

present. The retrospective analysis of Surveillance, Epidemiology, and End Results database demonstrated head and neck area to be the predominant location of MAC (74%), followed by trunk (9%), upper extremities (8.5%), and lower extremities (5.4%). Only 2 cases (0.9%) involved the vulva (labia majora); whereas 0.9% were categorized as skin not otherwise specified, no penile involvement was reported.¹ Further, we could not identify reports of MAC in the penile region. Syringomatous carcinoma may be considered a variant of MAC and is characterized by ductal proliferation with a tadpole pattern, extension into subcutaneous tissue, and perineural involvement without follicular differentiation. Cellular atypia and mitoses may be observed.²

Although syringomas most commonly develop in the periorbital region, presentation with confluent plaquelike morphology is rare and was first described by Kikuchi et al³ in 1979. To our knowledge, plaque-like syringoma on the penile shaft has only been described once.⁴ Furthermore, although only 1 case of vulvar syringoma with subcutaneous extension has been previously described,⁵ published cases of penile syringomas have been confined to the papillary and mid dermis.

Our rare case of plaquelike penile syringoma with extension of ductal structures to involve the reticular dermis is unusual and creates a potential for misdiagnosing this benign entity. However, consideration of clinical and histologic characteristics can help in distinguishing it from MAC/syringomatous carcinoma. In conclusion, adequate tissue sampling and clinical-pathological correlation are critical to avoid a potential diagnostic pitfall.

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Urticaria pigmentosa complicated with esophageal eosinophilia

To the Editor: Mastocytosis is defined as an accumulation of clonal mast cells, and it may involve only the skin (urticaria pigmentosa, UP) or it may have a systemic presentation with multiorgan involvement. We report a patient with UP associated with esophageal eosinophilia.

A 20-year-old man was referred to our hospital with an 8-year history of itchy eruptions. He had intense itching after rubbing the lesions. He had had no episodes of diarrhea, abdominal pain, dysphagia, or gastroesophageal reflux disease (GERD)-like symptoms. Physical examination revealed multiple round brownish macules of less than 1 cm in diameter on the trunk and extremities (Fig 1). Darier sign was positive. Skin biopsy showed dense mast cell infiltrates in the dermis. They were CD117 (c-KIT) positive. Laboratory analyses showed a white blood cell count of 4600/ μ L, eosinophils at 6.6%, IgE at 737 IU/L, a total tryptase level of 2.2 μ g/mL, and a histamine level of 1.52 ng/mL (normal range: 0.15 to 1.23).

To exclude systemic mastocytosis with multiorgan involvement, we performed computed tomography scans and endoscopic examinations. Upper gastrointestinal endoscopy revealed white linear furrows on the esophageal mucosa (Fig 2). Biopsy specimens from the esophageal mucosa showed dense eosinophilic infiltrates at more than 20 eosinophils per high-power field, and the specimens also contained sparsely infiltrating tryptase-positive mast cells. The infiltrating mast cells aberrantly expressed CD2. No activating germline KIT mutations were detected in the patient's peripheral blood cells. Under the diagnosis of UP complicated with esophageal eosinophilia, we have been following up the patient every half-year without medication.

Eosinophilic esophagitis (EoE) can be diagnosed based on clinical and histopathologic features.^{1,2} Clinically, symptoms of esophageal dysfunction are characteristic of EoE. Histopathologically,

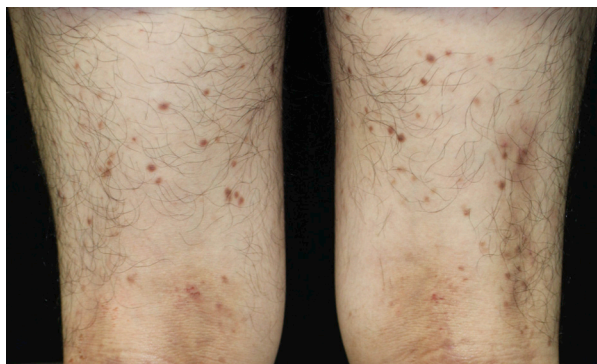


Fig 1. Urticaria pigmentosa. Multiple round brownish papules and macules on the legs.

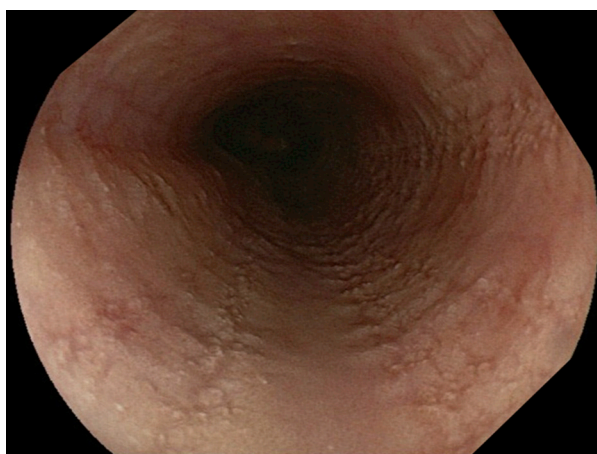


Fig 2. Esophageal eosinophilia. Concentric linear furrows on the esophageal mucosa.

eosinophil-predominant infiltration is seen in esophageal biopsy specimens.^{1,2} Various conditions presenting esophageal eosinophilia, including GERD, should be excluded. The present case could not be diagnosed as EoE, because the patient had no apparent symptoms of esophageal dysfunction, although dense eosinophilic infiltration was seen in the esophageal biopsy specimens and the endoscopic features were consistent with EoE. The patient had no other disorders that might cause esophageal eosinophilia, and we cannot completely exclude the possibility that the patient's condition will progress to EoE in the future.

Chemical mediators released from mast cells are well known to induce eosinophilic infiltration. TGF- β 1, which is expressed by both eosinophils and mast cells, has been recognized as an important molecular mediator of EoE.³ Abonia et al⁴ provided evidence for the involvement of KIT ligand in the pathogenesis of EoE. Niranjana et al⁵ reported that mast cells play a critical role in muscular cell hyperplasia and possibly in the esophageal

functional impairment of EoE. In this context, mast cell infiltration in the esophagus and/or the skin may have triggered the onset of esophageal eosinophilia in the present case, although the mast cell infiltration in the esophagus was sparse and the esophagus was not apparently involved in the mastocytosis. The present case might indicate an association between UP and esophageal eosinophilia.

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Absence of HLA-DR1 positivity in 2 familial cases of frontal fibrosing alopecia

To the Editor: Frontal fibrosing alopecia (FFA) is a variant of lichen planopilaris (LPP), a specific presentation of lichen planus involving the hair follicles.^{1,2} HLA-DR1 positivity has already been linked to cases of lichen planus and another variant