

### Case 11

A 12 month-old female infant

**Chief complaint:** Progressive whitish macules on pubis and inguinal area

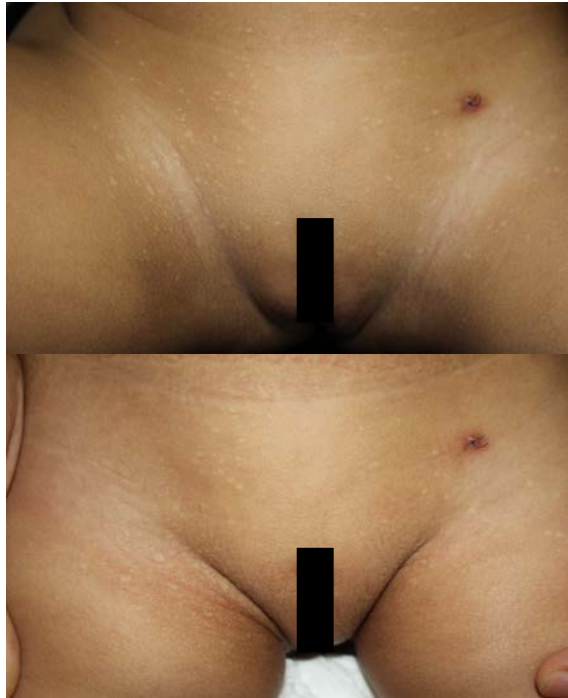


Fig 11.1

### Present illness:

3 months prior to visit the patient developed multiple asymptomatic whitish macules on both inguinal and genital areas. The lesions had been increasing in numbers

**Past history:** The lesions did not improve after treating with topical antifungal drugs.

**Underlying disease:** None

**Family history:** No family history of genetic disease. No similar lesions on her twin sister and relatives.

**Physical examination:** Unremarkable

**Dermatologic examination:** Multiple asymptomatic discrete hypopigmented oval-shaped macules and slightly elevated flat-topped papules on lower abdomen, pubis, inguinal and genital areas

**Lab investigation:** None

**Histopathology:** (S17-040040, skin, left lower abdomen) (Fig. 11.2, 11.3)

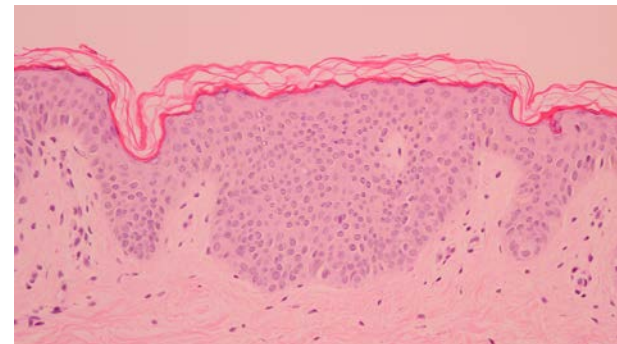


Fig 11.2

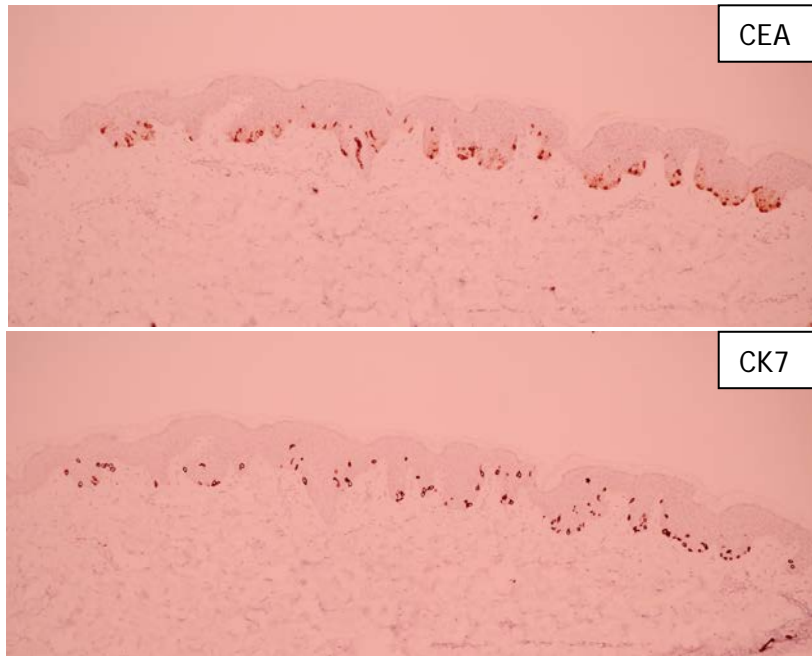


Fig 11.3

- Proliferation of clear cells with round centrally located nuclei and clear abundant cytoplasm along the basal cell layer of the epidermis
- Mild papillomatous epidermal hyperplasia
- CEA, CK7: positive

**Diagnosis: Clear cell papulosis**

**Treatment:**

- Observe

- Follow-up every 3 months

**Presenter:** Kanchana Leerunyakul, MD

**Consultant:** Pamela Chayavichitsilp, MD

**Discussion:**

Clear cell papulosis is a rare cutaneous manifestation that typically presents with multiple asymptomatic macules and flat-topped papules that are barely palpable. The number of lesions ranges from only a few to more than one hundred. Color varies from skincolor to hypopigmentation or depigmentation.<sup>1</sup>

Characteristically, lesions distribute along the milk line in children<sup>1, 2</sup>, predominantly on the abdomen, pubic area and axilla.<sup>3</sup> Patients mostly develop the disease in their early childhood, but not beyond 6 years of age. The etiology is still unknown, but it is believed to originate from eccrine secretory cells of the epidermis.<sup>4, 5</sup>

A skin biopsy is essential for diagnosis. The hallmark of microscopic finding is the presence of proliferating clear cells within the basal cell layer of the epidermis. Some may be scattered in the spinous or granular layers.<sup>1</sup> Clear cells are round-shaped with centrally located nuclei and abundant pale cytoplasm. No cellular atypia should be observed. Hyperkeratosis and acanthosis differ between cases. Immunohistochemistry staining is also used for confirmation. Clear cells are consistently positive for AE1, CEA and EMA.<sup>6</sup>

Clear cell papulosis is a benign condition. None of the reported cases developed malignant transformation and some patients showed spontaneous resolution. In the literature, 85.7% of patients had regression after being followed with a median duration of 11.5

years.<sup>1</sup> Hence, long term follow-up without any treatment is recommended.

Our patient shares a typical presentation and histological features of this rare entity. She has not received any treatment except long-term monitoring.

**Reference:**

1. Tseng FW, Kuo TT, Lu PH, Chan HL, Chan MJ, Hui RC. Long-term follow-up study of clear cell papulosis. *J Am Acad Dermatol.* 2010;63:266-73.
2. Kuo T, Chan H, Hsueh S. Clear cell papulosis of the skin. A new entity with histogenetic implications for cutaneous Paget's disease. *The American journal of surgical pathology.* 1987;11:827-34.
3. Wang D, Ho MS, Koh MJ, Giam YC. A Case Report of Clear Cell Papulosis and a Review of the Literature. *Ann Acad Med Singapore.* 2017;46:160-6.
4. Kim SW, Roh J, Park CS. Clear Cell Papulosis: A Case Report. *J Pathol Transl Med.* 2016;50:401-3.
5. Kuo TT, Huang CL, Chan HL, Yang LJ, Chen MJ. Clear cell papulosis: report of three cases of a newly recognized disease. *J Am Acad Dermatol.* 1995;33:230-3.
6. Seykora CM, Dalton SR, Brown P. Clear cell papulosis: report and review. *Dermatol Online J.* 2018;24.