Pediatric spitzoid lesions of the ear: a single-center experience and review of the literature

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Abstract

Spitzoid lesions are challenging melanocytic lesions comprising benign, intermediate, and malignant lesions. In this study, we aimed to analyze the diagnostic accuracy of clinical and dermatoscopical evaluations of pediatric spitzoid ear lesions. We collected and analyzed, clinically, dermatoscopically, and histologically, pediatric spitzoid ear lesions. We also conducted a systematic review of the literature. At the Pediatric Hospital Gaslini, excision and histopathological evaluation were performed on eight cases: 87.5% of the lesions were consistent with Spitz nevus (SN), and 12.5% with atypical Spitz tumor (AST). Notably, multiple (≥2) dermatoscopical irregularities were present in 5 of 7 SN (71%), yet none were found in AST (0%, 0/1) (Fisher's exact test, P=0.375). From systematic research in the literature, 9 patients were included in this review. At histology, 88.9% were SN and 11% AST. Remarkably, also in the literature, multiple dermatoscopical irregularities were present in most SN (75%, 6/8), but not in the identified AST (0%, 0/1) (P=0.3333). We present a monocentric study on pediatric spitzoid ear lesions. Importantly, dermatoscopical irregularities were not significantly associated with AST, neither in our series nor in the reviewed literature (respectively P=0.375 and P=0.3333), supporting the fact that relying only on the dermatoscopical aspect of spitzoid lesions is not accurate enough for the special site of the ear, where dermatoscopy could actually be misleading.

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Introduction

Spitzoid lesions represent a challenging dermatological entity, as they comprise benign Spitz nevi (SN), malignant melanoma (MM), and melanocytic proliferations with intermediate features between melanoma and nevus, the so-called atypical Spitz tumors (AST).[1-4]

The differential diagnosis of these spitzoid entities is clinically as well as histopathologically challenging, particularly at special body sites such as the ear, mucosae, and acral skin, where microanatomic diversity influences skin morphology.[4] While dermatoscopy was suggested as a diagnostic tool to avoid excision of well-recognized benign lesions, on special body sites such as the ear, clinical-dermatoscopic aspects may not correspond to histological characteristics. Moreover, spitzoid ear lesions are rare and therefore seldom reported in the literature, hindering the development of consensus guidelines on the management and treatment of these lesions.

Herein, we present a case series of pediatric spitzoid ear lesions from a single-center experience. Lesions were analyzed clinically, dermoscopically, and histologically, expanding current knowledge on this rare entity. Also, we conducted a systematic review on spitzoid lesions of the ear in pediatric age to compare known literature data with our monocentric experience and to evaluate the diagnostic accuracy of the clinical-dermatoscopic assessment.

Materials and Methods

We conducted a retrospective study on pediatric spitzoid lesions of the ear presenting at the Dermatology Unit of the Pediatric Hospital IRCSS Giannina Gaslini over the last 7 years (2015-2022). The study was conducted retrospectively following

local guidelines, employing electronic records of patients and histopathological reports to retrieve all relevant data. Written informed consent was obtained for publication of the documentation, including the photographic documentation.

All lesions clinically diagnosed as SN, AST, or MM of the ear were registered and dermoscopically evaluated by two observers. Surgical excision was performed, also in compliance with the International Dermoscopy Society (IDS) guidelines. All lesions were completely excised with an elliptical excision using a #15 scalpel blade and histopathologically analyzed (Table 1).

Also, literature about spitzoid pediatric ear lesions available on Google Scholar and PubMed was systematically reviewed in compliance with the preferred reporting items for systematic reviews and meta analysis.[5]

Combinations of the following keywords were used to retrieve all relevant articles: (((Spitz) OR (spitzoid) OR (Spitz nevus) OR (Spitz tumor) OR (spitzoid melanoma) OR (spitzoid lesion)) AND (ear) AND (pediatric)). We included studies and reviews reporting new cases of spitzoid lesions of the ear in pediatric age (≤18 years), dermatoscopically and clinically evaluated, and histopathologically confirmed (Table 2).[4,6-8]

Results

At the Dermatology Unit of the Pediatric Hospital IRCSS Giannina Gaslini, 8 ear lesions with spitzoid features or possibly representing MM were studied, excised, and histologically analyzed (Figure 1). Patients' features are reported in Table 1.

Patients had a mean and median age of, respectively, 7.4 and 6.5 years, male:female ratio of 1.7:1 and Caucasian ethnicity. The helix was the most frequently interested anatomic site (75%, 6/8), and most spitzoid lesions were clinically raised (87.5%, 7/8), well-circumscribed (88%, 7/8), small-sized (≤6 mm diameter) (75%, 6/8), and smooth-surfaced (75%, 6 /8).

Dermoscopically, 75% (6/8) of the lesions were pigmented. Of note, 75% (6/8) of lesions had at least one sign of dermoscopic irregularity, and 62.5% (5/8) of lesions had 2 or more signs of irregularity. Indeed, irregular peripheral streaks were described in 37.5% (3/8), dark structureless pigment in 50% (4/8), thickened pigmented pseudo-network in 12.5% (1/8), pigmented irregular globules in 25% (2/8), and inverted network in 25% (2/8) of cases. Of lesions without dermoscopic irregularities (25%, 2/8), one was histologically AST.

Excision was performed in all cases for histopathological evaluation (100%, 8/8). At histology, 87.5% (7/8) of lesions were consistent with SN, of which 1/8 was a compound SN, 1/8 was a congenital SN, and 1/8 was an angiomatoid SN. Only one lesion (12.5%, 1/8) was diagnosed as AST. At histology, no lesion was consistent with MM.

Multiple (≥2) dermatoscopical irregularities were present in 5 of 7 SN (71%), yet none were found in AST (0%, 0/1). The presence of multiple dermatoscopic irregularities was not statistically associated with AST more than with SN (Fisher's exact test, P=0.375).

Throughout the systematic research of the literature, a total of 2105 articles were initially retrieved, but, finally, only 4 articles describing 9 patients satisfied the strict inclusion criteria and were selected, as presented in Table 2.[4,6-8] A schematic illustration of the literature search and the study selection criteria is presented in Figure 2.

Also in the literature, the most frequently interested anatomic site was the helix (44.4%, 4/9), spitzoid lesions were mostly raised (66.6%, 6/9), clinically well-circumscribed (100%, 9/9), small-

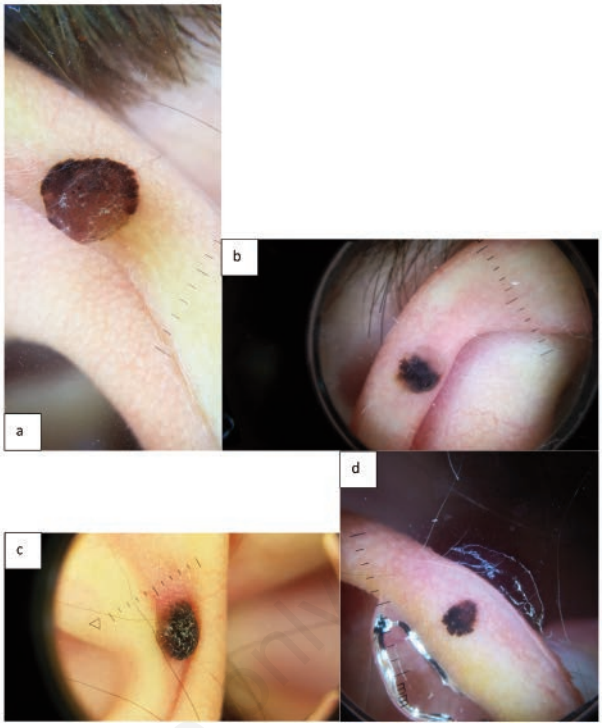

Figure 1. Dermatoscopical images. a) Compound Spitz nevus of the ear helix, with multiple dermatoscopic irregularities; b) Compound Spitz nevus of the helix, with multiple dermatoscopical patterns, thickened pigmented pseudo-network, pigmented streaks, dark structureless pigment, and a blue whitish veil; c) Atypical Spitz tumor, presenting as 4×4 mm pigmented papule of the anthelix, with globular cobblestone pattern, without dermatoscopic irregularities; d) Spitz-Reed nevus of the helix, with multiple dermatoscopic irregularities.

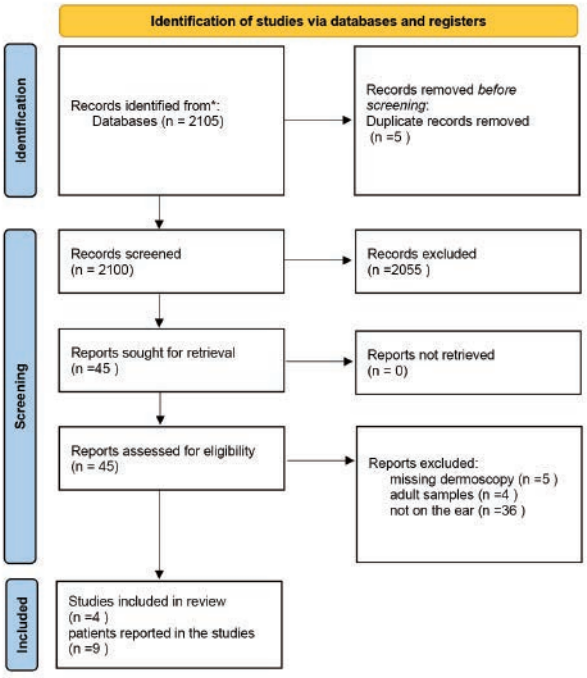

Figure 2. A schematic illustration of the literature search and the study selection criteria is presented in the preferred reporting items for systematic reviews and meta-analysis 2020 flow diagram.

sized (≤6 mm) (88.9%, 8/9), and smooth-surfaced (55.6%, 5/9).

Dermoscopically, 66.6% (6/9) of lesions were pigmented. Overall, 88.9% (8/9) had at least one dermatoscopic irregularity, and 66.6% (6/9) had multiple (≥2) dermatoscopical irregularities. Indeed, irregular peripheral streaks were described in 55.6% (5/9) of the cases, pigment forming a thickened pseudonetwork in 33.3% (3/9), structureless pigment in 22.2% (2/9), irregular globules and blotches in 33.3% (3/9), and a blue-whitish veil in 22.2% (2/9). Atypical vascular patterns were described in 22% (2/9) of the lesions, always combined with small, pigmented areas. Only one lesion showed no particular irregularity (11.1%, 1/9), forming red vascular globules organized in cobblestones. At histology, 88.9% (8/9) of lesions were SN, and 11% (1/9) were diagnosed as AST. No lesion was consistent with MM. Of note, also in the literature, multiple dermatoscopical irregularities were present in most SN (75%, 6/8), but not in the identified AST (0%, 0/1). Dermatoscopic irregularities were not statistically associated with AST more than SN, neither in our series nor in the literature (Fisher's exact test P=0.3333).

Discussion and Conclusions

Management of spitzoid lesions is controversial, and numerous succeeding guidelines have been developed by IDS to help clinicians handle the morphological overlap between SN, AST, and spitzoid melanoma.[9-10] Further to the valuable recommendations of IDS that are developed as a general guideline to follow in all possible settings, the clinician must establish a tailored approach for every patient to ensure the best diagnostic and therapeutic approach. The present series on pediatric spitzoid ear lesions, with almost a decade of data collected monocentrically from consecutive patients, encompasses a large case series, including 9 lesions from this rare entity. Notably, all spitzoid-looking lesions in consecutive patients were excised and histologically examined, reducing the inevitable bias toward adopting conservative management for some benign-looking spitzoid lesions.[11-13] Yet, the present study was conducted in compliance with IDS guidelines: 75% (6/8) of lesions had at least one sign of dermoscopic irregularity justifying surgical excision. Indeed, IDS guidelines suggest the excision of all asymmetric spitzoid lesions, intended as lesions with asymmetrically distributed spitzoid features (dotted vessels, reticular depigmentation, peripheral streaks/pseudopods). In fact, the distinction from melanoma is only histologically feasible in these cases.[9] Of the remaining two lesions analyzed in this study, one was raised and roughly nodular, rationalizing surgical excision. The other one was a raised, eroded, and bleeding papule, and IDS suggested a lower threshold for excision of non-pigmented lesions, supporting the prudential choice of surgical removal.[9,10]

In light of a risk-associated age-dependent strategy and taking into account concerns about anesthesiologic procedures in children, it is important to consider that IDS recommendations explicitly favor flexibility in children.[9] Still, it must be mentioned that pediatric dermatologists are trained to perform surgical excisions in children with high levels of surgical manageability, largely without general anesthesia, also on the head and neck. This difficult-to-treat site should by no means represent a diagnostic limit.

Considerable overlap exists between malignant and benign dermatoscopic features in spitzoid lesions,[14-17] especially in ear lesions, that tend to have clinical and dermatoscopic irregularities due to the anatomic morphology of this special site.[4]

This study evidenced the presence of multiple dermatoscopical irregularities in 71% of SN, which is consistent with literature

Table 1. Features of patients with pediatric spitzoid lesions of the ear presented in the current series.

Patient number	Representative figure	Age (years)	Gender	Ethnicity	Time at presentation since onset (months)	Anatomic site	Size (mm)	Primary lesion	Surface	Clinical aspect	Pigmentation	Color shades	Dermatoscopic pattern	Secondary dermatoscopic aspects	≥1 dermatoscopic irregularities	≥2 dermatoscopic irregularities	Intervention	Diagnosis
1	1a	7	M	Caucasian	8	Helix	6×4	Slightly raised papule	Smooth	Well-circumscribed	Pigmented	Black, dark brown, light brown	Pigmented cobblestones forming an inverted network	Irregular peripheral hyperpigmented streaks	Yes	Yes	Excision	Compound Spitz nevus
2	1b	5	M	Caucasian	6	Helix	4×4	Macule	Smooth	Irregular borders	Pigmented	Black, dark brown, light brown	Pigmented pseudo-network, thickened	Irregular peripheral hyperpigmented streaks, dark structureless pigment, blue-whitish veil, pigmented irregular globules	Yes	Yes	Excision	Compound Spitz nevus
3	1c	4	F	Caucasian	7	Anthelix	4×4	Papule	Smooth	Well-circumscribed	Pigmented	Brown	Pigmented cobblestones	None	No	No	Excision	AST
4	1d	9	F	Caucasian	6	Helix	3×2	Papule	Smooth	Well-circumscribed	Pigmented	Dark brown	Dark structureless pigment	Irregular peripheral hyperpigmented streaks	Yes	Yes	Excision	Spitz Reed nevus
5	NA	6	M	Caucasian	3	Helix	6×4	Papule	Eroded, bleeding	Well-circumscribed	Non-pigmented	Red	Vascular pattern: red homogeneous lacunae	None	No	No	Excision	Dermal Spitz nevus
6	NA	12	F	Caucasian	3	Posterior part of the ear	5×5	Papule	Smooth	Well-circumscribed	Pigmented	Brown, blue	Dark structureless pigment	Blue-whitish veil	Yes	Yes	Excision	Compound spitz nevus
7	NA	4	M	Caucasian	12	Helix	7×7	Papule-nodule	Smooth	Well-circumscribed	Hypo-pigmented	Red, brown	Vascular pattern: dotted vessels	Eccentric dark structureless pigment	Yes	No	Excision	Angiomatoid Spitz nevus
8	NA	12	M	Caucasian	12	Helix	8×8	Papule	Hyperkeratotic	Irregular borders	Pigmented	Brown, dark brown	Globular irregular	Inverted network	Yes	Yes	Excision	Spitz nevus

AST, atypical Spitz tumor; F, female; M, male; NA, not applicable.

Table 2. Features of patients with pediatric spitzoid lesions of the ear reported in the literature.

Author, publication year	Representative figure	N° of patients described	Age (years)	Gender	Ethnicity	Time at presentation since onset (months)	Anatomic site	Size (mm)	Primary lesion	Surface	Clinical aspect	Pigmentation	Color shades	Dermatoscopic pattern	Secondary dermatoscopic aspects	≥1 dermatoscopic irregularities	≥ 2 dermatoscopic irregularities	Intervention	Diagnosis
Ferrara *et al.*, 2015	NA	1	9	M	Caucasian	NA	Anti-tragus from figure	>10 (from figure)	Multinodular	Polypoid	Well-circumscribed	Hypo-pigmented	Red, brown	Atypical vascular pattern	Brown areas	Yes	No	Excision	AST
Liang *et al.*, 2022	NA	1	18	M	Asiatic	12	Anthelix	6×6	Papule	Verrucous	Well-circumscribed	Hypo-pigmented	Red, peripheral brown	Atypical vascular pattern	Peripheral pigmented globules and pseudo network	Yes	Yes	Excision	Compound Spitz nevus
Ikeda *et al.*, 2018	NA	1	2	M	Caucasian	24 (since birth)	Posterior part of the ear	4×6	Small nodule	Papillomatous, verrucous	Well-circumscribed	Non-pigmented	Red, pink	Milky-red cobblestone pattern	Glomerular vessels	No	No	Excision	Congenital Spitz nevus
Vaccaro *et al.*, 2021	NA	6	8	F	NA	NA	Lobule	2×1.4	Macule	Smooth	Well-circumscribed	Pigmented	Black, brown	Thickened pigmented pseudo network	Irregular peripheral streaks	Yes	Yes	Excision	Spitz nevus
Vaccaro *et al.*, 2021	NA		10	M	NA	NA	Helix	3.5×3	Macule	Smooth	Well-circumscribed	Pigmented	Black, brown, grey, white	Thickened pigmented pseudo network	Irregular peripheral streaks, grey-whitish area	Yes	Yes	Excision	Spitz nevus
Vaccaro *et al.*, 2021	NA		16	M	NA	NA	Helix	4×3	Macule	Smooth	Well-circumscribed	Pigmented	Black, brown, grey, white	Thickened pigmented pseudo network	Irregular peripheral streaks, irregular globules	Yes	Yes	Excision	Spitz nevus
Vaccaro *et al.*, 2021	NA		14	M	Caucasian	NA	Helix	2.8×2.4	Papule	Smooth	Well-circumscribed	Pigmented	Black, brown, blue-grey, white	Blue-grey structureless	Irregular peripheral streaks, irregular blotches	Yes	Yes	Excision	Spitz nevus
Vaccaro *et al.*, 2021	NA		9	M	NA	NA	Anthelix	3.8×3.2	Papule	Smooth	Well-circumscribed	Pigmented	Black, blue-grey, white	Pigment structureless	Irregular peripheral streaks, blue-whitish veil	Yes	Yes	Excision	Spitz nevus
Vaccaro *et al.*, 2021	NA		17	M	Caucasian	NA	Helix	5×5	Papule-nodule	Papillomatous	Well-circumscribed	Pigmented	Brown, grey, blue, white	Irregular globules	None	No	No	Excision	Spitz nevus

AST, atypical Spitz tumor; F, female; M, male; NA, not applicable.

data reporting multiple dermatoscopical irregularities in 75% of SN. However, this irregular dermatoscopical aspect did not correspond to the histological benignity of the great majority of these lesions. Moreover, 0% of identified ear ASTs had multiple dermatoscopical irregularities, which is in line with the general trend of the literature, where almost 20% of ASTs of the whole body are reported to have only typically benign features.[7]

It is worth noting that dermatoscopical irregularities were not significantly associated with AST, neither in our series nor in the reviewed literature (respectively p=0.375 and p=0.3333), supporting the fact that relying only on the dermatoscopical aspect of spitzoid lesions is not accurate enough for the special site of the ear and that dermatoscopy could actually be misleading.

In addition, though pediatric MM is rare (<1% of all melanomas) and no MM was reported in the present study, one cannot exclude the possibility of spitzoid melanoma arising in pediatric age, especially on the ear, keeping in mind that the spitzoid manifestation of MM is the most frequently reported in MM of pediatric age.[5,11,12]

In conclusion, considering the low correspondence of clinical-dermatoscopic and histological aspects in spitzoid lesions, especially at the challenging anatomic site of the ear, the authors recommend that spitzoid lesions of the ear should be handled prudently.

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