Protein-Losing Gastroenteropathy in a Patient with Pityriasis Rubra Pilaris

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Dear Sir,

Protein-losing gastroenteropathy (PLGE) is characterized by the leakage of serum protein from the gastrointestinal tract, resulting in hypoalbuminemia and generalized edema. PLGE can be considered as a clinical syndrome that is related to a large number of pathological conditions; however, it is an unusual manifestation of pityriasis rubra pilaris (PRP).

A 62-year-old woman receiving treatment for PRP was referred to our hospital because of repeated diarrhea (more than 10 episodes/day) and edema. She had previously experienced both symptoms and had recovered with prednisolone treatment. A photo of her skin is shown in figure 1. A laboratory study revealed hypo-

proteinemia: total protein, 3.9 g/dl; albumin, 2.2 g/dl; her antinuclear antibody was negative. Her α_1 -antitrypsin clearance rate was 596 ml/day, and her endoscopic findings were almost normal. We diagnosed her as having PLGE. Because her symptoms did not improve by fasting, prednisolone (40 mg/day) was initiated. The episodes of diarrhea decreased soon, and 2 months after the start of the prednisolone therapy, she was discharged without exacerbated diarrhea or eruptions.

PRP is a red scaly eruption characterized by the association of palmoplantar keratoderma, follicular plugging and erythematous perifollicular papules or erythroderma; it was first described in 1856.

Histological examination of the follicular papules reveals inflammation extending to the dermis and follicular ostia associated with follicular hyperkeratosis. The presence of PRP has occasionally been associated with immunodeficient states, such as an HIV infection, hypothyroidism, myasthenia gravis, celiac disease or acute stem cell leukemia; however, the underlying cause of this disease is unknown.

We have described a case of PLGE associated with PRP in which both diseases were successfully treated with prednisolone. To our knowledge, this is the first case report of a patient with PLGE and PRP.

Fig. 1. Patient's skin. Follicular papules are visible.