INTRODUCTION

Incontinentia pigmenti (IP) is a rare genodermatosis that is lethal in males. It is a multisystem disorder, which affects ectodermal structures including cutaneous, ocular, dental, cerebral, and skeletal manifestations. A comprehensive review of 600 published clinical reports classified the skin findings in IP into four different stages.

CASE REPORT

An 8-year-old female child was clinically diagnosed with IP at birth. Genetic testing revealed the mutation of IKBKG gene (inhibitor of nuclear factor kappa-B kinase subunit gamma), confirming the diagnosis. Her clinical course was of interest as she experienced reactivation of her IP rash in a blaschkoid pattern on the trunk every time she developed a viral infection from 11 months of age. The flares eventually subsided, with no exacerbations after the age of 2. Over the past 2 years, her mother noted an area of alopecia near the vertex of the scalp. She was unsure of the duration of the hair loss but felt that it has become increasingly noticeable. On examination, she had subtle areas of whorled pigmentation in a blaschkoid distribution on the lower trunk and groin. A linear to whorled area of scarring alopecia was evident near the vertex of the scalp. Hair texture and structure was normal under the dermatoscope. The nail, teeth, and rest of the skin showed no abnormality. Neurological and ophthalmological assessments revealed no systemic features of IP.

DISCUSSION

A review of all manifestations of IP identified whorled alopecia as a recognized feature of the condition, but diffuse alopecia was aborted. In a case series by Hadj-Rabia et al., alopecia was identified in 11 (28%) of the 40 patients occurring at the site of previous blistering and generally of the hair loss but felt that it has become increasingly noticeable. On examination, she had subtle areas of whorled pigmentation in a blaschkoid distribution on the lower trunk and groin. A linear to whorled area of scarring alopecia was evident near the vertex of the scalp. Hair texture and structure was normal under the dermatoscope. The nail, teeth, and rest of the skin showed no abnormality. Neurological and ophthalmological assessments revealed no systemic features of IP.
unnoticed. Abnormal hair tended to be lusterless, wiry, and coarse, mainly in the vertex. Woolly hair has also been reported, but the hair shaft was normal on microscopic examination. Thin or sparse hair early in childhood did not seem to correlate with the quality or quantity of hair in later life.

In 1990, Happle argued that the linear and patchy alopecia seen in the patients with IP can be a manifestation of functional X chromosome mosaicism. They appeared to correspond to Blaschko’s lines. Functional X chromosome mosaicism was recognized as a genetic mechanism underlying cutaneous anomalies seen in a number of X-linked diseases such as IP. It has been hypothesized that the segmental and streaked manifestations of IP may be consequential to tissue mosaicism from random X inactivation, with the normal X chromosome active in uninvolved skin and the IP X chromosome active in involved skin. A similar pathogenesis could also explain the whorled pattern of alopecia in our patient.

This phenomenon may be underreported and dermatologist should specifically seek this sign as it could be hidden under the scalp hair. The scarring alopecia tends to be permanent and can be a useful marker to ascertain affected adult women who may no longer have cutaneous manifestations. In our patient, with the gradual resolution of cutaneous features and in the absence of systemic findings, scarring alopecia may be the only indication of IP as an adult.

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Conflicts of interest
There are no conflicts of interest.

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