and 31 males). Their age range at the start of disease was 1–77 years (Fig. 1). Forty-one of the patients were admitted for investigations and treatment, while only 2 had outpatient treatment. Nine women and 24 men (mean age 58 years) had adult onset of disease (>15 years of age) and 3 girls and 7 boys (mean age 8 years) had juvenile onset (<15 years of age).

Diagnosis

The clinical picture is somewhat different for the various types of PRP. In its classical form, the skin eruption most often starts in the face and associated areas and then spreads within a few weeks to the rest of the body, leaving only small islands of unaffected skin (Fig. 2). The eruption is erythematous and scaling with follicular plugging. The palms and soles are yellowish and hyperkeratotic. Within 2–3 months erythrodermia may develop. Some patients also develop ectropion (1).

In our material it was not possible to distinguish between the various types of PRP described (2), because the clinical information in the medical reports was not sufficiently accurate.

Most patients were diagnosed from the clinical picture and the diagnosis was verified histologically. In 8 patients the diagnosis could not be verified by our pathologist. However, the clinical pictures were so characteristic that the diagnosis was maintained. Five of those patients were females. This means that in 42% of the females, the diagnosis of PRP could not be verified histologically, whereas this was the case in only 10% of the males ($p<0.05$).

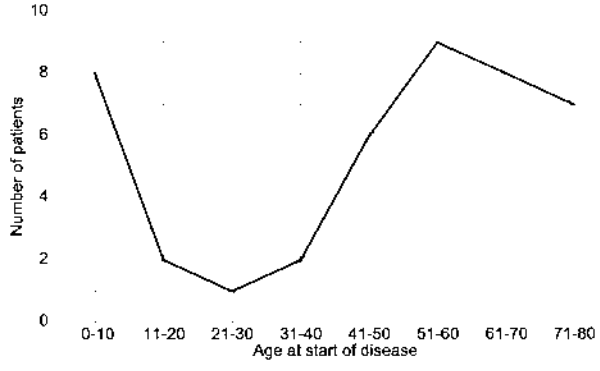

Fig. 1. Age distribution at the start of the disease.

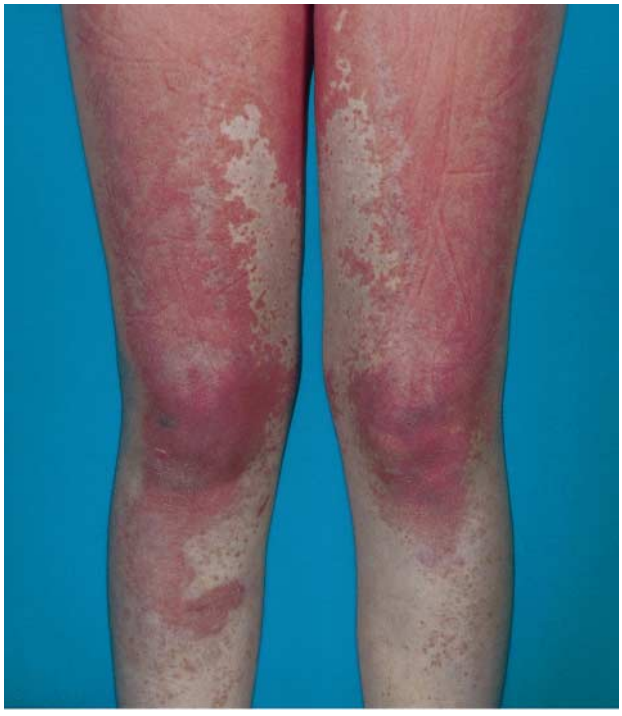

Fig. 2. An 11-year-old-girl with classical juvenile onset of pityriasis rubra pilaris.

Treatment and prognosis

The patients were treated with either acitretin or methotrexate. All received topical treatment with emollients and steroids. A few got photopheresis. Treatment with PUVA does not help these patients, as sunlight often worsens the condition.

Ten of the patients had an unknown course of PRP due to lack of follow-up. Nine patients are currently in treatment. One patient died. The remaining 23 patients had remission after approximately 2 years (range 2 months to 7 years). If they are divided in adults and juveniles, the adults had remission after approximately 2.5 years (range 5 months to 7 years). The juveniles had remission after approximately 1 year (range 2 months to 5 years).

DISCUSSION

The sex ratio is normally said to be 1 : 1 for PRP (1, 3), but in our material we had 28% females and 72% males. In Denmark it is known that women are more likely than men to see a doctor. All, but 2 of our patients were admitted because of severe skin disease, and there may be a selection bias, if patients with less severe PRP are not referred to us. It could indicate that women have less pronounced PRP than men, a hypothesis supported by our histological findings, where women had significantly less histological changes diagnostic of PRP than men.

PRP almost always appears suddenly without any preceding factors. However, in some children the disease is preceded by an infection (1). Two children had onset of PRP after influenza and what seemed like pneumonia. One patient had aggravation after tonsillitis and after appendectomy.

PRP appears sporadically, but there have been reports of autosomal dominant inheritance (4). None of our patients seemed to have inherited disease. Eleven of them, however, were predisposed to psoriasis, indicating that there could be a close association between these diseases.

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