A Rare Case of Idiopathic Hypereosinophilic Syndrome Involving the Oral Cavity Associated with the Esophagus and Gastrointestinal Tract

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Abstract

We report the rare case of HES involving oral cavity associated with esophagus, and gastrointestinal tract, which we succeeded in diagnosing precisely through a biopsy specimen obtained from the lip. A 64-year-old man had dysphagia, swelling of the oral mucosa and the posterior cervical muscles, accompanied by an abdominal pain and diarrhea. Peripheral blood cell count showed marked eosinophilia. Computed tomography showed thickening of posterior wall of the pharynx, esophagus, and gastrointestinal tract. Histologic specimen obtained from the lower lip demonstrated a moderate infiltration of eosinophils. His clinical condition was improved by oral prednisolone therapy.


Key words: HES, oral mucosa, digestive system, eosinophilia, lip biopsy, prednisolone therapy

Case Report

A 64-year-old man had a 9-day history of dysphagia and swelling of the oral mucosa and posterior cervical muscles. Also he had abdominal pain and diarrhea for 4 days.

He had bronchial asthma for the previous 1 year, which was controlled with regular inhalations of budesonide and oral theophylline and occasional inhalations of salbutamol. There were no recent infections or relevant travel history.

At admission, he was unwell with a low-grade fever up to 37.1°C, swelling of oral mucosa (Fig. 1), and frequent bowel movement. Because of dysphagia and severe swelling of the oral mucosa, he could not eat any food.

Laboratory examinations revealed an excess of white blood cells, 17,800/mm³, with marked eosinophilia, and 52.7% eosinophils. Serum IgE concentrations were also elevated at 1,785 IU/ml (normal <310 IU/ml). Radioallergosorbent tests for multiple allergens were negative.

Other normal laboratory values included antinuclear antibody, anti-DNA antibody, and complement titer. Drug lym-

Figure 1. Tooth marks on the inside of the oral cavity induced by a swelling of the oral mucosa (arrows).
phocyte stimulation tests of budesonide and theophylline were negative. Stool specimens were positive for occult blood but negative for parasites and ova.

An X-ray film of the chest on admission was normal. Ultrasonography revealed swelling of cervical soft tissue and thickening of gastric wall. Barium examinations of the esophagus, stomach, duodenum, and small intestine revealed mildly thickened edematous folds, and esophageal motility revealed on increase in simultaneous contraction (Fig. 2).

Upper gastrointestinal endoscopy showed decreased elasticity of the esophagus with insufficient insufflation and edematous mucosa. The antral and the duodenal mucosa were edematous with flare, and there was an open ulcer at the posterior wall of the antrum. Barium examinations of the

Figure 2. Barium examinations of the esophagus (A) and small intestine (B). Mildly thickened edematous folds are seen. Also esophageal motility revealed on increase in simultaneous contraction.
large intestine were normal. Endoscopic ultrasonography (EUS) disclosed thickening of the second to forth layers throughout the esophagus, stomach, and duodenum (Fig. 3). A computed tomography (CT) showed thickened walls of those three organs (Fig. 4). Retrospectively, the posterior wall of the pharynx was also thickened on CT on admission. Biopsied specimen obtained from the lower lip could only demonstrate a moderate infiltration of eosinophils (Fig. 5), although specimens from the esophagus, stomach, and duodenum failed to reveal dense infiltration of eosinophils.

Therapy with prednisolone, 60 mg daily, was started, and the dosage was gradually tapered over 1 week. Symptoms and peripheral eosinophilia rapidly resolved within 3 days, and he could begin to eat some food. One week after the initiation of prednisolone, CT showed slight improvement of wall thickening of those organs. Three weeks after the initiation of prednisone, all symptoms disappeared.

Based on the clinical course and the histologic findings of this case, swelling of the posterior cervical muscles, thickening of the posterior wall of the pharynx, whole esophagus, and gastrointestinal tract were also considered to be caused by eosinophilic infiltration.

Discussion

In 1968, Hardy and Anderson drew attention to the fact that persistent hypereosinophilia of any type could be associated with a range of similar complications and they grouped these together as the hypereosinophilic syndrome (HES) (1). Chusid et al confirmed this in 1975 but restricted the diagnosis of hypereosinophilic syndrome to patients in whom no underlying cause for the hypereosinophilia could be shown (2). So in their report they began to use the term idiopathic hypereosinophilic syndrome when the following criteria are fulfilled:

1. Eosinophilia >1.5×10⁹/l;
2. Persists for at least six months or fatal in a shorter time;
3. Results in organ system dysfunction;
4. Absence of a recognized cause for the eosinophilia.

In HES cases, the organs previously reported to be involved include heart, nervous system, lungs, liver, spleen, muscle, gastrointestinal tract, skin, kidney, and middle ear. Especially heart, lungs, skin, and nervous system were in-
In this case, prednisolone therapy was very effective, and the patient’s symptoms dramatically disappeared. Some investigators have reported that IFN-α therapy is effective in steroid-resistant cases (14, 15). When symptoms recur in a patient, another therapy must be chosen.

To our knowledge, only 5 cases of HES with mouth ulcers have been reported (16–19), and this is the first report describing a patient who developed a severe swelling of the oral mucosa. It is worth noting that HES can involve the mouth and cause a swelling of the oral mucosa.

**References**