Concordant Urticaria Pigmentosa in a Couple of Identical Twins
A Five-year Follow-up

Sir

A 5-year follow-up is reported of 2 cases of skin mastocytosis of the urticaria pigmentosa (UP) type, with synchronous onset and concordant manifestations and course in a couple of identical male twins. We observed synchronous UP in a couple of identical twin boys at the age of 4 months (Fig. 1), born at full term after normal labour (1). The cutaneous picture consisted of a few dozen diffuse maculo-papular and papulo-nodular lesions, reddish-brown in colour, with a round and sometimes oblong shape and undefined margins. Both twins had developed some bullous lesions, episodes of diarrhoea and generalized pruritus. Fever, flushing or muco-cutaneous involvement were absent. The progressive onset of the lesions had occurred almost synchronously about 50 days previously. Stroking of lesions was followed by erythematous wheals. The twins were almost identical in height, weight and phenotype. The parents were not consanguineous and there was no family history of mastocytosis. Laboratory tests did not reveal any abnormal parameter. Histologic examination showed in both cases the presence in the papillary and superficial dermis of a number of roundish mononuclear cells with clear cytoplasm, intermingled with eosinophils. Mast cells showed the typical metachromasia after toluidine blue staining. Both patients received symptomatic treatment with oxatomide, with good control of the pruritus, and were seen every 6 months; there were no complications. During the first 2 years the lesions appeared unchanged. During the 3rd and 4th years they appeared to be slowly and steadily diminishing. Five years after the diagnosis only a few asymptomatic, macular brownish lesions are present on the trunk in both twins, and there are no systemic symptoms.

Mastocytoses appear in the first 2 years of life in 55% of cases, at between 2 and 15 years in 10%, and after 15 years of age in the remaining 35% (2). The most common clinical pictures of cutaneous pediatric mastocytosis are mastocytoma and UP. Diffuse cutaneous mastocytosis is less frequent, whilst telangiectasia macularis eruptiva perstans is a very rare condition (3). UP heals in about 50% of cases before puberty (4).

As far as we are aware, 17 reports of identical twins with mastocytosis of the UP type have been published (5). Of these, 13 were with concordant mastocytosis, including a triplet. Macular or maculo-papular lesions were observed in 13 cases and papulo-nodular lesions in only 4 cases (6–9). The female sex seems to be prevalent in UP in twins (12 cases out of 19, of which 2 were heterozygotes), although in absolute terms no sex prevalence is demonstrated in sporadic UP. In concordant cases the intervening time in twins was generally no more than 2 months and the disorder normally had its onset in the first months of life (5). As a rule, no family history of mastocytosis is detectable in these cases. For concordant cases without a positive family history a dominant autosomal mechanism with incomplete penetrance has been hypothesized (10). In discordant cases a mutation has been suggested (11, 12).

Our observation, apart from the very rare occurrence of UP in identical twins, is noteworthy for the presence of papulo-nodular lesions in both cases, although not in all elements observed. Macular, papulo-nodular and bullous lesions coexisted in both twins despite the overall not extremely high number of lesions in each twin. Even the course was absolutely concordant, as the lesions slowly and steadily diminished and, after 5 years only a few asymptomatic, macular, slightly pigmented lesions can be observed on the trunk in both twins.

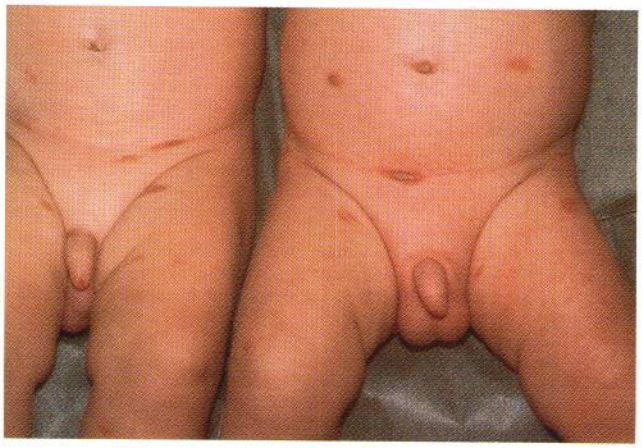

Fig. 1. Maculo-papular lesions of urticaria pigmentosa in identical twins.

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