Idiopathic Recurrent Palmoplantar Hidradenitis in Children

Report of 22 Cases

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**Background:** Idiopathic recurrent palmoplantar hidradenitis (IRPH) is characterized by tender erythematous plaques and nodules on the soles and, less often, the palms of young patients. To date, 10 cases of IRPH have been documented in the literature.

**Observations:** We describe 22 pediatric patients with characteristic cutaneous and histologic findings of IRPH. Their mean age was 6 years (age range, 1.5-15 years). The onset of the disease clustered in 2 peaks, in autumn and spring. All patients had complete resolution of their lesions within 3 weeks, in 16 cases without any treatment. Ten of the 22 patients experienced more than 1 episode of IRPH.

**Conclusions:** Our study represents the largest number of cases of IRPH collected to date. Based on our data, the clinical picture is so characteristic that a histopathologic examination may not be necessary in every case. We suggest that there is a seasonality in the occurrence of this always benign and limited disease.

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Since being described by Stahr et al in 1994, idiopathic plantar hidradenitis or idiopathic recurrent palmoplantar hidradenitis (IRPH) has been observed only in few cases, primarily in children. In contrast to neutrophilic eccrine hidradenitis (NEH), which was described by Harrist et al in 1982, IRPH is characterized by multiple tender erythematous nodules on the soles and, in some cases, also the palms of otherwise healthy young persons. Histologically, neutrophilic infiltrates localized to the eccrine coils in the deep reticular dermis are characteristic.

Over the last 3 years, we studied 22 children with clinical and histopathologic features consistent with those of the patients of Stahr et al and Rabinowitz et al. We observed seasonality in the occurrence of IRPH and emphasize the possible role of mechanical and/or thermal trauma in the pathogenesis of this probably common and, in every case, benign condition.

Neutrophilic eccrine hidradenitis was first described by Harrist et al in 1982 in a patient undergoing chemotherapy for acute myelogenous leukemia. Since then, similar dermatologic observations have been described in association with the use of various drugs and chemotherapeutic agents, cancer, and infections. The urticarial-like erythematous plaques and/or nodules of NEH develop on the trunk, the proximal aspect of the extremities (involvement of the hands and wrist was recorded only in a few patients), or the face, whereas exclusive involvement of the plantar skin has not been observed. Most lesions have resolved spontaneously over the course of several days.

Idiopathic recurrent palmoplantar hidradenitis, unlike NEH, appears almost exclusively in otherwise healthy children. The tender erythematous macules and nodules of IRPH are localized and appear unilaterally or bilaterally on the palms and/or soles. Our study represents the largest published collection of cases of IRPH, 2 of which involved sisters who became ill at the same time. Follow-up of our patients showed a resolution of their lesions within 3 weeks, in 16 cases without any treatment. Relapses occurred in 50% of the group and cleared spontaneously.

The clinical differential diagnosis (Table) of IRPH encompasses patients presenting with acral, tender, erythematous lesions, such as those of traumatic
PATIENTS AND METHODS

EPIDEMIOLOGICAL AND LABORATORY FINDINGS

Fifteen of our 22 patients were female, revealing an overall female-male ratio of 2.1:1. The mean patient age was 6 years (age range, 1.5-15 years) (Figure 1). Two patients were sisters who suffered from IRPH at the same time. The onset of the disease in our patients clustered in 2 peaks, in autumn and spring (Figure 2), suggesting seasonality.

Laboratory tests, which were performed in 21 of 22 cases, revealed elevated white blood cell counts (14-20×10⁹/L), erythrocyte sedimentation rates (24-28 mm/h; reference range, 0-10 mm/h), and serum C-reactive protein levels (11-12 mg/L; reference range, 0-10 mg/L) in 3 patients. The results of other routine blood chemistry tests were within normal limits.

CLINICAL SPECTRUM

The occurrence of multiple, painful, erythematous macules and indurated nodules ranging from 0.5 to 3 cm in diameter on the palms and soles of otherwise healthy children is the clinical hallmark of IRPH (Figure 3). Of 22 patients, 21 had exclusively tender plantar involvement, resulting in debilitation. The lesions were unilateral in 2 children. Bilateral, tender, palmar macules and nodules appeared in 50% of the group. An elevated body temperature (<38°C) was noted in 2 girls. All patients had complete resolution of the lesions (duration, 2-21 days after onset [median duration, 8 days]); 6 were treated with topical nonsteroidal anti-inflammatory agents, and 16 were not treated. Ten of our 22 patients experienced more than 1 episode of their eruption.

HISTOLOGIC FINDINGS

The microscopic findings in all patients (n=22) with IRPH were very similar. Each specimen showed a nodular infiltrate consisting predominantly of neutrophils that was localized to the eccrine apparatus, particularly to the coils (Figure 4), with slight extension to the periglandular tissue. In the middle and deep dermis, there was a mild, mixed perivascular infiltrate without the typical changes of a leukocytoclastic vasculitis. Stains were negative for microorganisms, including bacteria and fungi, in all cases.

The pathogenesis of IRPH is unknown. Based on our clinical and histologic observations, we believe that mechanical and/or thermal trauma can lead to a rupture of the palmoplantar eccrine glands. The release of glandular secretions into the surrounding tissue may activate a cytokine cascade (eg, tumor necrosis factor α, granulocyte colony-stimulating factor, and interleukin 8) that is capable of attracting neutrophils that can result in a transient inflammation clinically represented by tender, erythematous palmoplantar papules and nodules. Therefore, the high relapse rate observed in our patients is not surprising.

The majority of our patients did not need any distinct therapy. If the disease causes pain or low-grade fe-
Figure 3. Classic lesions of idiopathic recurrent palmoplantar hidradenitis. A, B, and C, Tender, erythematous macules and nodules on the plantar aspects of the feet. D, Painful, erythematous nodules on the palms.

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Patients' History</th>
<th>Skin Lesions and Symptoms</th>
<th>Laboratory Results*</th>
<th>Histologic Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Traumatic plantar urticaria</td>
<td>Mechanical stress, eg, running, dancing</td>
<td>Tender plantar macules and papules</td>
<td>Normal</td>
<td>Neutrophilic pressure urticaria; eccrine units uninvolved</td>
</tr>
<tr>
<td>Erythema multiforme</td>
<td>Herpes simplex infection; drug intolerance</td>
<td>Target lesions with central blistering; fever and arthralgia</td>
<td>Elevated ESR and CRP levels; leukocytosis</td>
<td>Less neutrophils; eccrine units uninvolved</td>
</tr>
<tr>
<td>Erythema nodosum</td>
<td>Infections, drugs; associated diseases</td>
<td>Confined to the lower legs; fever and arthralgia</td>
<td>Elevated ESR and CRP levels; leukocytosis</td>
<td>Septal panniculitis</td>
</tr>
<tr>
<td>Sweet syndrome</td>
<td>Similar to erythema nodosum; paraneoplastic</td>
<td>Erythematous plaques; fever and general malaise</td>
<td>Elevated ESR and CRP levels; leukocytosis</td>
<td>Bandlike infiltrate of mononuclear cells and numerous neutrophils</td>
</tr>
<tr>
<td>Behçet disease</td>
<td>Chronic disease with acute exacerbations</td>
<td>Oral and/or genital aphthae, postular skin lesions; thrombophlebitis</td>
<td>Increased chemotaxis of neutrophils</td>
<td>Vasculitis</td>
</tr>
<tr>
<td>Chilblains</td>
<td>Cold, damp environment; associated diseases</td>
<td>Burning, painful, livid patches and plaques, blisters, ulcers</td>
<td>Normal in most cases</td>
<td>Dense, perivascular lymphoid infiltrate; endothelial swelling</td>
</tr>
</tbody>
</table>

*ESR indicates erythrocyte sedimentation rate; CRP, C-reactive protein.
ver, treatment with nonsteroidal anti-inflammatory drugs is helpful. We do not see the necessity for further treatment or diagnostic procedures in this probably common and, in every case, harmless benign disease. On the other hand, if no improvement occurs within 1 or 2 weeks after the onset of the tender, erythematous palmoplantar nodules in young patients, histopathologic evaluation is necessary to rule out clinically similar diseases (Table).

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REFERENCES