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CASE REPORT / ПРИКАЗ БОЛЕСНИКА

Clinical and dermoscopic spectrum of age-dependent spitzoid lesions – when to react?

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SUMMARY
Introduction Spitzoid lesions represent a spectrum of melanocytic lesions comprising benign Spitz nevi, intermediate lesions known as atypical Spitz tumors, and Spitzoid melanoma. They tend to be more common in children and young adults, but all age groups can be affected. Due to complexity of their clinical, dermoscopic and histological differentiation, they are extremely difficult to manage, especially in pediatric population.
Outlines of cases In this report, we present a series of six cases with spitzoid lesions in different age groups with different outcomes.
Conclusion With the following case series, we report clinical and dermoscopic features of biologically various spitzoid lesions, appearing in different age groups. We believe that this article will increase knowledge of both physicians and dermatologists about when and how to react when dealing with a patient with spitzoid lesion.
Keywords: spitzoid lesions; Spitz nevus; atypical spitz tumor; spitzoid melanoma; management

INTRODUCTION

Spitzoid lesions present a distinct group of melanocytic lesions with overlapping clinical, dermoscopic and in some cases even histopathological features. They are classified into benign Spitz nevi (SN), intermediate lesions known as atypical Spitz tumors (AST), and spitzoid melanoma (SM) [1]. While typical SN is a pink to reddish-brown or purple-red papule, a highly pigmented variant of SN also exists and is referred to as Reed nevus (RN) [2]. Spitzoid lesions are extremely rare, as only 1–2% of all melanocytic lesions in both children and adults are diagnosed as SN, while AST and SM are even less common than SN [3, 4]. Most lesions are up to 1 cm in diameter, with a wide range of coloration from pink to black [1, 5]. Although more frequently found in childhood and young adulthood, they occur in individuals of all ages. Currently, no consensus guidelines exist for the management of spitzoid lesions, and due to complexity of their clinical, dermoscopic and histological differentiation, they are extremely difficult to manage.

REPORT OF CASES

Case 1

A six-month-old female infant presented to our dermatology department with a pigmented macule in the umbilical region which was discovered by accident during examination. The lesion was dark brown to black, flat, well demarcated, with a diameter of 5–6 mm. Dermoscopy revealed a melanocytic lesion with a starburst pattern typical of a pigmented Spitz nevus (Figure 1). Follow-up was suggested, every six months, without excision. At the age of two years, the lesion changed symmetrically maintaining the previously noted starburst pattern (Figure 2).

Case 2

A 30-year-old woman presented to our dermatology department with a pigmented lesion on the interior surface of her right upper leg. The patient reported the presence of the lesion since early childhood, but noted that the lesion started changing in color and diameter in the previous several months. Dermoscopic findings revealed an asymmetrical melanocytic lesion, without a clear pattern of pigment distribution. In the center of the lesion milky gray, homogenous pigmentation with subtle pin-point peppering, and remanences of streaks that resemble starburst pattern were found (Figure 3). Wide local surgical excision was suggested and histopathological findings showed features of an atypical spitz tumor, without necessary criteria for the diagnosis of an SM.

Case 3

A 40-year-old woman presented to our dermatology department with a newly acquired pigmented papule on the lateral aspect of her right knee. Dermoscopy showed a melanocytic lesion with homogenic structureless dark pigmentation in the center, surrounded by asymmetrical pseudo-pod-like structures resembling a

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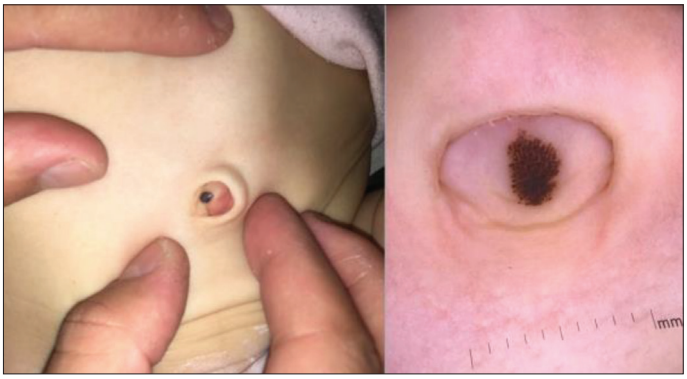

Figure 1. Spitz nevus in the umbilical region of a six-month-old infant

Figure 2. Spitz nevus in the umbilical region of a two-year-old child

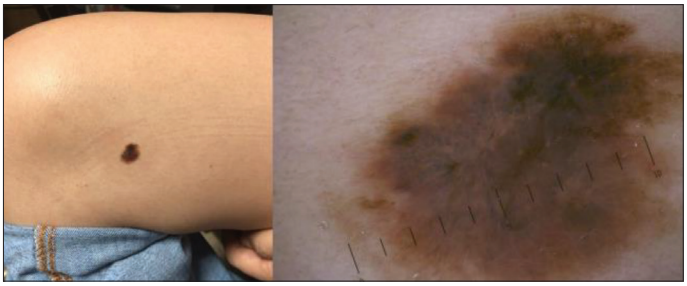

Figure 3. Atypical spitz tumor in a 30-year-old woman

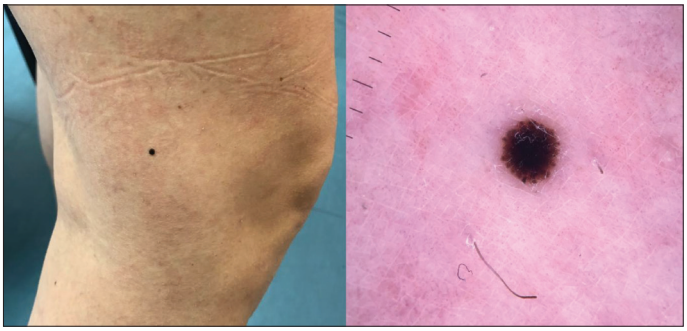

Figure 4. Spitzoid melanoma in situ in a 40-year-old woman

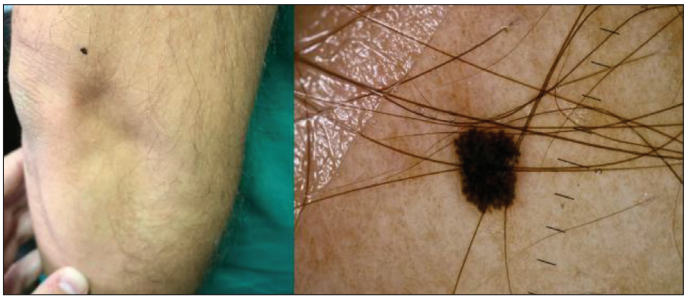

Figure 5. Spitzoid melanoma in a 47-year-old man

starburst-pattern of a Reed nevus (Figure 4). Having in mind the dermoscopic findings and the patient age, wide local surgical excision was suggested. Histopathology revealed atypical epithelioid melanocytes confined to the epidermis and epidermal adnexal structures. The diagnosis of SM *in situ* was made.

Case 4

A 47-year-old man presented to our dermatology department with a newly appeared pigmented papule on his left elbow. Dermoscopy showed asymmetry and radial streaming, pseudopods, and peripheral black dots and globules (Figure 5). Wide local surgical excision was suggested. Histopathology revealed a microinvasive, 0.2 mm-thick SM.

Case 5

A 3-year-old girl presented to our dermatology department with a flesh-colored nodule on the extensor surface of her left lower leg, which had grown over the past six months, more intensively over the past two months. Her mother stated she had the lesion for a longer time, and that it was flat and pigmented prior to the rapid growth phase. Dermoscopy showed a melanocytic lesion, centrally without pigment and with thin interlaced blood vessels. Towards the periphery of the lesion individual globules were present. In one most peripheral zone, the remains of a starburst or coffee bean-like appearance that looked like remnants of a spitzoid lesion (Figure 6). Wide local surgical excision was suggested and histopathology revealed a 2 mm-thick SM, Clark level 4.

Case 6

A 20-year-old man presented to our dermatology department with a pigmented nodular lesion on the extensor surface of his right lower leg. The patient reported a known presence of the observed lesion since his early childhood, also he noted that the lesion started changing in color, diameter and thickness in the past several months Dermoscopy showed remanences of a melanocytic lesion, with absence of color uniformity and pattern symmetry, but also presence of asymmetrically distributed brown dots, pseudopods and central blue–white veil (Figure 7). Wide local surgical excision was suggested and histopathology revealed a 2.3 mm-thick SM, Clark level 4.

All procedures were performed in accordance with the ethical standards of the institutional and national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. Written consent to publish all shown material was obtained from the patients.

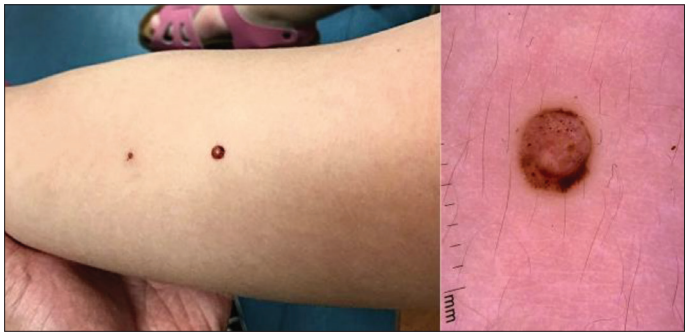

Figure 6. Spitzoid melanoma in a three-year-old girl

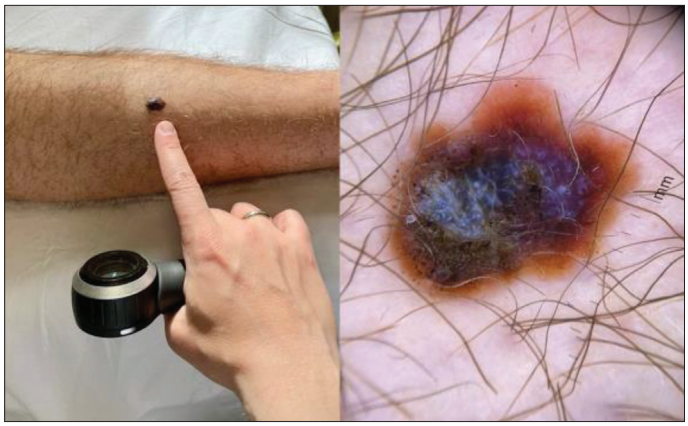

Figure 7. Spitzoid melanoma in a 20-year-old man

DISCUSSION

Spitz nevi present benign spitzoid lesions and are on average less than 6 mm in diameter. They are typically described as dome-shaped, but may as well be flat or polypoid. [3]. A pink to red color is most commonly observed, but flesh-colored and brown to black lesions have also been seen in SN, the latter of which are also referred to as Reed nevi [2, 3]. Some authors consider RN a single nosological entity, but whether SN and RN are distinct lesions, or unpigmented and pigmented variant of the same spectrum is still controversial, due to their both similarities and differences [2].

Atypical spitz tumors are currently defined as tumors of uncertain malignant potential. Their diameter ranges 5–10 mm and they present as a plaque or a nodule in variety of colors ranging from pink to black. The multicomponent pattern is characterized by an uneven distribution of colors and structures [3, 4, 5].

Spitzoid melanomas tend to be larger than AST, with a mean diameter of 1.05 cm, more rarely a SM less than 6 mm can also occur [6]. Spitzoid melanomas are more likely to be nodular, but may also be flat or slightly elevated [3]. Majority of SM lesions have a multi-color pattern as Carrera et al. [7] demonstrated a mean number of 2.7 colors per SM lesion, while the spectrum ranges from red/pink, grey to brown/black.

Dermoscopically, the majority of SN (53%) exhibit a starburst pattern. However, a notable percentage of cases also display globular, coffee-bean like appearance (22%) and atypical pattern (25%) while in hypopigmented SN the most characteristic feature is vascular pattern [2]. Atypical spitz tumors are more challenging to classify as their characteristics overlap with SN and SM [1]. They are more likely to show a non-specific or multicomponent pattern in comparison to SN, but also exhibit a dotted vessel pattern and shiny white lines as frequently as SM [7, 8]. Spitzoid melanomas tend to exhibit more asymmetry and more colors, pink Spitz-like pattern, milky red areas, shiny white lines, red/pink and white coloration, and polymorphous vascular pattern [7].

Age is one of the main criteria to distinguish Spitzoid lesions with indolent behavior from AST with greater risk of malignancy and SM [9]. However, it should not be overly relied on, as although melanomas are rare in pediatric population, SM represent the most common type regarding children, making it the most frequent malignant skin tumor in children [10, 11]. As shown both in the literature and in case of our patient No. 5, nodular spitzoid lesions with multicomponent dermoscopical pattern should always be excised, regardless of age. On the other hand, evolution of SN undergoes from rapid, often dramatic growth phase and in these cases many physicians and even dermatologists tend to raise a red flag, especially when dealing with pediatric population and react advising excision, though unnecessary. These lesions usually stabilize as shown in our case No. 1 and may gradually undergo an involution process [12].

Several studies involving both pediatric and adult patients reported a mean age that ranges from late teens to early 30s, and Lott et al. [14] reported a mean age of 22 years at the time of diagnosis of SN [1, 11, 13]. Very few studies reported an average age of AST diagnosis solely, one of them was conducted by Moscarella et al. [8] and they reported a mean age of 20.8 ± 13.8 years in 55 patients with AST in a multicenter retrospective case–control study. Our patient No. 2 also belongs to this age group, as she was diagnosed at the age of 30 years. The average age for SM diagnoses tends to be higher, with a mean age of 55 years (range 8–90 years) reported by Lott et al. [14] in a 54-cases study. Our adult patients with SM were within this age range as well.

A Study by Bartenstein et al. [6], which included only pediatric patients, demonstrated a median age of 7.4 and 7.2 for SN and AST, respectively, the age group being from three months to 19.7 years, which correlates with the age of our patient No. 1. Carrera et al. [7] reported a mean age of 12.5 years (range 2–20) for SM based on a sample of 15 patients collected in a multicentric retrospective study, as SM is relatively rare in the pediatric population. In the case of our patient No. 5, although by age she was at the lower end of this range, timely excision led to a better prognosis.

In a recent single center 10-year retrospective study comprising 250 spitzoid lesions in pediatric population, conducted by Herzum et al. [15], literature data was confirmed. The results showed that 82% of spitzoid lesions in pediatric patients were benign, though a not negligible

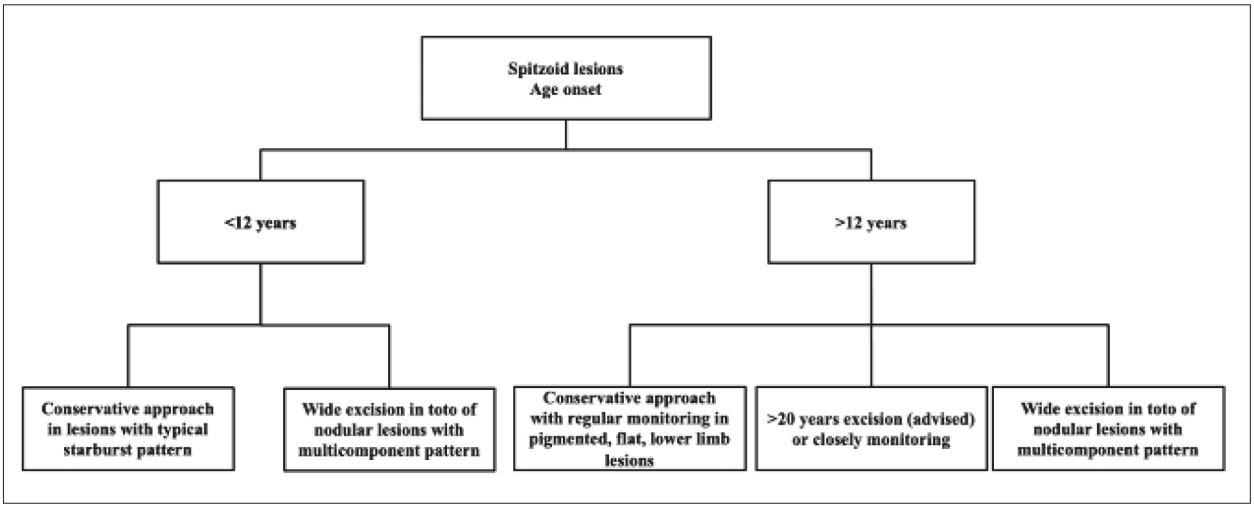

Figure 8. Proposed protocol for the management of spitzoid lesions

percentage of AST (17%) were retrieved, as well as 1% of SM [15]. These findings highlight the need for extreme caution when dealing with spitzoid lesions in childhood, as demonstrated in the case of our patient No. 5.

To date, no specific guidelines or standardized criteria exist for the management of spitzoid lesions, and due to complexity of their clinical, dermoscopic and histological differentiation, they are highly difficult to manage. Numerous guidelines have been developed by several dermatology societies, to help clinicians handle this problem [9, 16, 17]. The majority suggested approach to minimize unnecessary excisions, without raising the risk of overlooking concerning lesions [13, 17]. Based on the suggested algorithms, depending on the patients age, symmetrical flat lesions can be monitored until stabilization, symmetrical nodular lesions should be excised or scheduled for a close follow-up, while asymmetric Spitzoid tumors should always be excised [17].

Data from a newer study by Herzum et al. [15] also suggest that pediatric patients older than 12 years with clinically pigmented, flat, and lower-limb lesions had benign lesions in 97%, 98%, and 89%, respectively, and the simultaneous presence of all three criteria was always associated with benign histology (100%). This could thus probably represent a criterion for conservative management in pediatric patients older than 12 years [15].

Most recent studies also show that nodular spitzoid lesions at any age with dermoscopical multicomponent pattern should raise intense suspicion being highly associated with AST/SM and demand highest attentiveness [7, 15].

Newer studies also confirm that Spitzoid lesions in adult patients should always be closely monitored or excised as they tend to have a greater likelihood of malignancy [17].

After reviewing the most recent literature on this subject, as well as based on our own professional experience, we strongly advise outmost caution when dealing with a patient with spitzoid lesion, especially in those with nodular lesions and older than 12 years; in that light we developed our own protocol in managing spitzoid lesions (Figure 8).

With the following case series, we report clinical, as well as dermoscopic features of biologically various spitzoid lesions, appearing in different age groups. We believe that this case series may increase knowledge of physicians, pediatricians, and dermatologists about when to react when dealing with a patient with a Spitzoid lesion.

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Conflict of interest: None declared.

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Клинички и дермоскопски спектар спицоидних лезија у односу на узраст пацијента – када реаговати?

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САЖЕТАК

Увод Спицоидне лезије представљају спектар меланоцитних лезија које обухватају бенигне спиц невусе, интермедијарне лезије познате као атипични спиц тумори и спицоидни меланом. Иако чешћи код деце и у млађој популацији, могу се јавити у било којој узрасној групи. Због сложености њихове клиничке, дермоскопске и хистопатолошке слике, изузетно су тешки за лечење, поготово у педијатријској популацији.

Приказ болесникâ У овом приказу представљамо серију од шест болесникâ са спицоидним лезијама у различитим старосним групама и са различитим исходима.

Закључак У раду приказујемо клиничке и дермоскопске карактеристике биолошки различитих спицоидних лезија, које се јављају у различитим старосним групама. Верујемо да ће овај чланак повећати знање како дерматолога, тако и других лекара о томе када и како реаговати када се ради о пацијенту са спицоидном лезијом на кожи.

Кључне речи: спицоидне лезије; спиц невус; атипични спиц тумор; спицоидни меланом; лечење