

A Pragmatic Approach: *Pediatric Spitz-like Lesions*

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ABSTRACT

Nurse practitioners serving primary care pediatric patients frequently lack referral access to dermatology specialists. Deciding whether or not to biopsy Spitz-type lesions in children is particularly complex, with significant potential sequelae. Forming an accurate prebiopsy differential through dermoscopic examination, palpation, and history is essential. When atypical Spitz-like features warrant biopsy, diagnostic accuracy requires proper technique and expert dermatopathology assessment. The implications of misdiagnosing malignant melanoma in a Spitz tumor and vice versa are profound for young patients and parents, clinicians, pathologists, and society. Information regarding Spitz-like lesions, including clinical characteristics, biopsy rationale, histology, lymph node assessment, and malpractice litigation, is reviewed.

Keywords: biopsy, dermoscopy, lymph node, Spitz, spitzoid melanoma
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It has been estimated that more than 40% of patients in the United States lack access to dermatology specialist care,¹ which places increased responsibility on primary care providers (PCPs). Spitz nevi, which most commonly arise in patients under 20, require knowledgeable assessment to prevent inappropriate treatment. Nurse practitioners (NPs) caring for pediatric patients may find the assessment and treatment of these melanocytic lesions to be particularly challenging because of a lack of clear guidelines.² Fully describing the histologic characteristics of Spitz nevi and distinguishing these benign lesions from malignant melanoma has even challenged expert dermatopathologists.³ This article is intended to provide pediatric and family NPs with a pragmatic, evidence-based approach to caring for children with Spitz-like lesions.

BACKGROUND

In 1948 Sophie Spitz described a melanocytic lesion with large epithelioid or spindle cells in children, which she labeled *benign juvenile melanoma*.⁴ Spitz hypothesized that it was the hormonal state of pediatric patients that afforded them protection from the widespread metastasis and death associated with the cytologically similar spitzoid melanomas of adults.

Although Spitz's hormonal control theory has not been proven, melanoma remains extremely rare in pediatric populations, occurring in 1 per million patients under 16 years old.⁵ Nevertheless, 1 of the 13 patients in Spitz's original study died of malignant melanoma at age 12, demonstrating the need for improved diagnostic accuracy.⁶

False-negative melanomas diagnosed as Spitz nevi are at the top of the most frequent pathology malpractice claims list, according to recent risk-management studies.⁷⁻⁹ Underdiagnosing the 3%-4% of melanomas occurring in patients younger than 20 could result in what malpractice calls significant "lost years of life per case fatality."⁹ Medicolegal issues involving spitzoid melanomas misdiagnosed as benign Spitz nevi have caused some pathologists and clinicians to err on the side of overdiagnosis and aggressive treatment in equivocal cases.^{9,10} According to Weedon,¹¹ indiscriminant use of the label *minimal deviation melanoma of Spitz-nevus like type* has been used as an "insurance policy" by some clinicians against misdiagnosis.

Over the past 60 years, dermatologists, researchers, oncologists, and dermatopathologists have debated exactly where along the continuum, between benign Spitz nevus and spitzoid melanoma, sufficient atypia

exists to warrant aggressive interventions.^{1,10-21} Whether Spitz nevi and spitzoid melanoma are separate entities has yet to be sorted out, despite enormous efforts involving the most recent advances in pathology and imaging. Diagnostic ambiguity has resulted in wide excisions consistent with a diagnosis of melanoma and sentinel lymph node biopsies.

SENTINEL LYMPH NODE BIOPSY

Unfortunately, atypical Spitz nevi cells in children frequently accumulate in sentinel lymph nodes, leading surgeons to perform complete lymphadenectomy and oncologists to prescribe aggressive chemotherapy.¹⁷ Research, however, has consistently demonstrated that positive sentinel lymph nodes in atypical Spitz nevi do not share the same poor prognosis as exists in spitzoid melanoma. There exist no data associating atypical Spitz-positive sentinel lymph nodes with increased mortality. It has been hypothesized that even typical benign Spitz nevi may normally accumulate in the lymph nodes, but this theory cannot be studied for obvious ethical reasons.

Overdiagnosis of childhood Spitz nevi as spitzoid melanoma can lead to unwarranted wide excisions, sentinel and complete lymphadenectomies, or aggressive chemotherapy regimens, causing lifelong disfigurement, anxiety, morbidity, and socioeconomic burden.²² It is therefore imperative that NPs caring for children use every tool available to optimize initial diagnosis.

DEVELOPING A PEDIATRIC SPITZ ALGORITHM

The prevalence of pediatric Spitz nevi has yet to be determined; however, they are frequently confused with other lesion types and represent approximately 1% of all melanocytic nevi biopsied in pediatric populations.^{11-13,23} Although dermatopathologists can distinguish between Spitz nevi and Reed nevi, many consider Reed nevi to be a Spitz variant, which is how this article will approach them for clinical simplicity. Patients typically lack a family or personal history of melanoma. Spitz nevi are uncommon in children of darker skin types,²⁴ and pediatric cases demonstrate no gender predilection. Because children under age 10 are more likely to have typical benign Spitz nevi, whereas lesions of older pediatric patients more commonly demonstrate atypia, it is important to be familiar with indications for watchful monitor-

ing.¹⁰⁻²⁷ Spitz lesions commonly display a period of rapid growth similar to melanoma, so protocols and techniques for effective monitoring are imperative. Fabrizi and Massi²⁸ found that teenagers with spitzoid melanomas shared the same poor prognosis as those with other types of melanoma.

Spitz nevus should be considered in the differential for pediatric patients presenting with a solitary, 3-10 mm, dome-shaped papule or nodule, often with surface telangiectasia and relatively uniform pink, red, tan, brown, or black color, most commonly on the head, neck, or extremities. They are commonly mistaken for dermatofibromas, hemangiomas, and pyogenic granulomas. A light halo surrounding Spitz nevi is not extremely rare but may complicate pathology diagnosis if biopsied inadequately or interpreted by a less experienced pathologist.²⁹

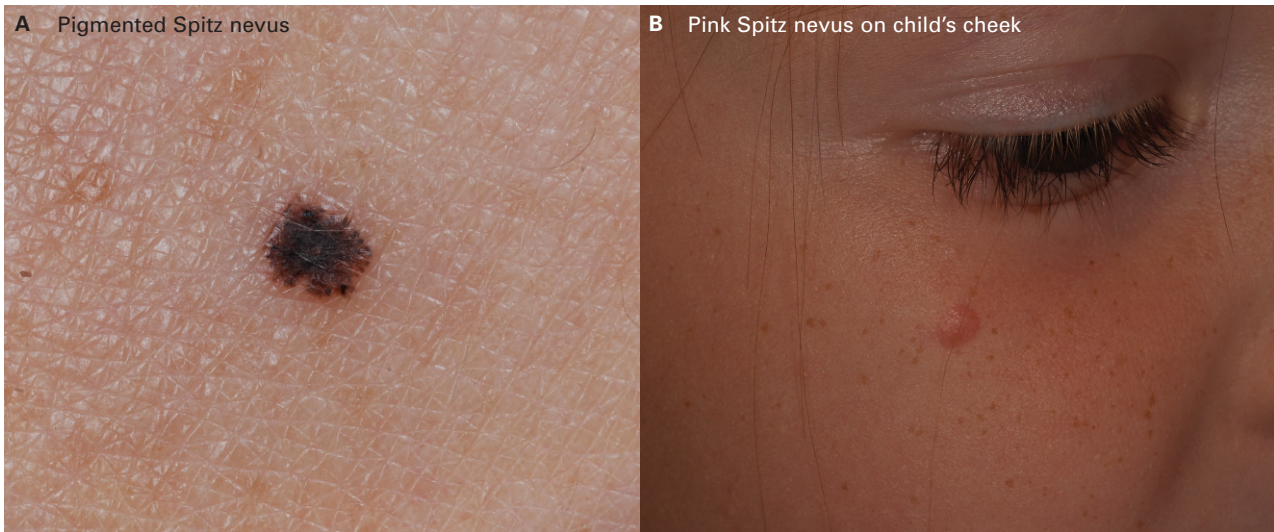
Figure 1 offers examples of Spitz nevi. Lesions may be congenital or acquired and can be either soft or firm to palpation. Smooth or verrucous is equally common, and both types have well demarcated borders. Hurwitz³⁰ recommends diascopy, which simply entails compressing the lesion with a glass slide and observing brown pigmentation in order to eliminate nonmelanocytic growths from the differential.

Less common Spitz tumors may occur in other body locations, including the genitals and oral mucosa on rare occasion; may be ulcerated, less evenly pigmented, polypoid, macular, or plaque-like; between 11-30 mm wide; agminated or disseminated; even occasionally occurring as multiple nevi within a congenital hyperpigmented macular patch. Clinicians must be aware that less common features, older age of presentation, uncommon body location, and increased size are all relevant indications when determining whether to monitor or immediately biopsy a lesion. Figure 2 contains an assessment and treatment algorithm summarizing this information, as well as the next steps in the process.

ASSESSMENT AND MONITORING

Methods for monitoring lesions must include clear photographs of the lesion, including lesion measurements and noting topographical landmarks in relation to the lesion. Familiarity with the dermoscopic features of Spitz nevi can be very helpful in establishing the need to biopsy, and images

Figure 1. Examples of Spitz Nevi



Images courtesy of Dr. Ashfaq Marghoob, Memorial Sloan Kettering Cancer Center, who retains the copyright on these images.

photographed using dermoscopy (epiluminescence microscopy, dermatoscopy) are invaluable for ongoing monitoring.³¹⁻³⁴

Simple, affordable, hand-held dermascopes (polarized and nonpolarized) are readily available and considered essential dermatology assessment tools by many. PCPs in countries with high rates of melanoma, such as Australia, use dermoscopy as their standard of care. This tool allows clinicians to noninvasively see otherwise undetectable common features, such as the circumferential *starburst pattern*, peripheral *brown globules*, or symmetrical *radial streaming* common in pigmented Spitz nevi and the *dotted vessels* and reticular depigmentation in nonpigmented Spitz nevus variants, as shown in [Figure 3](#).^{32,33}

Manufacturers of dermascopes and the International Dermoscopy Society offer free online training, and many introductory and advanced dermoscopy courses and textbooks are offered in the US. It is a mystery why dermascopes have not become as familiar to PCPs as have stethoscopes and otoscopes, but this practice can change as NPs assume a greater role in dermatology assessment and gain insights and experience through routine dermoscopy use.

LESIONAL BIOPSY

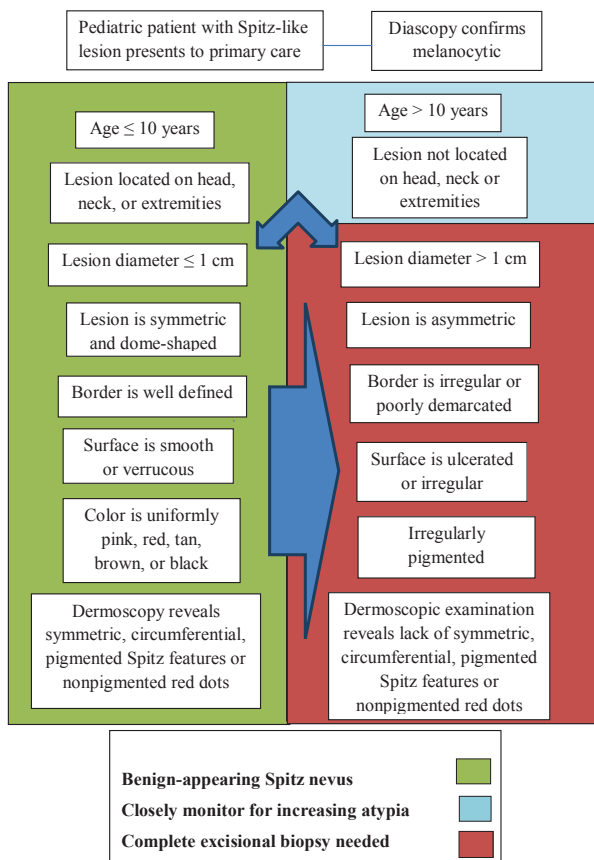
It is never acceptable to choose watchful monitoring as an option in cases where atypical features exist, the

plan and differential are not clearly understood by the parent, or the patient is unlikely to follow up. Appropriate biopsy technique for Spitz-like lesions is predicated upon the fact that accurate assessment requires an adequate specimen. Adequacy is achieved only by removing the entire intact lesion, plus a surrounding clear margin of at least 1 mm of normal skin.^{27,35} Note that halos surrounding lesions are not calculated as normal skin; therefore, margins are measured beginning after the halo edge.

According to Gelbard,³⁶ even experienced clinical dermatologists report confusion about the need to consistently avoid partial biopsy of Spitz nevi, despite consensus and pleas for diagnostic assistance by their dermatopathologist colleagues. The term *benign* may be the source of that confusion. Unless atypia is suspected, typical benign Spitz nevi in children under 10 years of age can simply be monitored. Because the architectural features of symmetry, maturation, and circumscription are the primary characteristics distinguishing atypical Spitz nevi from spitzoid melanoma, partial biopsies fail to provide sufficient diagnostic information. [Figure 4](#) illustrates these diagnostically critical architectural features of Spitz nevi.

A deep scoop biopsy penetrating at least 2 mm into the dermis and including at least 1 mm of normal tissue around the entire perimeter of the lesion or an excisional biopsy down to fat, including at least 1 mm

Figure 2. Assessment and Treatment Algorithm for Pediatric Spitz-Like Lesions



of normal tissue around the lesion, is recommended. Punch biopsy may be performed only in extremely rare cases of unusually small lesions that are well-contained within the diameter of the punch, including a surrounding 1 mm border of normal tissue.

All previously cited dermatopathologists concur that melanocytic lesions should never be removed by curettage, which destroys any chance of architectural assessment. Partial biopsying of Spitz nevi not only creates the need for a second excisional procedure, but also results in a less typical-appearing lesion with former normal architecture destroyed in the biopsy process. Poor clinical technique contributes to the need for subsequent biopsies and increased likelihood of misdiagnosis. Misguided clinicians may be performing partial biopsies in an effort to minimize trauma to young patients and their families.

By initially providing careful assessment, taking a thorough history, palpating, measuring and using

dermoscopy and dermoscopy to examine the lesion, unnecessary biopsying can be avoided in low-risk situations (Figure 2). By explaining why complete excision provides the proper specimen for ruling out melanoma in cases of atypical Spitz, clinicians optimize parental support. Complete excision not only spares patients unwarranted treatments but hastens the diagnosis process, reducing anxiety and costs. If the lesion is too large or located in a difficult area of a child's head or neck, surgical referral is recommended. NPs' ability to provide the surgeon with complete documentation and an informed clinical differential will expedite that process.

If an NP is capable of performing the necessary complete excision, diagnostically significant information to provide on the accompanying lab requisition includes:

- Patient age and gender
- Any family history of melanoma, pancreatic, or breast cancer
- Patient's Fitzpatrick skin type (1-6 based on amount of skin pigmentation)
- Gross lesion description
- Clinical differential diagnosis
- Prebiopsy lesional photographs
- History of lesion growth or changes
- Exact location on patient's body
- Prior lesion excisional history
- Comorbidities

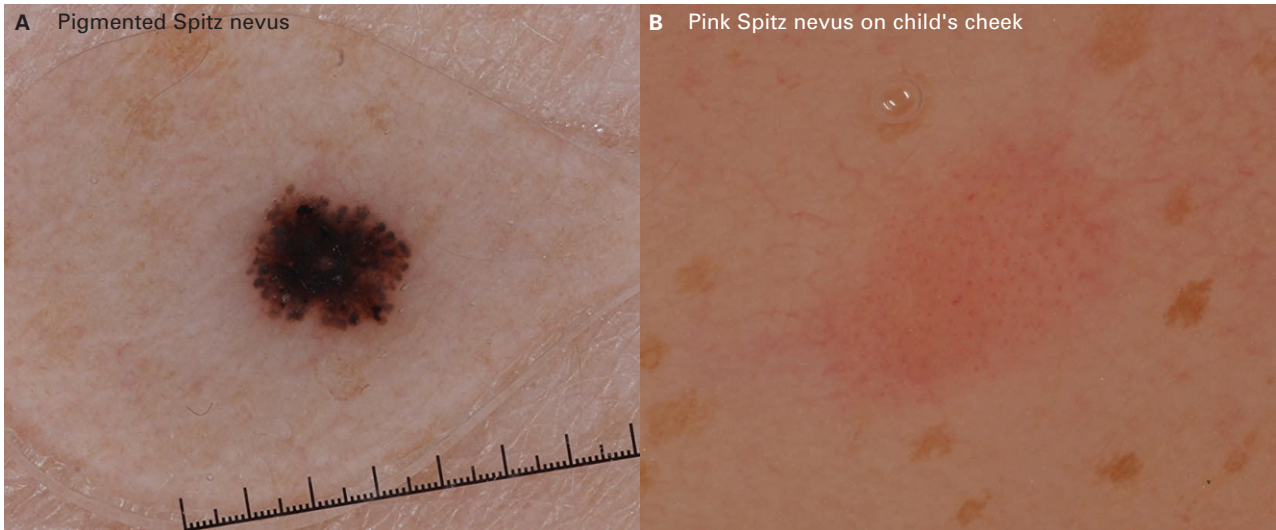
CONSULTATION

Because Spitz nevi have been the subject of so much diagnostic discordance, litigation, and challenge, they should always be sent directly to a dermatopathologist with expertise in Spitz nevi. The training of dermatopathologists is quite different from that of general pathologists, who lack clinical dermatology education and experience. NPs who biopsy and send their specimens directly to expert dermatopathologists not only have their slides optimally prepared and expertly interpreted; they obtain the assistance of that consulting dermatologist, who can discuss treatment options and serve as an ongoing resource.

It is standard practice for Spitz lesions sent to general pathology labs to be mounted on slides, stained, and read, with charges being incurred by

Figure 3. The Same Spitz Nevi from Figure 1 as Viewed Under Dermoscopy

Note: Dermoscopically visible circumferential starburst pattern, symmetrical radial streaming, and peripheral brown globules in the pigmented Spitz nevus (left); the pink Spitz nevus features dotted vessels and reticular depigmentation (right).

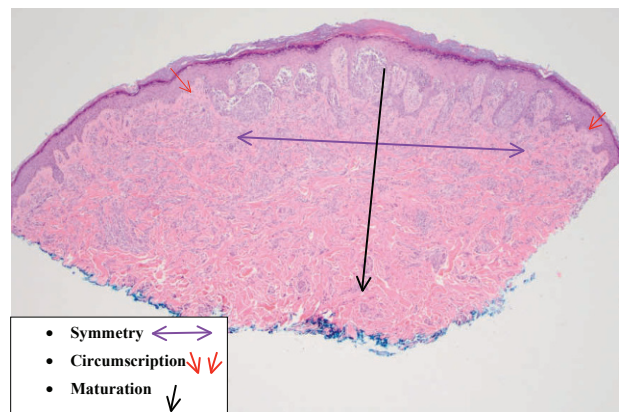


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the insurer or patient; then they are sent to dermatopathologists for secondary consultation. This process often necessitates not only a second charge, but time spent with dermatopathologists requesting and obtaining the remaining portion of the tissue block for additional slide preparation. That unnecessarily costly process is avoided by experienced clinicians who initially send Spitz-like melanocytic specimens to appropriate experts in dermatopathology interpretation. With numerous pathology and dermatology publications stressing the importance of expert dermatopathology interpretation as the standard of care for Spitz-like lesions, clinician liability incurred from misdiagnosis of lesions by non-experts should be considered.³⁷

It is also the responsibility of biopsying clinicians to question biopsy reports that seem highly inconsistent with the clinical diagnosis. Errors can be made, specimens can be mislabeled, and requests for second opinions are part of clinicians' responsibilities in complex cases. NPs only require pathology knowledge sufficient to understand the potential for Spitz lesion ambiguity in order to assertively justify, to collaborating physicians and insurers, their request for an expert dermatopathology evaluation of these cases.

Figure 4. Diagnostically Critical Architectural Features of Spitz Nevi



Complete excisional biopsy of this Spitz nevus demonstrates overall lesional symmetry, clearly circumscribed lateral edges, and maturation of the melanocytes from large atypical to small with descent into the dermis, all of which are required to distinguish the benign nature of this nevus in contrast to spitzoid melanoma. Slide image courtesy of Dr. Matthew Kuhar, Strata Pathology Services, Inc., Lexington, MA; no reprints permitted.

CONCLUSION

Spitz nevi present unique challenges to PCPs serving pediatric populations. Determining which lesions are most at risk for atypical behavior or

malignant transformation can be accomplished by thorough evaluation. It is important for NPs to develop dermoscopy skills to effectively examine the skin of their patients. These skills can reduce unnecessary procedures, while facilitating appropriate referrals and interventions. If NPs perform biopsies of Spitz-like lesions, complete excisions with appropriate clear margins are needed to provide accurate assessment.

Expert dermatopathology interpretation is the standard of care for Spitz-like lesion interpretation and should be directly accessed to reduce health care system waste. Clear communication with and support from dermatopathologists is always available to NPs providing dermatology services to their primary care patients.

The parents of pediatric patients presenting with Spitz-like lesions need to be fully informed of the assessment challenges created by these unique tumors in order to work together for optimal assessment and appropriate treatment. It is hoped that the simple algorithm included in this article will support primary care NPs in providing dermatopathologists with appropriate specimens, correctly biopsied, and only from pediatric patients warranting that degree of invasive intervention. JNP

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