Acquired hyperpigmented lesion on the foot

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Pouran Layegh, MD[1]
Nona Zabolinejad, MD[2]
Naghmeh Zabolinejad, MD[1]
Akram Momenzadeh, MS[3]

1. *Department of Dermatology, Mashad University of Medical Sciences, Mashad, Iran.*
2. *Department of Pathology, Mashad University of Medical Sciences, Mashad, Iran.*
3. *Research Center for skin diseases & cutaneous Leishmaniasis, Mashad, Iran.*

Correspondence author:
Pouran Layegh, MD
Associate Professor of Dermatology, Qaem Hospital, Mashad University of Medical Sciences
Email: Pouran_Layegh@yahoo.com

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CASE

A 3.5-year-old girl presented with a 1-year history of a slow growing pigmented lesion on the dorsal aspect of her right foot. Physical examination revealed the presence of a pigmented patch with color distribution from pink to tan to dark brown, relatively well circumscribed and approximately 1.5 × 2cm with irregular borders, and especially a single dark central papule superimposed in the middle of it (Figure 1). The lesion had a soft consistency and the child did not have any other symptoms. During the last year, it had increased in dimensions and thickness.

The remainder of her physical examination was insignificant. Also, there was no positive family history of a particular disease or any history of trauma or drug usage. A 3 mm punch biopsy was obtained from the central dark papule and the specimen was sent for histopathologic examination.

What is your diagnosis?

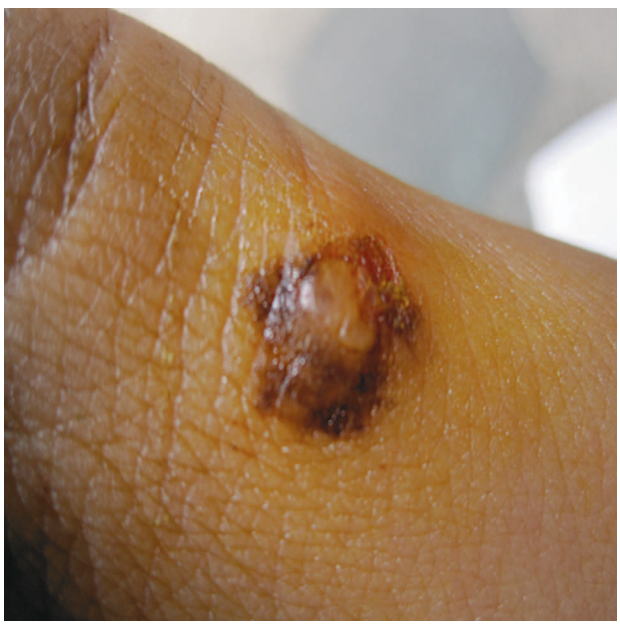

Figure 1. A pigmented plaque with color distribution from pink to dark brown on the dorsal aspect of the left foot.

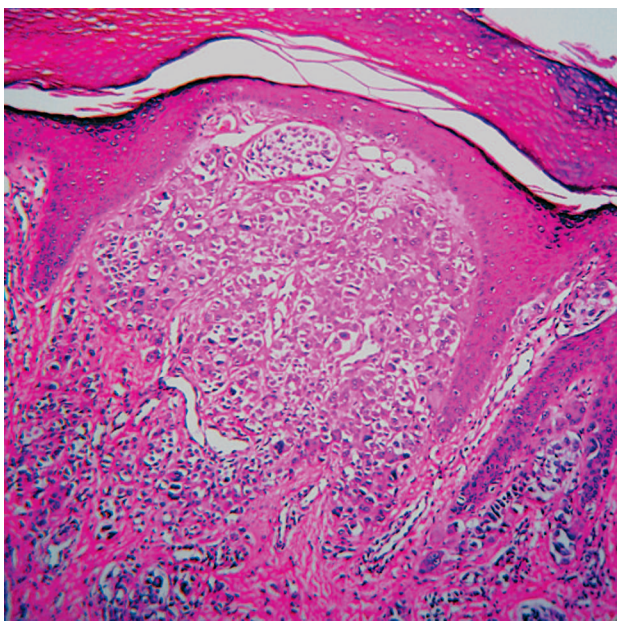

Figure 2. The histopathology view: intradermal nests of large epithelioid cells with a polygonal shape, occasional multinucleation, and a strongly eosinophilic cytoplasm (H&E × 10)

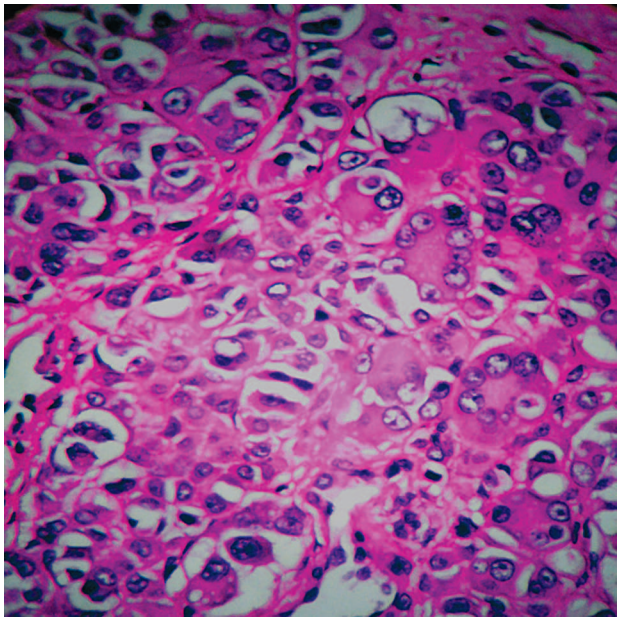

Figure 3. The nuclei were large with smooth nuclear membranes and prominent nucleoli (H&E*40)

DIAGNOSIS

Spitz nevus

Microscopic findings

The histopathology of the specimen revealed a well circumscribed and symmetrical lesion which was composed of intradermal nests of large epithelioid cells with polygonal shape, occasional multinucleation, and a strongly eosinophilic cytoplasm (Figure 2). The nuclei were large with smooth nuclear membranes, and prominent nucleoli (Figure 3). There was no mitosis or necrotic cell. The clinical and histopathological features were consistent with diagnosis of spitz nevus.

DISCUSSION

The Spitz nevus, named after Sophie Spitz who first described it in 1948, is also known as benign juvenile melanoma and spindle and/or epithelioid cell nevus. The lesion was originally believed to occur largely in children, but it is now well recognized in young to early middle-aged adults [1]. The prevalence of Spitz nevus has not been accurately documented in the general population. However, Spitz nevus account for approximately 1% of melanocytic lesions. Spitz nevi are mostly acquired, but congenital ones have been reported as well [2]. They are seen in all age groups but are uncommon beyond the age of 40 to 50 years. The lesions in adults are more pigmented than in children [2,3].

No particular etiologic factors have been found [2]. They may be derived from the same progenitor cells that give rise to epidermal melanocytes and nevomelanocytes [3]. Amplifications of chromosome 11p and H-RAS and activating mutations of H-RAS have been noted in a subset of Spitz nevi [4]. Spitz nevi vary in size from 2 mm to 2 cm or more, with an average diameter of approximately 8 mm. Most commonly, they are well circumscribed, dome-shaped papules or nodules varying in color from pink to tan to dark brown. Generally, the color is homogeneous and the margins are well defined. Relatively flat polypoid and pedunculated morphologies have also been described. Occasionally, lesions may have erosions and scale-crust. Telangiectasia is seen frequently [2]. The head and neck area is probably the most common site, accounting for 42% of lesions in one series. Other parts of the body can be affected less frequently. It often has a recent onset, but a small percentage of nevi may be present for many years [2].

Atypical Spitz nevi refer to lesions demonstrating one or more (usually a constellation of) features that deviate from conventional Spitz nevi. The features may include a large size (e.g. > 1 cm in diameter), asymmetry, deep involvement of the dermis or subcutis, ulceration, easily found dermal mitoses (>2-3 mitoses/mm2), being specially deep, a significant pagetoid spread, prominent confluence and high cellular density of melanocytes in the dermis, and lack of maturation [2].

Histologically, these lesions display striking nests of large epithelioid cells, spindle cells or both, usually extending from the epidermis into the reticular dermis in an inverted-wedge configuration. The closely apposed nests of cells within a uniformly hyperplastic epidermis often contribute to a so-called 'raining-down' appearance. Both mononuclear and multinucleate giant epithelioid cells are frequently observed. These cells extend into the subjacent dermis as both single cells and as nests or fascicles. Occasional bizarre cytologic features, necrotic cells and mitotic figures are found within even the most banal lesions [2,5]. Differentiation from a melanoma can often be very difficult and occasionally even impossible. Some

features that favor the diagnosis of Spitz nevus are a symmetric shape, sharp lateral demarcation maturation in depth, tadpole and multinucleated giant cells and lack of upward epidermal spread.

The clinical differential diagnosis of Spitz nevi is wide and includes other melanocytic nevi, particularly dermal nevi, hemangiomas, pyogenic granuloma, verrucae, molluscum contagiosum, juvenile and adult xanthogranulomas, dermatofibroma, mastocytoma, clear cell acanthoma, insect bite reactions, seborrehic keratoses, epidermal nevus and adnexal tumor [2,3]. The most important diagnostic problem is the histologic differentiation of Spitz nevus from cutaneous melanoma [2]. Complete excision with margins free of tumor is recommended for all Spitz nevi. However, there are clinicians who reserve this recommendation for lesions with any atypical feature (clinically or histologically) or Spitz nevi in adults [2]. Patients with atypical lesions should be evaluated every 6 to 12 months.

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