

## Nevo de Spitz desmoplásico: reto diagnóstico en paciente pediátrico

### Desmoplastic Spitz nevus: diagnostic challenge in pediatric patient.

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#### ABSTRACT

Spitz nevus (SN) has an overall incidence ranging from 1.4 to 7 new cases per 100,000 people per year, 60 % are infants and young people under 20 years of age, and the desmoplastic SN variant is the most frequent subtype in late adolescence and adulthood. This clinical case report aims to recognize risk factors associated with SN development, describe the clinical and dermatoscopic manifestations, and analyze the desmoplastic SN variant's histopathological and immunohistochemical findings. Dermoscopy, skin biopsy, histopathological, and immunohistochemical studies were performed. The patient presented a dermatosis localized to the right upper limb affecting the arm, on the external face, middle third and was characterized by an exophytic nodular neof ormation, surrounded by an erythematous halo of 0.8 x 0.8 cm, in the dermoscopy a multicomponent pattern accompanied by a punctate vascular pattern was observed. The biopsy showed nests of fusocellular melanocytes distributed in the papillary and reticular dermis, with fibrous stroma, deep hyaline changes, and accompanying small vessels. The immunohistochemical study showed positive SOX10 and K167 expression, the diagnosis was desmoplastic Spitz nevus. The main treatment was surgical incision, with subsequent surveillance and sun care. The diagnosis of SN represents a difficulty for medical personnel since it has clinical, dermatoscopic, and even histopathological similarities with atypical spitzoid lesions and melanomas, which have a higher risk of mortality, which makes it essential to recognize the wide clinical morphology of the disease.

**KEY WORDS:** Desmoplastic Spitz's nevus, Melanoma.



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## RESUMEN

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El Nevo de Spitz (SN) tiene una incidencia general que oscila entre 1.4 y 7 casos nuevos por cada 100.000 personas por año, un 60 % son infantes y jóvenes menores de 20 años, la variante SN desmoplásico es el subtipo más frecuente en la adolescencia tardía y adultos. El objetivo del presente caso clínico es reconocer factores de riesgo asociados al desarrollo del SN, describir las manifestaciones clínicas, dermatoscópicas, analizar los hallazgos histopatológicos e inmunohistoquímicos propios de la variante SN desmoplásico. Como diagnóstico se realizó dermatoscopia, biopsia cutánea, estudio histopatológico e inmunohistoquímico. La paciente presentaba una dermatosis localizada a miembro superior derecho que afectó brazo, en cara externa, tercio medio y se caracterizó por neoformación nodular exofítica, rodeada por halo eritematoso de 0.8 x 0.8 cm, en la dermatoscopia se observó patrón multicomponente acompañado de patrón vascular punteado. La biopsia mostró nidos de melanocitos fusocelulares distribuidos en dermis papilar y reticular, con estroma fibroso, cambios hialinos profundos y vasos pequeños acompañantes. El estudio inmunohistoquímico mostró SOX 10 positivo y expresión de K167, el diagnóstico fue Nevo de Spitz desmoplásico. El tratamiento principal fue la incisión quirúrgica, con posterior vigilancia y cuidado solar. El diagnóstico del SN representa una dificultad para el personal médico ya que guarda similitud clínica, dermatoscópica e incluso histopatológica con lesiones spitzoides atípicas y melanomas los cuales tienen un mayor riesgo de mortalidad, lo que vuelve imprescindible reconocer la amplia morfología clínica de la enfermedad.

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**PALABRAS CLAVE:** Nevo de Spitz desmoplásico, Melanoma.

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### Introduction

Spitz nevus (SN) is a benign melanocytic neoplasm composed of epithelioid and spindle cells, first described in 1910 by Darier and Civattese. Its general incidence ranges from 1.4 to 7 new cases per 100,000 people per year, occurring more frequently in Caucasian children and young individuals, typically before the age of 20, with the first two years of life being the most common period of onset. A 12-year clinical-epidemiological study conducted in Mexico reported an SN incidence of 9.75 cases per year, with 84 % of patients being under 20 years old. SN development is linked with many risk factors, including genetic, hormonal, immunological, and environmental influences. Among genetic factors, oncogenic alterations in the MAPK pathway stand out, including mutations in the HRAS gene and rearrangements in kinases such as BRAF, MAP3K8, ROS1, and ALK, which facilitate the clonal expansion of melanocytes. Hormonal influences may contribute to the higher prevalence in women between the ages of 15 and 30. Immunosuppression may also promote SN development by allowing the proliferation of mutated melanocytes, which the immune

system would otherwise control. A relationship between SN and immunogenic lesions such as lichen striatus has been reported. Environmental factors, particularly ultraviolet radiation, play a role in its growth (Palacios *et al.*, 2012; Orozco *et al.*, 2023).

SN shares clinical and pathological characteristics with melanoma, which is considered the fifth leading cause of cancer-related death worldwide, this similarity makes SN a controversial entity, as no consensus currently defines its diagnostic features or a clear clinical management strategy, this increases the risk of overlooking malignant lesions that mimic SN (Sepeh *et al.*, 2011; Sainz-Gaspar *et al.*, 2020; Orozco *et al.*, 2023).

Spitzoid melanocytic neoplasms encompass a spectrum ranging from benign to malignant lesions, classified into three main types: conventional SN with its respective clinical variants, atypical Spitz tumor (AST), and spitzoid melanoma. AST may present histological features insufficient for a melanoma diagnosis yet still carry an uncertain malignant potential. Diagnosis is based on dermoscopy and clinical evolution. However, dermoscopy has limitations, as it is not effective for acral surfaces or the diagnosis of amelanotic melanomas, consequently, histopathological examination of a skin biopsy is considered the gold standard. When the histopathological diagnosis of SN is inconclusive, immunohistochemical markers are used as complementary tools to distinguish SN from AST and melanoma. While these markers help identify malignant features, their interpretation can be subjective among pathologists (Sainz-Gaspar *et al.*, 2020; Orozco *et al.*, 2023).

There are multiple clinical variants of SN, including desmoplastic SN, which is described as a rare subtype more commonly observed in late adolescence or adulthood. It presents as a firm or indurated papule with minimal or no pigmentation, resembling a scar with poorly defined borders and a woody consistency, typically located on the extremities. Histopathological examination reveals an intradermal proliferation of spindle-shaped melanocytes within a stroma characterized by hyperplasia of eosinophilic collagen fibers. A central blue-white area and a peripheral network of atypical pigments may be observed. Kamino bodies are absent (Orozco *et al.*, 2023).

Although SN is typically benign, some cases may progress to Spitz melanoma, particularly in adults. Treatment involves surgical excision with wide margins to ensure complete removal of cancerous tissue if malignancy is confirmed. Depending on the extent of the lesion, adjuvant therapies such as immunotherapy or radiotherapy may be considered in more advanced cases (Bologna, 2017).

This report presents the case of a 16-year-old female patient from Cuernavaca, who visited a dermatology clinic due to the abnormal growth of a suddenly appearing lesion on the inner surface of the mid-third of her right upper limb. The lesion had been evolving for 9 months, showing progressive growth throughout this period. At six months, the patient noticed the development of an erythematous halo, occasional stinging, and color changes, becoming erythematous during physical activity. She had no relevant personal or family medical history.

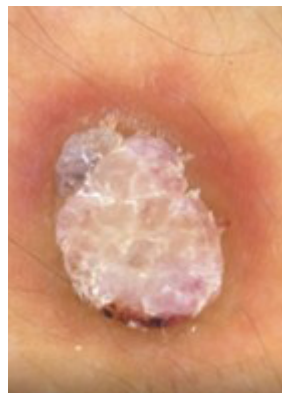
## Clinical findings

Physical examination revealed a localized dermatosis on the right upper limb, affecting the external surface of the mid-third of the arm. It was characterized by an exophytic nodular neof ormation, non-mobile, surrounded by an erythematous halo measuring 0.8 × 0.8 cm (Figure 1). Dermoscopy revealed a multicolored lesion with areas of hemorrhagic pinpointing and confluent oval pigment points, which exhibited whitish halos at the periphery extending into regression zones. A multicomponent pattern was identified, accompanied by a dotted vascular pattern. No additional dermatological or general examination abnormalities were found. Based on these characteristics, a possible pyogenic granuloma was initially suspected, as it typically presents as a rapidly growing papule or nodule with an angiomatous appearance, prone to bleeding and ulceration. This suspicion was reinforced by the patient's medical history. However, a spitzoid melanoma was also considered a differential diagnosis due to the observed multicomponent pattern (Figure 2).



**Figure 1. Asymmetric hypochromic melanocytic melanocytic neof ormation with 9 months of evolution.**

Source: Own elaboration



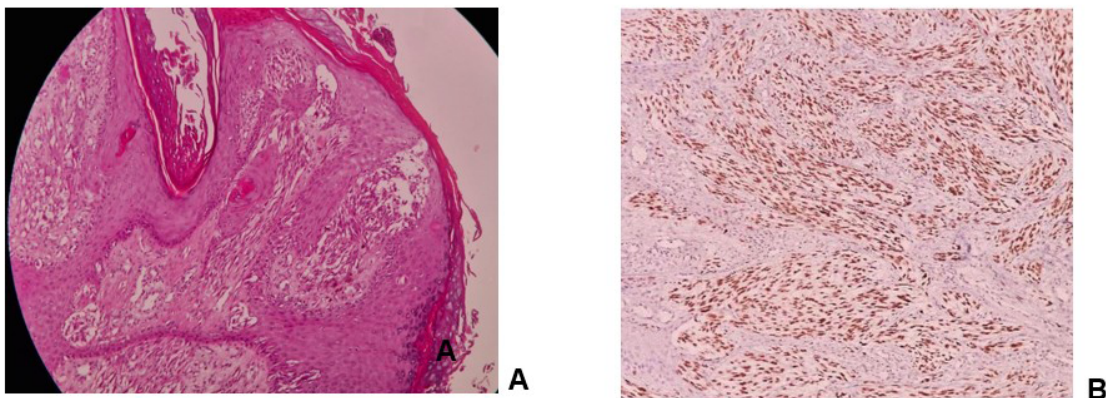
**Figure 2. Dermoscopy of desmoplastic nevus.**

SN with multicomponent pattern accompanied by a punctate vascular pattern.

Source. Own elaboration

A skin biopsy confirmed the diagnosis, followed by a histopathological study. The findings revealed skin with hyperkeratosis and acanthosis, along with randomly distributed nests of spindle-shaped melanocytes at the dermo-epidermal junction extending into the papillary and reticular dermis. These cells exhibited oval nuclei, prominent nucleoli, and minimal mitotic activity.

The stroma was fibrous, with deep hyalinized changes and small, thin-walled blood vessels. Additionally, immunohistochemical analysis showed SOX10 positivity, Ki-67 expression at 1 %, and negative staining for CD68 and CD34, leading to a final diagnosis of completely excised desmoplastic Spitz nevus (Figure 3).



**Figure 3. Histopathological and immunochemical examination results.**

**A.** Desmoplastic Spitz nevus. **B.** SOX 10, positive.

Source. Own elaboration

The patient's prognosis is favorable both in terms of overall health and functional outcomes, thanks to the opportune diagnosis and the inherently benign behavior of Spitz nevus in pediatric and adolescent populations.

### **Therapeutic intervention**

The lesion was analyzed to determine the most appropriate medical intervention, assessing the risk of atypical spitzoid lesions or melanoma. Given that the patient was over 12 years old and the lesion exhibited asymmetry, she was considered high-risk. Consequently, the best therapeutic option was deemed to be complete surgical excision with 1 cm margins. Postoperatively, prophylactic treatment was prescribed to prevent infection, and the use of sunscreen was strongly recommended, as UVB exposure is a known risk factor for skin cancer development.

## Follow-up and outcomes

Three weeks after the surgical procedure, the patient returned for suture removal. There were no signs of infection, although one suture showed early dehiscence and was removed ahead of schedule. One-month post-excision, continued improvement in wound healing was observed (Figure 4). Given that this case is still within the early post-treatment phase (less than one month), ongoing medical monitoring was recommended as follows: a follow-up visit at three months to assess the scar and two evaluations per year thereafter. This active surveillance strategy aims to ensure early detection of any recurrent or similar lesions.



**Figure 4. Outcome of surgical treatment.**

**A.** Postoperative suture. **B.** Postoperative scar, 1 month old.

Source. Own elaboration

## Discussion

In this clinical case, the delay in seeking medical attention and the patient's inability to accurately describe the lesion's progression introduced a significant bias in the information provided during the anamnesis, complicating the early identification of Spitz nevus. While the patient reported color changes in the lesion, describing it as erythematous only during physical

activity and sun exposure, features not commonly documented for Spitz nevi, this led the physician to consider a broader differential diagnosis, including melanoma, rather than initially focusing on pyogenic granuloma or Spitz nevus. Additionally, the lack of initial measurements of the lesion size hindered proper monitoring of its growth in the early months. This contrasts with the expected growth pattern of Spitz nevi, which can reach up to 1 cm in six months, as described in the literature. However, despite these limitations, identifying risk factors significantly contributed to the clinical diagnosis (Sainz-Gaspar *et al.*, 2020; Brown *et al.*, 2021).

Unprotected sun exposure and an age under 20 years are relevant factors in the development of benign skin lesions such as Spitz nevi. Furthermore, puberty has been identified as an associative factor in studies documenting a higher incidence of these nevi in adolescents, particularly in females over 15 years of age, reinforcing the relevance of these variables in clinical suspicion. A crucial aspect in guiding the diagnosis was the use of dermatoscopy, which helped identify a multicomponent pattern characterized by an irregular distribution of colors and white lines, along with a dotted vascular pattern featuring punctate, monomorphic vessels evenly distributed over a pink background. Although this pattern is also characteristic of spitzoid melanoma, it did not completely rule out melanoma but helped narrow the suspicion. However, the absence of the starburst pattern, which appears in more than 50 % of Spitz nevi cases, supported the continued suspicion of a Spitz nevus (Requena *et al.*, 2009; Ferrara *et al.*, 2013; Kerner *et al.*, 2013; Moscarella *et al.*, 2015; Dika *et al.*, 2017; Liebmann *et al.*, 2023; Orozco *et al.*, 2023).

Despite the low incidence and mortality rate of spitzoid melanoma in pediatric patients, the potential malignancy of atypical spitzoid lesions justified the decision to perform surgical excision. The histopathological and immunohistochemical analysis of the excised lesion revealed findings characteristic of desmoplastic Spitz nevus, such as the presence of spindle-shaped melanocytes with oval nuclei and prominent nucleoli, low mitotic activity, and superficial melanocytes with cytoplasmic pigment. Immunohistochemical results, including SOX10 positivity, CD68 and CD34 negativity, and KI67 expression at 1 %, confirmed the diagnosis of desmoplastic Spitz nevus, as the lesion primarily consisted of melanocytes with low proliferative activity and no involvement of other cell types, such as macrophages or fibroblasts (Elder *et al.*, 2015; Bartenstein *et al.*, 2019; Davies *et al.*, 2020).

Due to clinical suspicion and dermatoscopic findings, a complete excision with 1 cm margins was performed to minimize the risk of recurrence or malignant transformation. The decision for surgical excision was based not only on clinical and dermatoscopic findings but also on the patient's age, as previous studies suggest that the risk of melanoma increases with age (Orozco *et al.*, 2023).

The delay in seeking medical attention affected the accuracy of the reported clinical evolution from onset to progression, influencing the diagnostic process. For an accurate and timely diagnosis, dermatoscopy performed by an experienced physician was crucial in guiding a presumptive diagnosis, complemented by histopathological and Immunohistochemical confirmation. It was identified that patterns associated with spitzoid melanoma, such as the multicomponent pattern, are not exclusive to malignant lesions. The utility of dermatoscopy as a

diagnostic tool is emphasized, particularly in its ability to enhance the suspicion of malignancy in spitzoid lesions, especially in patients over 12 years of age (Lallas *et al.*, 2017).

## Conclusions

This clinical case is relevant because it presents an atypical form of Spitz nevus, highlighting the importance of complementary studies such as dermoscopy and histopathology to achieve an accurate diagnosis. It also demonstrates how a thorough evaluation can differentiate benign lesions from those with a potential risk of malignancy. Compared to other documented cases, this one reinforces the idea that, although spitzoid melanomas are rare in the pediatric population, their potential occurrence remains relevant and does not eliminate the priority of achieving a timely diagnosis. Therefore, it should be considered in the differential diagnosis of atypical cutaneous lesions (Bartenstein *et al.*, 2019; Davies *et al.*, 2020).

Complete surgical excision is indicated in treatment when the patient meets any of the following criteria suggesting the risk of developing an atypical spitzoid lesion or melanoma: age over 12 years, asymmetric lesion, and a multicomponent pattern observed in dermoscopy.

## Recommendations

It is important to highlight that studies indicate that the use of lasers on pigmented lesions can complicate the diagnosis of melanoma, hinder clinical management, and potentially worsen the patient's prognosis. Therefore, it is recommended to avoid laser treatment without prior histological confirmation to rule out malignancy (Delker *et al.*, 2017).

## Declarations and ethical considerations

### Informed consent

Informed consent was obtained from all subjects involved in the study. Given the nature of this case report, the patient was informed about data management, and consent was obtained for handling the information. No experimental research procedures were involved; however, consent was collected following Guideline 12 of the International Ethical Guidelines for Health-Related Research Involving Humans, ensuring confidentiality at all times, following the latest update (Ginebra, 2016).

## Conflicts of interest and relevance

The authors declare no conflicts of interest.



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