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Letter to the Editor
Regression of a pigmented Spitzoid lesion in an adolescent

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To the Editor

Spitzoid melanocytic lesions are a controversial group of tumors due to their overlapping clinical features, biological behavior, and challenging treatment. Although Spitz/Reed nevi are benign tumors, their clinical and dermoscopic overlap with Spitzoid melanoma makes the management of these lesions particularly demanding (1).

A 15-year-old boy, accompanied by his father, presented to a dermatology clinic in March 2021 for a preventive mole examination. The family history for cancer was negative. On clinical examination, a darker skin phototype (between 3 and 4) was observed, and approximately 10 brown nevi of the globular type and age-appropriate appearance were found. A darkly pigmented nevus on the lateral aspect of the left upper arm, 8 mm in size, stood out in appearance: the ugly duckling sign (Fig. 1). Dermoscopically, it was an asymmetric Spitzoid lesion with a central inverse network and radial streaks in the periphery, with central blue-gray clods (melanin in dermal macrophages) and a few black dots (melanin in the epidermis; Fig. 2). The differential diagnosis was Spitz/Reed nevus, Clark nevus, atypical Spitz tumor, and Spitzoid melanoma. The father reported that a "darker mole on the upper arm" had been present from the age of six. On the basis of the clinical and dermoscopic picture and the history, a diagnosis of Spitz/Reed nevus was made. We decided on a follow up with digital dermoscopic imaging (FotoFinder) and scheduled the next checkup in 6 months, which the patient did not come to. He and his father returned in August 2022, 17 months later. The lesion had almost completely regressed by that time (Fig. 3). On re-examination in November 2022, we observed continuation of the regression process (Fig. 4, Fig. 5).

Spitzoid melanocytic lesions range from benign to malignant tumors. They are divided into three subgroups that are difficult to distinguish clinically, dermoscopically, and histopathologically, making their management in daily practice often very challenging (1): Spitz/Reed nevus, which has a benign course; atypical Spitz tumor, which has a nodular appearance, atypical pathohistological features that are insufficient for the diagnosis of melanoma, and unclear malignant potential; and Spitzoid melanoma, whose prognosis is not different from other types of melanoma of comparable thickness (2, 3).

Spitz nevus is a type of melanocytic nevus presenting with epithelioid and/or spindle-shaped melanocytes (2). It was first described in 1948 by the histopathologist Sophie Spitz, who named it *melanoma of childhood* or *juvenile melanoma*. This term soon turned out to be unsuitable because of its occurrence in adults and its benign nature. In 1975, Reed described a highly pigmented benign melanocytic lesion occurring mainly on the lower limbs in young adults. Some authors consider Reed nevus to be a separate entity, whereas others refer to it as a pigmented variant of Spitz nevus because histopathologically they both show proliferation of large epithelioid and/or spindle-shaped melanocytes (4). Spitz/Reed nevi are rare, with an incidence rate between 1.4 and seven cases per 100,000 population per year. They mostly occur in children and young adults: 40% of nevi occur under 15 years of age and 77% under 30 years of age. They constitute less than 1% of all melanocytic tumors excised in childhood (2, 5). A Yale University cohort of patients with 484 Spitz nevi and 54 Spitzoid melanomas between 1991 and 2008 confirms that Spitz nevus is more common in young people, with a median age at diagnosis of 22 years, whereas the median age at diagnosis for Spitzoid melanoma was 55 years. Spitz nevi are therefore the hallmark of childhood and young adulthood, whereas increasing age increases the risk of melanoma diagnosis (6).

Dermoscopically, four patterns are characteristic of Spitz/Reed nevi. The starburst pattern is the most common and occurs in half of cases. It is characterized by central blue-gray to black pigmentation around which there are radial streaks. This is followed by a globular pattern with reticular depigmentation: an inverse network in the center and large brown globules at the periphery, a pattern of dotted vessels with an inverse network in flat non-pigmented lesions, and a pattern of globular or coiled vessels with an inverse network in nodular non-pigmented lesions. Approximately 20% of Spitz/Reed nevi dermoscopically show a multi-component pattern: an asymmetric arrangement of structures and colors, and a blue-whitish veil may be present. This form is problematic because it cannot be distinguished from melanoma (1, 7). In fact, there are no clinical or dermoscopic features that can reliably distinguish Spitz/Reed nevus from Spitzoid melanoma. Even a perfectly symmetrical lesion that completely mimics a Spitz/Reed nevus may be a melanoma (1).

The distinctive feature of Spitz/Reed nevus is its transformation, which takes place in three stages. After a phase of rapid, often dramatic, growth, it usually stabilizes and gradually undergoes an involution process. As many as 80% of them spontaneously disappear or change into another more common type of melanocytic nevus with age (7). This explains why Spitz/Reed nevi are rarely found in the elderly. For this reason, we should pay extra attention when Spitzoid lesions occur in the elderly (8).

The likelihood of a Spitzoid lesion being a melanoma increases linearly after puberty, whereas it is very low below 12 years of age. Lallas et al. (9) calculated, on the basis of 333 nevi and 51 melanomas, that there is a 13.3% probability of melanoma in those 12 years and older with a symmetrical Spitzoid lesion. This probability is 2%

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between 12 and 20 years, 7% between 12 and 30 years, and 50% or higher after age 50. Age 50 is the cutoff point above which the risk of melanoma is equal to or greater than the chance of a nevus. After 70 years of age, any Spitzoid lesion should be considered melanoma (9).

The International Dermoscopy Society has made recommendations for the management of Spitzoid lesions based on the probability calculations described above, with the aim of missing as few melanomas as possible while avoiding unnecessary excision of benign nevi. This is particularly important in children, in whom the majority of Spitzoid lesions occur and in whom excision is burdensome and difficult (1). Because melanoma can be clinically and dermoscopically identical to Spitz/Reed nevus, the only safe strategy is excision of all Spitzoid lesions in patients older than 12, excision of dermoscopically asymmetric Spitzoid lesions even in patients younger than 12, excision of dermoscopically symmetric Spitzoid nodular lesions also in patients under 12 or close follow-up, and follow-up of dermoscopically symmetrical flat Spitzoid lesions in children under 12 until stabilization.

In any case, each situation should be individually weighed and decided on regarding the overall clinical context (anatomical site, family environment, etc.). Because no case of melanoma or atypical Spitz tumor with a symmetrical starburst pattern has been described in children before puberty, and because a large proportion of initially symmetrical Spitz/Reed nevi do not follow a symmetrical growth pattern, monitoring these lesions may lead to unnecessary parental concern and unnecessary excisions. Therefore, a modified recommendation was made: after a diagnosis of a flat pigmented dermoscopically symmetrical Spitz/Reed nevus before puberty, further follow-up is not needed (10).

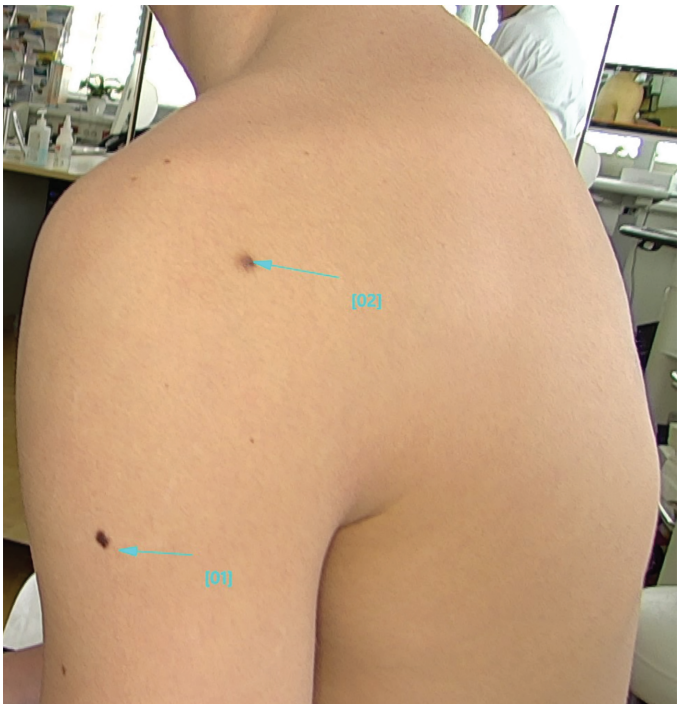

Figure 1 | Spitz/Reed nevus on the left upper arm, March 2021.

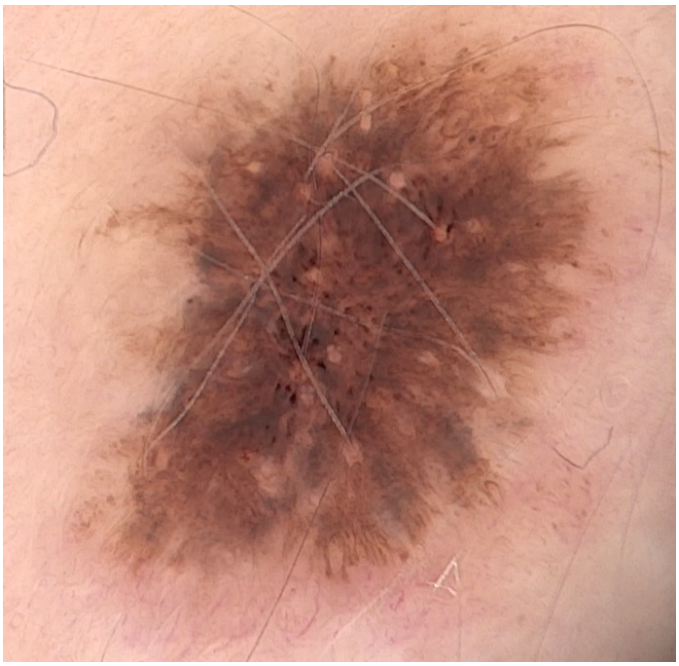
Figure 2 | Dermoscopic image, March 2021.

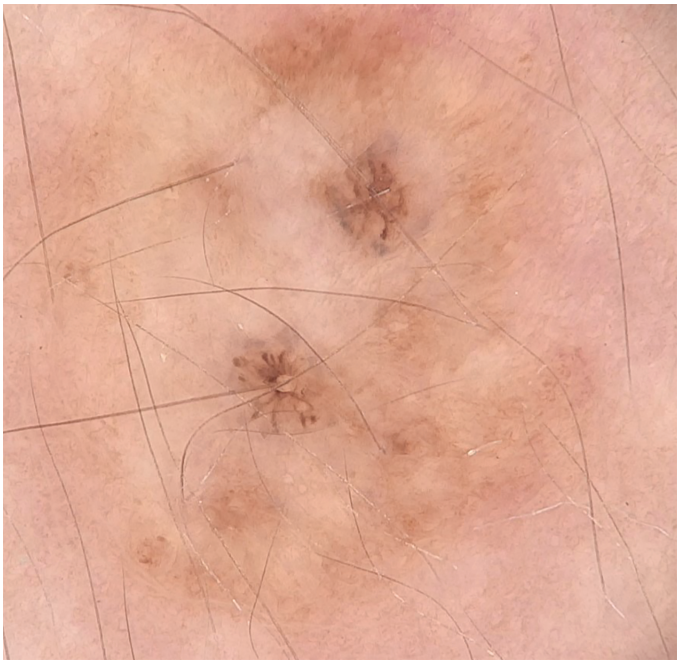
Figure 3 | Dermoscopic image, August 2022.

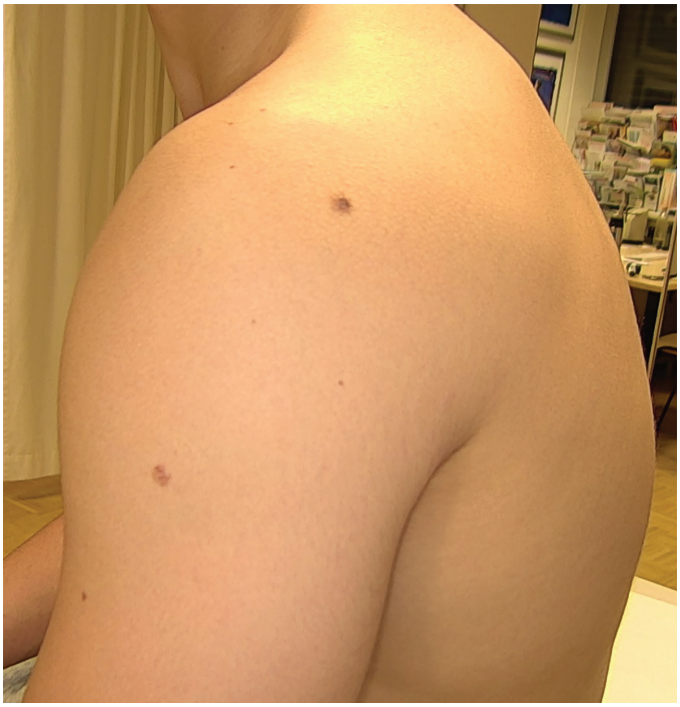
Figure 4 | Nevus regression, November 2022.

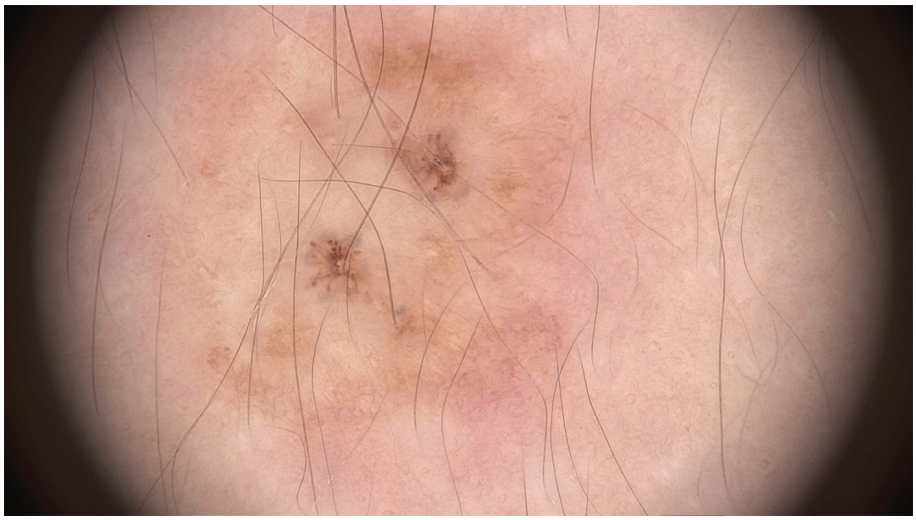

Figure 5 | Dermoscopic image, November 2022.

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