Nonhealing Perianal Ulcers

Jen-Chin Wu      Tien-Yi Tzung

CASE REPORT

A 67-year-old heterosexual Chinese male presented with a one-year history of persistent multiple painful well-defined punched-out ulcers around the anus and in the perineum without involvement of anal mucosa (Fig. 1). There was no lymphoadenopathy on the groin. He had been treated as herpes simplex viral infection with topical and oral antiviral agents without improvement. In addition, he was known to have central diabetes insipidus for 14 years and received nasal desmopressin management. Cultures were obtained for viruses, fungi, and acid-fast bacilli, and all were negative. The results of complete blood cell count, differential count, chest and whole body bone X-ray, abdominal sonography, enhanced computed tomography of the brain with thin slice sections of the sella tursica were unremarkable.

A biopsy specimen was obtained for histopathologic and electron microscopic analysis (Fig. 2 and 3).

Fig. 1
Well-defined punched-out ulcers around the anus.

Fig. 2
Skin biopsy specimen (H&E, x400).

Fig. 3
Electron microscopy, x20000.
DIAGNOSIS: *Eosinophilic Granuloma.*

**MICROSCOPIC FINDINGS AND CLINICAL COURSE**

The skin biopsy specimen demonstrated an ulcer surrounded by a dense cellular infiltrate composed of eosinophils, lymphocytes and histiocytes with abundant eosinophilic cytoplasm, vesicular nuclear chromatin and nuclear grooves. These histiocytes showed positive S-100 staining. On electron microscopy, a population of histiocytes with Birbeck granules (Langerhans histiocytes) was identified. Tacrolimus ointment 0.1% for 3 weeks and later 30 sessions of high dose (130 J/cm²) UV A1 phototherapy had been given to the patient. Despite treatment, the skin condition remained stationary.

**DISCUSSION**

Langerhans cell histiocytosis (LCH), formerly known as histiocytosis X, is a rare disorder with variable clinical forms and uncertain outcome. The key to diagnosis of LCH is identifying the typical Langerhans histiocytes in the appropriate surroundings. Eosinophilic granuloma is a chronic localized form of LCH, primarily occurring in bone. Skin involvement in LCH is not common. When present, infiltrative and ulcerative lesions of the oral, genital and anal areas can be found alone or with diabetes insipidus (DI), skeletal or visceral lesions. DI is the most common manifestation of central nervous system involvement of LCH. The incidence of DI secondary to LCH varies among different reports, ranging from 9.5% to 50%. In some patients, DI can be the first symptom up to 10 years preceding the diagnosis of LCH. In patients of LCH with DI, one or more lesions in the skull or brain can usually be found. The possibility of spontaneous healing even in systemic LCH is well documented. Treatment for benign and self-limited cases should be as minimally aggressive as possible. Topical nitrogen mustard, PUVA, CO2 laser, thalidomide, and isotretinoin have been used to treat cutaneous LCH with fair results. Based on the facts that tacrolimus can inhibit the stimulatory capacity of Langerhans cells and that UV A1 radiation can reduce the numbers of dermal Langerhans cells, our patient had been treated with topical tacrolimus ointment and later with UV A1 phototherapy. However, both treatment modalities were not effective in our case. Concerning DI secondary to LCH, it is usually not reversible by chemotherapy and radiotherapy but can be controlled by vasopressin.

**REFERENCES**