Keratodermia palmoplantaris of the Unna-Thost Type in Slovenia

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Keratodermia palmoplantaris of the Unna-Thost type was studied in the population of Slovenia. Althogether 157 patients were recorded: 106 were examined by the authors; for the other 51, anamnestic data were used. Special attention was paid to the accompanying symptoms which are more or less rarely mentioned in the literature: hyperkeratosis of the elbows and knees, transgredience of the hyperkeratosis, hyperkeratotic changes of the nails and changes of the teeth. It is estimated that there are in Slovenia 8.44 PPK UT patients per 100000 inhabitants. *Key words: Keratodermia palmoplantaris Unna-Thost; Population study Slovenia.* (Received January 26, 1983.)

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Palmoplantar kertodermas (PPK) represent one of the most frequently encountered groups of genodermatoses. Within this group various types of PPK (nosologic entities) may be distinguished, e.g. palmoplantar keratoderma of the Unna-Thost type (PPK UT), PPK of the Brauer type, PPK of the Mljet type (mal de Meleda), PPK of the Jadassohn–Lewandowsky type (pachyonychia congenita), PPK of the Papillon–Lefèvre type (PPK cum periodontopathia) as well as others. Of all these types, PPK UT is by far the most frequent. No reliable data are available in the literature about the prevalence of PPK UT in various populations. Rook and Ebling (1) mentioned that in Northern Ireland, one case per 40000 inhabitants is seen, and assume the same frequency in other populations.

Slovenia with its 1.86 million inhabitants and well-organized health service offers good opportunities for genetic studies. On the basis of these premises the authors decided to investigate the prevalence of different types of PPK in the population of Slovenia.

MATERIALS AND METHODS

The data on PPK UT patients were gathered in the following ways: 1) new cases were discovered by routine clinical procedures in the University Dermatologic Clinic in Ljubljana; in the dermatological departments of the General Hospitals of Maribor, Celje and Novo mesto; as well as in several dermatological outpatient clinics. 2) Additional cases were identified by retrospective review of clinical records in the above-mentioned institutions; a 30-year period was covered. 3) In all instances relatives of the already recorded and of newly detected patients were invited to a check-up. It soon became clear that a large number of relatives would not cooperate. 4) It was decided therefore to visit patients at their homes at weekends. This time-consuming procedure enabled the authors to investigate a large number of patients' kin and to draw up reliable genealogical trees.

The criteria on which the diagnosis was made were as follows: 1) hyperkeratosis of the palms and soles; 2) hyperkeratoses of the knees and elbows; 3) the red edge of the palms and soles; 4) hyperhidrosis of the areas, and 5) anamnestic and genealogical data. Such a complex evaluation enabled us in many instances to detect latent cases which otherwise would have remained unobserved. The main difficulty in detecting cases of PPK UT, especially among the rural population, is the relative frequency of patients with acquired (non-genetic) hyperkeratoses of the palms and soles.

We only hope that sooner or later some laboratory assay will be discovered with which to confirm the diagnosis of genetically determined PPK.

Only those patients were recorded who were permanent residents of Slovenia or were holding a permanent job there.

RESULTS

During this study 157 patients with PPK UT were recorded. Of this number, 106 were examined by the authors. For the other 51, anamnestic data were used inasmuch as the patients could not be examined because of death or prolonged absence.

Regional distribution of patients

In general, PPK UT patients were detected throughout the territory of Slovenia. It is interesting to note, however, that they were found clustered in larger numbers in certain restricted localities. The greatest number was detected in two rather small, until recently predominantly rural communities: Velenje (Celje area) and Ajdovşçina (Nova Gorica area). In one kinship in the vicinity of Velenje, 23 cases (all previously unknown) were recorded. In one further kinship in the Celje area, 16 cases and in one kinship in Ajdovşçina, 13 cases were registered. In other families, fewer cases were detected, mostly 2 to 6. In addition to the cases with a positive family history, a few patients with a negative family history were observed: 2 in the Novo mesto area, 5 in Nova Gorica, 4 in Ljubljana area. The detection of PPK UT cases among gypsies presented a difficult problem. A few such cases were observed in the gypsy settlement of Çernelavci (Murska Sobota area). During a visit to this settlement, 3 female patients with PPK UT were observed. A few individuals with faintly expressed signs and without a positive family history were not included into this study.

Skin symptoms

Skin symptoms were evaluated in 106 patients. A typical hyperkeratosis of the palms and soles was expressed in all, except in a few patients who were included in this group on the basis of hyperkertoses of the elbows and (or) knees, some other minor symptoms and a positive family history. These are the latent cases. Altogether 10 such patients were observed (Table I).

Hyperkeratoses of the edges of the palms (or fingers) and soles were seen in 41 patients (42.7%). In 20 patients (20.8%) a hyperkeratosis was observed on the back of the hands or feet, too. Such hyperkeratoses were, however, less pronounced than in PPK cases of the Mljet type. A marked hyperhidrosis of the palms and soles is a constant feature of the PPK UT. 28 patients (29.1%) showed a hyperkeratosis on the knees and 23 (23.9%) on the elbows. A typical red edge surrounding the hyperkeratoses of the palms and soles was present in 36 patients (37.5%). The observation was also made that in elderly patients the hyperkeratoses were mildly expressed or even tended to disappear.

Changes of the nails: 18 patients (18.7%) displayed a subungual hyperkeratosis and a thickened nail plate.

Teeth with accompanying structures (organon dentale): 27 persons were observed with changes which were diagnosed as paradentosis. The inflammation of the gums appeared early, as also did the pockets around the roots. The majority lost their teeth early, on average at the age of 30–35 years. The shape and arrangement of the deciduous and permanent teeth in these patients did not differ from those in normal persons. The changes on the teeth and the accompanying structures will be described in detail elsewhere.

Mental status was roughly evaluated on the basis of an unstructured interview and a

Table I. *Symptoms observed in 96 patients with palmoplantar keratoderma of the Unna-Thost type*

	Investigated	Patients with symptoms		Patients without symptoms
		No.	%	
Hyperkeratosis of palms and soles	96	96	100.0	0
Hyperkeratosis of the hands				
Margins	96	41	42.7	55
Dorsa	96	20	20.8	76
Hyperkeratosis				
Elbows	96	23	23.9	73
Knee	96	28	29.1	68
Erythematous band	96	36	37.5	60
Thickened nails	96	18	18.7	78
Paradentosis	96	27	28.1	69
Mental retardation	96	5	5.2	91

review of scholastic achievements. It was considered to be slightly below normal in 5, and normal in all the other examinees.

Latent cases are very important for genetic studies, inasmuch as they may represent the missing link in genealogical trees. Out of 106 patients examined, 10 were labelled as latent cases. A slight hyperkeratosis of the palms and soles with somewhat deepened furrows, hyperkeratoses at the edges of the hands and feet or in the region of the Achilles tendon or malleoli, hyperkeratoses of the knees or elbows as well as hyperhidrosis of the palms and soles may be the only manifestations. Such signs might easily be overlooked unless such persons are examined as members of a PPK UT kinship. At least in one kinship the observation was made that in the younger generations the signs were less pronounced than in the older ones (5).

Inheritance. Our observations confirm the already established fact that the mode of inheritance in PPK UT us autosomal dominant. However, some observations are not easy to explain. In family Ş. the female patient II/2 showed no signs of PPK UT, although her late father, her sister, her two sons and her daughter are all clear cases. A similar observation was made in family HS.: the female patient III/2 has normal skin throughout, while both her daughter and son have mild signs of PPK UT.

DISCUSSION

The geographic distribution of patients with PPK UT shows clearly that they are present in all regions of Slovenia. As no family links have been established between different kinships, the assumption is justified that the genetic anomaly which causes PPK UT is not uncommon. The evaluation of single signs appears to be very important.

Hyperkeratosis of the knees was detected in 29.1% and that of elbows in 23.9% of the examinees. These manifestations are mentioned as part of the PPK TU syndrome by Baden (2) but not in other standard textbooks (1, 3, 4). The hyperkeratoses are often not completely symmetrical; they may be expressed only on elbows or only on the knees. It was shown earlier that they may be inherited within the PPK UT without a consistent pattern (5). Their relatively frequent occurrence indicates that there is no reason for singling out the so-called PPK of Greither's type (5, 6). Restraint about singling out this

type as a special nosologic entity was expressed long ago by Schnyder and Klunker (3) and by Rook and Ebling (1). According to the authors' experience, these signs are very useful in the detection of latent cases.

The hyperkeratoses on the edges of the hands and feet as well as hyperkeratoses on the dorsal surfaces of the hands are usually not mentioned as part of the PPK UT in the standard textbook, with exeption of Gahlen (7). The regions between the 1st and 2nd or between the 2nd and 3rd finger have to be examined carefully. Hyperkeratotic changes of the nails which we observed in 18.7% of out patients, have already been mentioned by Moncorps (8) and later by other authors (1, 2).

The changes of the teeth and the accompanying structures in PPK TU, which we have observed in 28.1%, are hardly mentioned in the literature. Most probably this was the reason why some authors misdiagnosed PPK UT with changes of the teeth as PPK of the Papillon–Lefèvre type. These symptoms, although qualitatively different, may represent an overlap between those two types in a similar way as nail changes may represent an overlap between the PPK UT and the pachyonychia congenita. Unluckily, there is at present no reliable laboratory test by which the diagnosis of congenital PPK can be confirmed or even differentiated into various types.

Considering the number of patients who were recorded in our study it is estimated that in Slovenia there must be 8.44 PPK UT patients per 100000 inhabitants. This would indicate that in Slovenia the syndrome appears 3.4 times as frequently as is the case in Northern Ireland, where a prevalence of 2.5 cases per 100000 inhabitants has been reported.

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