# Update on dermoscopy of Spitz/Reed naevi and management guidelines by the International Dermoscopy Society

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# Summary

Spitzoid lesions represent a challenging and controversial group of tumours, in terms of clinical recognition, biological behaviour and management strategies. Although Spitz naevi are considered benign tumours, their clinical and dermoscopic morphological overlap with spitzoid melanoma renders the management of spitzoid lesions particularly difficult. The controversy deepens because of the existence of tumours that cannot be safely histopathologically diagnosed as naevi or melanomas (atypical Spitz tumours). The dual objective of the present study was to provide an updated classification on dermoscopy of Spitz naevi, and management recommendations of spitzoid-looking lesions based on a consensus among experts in the field. After a detailed search of the literature for eligible studies, a data synthesis was performed from 15 studies on dermoscopy of Spitz naevi. Dermoscopically, Spitz naevi are typified by three main patterns: starburst pattern (51%), a pattern of regularly distributed dotted vessels (19%) and globular pattern with reticular depigmentation (17%). A consensus-based algorithm for the management of spitzoid lesions is proposed. According to it, dermoscopically asymmetric lesions with spitzoid features (both flat/raised and nodular) should be excised to rule out melanoma. Dermoscopically symmetric spitzoid nodules should also be excised or closely monitored, irrespective of age, to rule out atypical Spitz tumours. Dermoscopically symmetric, flat spitzoid lesions should be managed according to the age of the patient. Finally, the histopathological diagnosis of atypical Spitz tumour should warrant wide excision but not a sentinel lymph-node biopsy.

## What's already known about this topic?

- Spitzoid lesions represent a challenging and controversial group of tumours, in terms of clinical recognition, biological behaviour and management strategies.
- Dermoscopy improves the clinical recognition of Spitz naevi, but a morphological overlap between Spitz naevi and spitzoid melanoma does exist.
- Histopathologically, some tumours cannot be safely diagnosed either as Spitz naevi or as melanomas.
- These controversies result in a significant inconsistency of management of spitzoid tumours among clinicians.

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#### What does this study add?

- Three dermoscopic patterns may be considered as suggestive of a Spitz naevus: starburst pattern, regularly distributed dotted vessels and globular pattern with reticular depigmentation.
- Dermoscopically asymmetric lesions with spitzoid features (both flat/raised and nodular) should be excised to rule out melanoma.
- Dermoscopically symmetric spitzoid nodules should also be excised or closely monitored, irrespective of the age, to rule out atypical Spitz tumours.
- Dermoscopically symmetric flat spitzoid lesions should be managed according to the age of the patient.

Since its first description by Sophie Spitz, the morphological and biological spectrum of Spitz naevus has been extensively investigated.<sup>1,2</sup> Although initially described as 'juvenile melanoma', the benign nature of Spitz naevus was very soon understood. For several decades, this tumour was considered to be completely benign and cases of metastatic disease were assumed to represent incorrectly diagnosed melanomas mimicking a Spitz naevus.<sup>3</sup>

However, in the late 1990s, cases of spitzoid lesions without malignant histopathological criteria but with the potential of nodal metastasis were described.<sup>4</sup> This observation initiated a new era of controversies on the true biology of spitzoid neoplasms, characterized by the introduction of several terms attempting to classify spitzoid tumours with intermediate histopathological features between Spitz naevus and spitzoid melanoma (Spitz naevus with atypia and metastasis, metastasizing Spitz tumour, atypical Spitz tumour, atypical Spitz naevus, melanocytic tumour of unknown malignant potential, melanocytoma).<sup>5–8</sup> The controversies on terminology, classification and management strategies of these tumours continue to the present day.

The introduction of dermoscopy significantly improved the clinical recognition of Spitz naevi, as they were shown to exhibit a peculiar and characteristic pattern of dermoscopic structures.<sup>9</sup> Pigmented variants were first investigated and shown to display the so-called 'starburst' pattern, consisting of a central area of homogeneous black-blue pigmentation and symmetrically distributed peripheral streaks or pseudopods.<sup>9–11</sup> Several additional patterns were later found to be associated with pigmented Spitz naevus, including globular, homogeneous, reticular and multicomponent pattern.<sup>12</sup>

Another criterion initially reported to characterize melanoma but later shown to be rather predictive of Spitz naevus was the so-called negative pigment network or reticular depigmentation, consisting of white intermingled lines surrounding pigmented globules.<sup>13</sup>

Studies on the dermoscopic morphology of nonpigmented Spitz naevi were subsequently conducted and suggested that these tumours more frequently display dotted vessels in a regular distribution.<sup>14,15</sup> The aforementioned negative pigment network was found to be present also in nonpigmented Spitz naevi, with the only difference being that the white lines surround vessels instead of pigmented globules.<sup>13,16</sup>

Several infrequent dermoscopic patterns have been also reported in Spitz naevi and suggested to correspond to peculiar histopathological variants such as angiomatoid or desmoplastic Spitz naevus.<sup>17</sup>

Although dermoscopy provided further insights into the morphology of Spitz naevus, management of spitzoid lesions remains controversial. The main source of controversy is the fact that spitzoid melanoma might perfectly mimic a Spitz naevus, displaying one of the aforementioned dermoscopic patterns.<sup>18</sup> This has been highlighted by studies reporting a high variability in clinicians' beliefs and behaviour.<sup>19</sup> Two clinically relevant questions remain to be addressed: how to manage a spitzoid-looking lesion and how to manage a tumour histopathologically diagnosed as 'atypical spitzoid naevus or tumour'.

Our study had three aims: (i) to provide an updated dermoscopic classification of Spitz naevi based on a detailed search and review of the literature; (ii) to provide recommendations on the management of spitzoid lesions according to their clinical and dermoscopic morphology; and (iii) to provide recommendations on the management of histopathologically ambiguous spitzoid tumours.

### Materials and methods

#### Search strategy

To identify eligible studies, the main search was conducted in the electronic databases MEDLINE, Embase, and Cochrane Central Register of Controlled Trials (CENTRAL), using any one of the terms 'dermoscopy' (MeSH), 'dermatoscopy' (MeSH), 'epiluminescence microscopy' (MeSH), 'videodermoscopy' (MeSH) and any one of the terms 'Spitz nevus' (MeSH), 'Spitzoid melanoma' (MeSH), 'atypical Spitz' (MeSH), 'spitz\*', 'atypical melanoma' (MeSH), 'Nevus, Spindle Cell and Epithelioid' (MeSH), 'atypical epithelioid melanocytic proliferation', 'melanocytic neoplasm', 'pigmented epithelioid melanocytoma' and 'melanocytic tumour of uncertain malignant potential', without language restriction. The manual search was concluded by perusal of the reference sections of all relevant trials or reviews and contact with experts on the subject in an effort to identify relevant unpublished data. Studies describing patients with 'spitzoid melanoma' were thoroughly reviewed to avoid missing cases of Spitz naevi.

The main search and the screening of titles and abstracts were completed independently by two reviewers (A.K. and A.L.) with expertise in conducting systematic reviews (Fig. 1).

Studies of patients with Spitz naevi that met the following two criteria were included in the analysis: (i) the diagnosis of Spitz naevus was histopathologically confirmed and (ii) adequate information on the dermoscopic criteria was provided either in the text or in tables. Case reports or case series aiming to describe uncommon or peculiar dermoscopic findings of Spitz naevi were excluded.

The primary outcome was the dermoscopic criteria of the lesions in the included studies. Other variables recorded were patients' age and sex.

In this manuscript, the 'metaphorical' dermoscopic terminology was used. The respective equivalents of these metaphorical dermoscopic terms in descriptive terminology, as well as their definitions, are provided in Table 1.

A web-based consensus was performed among the authors and other experts in the field. Available literature data were insufficient to provide evidence-based recommendations. Although all available evidence was taken into consideration, the recommendations provided in this manuscript should rather be considered as expert-consensus guidelines.

#### **Results**

As shown in Table 2, 15 eligible studies were finally included in the data synthesis. The 15 eligible studies included six multicentre case–control studies, three single-centre case–control studies, one multicentre case series study and five single-centre case series studies.  $^{10,11,13,14,16,18,20-28}$ 

Overall, 896 patients with Spitz naevi were included in our analysis. Mean age was 27.7 years and the male-to-female ratio was 1 : 1.65. However, it should be mentioned that the largest case–control study included patients aged > 12 years.

Table 3 shows the results of the dermoscopic analysis. The starburst pattern was the most frequently observed dermoscopic pattern of Spitz naevi (453 of 896 cases, 50.6%), followed by the pattern of dotted vessels (173 of 896, 19.3%) and the globular pattern (152 of 896, 17%). Finally, a multicomponent/atypical pattern characterized 81 of 896 Spitz naevi (9.0%). No significant correlation was found between age or sex and the dermoscopic naevus pattern.

#### Discussion

Our results confirm that Spitz naevi display characteristic dermoscopic patterns that facilitate their recognition. Based on the findings of our review of the literature, we propose an updated dermoscopic classification of Spitz naevi.

From a morphological point of view, a 'spitzoid' pattern by definition presupposes a symmetric arrangement of colours and structures. In contrast, the same dermoscopic structures, when nonsymmetrically distributed, represent melanoma-specific criteria. For instance, asymmetrically distributed peripheral streaks, pseudopods or globules are considered features suggestive of melanoma, and are often combined, forming the so-called 'multicomponent pattern'.<sup>29</sup> Although the latter pattern is generally suggestive of melanoma, our results highlight that it can be also seen in a proportion of Spitz naevi. However, a Spitz naevus exhibiting asymmetrically distributed features is impossible to differentiate from melanoma and, accordingly, we strongly suggest clinicians excise such lesions irrespective of age or clinical morphology.



Fig 1. The starburst pattern of pigmented Spitz (Reed) naevi. The pattern consists of a hyperpigmented centre and symmetrically distributed peripheral pseudopods (a, b) or streaks (c) or a combination of pseudopods and streaks (d).

Metaphorical term		
used in the text	Equivalent in descriptive terminology	Definition
Starburst pattern	Pattern of circumferential radial lines/ pseudopods, with central structureless area	Pigmented streaks or pseudopods in a radial arrangement at the periphery of the lesion, combined with a central area of
		homogeneous black-blue pigmentation
Globular pattern	Pattern of clods	Numerous, variously sized, round to oval structures with various shades of brown and grey-black
Homogeneous pattern	Structureless pattern	Diffuse, brown, grey-blue to grey-black pigmentation in the absence of other distinctive local features
Reticular pattern	Pattern of reticular lines	Pigment network covering most parts of the lesion
Multicomponent pattern	Combined patterns	A pattern composed of more than one of the patterns described above
Streaks	Peripheral radial lines	Finger-like projections seen at the edge of a lesion
Pseudopods	Pseudopods	Finger-like projections with a bulbous ending seen at the edge of a lesion
Reticular depigmentation	Hypopigmented reticular lines	A network of hypopigmented/white lines surrounding either globules of vascular structures
Dotted vessels	Dotted vessels	Red dots
Glomerular vessels	Coiled vessels	Tortuous capillaries (variation of dotted vessels)
Hairpin vessels	Looped vessels	Vascular loops sometimes twisted and bending
Linear irregular vessels	Linear irregular vessels	Linear and irregularly shaped and sized vascular structures

Table 1 Metaphorical and descriptive dermoscopic terms used in the text and their definitions

Table 2 Studies included in the data synthesis

Study	No. of cases	Pigmented	Nonpigmented
Argenziano 1999 <sup>10</sup>	36	36	0
Pellacani 2000 <sup>20</sup>	26	17	9
Rubegni 2001 <sup>21</sup>	43	43	0
Argenziano 2001 <sup>22</sup>	57	57	0
Argenziano 2004 <sup>14</sup>	18	0	18
Ferrara 2005 <sup>11</sup>	69	58	11
Nino 2009 <sup>23</sup>	8	8	0
Pellacani 2009 <sup>24</sup>	40	40	0
Argenziano 2011 <sup>25</sup>	64	25	39
Botella-Estrada 2012 <sup>28</sup>	9	0	9
Pizzichetta 2013 <sup>16</sup>	40	N/A	N/A
Zalaudek 2013 <sup>13</sup>	26	0	26
Lallas 2015 <sup>18</sup>	333	286	47
Moscarella 2015 <sup>17</sup>	110	78	22
Guida 2016 <sup>27</sup>	17	15	2
Total	896	713	183

Table 3 Dermoscopic criteria of Spitz naevi from 15 studies included in the data synthesis (N = 896)

Dermoscopic pattern	Frequency, n (%)	
Global pattern		
Starburst	453 (50.6)	
Globular	152 (17.0)	
Multicomponent/atypical	81 (9.0)	
Homogeneous	15 (1.7)	
Reticular	22 (2.5)	
Dotted vessels <sup>a</sup>	173 (19.3)	
Additional features		
Reticular depigmentation	158 (17.6)	
Superficial black network	21 (2.3)	
Blue-white veil	14 (1.6)	

<sup>a</sup>Several authors mention that in elevated nodular nonpigmented Spitz naevi, the vessels are not precisely dotted but may project as red globules, coiled or tortuous vessels.

In fact, the starburst pattern has been suggested to typify Reed naevus, while the globular pattern is associated with pigmented Spitz naevi.<sup>31</sup> Whether Reed and pigmented Spitz naevi represent different tumours has not been fully elucidated to date. However, their morphological and epidemiological characteristics rather support the theory that they represent distinct entities within the same spectrum.

Spitz naevi with a different dermoscopic aspect do exist (homogeneous, reticular, multicomponent). The homogeneous and reticular patterns have been suggested to represent later evolution phases of the starburst pattern. In any case, the latter patterns cannot be considered as specific for spitzoid

For dermoscopic patterns of pigmented Spitz (Reed) naevus, our results show that the vast majority are dermoscopically typified either by a starburst or by a globular pattern, the latter usually associated with reticular depigmentation (Figs 1 and 2). It must be mentioned that a globular pattern not associated with reticular depigmentation cannot be considered as specific for Spitz naevus, as it is well known that the majority of naevi, especially in children, display a pattern of globules.<sup>30</sup>



**Fig 2.** Pigmented Spitz naevi typified by a globular pattern combined with reticular depigmentation (inverse network).

tumours, as they can be found much more frequently in other naevus types, while the multicomponent pattern, as discussed above, should be considered as suggestive of melanoma. Therefore, two dermoscopic patterns can be considered highly suggestive of pigmented Spitz naevus: the starburst pattern (Reed naevus) and the globular pattern with reticular depigmentation (pigmented Spitz naevus).

With regard to nonpigmented Spitz naevus, regularly distributed dotted vessels represent the dermoscopic hallmark. Reticular depigmentation, namely white crossing lines surrounding the vessels, represents a frequent additional feature. Although underreported in the literature, clinicians should expect that, in raised and nodular Spitz naevi, the vessels might not project as small dots but as larger red globules, coiled vessels or even hairpin or corkscrew vessels (Figs 3 and 4). However, their distribution should always be symmetric all over the lesion. If the vascular structures are asymmetrically distributed, as discussed above, the dermoscopic pattern should no longer be considered as spitzoid but instead as suggestive of melanoma.

With respect to ASTs, a recent study investigated dermoscopic findings in a series of 55 ASTs compared with 110 Spitz naevi.<sup>26</sup> According to the results, the vast majority of ASTs are dermoscopically typified by a multicomponent pattern and would thus be clinically interpreted as suspicious for melanoma. However, approximately 17% of ASTs were nodules displaying a typical nonpigmented Spitzoid pattern, namely regularly distributed dotted vessels with or without reticular depigmentation (Fig. 5). The latter result highlights a clinically relevant morphological overlap between Spitz naevi and ASTs, which is further discussed below.

For spitzoid melanoma, the dermoscopic morphology usually follows the basic rule of asymmetry: it is typically characterized by an asymmetric distribution of spitzoid features



Fig 3. The typical dermoscopic pattern of nonpigmented Spitz naevi consists of regularly distributed dotted vessels, usually associated with an inverse white network (a). In elevated or nodular naevi, the vessels might project as coiled (b), hairpin (c) or linear irregular (d). The pattern should be interpreted as spitzoid only if the vessels are distributed regularly and are surrounded by whitish halos or lines (inverse network).

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(streaks, globules, vascular structures), often combined among them to form the so-called multicomponent pattern. However, less frequently, melanoma might perfectly mimic a pigmented or nonpigmented Spitz naevus (Fig. 6). This was demonstrated by a recent study evaluating 384 dermoscopically symmetric spitzoid-looking lesions in patients greater than 12 years of age.<sup>18</sup> Approximately 13% of these tumours were proven to be melanomas, with the probability of melanoma increasing significantly with age. For example, a perfectly symmetric spitzoid-looking lesion developing in an individual after the age of 50 years is associated with a 50% probability of being melanoma, whereas this probability decreases to 7% for patients less than 30 years of age. Nodular spitzoid lesions are associated with a higher possibility of melanoma (32%), but also the risk of flat lesions remains considerable (12%). In the light of the latter significant morphological overlap between Spitz naevus and melanoma, the management of spitzoid tumours should be adjusted accordingly, as discussed below.

For the management of spitzoid-looking lesions, taking into account the aforementioned morphological overlap among Spitz naevus, ASTs and spitzoid melanoma, we recommend two possible strategies. These recommendations are based on the notion that the dual goal of our clinical practice is to minimize the risk of missing melanoma and avoid, as much as possible, unnecessary excisions of benign moles.<sup>32</sup> The safest strategy would be to excise any lesion with spitzoid features (symmetrically or asymmetrically distributed) irrespectively of the age of the patient. Obviously, this strategy is associated with no risk of missing melanoma, but will also result in a relatively high number of excisions of benign moles.

Given that the majority of Spitz naevi develop in children and therefore their excision would not be so easy and might require general anaesthesia, rather, a more flexible, risk-associated, age-dependent strategy is recommended. The proposed algorithm is presented in Figure 7 and includes the following three sequential steps.

- 1 Overall dermoscopic symmetry should be assessed. Lesions displaying spitzoid features (peripheral streaks/ pseudopods, dotted vessels, reticular depigmentation) asymmetrically distributed should be excised to rule out melanoma. All the aforementioned features, when not symmetrically distributed, should be considered as criteria suggestive of melanoma.
- 2 Dermoscopically symmetric spitzoid-looking lesions developing after the age of 12 years should also be managed with caution because, as mentioned above, there is a considerable probability that these lesions form a melanoma.<sup>18</sup> The recommended management is excision. An acceptable alternative option is close digital monitoring until stabilization. In case monitoring is selected, the recommended plan is as follows.
  - (a) For nodular lesions, conduct follow-up visits with dermoscopic documentation every 15 days. Detection of growth between two sequential visits should warrant excision. If the naevus is not growing after two visits, the interval of follow-up visits might be prolonged to 4 and later 8 weeks. The follow-up might be discontinued when there is documented evidence of no growth for at least 6 months. If the naevus undergoes involution, follow-up should continue until stabilization (documented evidence of no change for at least 6 months) or complete disappearance.
  - (b) For flat/raised lesions, conduct follow-up visits with dermoscopic documentation every 2–3 months. Lesions displaying a starburst pattern are expected to grow, reach stabilization and, very probably, enter an involution phase. Ideally, the naevus will grow symmetrically to all directions and gradually acquire a homogeneous aspect of blue-black colour (Fig. 8). Subsequently, the darkly pigmented area will be gradually restricted to the centre of the lesion, while the



Fig 5. The majority of atypical Spitz tumours dermoscopically exhibit an atypical/ multicomponent pattern suggestive of melanoma (a). However, approximately 20% of them might dermoscopically mimic a nonpigmented Spitz naevus, showing dotted/glomerular vessels and white inverse network (b).



**Fig 6.** Dermoscopy of spitzoid melanoma typically reveals an atypical/multicomponent pattern with evident presence of melanomaspecific criteria (a, b). However, rarely, melanoma might perfectly mimic a pigmented (c) or nonpigmented (d) Spitz naevus.

peripheral part of the naevus will exhibit regular network. At this stage, the naevus might look like the socalled black naevus found in dark skin type individuals. Finally, an involution process is very likely to begin and more than 50% of naevi will disappear completely.<sup>25</sup> Unfortunately, several 'starburst' naevi do not follow the aforementioned ideal symmetric morphological evolution (Fig. 9). In such cases, excision is highly recommended. Monitoring should continue until stabilization, defined as documented evidence of no growth for at least 6 months. If the naevus undergoes involution, follow-up should continue until stabilization (documented evidence of no change for at least 6 months) or complete disappearance. Lesions displaying a globular pattern or a pattern of dotted vessels should be monitored until stabilization, defined as documented evidence of no growth for at least 6 months. Detection of asymmetric growth should warrant excision. If the naevus undergoes involution, follow-up should continue until stabilization or complete disappearance.

- 3 Below the age of 12 years, the recommended management of dermoscopically symmetric spitzoid-looking lesions is as follows:
  - (a) Nodular lesions: The recommended management is excision, mainly because the possibility of AST cannot be excluded on the basis of dermoscopic morphology. An acceptable alternative option is close digital monitoring until stabilization. In case that monitoring is selected, the recommended plan is identical as in 2(a).
  - (b) Flat/raised lesions: The recommended management is follow-up until stabilization. Monitoring is highly recommended for lesions displaying a starburst pattern (Reed naevi). The morphological evolution of



Fig 7. A proposed algorithm for management of spitzoid-looking lesions.

Spitz naevi displaying a pattern of globules or dotted vessels is less elucidated and monitoring them might induce anxiety in clinicians and the parents of the patient. Effectively, clinicians should take into account the overall clinical context (anatomic site, family environment, etc.) and select either to monitor or excise the tumour. In any case, they should keep in mind that the possibility of melanoma in a flat/raised dermoscopically symmetric spitzoid-looking lesion below the age of 12 is extremely low. In case that monitoring is selected, the recommended plan is identical as in 2(b).

Interestingly, a survey among paediatric dermatologists highlighted that clinicians were more prone to follow up nodular nonpigmented spitzoid lesions compared with pigmented ones, which were more frequently excised.<sup>19</sup> However, recent data on the morphology of AST showed that it might be clinically and dermoscopically indistinguishable from a nonpigmented Spitz naevus, namely, a nodule with dotted vessels with or without reticular depigmentation.<sup>26</sup> In contrast, flat/ raised pigmented lesions dermoscopically characterized by a starburst or a globular pattern were almost always naevi. Effectively, we recommend a lower threshold for excision of nonpigmented nodules, while flat/raised lesions might enter follow up in children aged below 12 years.

For histopathologically ambiguous spitzoid tumours, the spectrum of spitzoid lesions ranges between Spitz naevus, which represents a benign tumour with no difference in prognosis compared with other types of naevi, and spitzoid melanoma. Subsequently, in the hypothetical scenario that all Spitz naevi and all spitzoid melanomas could be accurately diagnosed as such, the optimal management of naevi would



**Fig 8.** Monitoring of a flat pigmented Spitz naevus typified by a starburst pattern (a). The naevus grows symmetrically in all directions (b) and finally reaches stabilization, which is dermoscopically recognized by the loss of peripheral pseudopods (c).



Fig 9. Monitoring of a flat pigmented Spitz naevus typified by a starburst pattern (a). This naevus grew asymmetrically, acquiring an atypical pattern (b). When finally stabilized it acquired a pattern consisting of central hyperpigmentation and peripheral network (c). However, the interpretation of the morphological evolution in the intermediate stage was highly equivocal. Lesions evolving like this should be excised.

require no action, whereas spitzoid melanoma should be managed as any other melanoma type.

The controversy arises from the fact that between these two extremes are tumours that cannot be safely diagnosed histopathologically either as naevi or as melanomas. This is because their histopathological alterations extend beyond the expected atypia of Spitz naevi but without fulfilling the criteria of melanoma. Among the several terms that histopathologists have used to classify these tumours, 'atypical Spitz tumour' (AST) has been more globally adopted.33 The fact that AST represents an intermediate entity in terms of histopathological morphology does not necessarily equate to an intermediate biological course. Sentinel lymph node biopsy (SLNB) has been suggested as a possible solution to this diagnostic problem, based on the assumption that detection of melanocytic cells in the SLN could predict aggressive biological behaviour.<sup>34</sup> However, recent meta-analytic data including all published cases worldwide demonstrated that AST is associated with a highly favourable prognosis, even in cases of SLNB positivity.<sup>35</sup> Accordingly, evidence suggests that the optimal management of AST is wide surgical excision, while SLNB should be considered inappropriate. Although data suggesting optimal margins for wide local excision are lacking, the most frequently used margins are 1 cm, which is also our suggestion. Clinical follow-up with superficial node palpation is recommended at least once per year for 3 years. Work-up with ultrasound is required in case of palpable nodes or doubt.

Limitations of this review are that the studies included in the data synthesis were heterogeneous concerning their aims, design, dermoscopic criteria used in the evaluation, and reporting of results. To minimize this problem, authors of several studies were contacted to provide information on the raw data of their studies.

In addition, readers should take into account that clinicians often choose not to excise Spitz naevi, but instead monitor them until stabilization. In the present study, Spitz naevi lacking a histopathological diagnosis were excluded. Accordingly, the percentages of naevi displaying one of the 'symmetric' patterns (starburst, globular or dotted vessels) might be underestimated in this study, whereas the percentage of Spitz naevi with atypical/multicomponent dermoscopic pattern might be lower than reported herein.

Furthermore, although only cases with a definite histopathological diagnosis were included in this study, it must be mentioned that the accurate histopathological characterization of a spitzoid lesion might be highly challenging. This means that we cannot exclude the possibility that some of the tumours included in the studies enrolled in our review had been histopathologically incorrectly classified.

Finally, as explained above, available evidence was insufficient to allow us to provide evidence-based guidelines. Therefore, readers should consider the provided guidelines as expert consensus-based recommendations.

In conclusion, spitzoid lesions are dermoscopically typified by three main patterns: starburst pattern, a pattern of regularly distributed dotted vessels and globular pattern with reticular depigmentation. They represent a fascinating and challenging group of tumours. Dermoscopically asymmetric lesions with spitzoid features should be excised to rule out melanoma. The management of dermoscopically symmetric spitzoid lesions requires updated knowledge and integration of clinical and dermoscopic information.

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#### References

- 1 Spitz S. Melanomas of childhood. Am J Pathol 1948; 24:591-609.
- 2 Luo S, Sepehr A, Tsao H. Spitz nevi and other Spitzoid lesions part II. Natural history and management. J Am Acad Dermatol 2011; 65:1087–92.
- 3 Kernen JA, Ackerman LV. Spindle cell nevi and epithelioid cell nevi (so-called juvenile melanomas) in children and adults: a clinicopathological study of 27 cases. *Cancer* 1960; **13**:612–25.
- 4 Smith KJ, Barrett TL, Skelton HG et al. Spindle cell and epithelioid cell nevi with atypia and metastasis (malignant Spitz nevus). Am J Surg Pathol 1989; **13**:931–9.
- 5 Barnhill RL, Flotte TJ, Fleischli M, Perez-Atayde A. Cutaneous melanoma and atypical Spitz tumors in childhood. Cancer 1995; 76:1833–45.
- 6 Barnhill RL, Argenyi ZB, From L et al. Atypical Spitz nevi/tumors: lack of consensus for diagnosis, discrimination from melanoma, and prediction of outcome. Hum Pathol 1999; **30**:513–20.
- 7 Elder DE, Xu X. The approach to the patient with a difficult melanocytic lesion. Pathology 2004; 36:428–34.

- 8 Cerroni L, Barnhill R, Elder D et al. Melanocytic tumors of uncertain malignant potential: results of a tutorial held at the XXIX Symposium of the International Society of Dermatopathology in Graz, October 2008. Am J Surg Pathol 2010; 34:314–26.
- 9 Steiner A, Pehamberger H, Binder M, Wolff K. Pigmented Spitz nevi: improvement of the diagnostic accuracy by epiluminescence microscopy. J Am Acad Dermatol 1992; 27:697–701.
- 10 Argenziano G, Scalvenzi M, Staibano S et al. Dermatoscopic pitfalls in differentiating pigmented Spitz naevi from cutaneous melanomas. Br J Dermatol 1999; 141:788–93.
- 11 Ferrara G, Argenziano G, Soyer HP et al. The spectrum of Spitz nevi: a clinicopathologic study of 83 cases. Arch Dermatol 2005; 141:1381-7.
- 12 Peris K, Ferrari A, Argenziano G et al. Dermoscopic classification of Spitz/Reed nevi. Clin Dermatol 2002; 20:259–62.
- 13 Zalaudek I, Kittler H, Hofmann-Wellenhof R et al. "White" network in Spitz nevi and early melanomas lacking significant pigmentation. J Am Acad Dermatol 2013; 69:56–60.
- 14 Argenziano G, Zalaudek I, Corona R et al. Vascular structures in skin tumors: a dermoscopy study. Arch Dermatol 2004; 140:1485–9.
- 15 Zalaudek I, Kreusch J, Giacomel J et al. How to diagnose nonpigmented skin tumors: a review of vascular structures seen with dermoscopy: part II. Nonmelanocytic skin tumors. J Am Acad Dermatol 2010; 63:377–86.
- 16 Pizzichetta MA, Talamini R, Marghoob AA et al. Negative pigment network: an additional dermoscopic feature for the diagnosis of melanoma. J Am Acad Dermatol 2013; 68:552–9.
- 17 Moscarella E, Al Jalbout S, Piana S et al. The stars within the melanocytic garden: unusual variants of Spitz naevi. Br J Dermatol 2015; 172:1045–51.
- 18 Lallas A, Moscarella E, Longo C et al. Likelihood of finding melanoma when removing a Spitzoid-looking lesion in patients aged 12 years or older. J Am Acad Dermatol 2015; 72:47–53.
- 19 Tlougan BE, Orlow SJ, Schaffer JV. Spitz nevi: beliefs, behaviors, and experiences of pediatric dermatologists. JAMA Dermatol 2013; 149:283–91.
- 20 Pellacani G, Cesinaro AM, Seidenari S. Morphological features of Spitz naevus as observed by digital videomicroscopy. Acta Derm Venereol 2000; 80:117–21.
- 21 Rubegni P, Ferrari A, Cevenini G et al. Differentiation between pigmented Spitz naevus and melanoma by digital dermoscopy and stepwise logistic discriminant analysis. *Melanoma Res* 2001; 11:37– 44.
- 22 Argenziano G, Soyer HP, Ferrara G et al. Superficial black network: an additional dermoscopic clue for the diagnosis of pigmented spindle and/or epithelioid cell nevus. Dermatology 2001; 203:333– 5.
- 23 Nino M, Brunetti B, Delfino S et al. Spitz nevus: follow-up study of 8 cases of childhood starburst type and proposal for management. Dermatology 2009; 218:48–51.
- 24 Pellacani G, Longo C, Ferrara G et al. Spitz nevi: In vivo confocal microscopic features, dermatoscopic aspects, histopathologic correlates, and diagnostic significance. J Am Acad Dermatol 2009; 60:236– 47.
- 25 Argenziano G, Agozzino M, Bonifazi E et al. Natural evolution of Spitz nevi. Dermatology 2011; 222:256–60.
- 26 Moscarella E, Lallas A, Kyrgidis A et al. Clinical and dermoscopic features of atypical Spitz tumors: a multicenter, retrospective, casecontrol study. J Am Acad Dermatol 2015; 73:777–84.
- 27 Guida S, Pellacani G, Cesinaro AM et al. Spitz naevi and melanomas with similar dermoscopic patterns: can confocal microscopy differentiate? Br J Dermatol 2016; 174:610–16.

- 28 Botella-Estrada R, Requena C, Traves V et al. Chrysalis and negative pigment network in Spitz nevi. Am J Dermatopathol 2012; 34:188– 91.
- 29 Argenziano G, Fabbrocini G, Delfino M. Epiluminescence microscopy. A new approach to in vivo detection of Sarcoptes scabiei. *Arch Dermatol* 1997; **133**:751–3.
- 30 Zalaudek I, Grinschgl S, Argenziano G et al. Age-related prevalence of dermoscopy patterns in acquired melanocytic naevi. Br J Dermatol 2006; 154:299–304.
- 31 Bar M, Tschandl P, Kittler H. Differentiation of Spitz nevi and Reed nevi by integration of dermatopathologic and dermatoscopic findings. Dermatol Pract Concept 2012; 2:13–24.
- 32 Lallas A, Zalaudek I, Apalla Z et al. Management rules to detect melanoma. Dermatology 2013; 226:52–60.
- 33 Ferrara G, Cavicchini S, Corradin MT. Hypopigmented atypical Spitzoid neoplasms (atypical Spitz nevi, atypical Spitz tumors,

Spitzoid melanoma): a clinicopathological update. Dermatol Pract Concept 2015; **5**:45–52.

- 34 Kelley SW, Cockerell CJ. Sentinel lymph node biopsy as an adjunct to management of histologically difficult to diagnose melanocytic lesions: a proposal. J Am Acad Dermatol 2000; **42**:527–30.
- 35 Lallas A, Kyrgidis A, Ferrara G et al. Atypical Spitz tumours and sentinel lymph node biopsy: a systematic review. Lancet Oncol 2014; 15:e178–83.

# **Supporting Information**

Additional Supporting Information may be found in the online version of this article at the publisher's website:

Video S1. Author video.