

## Case report

## Acquired ichthyosis in ulcerative colitis

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### SUMMARY

Ichthyosis can be either congenital or acquired. Acquired ichthyosis in inflammatory bowel disease has never been reported so far. A 65-year-old woman suffering for three years from ulcerative colitis presented with a 4-month history of a non-pruritic, photosensitive eruption on the abdomen and on the upper and lower extremities. Laboratory tests were all within normal limits. Skin punch biopsies of ichthyosiform plaques showed compact lamellar orthohyperkeratosis and hypogranulosis, which were consistent with familiar ichthyosis or acquired ichthyosis. Local preparations with 5% urea and 5% NaCl were administered to the patient with satisfactory results. To the best of our knowledge this is the first documented case of acquired ichthyosis in a patient with inflammatory bowel disease. This case raises the question of whether acquired ichthyosis was secondary to ulcerative colitis, representing another rare type of inflammatory bowel disease extraintestinal manifestation, or whether each of these diseases developed independently.

**Key words:** ichthyosis, acquired ichthyosis, inflammatory bowel disease, ulcerative colitis, Crohn's disease, cutaneous manifestation

bowel disease can be categorized as specific lesions (perianal fissures, metastatic Crohn's disease), as reactive lesions (erythema nodosum, pyoderma gangrenosum, hidradenitis suppurativa) and finally, as cutaneous manifestations (dermatoses) such as epidermolysis bullosa acquisita and acne fulminans, which are connected, with high probability, with inflammatory bowel disease.<sup>1-3</sup>

Ichthyosis or sauriasis is the name given to a rare group of skin disorders with generalized disturbance in desquamation that appear to have different etiologies but a common pathogenesis and similar appearance. The entire skin surface shows abnormal desquamation, but the degree of scaling is worse on the extensor surfaces, particularly the legs. There is also considerable variation in severity between patients. Mildly affected individuals may just show slight roughness of the skin surface. Biochemical abnormalities and pathogenetic mechanism are poorly understood in ichthyosis. The epidermis and stratum corneum are of normal thickness and appearance apart from the absence (or great reduction) of the granular-cell layer. An increased rate of incorporation of lipid precursor compounds into all classes of lipids has been found in familiar ichthyosis, and an abnormality of lipid metabolism has therefore been suggested.<sup>4</sup>

Ichthyosis can be either congenital (familial ichthyosis or ichthyosis vulgaris) or acquired, the last being named also as "secondary",<sup>5</sup> "new-onset"<sup>6</sup> or even "paraneoplastic"<sup>7</sup> ichthyosis, although many of the reported cases represent quite benign conditions.

Acquired ichthyosis, clinically and histologically similar to familial ichthyosis, may develop in patients of any age with certain forms of malignant disease, with other dermatoses, in gross nutritional deficiencies, or in bone marrow transplant patients.<sup>4</sup>

Co-existence of acquired ichthyosis with gastrointestinal tract diseases has occasionally been reported, in-

### INTRODUCTION

Dermatological lesions in patients with inflammatory

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cluding cases of co-existence with dermatomyositis in a patient with hepatocellular carcinoma,<sup>8</sup> with coeliac disease,<sup>9</sup> with esophageal<sup>10</sup> and gastric carcinoma,<sup>11</sup> with eosinophilic fasciitis<sup>12</sup> and finally, with mesenteric cell sarcoma.<sup>13</sup>

Herein we report the first case of acquired ichthyosis in a patient with ulcerative colitis.

## CASE REPORT

A 65-year old woman presented with a 4-month history of a non-pruritic, photosensitive eruption on the abdomen and on the upper and lower extremities (Figure 1). Her past medical history contained a diagnosis of left-sided ulcerative colitis 3 years earlier. The patient was on maintenance therapy with 1.5g of sulfasalazine P.O.

A detailed medical history, including the use of retinoids, and other skin treatments, was obtained. There was no personal or family history of atopy or ichthyosis and no keratosis pilaris or hyperlinear palms. Physical examination revealed a poikilodermatous eruption on the abdomen and the four extremities. Also notable were



**Figure 1.** Acquired ichthyosis in a patient with ulcerative colitis before treatment.

large-rhomboid-shaped, grayish brown scales on the extension surface of the legs and on the abdomen, which were consistent with the diagnosis of acquired ichthyosis. According to the patient, the ichthyosis had developed over a 2-month period. No neurological, muscle, nail or hair involvement was evident.

The results of laboratory investigation including routine hematological, biochemical immunological tests, including neoplastic serum markers were all within normal limits. Hepatoviruses antigens and antibodies, HIV antibodies, and antibody for Lyme disease (*B. burgdorferi*) were also negative.

Whole body computed tomography and upper gastrointestinal endoscopy were unremarkable. Total colonoscopy showed a left-sided colitis in remission, which was also confirmed by multiple bowel biopsies. There was no evidence of dysplasia or bowel epithelial cell atypia on biopsies.

Skin punch biopsies (2mm) of ichthyosiform plaques showed compact lamellar orthohyperkeratosis and hypogranulosis, which are consistent with familiar ichthyosis or acquired ichthyosis.

Local preparation with 5% urea and 5% NaCl was administered to the patient with satisfactory results after one week of treatment. After two months the skin lesions were remarkably improved (Figure 2) and there was no evidence of ulcerative colitis relapse during a six month follow up.

## DISCUSSION

Acquired ichthyosis in inflammatory bowel diseases has not been reported to date, although one sporadic case of familial type of ichthyosis in a patient with Crohn's disease has been reported.<sup>14</sup> Onset of ichthyosis early after childhood is an indication for detailed investigation, as acquired ichthyosis is mainly regarded as a cutaneous paraneoplastic syndrome rather than a benign skin disease.

There have been reports of acquired ichthyosis co-existing with other dermatoses such as dermatomyositis in a case of ovarian tumor,<sup>15</sup> atopic dermatitis<sup>4</sup> with alopecia and loss of hair pigment in leiomyosarcoma,<sup>16</sup> acrokeratosis paraneoplastica (Bazex Syndrome), and laryngeal neoplasm's, pityriasis rotunda in IgG myeloma, hypergammaglobulinemic purpura, renal tubular damage and epidermodysplasia verruciformis in cases of congenital deficiencies of factors V and VIII.<sup>4,17</sup> An interesting case of acquired ichthyosis and pyoderma gangrenosum



**Figure 2.** Acquired ichthyosis in a patient with ulcerative colitis after treatment.

in a patient with systemic lupus erythematosus has also been reported<sup>18</sup>. However, no association of acquired ichthyosis with other dermatologic or extraintestinal manifestations of inflammatory bowel disease has been reported to date.

There have been also reports of acquired ichthyosis developing in patients with malignancies and other types of lymphomas, IgE myelomas, polyerythemia rubra vera, lymphosarcoma, Kaposi's sarcoma and acquired immunodeficiency syndrome, mucosis fungoides, disseminated carcinomatosis, bladder and kidney neoplasia, breast cancer and rhabdomyosarcoma.<sup>4,19-23</sup> Our clinical and laboratory investigations, as well as patient follow-up, failed to prove an underlying malignancy complicating ulcerative colitis course.

Acquired ichthyosis in non-neoplastic diseases includes cases with impaired vitamin A absorption. As skin changes of acquired ichthyosis are clinically and pathologically indistinguishable from familial ichthyosis, family history, age of ichthyosis onset and thorough in-

vestigation for other underlying causes is of great importance in differential diagnosis. Acquired ichthyosis must also be carefully separated from other cases of scaling disorders, which are characterized by an increase in intracorneal cohesion such as atopic dermatitis, pityriasis rotunda, ichthyosiform scaling secondary to megavoltage radiotherapy, ichthyosiform sarcoidosis and the acne itch-like disease. It is of interest that no cutaneous or other extraintestinal manifestation<sup>24,25</sup> was evident in our patient. In this distinguished group of diseases accurate clinical diagnosis requires patience and experience. Many new or unusual cases may represent a separately identifiable entity as the biochemical abnormalities and the pathogenetic mechanisms involved have not been described in detail in these diseases. Furthermore, in patients with longstanding inflammatory bowel disease, the possibility of an underlying intestinal or extraintestinal neoplasm should never be overlooked.

This case raises the question of whether acquired ichthyosis was secondary to ulcerative colitis or whether each of these diseases developed independently as co-existence cannot be excluded. As the only therapy we used for the acquired ichthyosis were topical regimens, we cannot exclude a possible effect of a p.os administered drug for acquired ichthyosis treatment on long-term outcome of the ulcerative colitis.

To the best of our knowledge, this is the first case of documented acquired ichthyosis in a patient with inflammatory bowel disease.

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