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

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**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 skin color to hypopigmentation or depigmentation.\(^1\)

Characteristically, lesions distribute along the milk line in children\(^1\),\(^2\), predominantly on the abdomen, pubic area and axilla.\(^3\) 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.\(^4\),\(^5\)

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.\(^1\) 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.\(^6\)

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.\textsuperscript{1} 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.

\textbf{Reference:}