(Fig. 1). The lesions consisted of dark-brown papules coalescing to form a plaque of approximately 3×5 cm on both cheeks. The lesions showed a bright yellow fluorescence in Wood's light and microscopy (Scotch tape stained with methylene blue) showed numerous clusters of *P. orbiculare*, but only in the yeast form.

Skin scrapings for culture were taken with a curette on a peptone–glucose–yeast extract medium containing glycerol monostearate (2.5 g/l) and Tween (2 ml/l), overlaid with olive oil. Culture showed abundant growth of *P. orbiculare*. Histology showed atrophy of the malpighian layer and a non-specific perivascular infiltrate of lymphocytes and granulocytes in the dermis (Fig. 2). Periodic acid-Schiff stain (PAS) showed some *P. orbiculare* yeasts in the stratum corneum (Fig. 3).

The patient was treated with miconazole cream 2% and the lesions disappeared after 2 weeks, as judged clinically, in Wood's light, and microscopically. They reoccurred however on the right cheek after further 4 weeks, but disappeared again after the same treatment. After 4 weeks they reoccurred, now on the left cheek, but again they disappeared after treatment with miconazole cream.

DISCUSSION

The aetiology of CRP is still unknown. The disease is most often seen in females shortly after puberty and an endocrine disturbance has been suggested, but most of the patients have been in good health. Roberts and Lachapelle associated *P. orbiculare* with CRP (4), but the role of *P. orbiculare* in CRP is still unknown. That the presence of *P. orbiculare* is significant in CRP is suggested by the beneficial effect of antifungal agents both in this and in other reports (4, 6). CRP is usually localized to seborrheic areas on the trunk and more seldom to the face, arms or legs. In this case lesions were only seen on the cheeks and the histological picture did not show hyperkeratosis or papillomatosis. The clinical picture in connection with fluorescence in Wood's light and the presence of *P. orbiculare* microscopically confirmed the diagnosis of CRP. The lesions reoccurred after a short period but each time they disappeared after antifungal treatment.

P. orbiculare links CRP to tinea versicolor. Even the distribution primarily to seborrheic areas, the pigmentary disturbances, and the clearance of lesions with antifungal agents show similarities. In tinea versicolor the conversion of *P. orbiculare* from the yeast form to the mycelial form is responsible for the disease (3, 5). In CRP this conversion is not seen and an abnormal host response to the fungus rather than a strictly infectious reaction may better explain the role of *P. orbiculare* in this disease.

REFERENCES

1. Faergemann, J.: Experimental tinea versicolor in rabbits and humans with *Pityrosporum orbiculare*. J Invest Dermatol *72:* 326, 1979.
2. Gougerot, H. & Carteaud, A.: Papillomatose pigmentée innominée. Bull Soc Franc Derm Syph *34:* 719, 1927.
3. McGinley, K. J., Lantis, L. R. & Marples, R. R.: Microbiology of tinea versicolor. Arch Dermatol *102:* 168, 1970.
4. Roberts, S. O. B. & Lachapelle, J. M.: Confluent and reticulate papillomatosis (Gougerot–Carteaud) and *Pityrosporum orbiculare*. Br J Dermatol *81:* 841, 1969.
5. Tosti, A., Villardita, S. & Fazzini, M. L.: The parasitic colonization of the horny layer in tinea versicolor. J Invest Dermatol *59:* 233, 1972.
6. Yesudian, P., Kamalam, S. & Razak, A.: Confluent and reticulated papillomatosis (Gougerot–Carteaud). Acta Dermatovener (Stockholm) *53:* 381, 1973.

Eosinophilic Fasciitis (Shulman Syndrome) in a 13-year-old Girl

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Received October 27, 1979

Abstract. Symptoms compatible with the diagnosis of eosinophilic fasciitis (morphea-like changes of the skin, restricted joint movement, eosinophilia, elevated immunoglobulin levels in serum) in a 13-year-old girl and the response to systemic corticosteroid treatment are reported. The literature is reviewed.

Key words: Eosinophilic fasciitis; Shulman syndrome; Localized scleroderma; Eosinophilia

In 1974 Shulman described in two male patients what he thought to be a new syndrome, characterized by localized cutaneous morphea-like changes (typically of the extremities, with varying degrees of mobility restriction of the joints), severe inflammation of the deep fasciae, eosinophilia, and selective elevation of IgG levels (12).

Up till now 35 cases of this "eosinophilic fasciitis" (EF) have been reported, almost exclusively in the US (Table I), but to our knowledge none have been described in Scandinavia. In spite of the rather alarming symptoms, early systemic treatment with corticosteroids seems effective. For this reason we

Table I. *Clinical findings and evaluated effect of corticosteroid treatment in 36 patients with EF*

+ indicates positive findings or instituted or effective treatment. – indicates negative findings or no or ineffective treatment. 0 indicates no information on symptoms or treatment

Author/year	Sex		Age	Eosinophilia			Elevated IgG			Cutaneous changes			Fasciitis			Decreased mobility		
	M	F		+	0	–	+	0	–	+	0	–	+	0	–	+	0	–
Shulman, 1974/75 (12)	3	1	19–53	4			4			4			4			4		
Rodnan, 1975 (9)	5	2	11–60	7			6	1		7			7			7		
Caperton, 1975 (4)	3	2	14–46	5			5			5			5				5	
Schumacher, 1976 (10)	1		28	1				1		1			1			1		
Talbot, 1976 (13)	1		57			1	1		1	1			1			1		
Athernon, 1976 (2)	1		49	1			1			1				1		1		
Ansell, 1976 (1)		1	14	1			1			1			1			1		
Camus, 1976 (3)	2		41–52	2			2			2			2			2		
Krauser, 1977 (8)		1	22	1			1			1			1			1		
Gray, 1977 (7)	1		36	1			1			1			1			1		
Torres, 1977 (14)	1		57			1	1			1			1			1		
Chanda, 1978 (5)	1	1	48	2			2			2			2			2		
Shewmake, 1978 (11)	1		36	1					1	1			1			1		
Weinstein, 1978 (15)		1	55	1					1	1			1			1		
Fleischmajer, 1978 (6)	4	2	39–88	6				4	2	6			3	3		1	5	
Present case, 1978		1	13	1			1			1			1			1		
All patients	24	12	11–88	34	0	2	26	6	4	36	0	0	32	4	0	26	10	0

think it of interest to report another case of EF successfully treated with corticosteroids.

CASE REPORT

A 13-year-old girl was in good health until February 1978 when she developed an increasing dull pain and fatigue in her left leg. Simultaneously she noticed decreasing mobility of her left knee and ankle joint and scattered patches on the skin above and below the left knee. She felt well otherwise and, in particular, denied dysphagia, gastrointestinal and pulmonary inconvenience, joint pains or morning stiffness, or symptoms of Raynaud's phenomenon. Some weeks before the debut of symptoms she had extended her horseriding activities considerably.

On admission in May 1978 physical examination showed a well nourished, normally developed girl. There was nothing remarkable except for the left leg where patches of yellow-brown, shiny atrophic, depressed morphea-like macules were scattered on the shin and distal thigh. On palpation, a peculiar firm tautness of the skin and subcutaneous tissue was felt, and the mobility of the knee and ankle joint was considerably reduced. No joint effusion or tenderness was found.

Laboratory evaluation showed a normal WBC except for an elevated eosinophilic count, 540–610/mm^3. The ESR (Westergren) was 29 mm/h. IgG was elevated (21.4 g/l), IgA-M-E within normal range. The following laboratory tests were negative or normal: hemoglobin concentration, thrombocyte count, total serum protein, albumin, haptoglobin, and complement C_3 and C_4 levels, serum values of creatinine, electrolytes, SGOT, LDH, creatine phosphokinase and aldolase; titres of rheumatoid factors, ANA, *Yersinia enterocolitica* (types 3 and 9), antistreptolysin titre, M-components and cryoglobulins; ECG, pulmonary function tests and X-ray of chest, oesophagus, stomach and intestine, left knee and ankle joint.

Histological examination of skin biopsy material showed a normal epidermis without atrophy. In the dermis the cutaneous appendages were preserved, but surrounded by condensed collagenous fibres. Only discrete focal perivascular inflammation—predominantly lymphocytic—was seen. The connective tissue trabeculae of the subcutaneous tissue and the fascia were widened and revealed marked inflammatory changes extending into the surrounding subcutaneous tissue and consisting of histiocytes, lymphocytes, and occasional plasma cells. No eosinophils were seen. Biopsy of the left anterior tibial muscle showed normal muscle fibres, but in the surrounding interstitium and connective tissue, inflammatory changes with lymphocytes and plasma cells appeared.

A regimen of prednisone 60 mg every second day was instituted, and the patient was followed at monthly control visits.

The mobility of the left leg improved almost parallel with normalization of the IgG level, which was within normal range 3 months after commencement of treatment. Prednisone was tapered off slowly to a level of 25 mg every second day by the 6-month control. At that time only slight reduction of the mobility of the left ankle joint remained (probably due to insufficient training as the joint could be redressed freely). The cutaneous changes were barely visible, and the tautness of the subcutis had disappeared.

DISCUSSION

Our findings in this 13-year-old girl fulfil the criteria of Shulman's syndrome or EF: cutaneous

Steroid treatment			Treatment effective		
+	0	−	+	0	−
4			4		
6		1	6		
5				5	
1			1		
1			1		
1			1		
1			1		
1		1	1		
	1				
1			1		
1			1		
2				2	
1			1		
1			1		
1	5		1		
1			1		
28	6	2	21	5	2

morphea-like changes, inflammatory changes of trabeculae from the deep fascia, reduced mobility of the affected extremity, eosinophilia of peripheral blood, and selective elevation of IgG. The syndrome is not particularly rare and is probably recognized increasingly often (cf. the 19 cases reported during the years 1976–78; Table I). The question of pathogenesis, however, is still unresolved, as are the criteria for a strict delineation of the syndrome.

In some cases an alleged correlation in time to physical exercise (7, 10, 11, 12, 13, 15) is described. In two patients the symptoms developed following "tissue lesions" (trauma, acute myocardial infarction) (6). This might suggest some autoimmune pathogenesis, which is corroborated by the occasional finding of positive titres of rheumatoid factors and ANA (4, 5, 10, 13). Immunofluorescence has demonstrated immunoglobulins and complement in the diseased fascia and skin (4, 9, 12). Confirmatory of involvement of the immunological factors is the constant elevation of eosinophilic counts and the elevated IgG levels (Table I). So far, however, no antigen has been isolated. Clinically the symptoms mimic those of localized scleroderma and scleredema, as emphasized by Atherton and Fleischmajer (2, 6), but the sex ratio males to females in EF of 2 : 1 (Table I) does not tally with the clear preponderance of females in the former disease.

Fleischmajer investigated 88 patients with cutaneous sclerodermoid changes (6). Sixty-seven had systemic sclerosis (positive Raynaud's phenomenon, acrosclerosis and internal organ involvement), 15 had localized scleroderma (induration of the skin without Raynaud's phenomenon or involvement of another organ system). The symptoms of six of Fleischmajer's patients justified the diagnosis of EF, thus comprising 7% of the series. However, the diagnosis was based on cutaneous changes, accompanied by eosinophilia, while information on restricted joint mobility was given in 1 patient only, and elevated IgG in none. Fascial biopsy was done in 3 of the 6 patients. The occurrence of EF in 7% of the 88 patients with sclerodermoid cutaneous changes is probably not the true figure and is not claimed to be so by the author, but it appears that the differential diagnosis is difficult. The ability to establish a correct diagnosis is by no means of pure academic interest, as corticosteroid treatment appears to be beneficial in EF (Table I) even though spontaneous cure in 1 patient has been reported (9), though to our knowledge the only one so far. On the other hand, Chanda (5) saw only a doubtful effect of prednisone in 2 patients with EF (40 and 60 mg per day respectively), but it should be noted that the duration of symptoms in these 2 patients before treatment was about one year, compared with a couple of months in all other reported cases.

If EF really does become more resistant to therapy when left untreated this will urge an early diagnosis.

ACKNOWLEDGEMENTS

We are grateful to Mrs Bitten Kjeldsen for her able technical assistance.

REFERENCES

1. Ansell, B. M., Nasseh, G. A. & Bywaters, E. G. L.: Scleroderma in childhood. Ann Rheum Dis *35:* 189, 1976.
2. Atherton, D. J. & Wells, R. S.: Scleredema of Buschke? Eosinophilic fasciitis? Br J Dermatol *95*, Suppl. 14: 36, 1976.
3. Camus, J. P., Crouzet, J. & Prier, A.: Syndrome de Shulman (pseudo-sclérodermie à éosinophiles). Ann Med Interne (Paris) *127:* 391, 1976.
4. Caperton, E. M. & Hathaway, D. E.: Scleroderma with eosinophilia and hypergammaglobulinemia. The Shulman syndrome. Arthritis Rheum *18:* 391, 1975.
5. Chanda, J. J., Callen, J. P. & Taylor, W. B.: Diffuse

fasciitis with eosinophilia. Arch Dermatol *114:* 1522, 1978.
6. Fleischmajer, R., Jacotot, A. B., Shore, S. & Binnick, S. A.: Scleroderma, eosinophilia, and diffuse fasciitis. Arch Dermatol *114:* 1320, 1978.
7. Gray, R. G. & Poppo, M. J.: Eosinophilic fasciitis: A scleroderma-like illness. JAMA *237:* 529, 1977.
8. Krauser, R. E. & Tuthill, R. J.: Eosinophilic fasciitis. Arch Dermatol *113:* 1092, 1977.
9. Rodnan, G. P., DiBartolomeo, A. G., Medsger, T. A. & Barnes, E. L.: Eosinophilic fasciitis: Report of 7 cases of a newly recognized scleroderma-like syndrome. Arthritis Rheum *18:* 422, 1975.
10. Schumacher, H. R.: A scleroderma-like syndrome with fasciitis, myositis, and eosinophilia. Ann Intern Med *84:* 49, 1976.
11. Shewmake, S. W., Lopez, D. A. & McGlamory, J. C.: The Shulman syndrome. Arch Dermatol *114:*556, 1978.
12. Shulman, L. E.: Diffuse fasciitis with eosinophilia: A new syndrome? Trans Assoc Am Physicians *88:* 70, 1975.
13. Talbott, J. H. & Cusani, B. A.: Faciitis (eosinophilic) with scleroderma-like changes in the skin. J Fla Med Assoc *63:* 702, 1976.
14. Torres, V. M. & George, W. M.: Diffuse eosinophilic fasciitis. Arch Dermatol *113:* 1591, 1977.
15. Weinstein, D. & Schwartz, R. A.: Eosinophilic fasciitis. Arch Dermatol *114:* 1047, 1978.

Is the Use of Ro 10-9359 (Tigason®) in Children Justified?

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Received December 21, 1979

Abstract. In 5 children ranging in age from 8 to 12 years, treatment with Ro 10-9359 for either psoriasis or etythrokeratodermia variabilis for periods of between 11 and 17 months did not cause marked growth retardation and gave excellent therapeutic results.

In many skin diseases characterized by abnormal keratinization, for instance psoriasis vulgaris, several forms of ichthyosis, erythrokeratodermia variabilis, and Darier's disease, Ro 10-9359 has opened up important new therapeutic prospects. Because these conditions often manifest themselves in childhood, the question arises whether the use of this new drug is warranted in this age group. This question must be asked because in rats (but not in other experimental animals) retinoid may impair bone growth (Loefler, personal communication). Furthermore, several authors (1, 2) state that they are concerned that Ro 10-9359 might interfere with bone growth in children.

In 5 children whom we treated successfully with Tigason® we monitored growth over a relatively long period by regular measurement of not only height but also the total 24 h urine hydroxyproline excretion. The latter parameter is considered to be a good indicator for an abnormal growth rate in children (3). In all cases we obtained informed consent from the parents for the treatment.

PATIENTS AND RESULTS

Relevant data concerning the patients are given in Table I. Regularly performed laboratory tests, including renal and liver function tests and determination of hydroxyproline excretion, gave normal results. The growth rate is shown in Table II.

DISCUSSION

It is well known that when growth is retarded in children due to illness or under the influence of drugs, catch-up growth will occur after cure or cessation of drug treatment. If time permits and growth inhibition has not been too severe or prolonged, adult height will lie within normal limits.

In the first 5 children we treated with Ro 10-9359, we regularly measured the height as well as the hydroxyproline excretion. A decrease in the latter could indicate growth retardation. In no case were pathologic values measured in the urine; the growth rate in all cases was within the normal range.

Our data do not exclude the possibility of growth retardation caused by the use of Ro 10-9359. We therefore recommend monitoring of the height every 3 months during treatment. When growth retardation is encountered in a child receiving Ro 10-9359, catch-up growth after cessation of treatment will prove that the retardation has been due to this therapy. In view of the excellent results obtained with Ro 10-9359 in the current therapeutic

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