# Giant blaschkoid-oriented nevus spilus with vulvar involvement

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## Introduction

Nevus spilus is a lesion composed of hyperpigmented macules or "speckles" with a café-au-lait-like patch. 1-2 We report a case of a blaschkoid-oriented nevus spilus within vulvar involvement.

#### Case

A 43-year-old woman with history of prediabetes, hypertension, asthma, eczema, and seborrheic dermatitis presented for follow-up of onychomycosis. On physical examination, she was noted to have multiple, hyperpigmented macules within an irregularly shaped, tan brown patch on the right calf. This patch followed a linear, blaschkoid distribution and extended proximally to the right labium majus. She reported that this lesion had been present since childhood; she denied any similar skin findings in family members. A diagnosis of nevus spilus was rendered. The patient also reported that she was going to see her gynecologist since she had noticed that the lesion involved her vulva.



Figure 1: Several hyperpigmented macules distributed within an irregularly shaped, tan brown patch (following a linear, blaschkoid distribution) involving the right calf and extending proximally to the right labium majus.

### Discussion

- Often congenital, nevus spilus can be associated with a variety of genetic mutations (e.g. BRAF, HRAS), with large/giant ones showing mutations at codon 61 of NRAS.<sup>3</sup>
- To our knowledge, only a few cases have been reported of nevus spilus involving oral mucosa and in a blaschkoid distribution, and none with genital involvement.<sup>4</sup>
- Most importantly, nevus spilus-type congenial melanocytic nevus (CMN) has a small risk of developing into melanoma.<sup>5</sup>
- In summary, this case highlights that nevus spilus can rarely affect the vulva.

#### References

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