

CASE REPORT

Agminated Spitz Nevi on a Hyperpigmented Macule

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Abstract. We present the case of a 2-year-old boy with multiple Spitz nevi clustered on a hyperpigmented macule that obeyed a quadrant distribution. Multiple Spitz nevi are rare and can be disseminated or clustered. A third of the cases of agminated lesions appear on hyperpigmented macules.

Key words: nevus, Spitz nevus, spindle and epithelioid cell nevus, mosaicism.

NEVOS DE SPITZ AGRUPADOS SOBRE UNA MÁCULA HIPERPIGMENTADA

Resumen. Presentamos el caso de un niño de 2 años con múltiples nevos de Spitz agrupados sobre una mácula hiperpigmentada, que seguía un patrón en cuadrante. Los nevos de Spitz múltiples son raros y pueden presentarse de forma diseminada o agrupada. Un tercio de las lesiones agrupadas puede aparecer sobre una mácula hiperpigmentada.

Palabras clave: nevo, nevo de Spitz, nevo de células epitelioides y fusiformes, mosaicismo.

Introduction

Spitz nevus, or spindle and epithelioid cell nevus, normally presents as a single lesion that tends to appear in the first 2 decades of life. It consists of a shiny red-to-brown papule. Multiple Spitz nevi are much less frequent and can occur in disseminated or agminated form. They have been classified into 4 groups: disseminated, agminated on normal skin, agminated on a hypopigmented area, and agminated on a hyperpigmented area.¹ Of these, the disseminated form is the least frequent and tends to occur in adults.^{2,3}

Less than 20 cases of agminated Spitz nevi appearing on a pigmented macule have been described in the literature. We present the case of a patient with multiple Spitz nevi agminated on a mildly hyperpigmented, quadrant-shaped macule.

Case Description

We describe a 2-year-old boy, with no significant medical history, admitted to our center presenting a wide pigmented

lesion on the shoulder, scapula, and right arm. A large pigmented macule had been observed for the first time at this site when the patient was 3 months old, and at 13 months multiple papules began to gradually appear on the macule. Physical examination showed a light brown homogeneous macule with a quadrant-shaped or flag-shaped outline, with well-delimited irregular edges on the shoulder, scapula, and external side of the right arm: it was roughly square-shaped with an inner edge at the midline of the back and perpendicular upper and lower edges at the height of the shoulder and subscapular region, respectively. The macule extended to the outer anterior aspect of the arm with a perpendicular lower edge 2 cm below the cubital fossa. More than 20 papules, ranging from pink to light or dark brown, 1- to 5-mm diameter, were diffusely distributed over the macule. Larger lesions were lighter, somewhat shiny, hard to the touch, and dome-shaped (Figure 1 and Figure 2). The lesion was completely asymptomatic. A biopsy was performed on one of the papules showing a predominantly intradermal proliferation of symmetrical, deep, mature, epithelioid and spindle melanocytes (Figure 3 and Figure 4). The histopathological diagnosis was intradermal Spitz nevus. Subsequently, the patient underwent surgical excision of 10 more papules that were all diagnosed as Spitz nevi. Histopathology of the underlying hyperpigmented macule only demonstrated basal layer hyperpigmentation without melanocyte proliferation (Figure 5).

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Figure 1. Macule in the form of a quadrant with brown papules of various hues within its margins.



Figure 2. E lesion extending to the right arm.

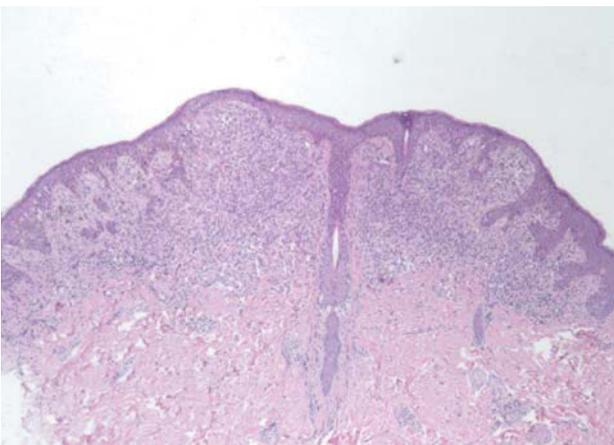


Figure 3. Symmetrical, well-defined, melanocytic lesion (hematoxylin-eosin stain, $\times 10$).

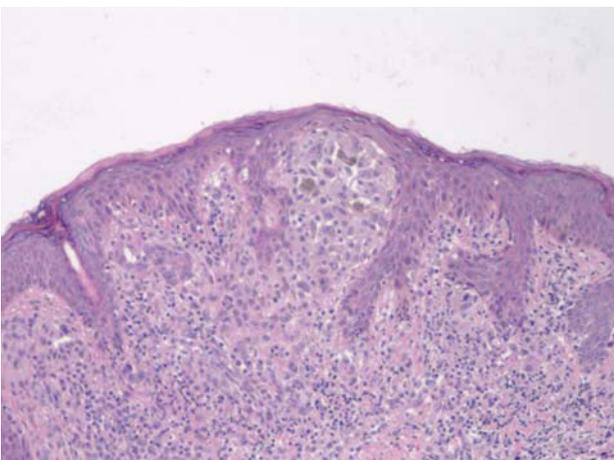


Figure 4. Epithelioid and spindle cell proliferation (hematoxylin-eosin stain, $\times 100$).

Discussion

It has been suggested that all nevi reflect mosaicism and different clinical cutaneous patterns have been described that reflect the presence of genetic mosaicism.⁴ Quadrant or flag-shaped outlines, or checkerboard patterns when there are several alternating lesions, appear in some melanocytic or vascular lesions that, as in our case, affect a well-defined rectangular area on 1 side of the body without crossing the midline. The agminate pattern appears in some melanocytic lesions or collagenous tissue lesions that are closely grouped together, and is therefore also known as a speckled pattern.

The term agminate is frequently used in the literature regarding most clustered Spitz nevi.⁵ This is not a good choice to describe cases with more dispersed lesions on

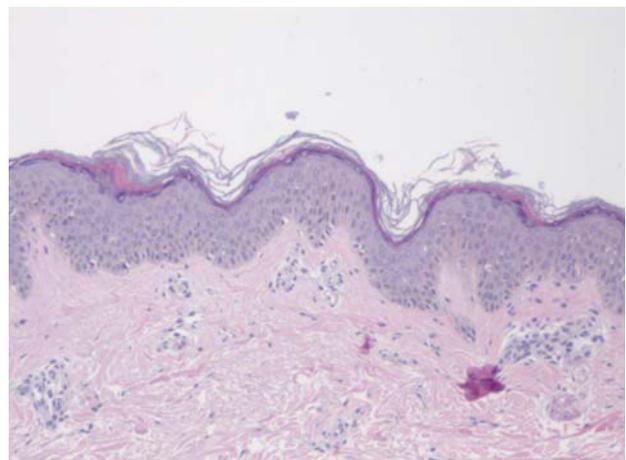


Figure 5. Basal layer hyperpigmentation with no increase in the number of melanocytes (hematoxylin-eosin stain, $\times 40$).

normal or hyperpigmented skin because these take the form of a quadrant, as in the case presented.^{1,5-8}

Around 40 cases of agminated Spitz nevi have been reported in journals published in English.^{9,10} These can appear on apparently healthy skin, hyperpigmented skin or, less frequently, hypopigmented skin. The number of Spitz nevi present in these lesions range between 2 and several hundred. There seems to be no predominance by sex, more than 50% of the cases occur in patients under 5 years, and the most frequent site is the face, followed by the arms.⁹

A third of the cases of multiple Spitz nevi appear on a hyperpigmented macule.^{9,11} This type of macule has been clinically described as a “café au lait macule,”⁹ a “brown macule,”¹¹ or a “nevus-spilus-like macule.”¹² In other cases, the appearance of agminated Spitz nevi on an existing nevus spilus has been described.¹³⁻¹⁵ Many of the published cases lack a histological study of the hyperpigmented macule,^{16,17} but in some cases this has been regarded as a junctional nevus,¹ basal hyperpigmentation, with or without melanocyte proliferation,^{7,16} or nevus spilus.¹³ It is quite likely that the histological differences between the hyperpigmented macule of a nevus spilus and a café au lait macule are only minor,⁹ and thus an agminated Spitz nevus on a hyperpigmented macule could be regarded as a spilus nevus subtype.¹⁷

The cause of multiple Spitz nevi is unknown. Multiple etiologies have been suggested that have been associated with sunlight exposure and burns,¹⁸ previous excision of an isolated lesion,¹⁰ pregnancy,¹⁵ and previous radiation therapy.¹⁹ It has been suggested that external stimuli could induce these lesions in a genetically predisposed individual.²⁰ Nevertheless, very often there are no associated predisposing factors, as in our case. It seems reasonable to assume that there may be a mutation in an oncogene or suppressor gene some time during embryogenesis giving rise to a clone of cells with greater potential for developing Spitz nevi.¹⁶

Due to the difficulty in differentiating melanoma from Spitz nevus, there is a trend toward excising isolated lesions,²¹ but this could be very difficult in individuals with multiple lesions. No case of conversion of multiple Spitz nevi to malignant melanoma has ever been reported. Thus, conservative treatment may be an option.^{22,23} However, the known possibility of a malignant transformation of a nevus spilus and limited experience of agminated Spitz nevi means that there should at least be strict clinical follow-up of these lesions and biopsy of atypical elements, in addition to monitoring should any suspicious change occur.

Conflicts of Interest

The authors declare no conflicts of interest.

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