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Case report

MELANOMA ARISING IN A CHILD WITH A MEDIUM-SIZED CONGENITAL MELANOCYTIC NEVUS

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Abstract

Pediatric melanomas are rare and some of them may arise on giant congenital melanocytic nevi. The risk of developing melanoma on a medium-sized nevus is not clear but is thought to be very rare. Proliferative cellular nodules which mimic malignant melanoma may pose significant diagnostic challenges. We report the case of a 9-year-old patient who developed a melanoma on a medium-sized congenital melanocytic nevus on the tip of the nose, requiring a complex surgery with excellent aesthetic results.

Key words: pediatrics, melanoma, melanocytic nevus, skin cancer, plastic surgery

Resumen

Melanoma sobre un nevo melanocítico congénito mediano en pediatría

Los melanomas en pediatría son raros y algunos de ellos pueden surgir sobre nevos melanocíticos congénitos gigantes. El riesgo de desarrollar melanoma en un nevo de tamaño mediano no está claro, pero se cree que es muy raro. Los nódulos proliferativos celulares, que imitan al melanoma, pueden plantear importantes desafíos diagnósticos. Presentamos el caso de una paciente de 9 años que desarrolló un melanoma sobre un nevo melanocítico congénito de tamaño mediano en la punta de la nariz, que requirió un procedimiento quirúrgico complejo con excelentes resultados estéticos.

Palabras clave: pediatría, melanoma, nevo pigmentado, neoplasias cutáneas, cirugía plástica

Pediatric melanomas are rare but increasing in incidence, representing 1% to 3% of pediatric malignancies[1,2]. It is the deadliest form of skin cancer in children and adolescents and may present with atypical characteristics compared to adults[3]. It can be divided into four age groups: congenital, infantile (up to 1 year), childhood (1 year to puberty), and adolescent (up to 20 years, post puberty). Although most pediatric melanomas arise *de novo*, approximately 30% are associated with a giant congenital melanocytic nevus (CMN) and 20% with other melanocytic lesions, such as small-sized and medium-sized CMN or acquired melanocytic nevi[1-6].

Congenital melanocytic nevi are relatively frequent with an incidence of 1 in 100 newborns[4]. Their clinical relevance relies not only on their potential association with melanoma but

with the central nervous system involvement in the form of neurocutaneous melanosis. The risk of developing melanoma from a CMN is believed to be directly proportional to the size of the nevus: for small (<1.5 cm) and medium (1.5-20 cm) CMN is still unclear, but it has been reported to be nearly 1% to 4.9%[2]. The risk for large or giant CMN (≥20 cm) is 5% to 10%[2,3,5]. Prophylactic excision of CMN does not eliminate completely the risk of melanoma[3].

Proliferative cellular nodules may develop on CMN and pose significant diagnostic challenges because they mimic malignant melanoma clinically and histologically. They are dermal nodular proliferation of melanocytes, characterized histologically by increased cellularity and distinct morphology (usually larger cell size) compared with the background nevus. Typically, they have benign behavior and may regress spontaneously. Occasionally, proliferative nodules may exhibit worrisome clinical features such as rapid growth, changes in shape and color, hemorrhage, and ulceration, and atypical histology as nuclear pleomorphism and expansive growth, making the distinction between melanoma extremely important[7,8].

Examination with dermatoscopy is essential as it is a useful, non-invasive tool. Dermatoscopy evaluation most commonly reveals globular pigmentation patterns, while structureless, reticular or mixed patterns may occur. Small and medium congenital nevi are fairly homogeneous, with most lesions showing either a reticular or a globular pattern. Melanoma arising in such lesions can be identified as a disruption of these benign patterns or by identification of melanoma-specific structures. Tissue biopsy may be used for ruling out malignant differential diagnoses through histopathological assessment[1,9,10].

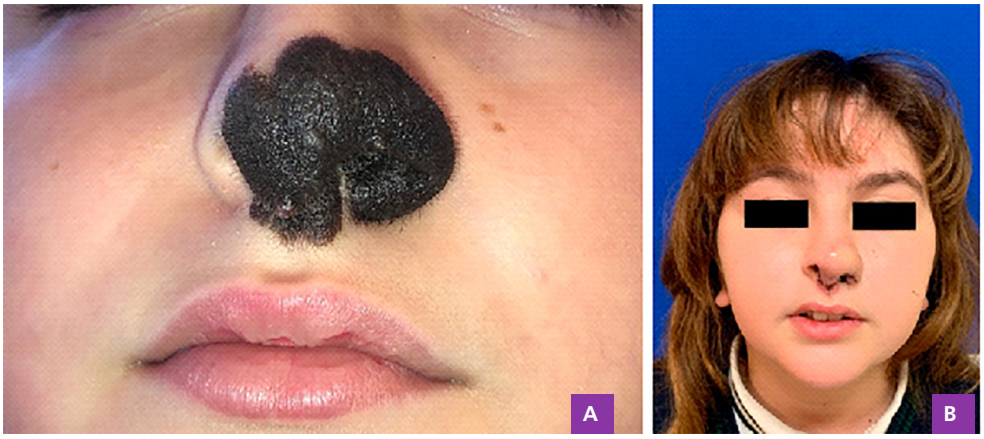
The head and neck are not the most common locations for CMN[11]. Because of aesthetic reasons, their extirpation is required by both pediatric patients and their parents. The surgical management of facial CMN needs to be practiced by a trained professional team and includes excision or staged excision, skin grafting, dermabrasion, tissue expansion or multiple re-expansion combined with flaps transplantation. Proper surgical strategy should be made based on the size and location of the nevi[12]. We report the case of a melanoma on a medium-sized congenital melanocytic nevus on the tip of the nose in a female child that was a complex surgery challenge which had excellent aesthetic.

Clinical case

A 9-year-old female patient was in follow-up due to a medium-sized congenital melanocytic nevus on the tip of the nose (Fig. 1A), with no family history of skin cancer. After dermoscopy detected three rapidly growing lesions in the columella, a biopsy was performed on suspicion of neuroid proliferation or a malignant process. The pathological anatomy report was consistent with a compound melanocytic nevus with congenital features and mild to moderate dysplasia at the junctional level.

Because of the profound psychological stress that the lesion caused to the patient, in conjunction with the de-

Figure 1 | A: Melanocytic nevus with central darker shaded round areas corresponding to neuroid growth. B: Final reconstruction´s results



partments of Plastic Surgery and Pediatrics it was decided to perform a complete surgical resection. Due to the extension of the nevus, that compromised the columella, rhinal ala and tip with invasion of the contralateral side, a three-stage surgery was scheduled.

The first step consisted of the placement of a tissue expander in the frontal region for the design of a pre-expanded midfrontal flap to be used in the final reconstruction. Secondly, the plastic surgeons performed the resection with margins of the lesion and the first stage of nasal reconstruction with a midfrontal flap and an auricular shell graft.

The final pathology report was consistent with a focus of superficial extensive melanoma in vertical growth phase associated with a compound melanocytic nevus with congenital features, Clark level II, Breslow of 1 mm, low mitotic index (without identified mitosis), without ulceration, neurotropism nor lymphovascular invasion, no regression, without microsatellitosis, with lesion-free margins at 1 cm (Fig. 2). According to the immunohistochemical analysis, the tumor cells were positive for melanoma-specific markers Melan A, HMB-45 and S-100, and Ki 67, used as an indicator of cell proliferation, showed positivity of 7%.

Given this diagnosis of melanoma arising from a nasal congenital nevus in a complex area, the case was discussed thoroughly at the Skin Cancer Center of our hospital. It was considered that the safety margins of the malignant lesion were sufficient as far as negative histological margins were concerned. No sentinel node biopsy was performed due to altered lymphatic drainage. Staging studies and magnetic resonance imaging ruled out any possibility of secondary involvement.

Finally, the third stage of the nasal reconstruction was performed with a midfrontal flap, pedicle section and nasal tip refinement, with excellent aesthetic results (Fig. 1B). She is currently doing well 36 months after diagnosis with no signs of recurrence.

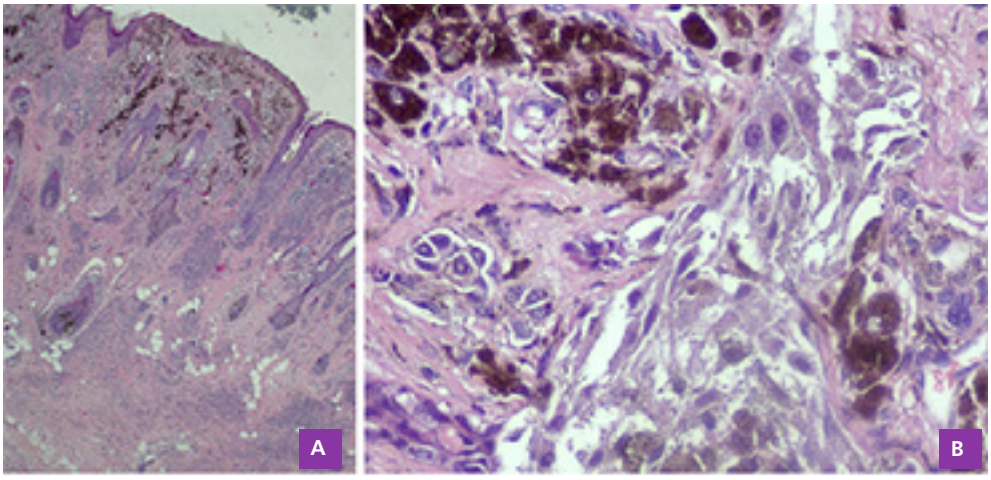
Consent for publication was obtained from legal guardians.

Discussion

Melanoma in childhood and adolescence is rare. In 2020, of 324 635 registered melanomas resulting in 57 043 deaths (0.73%), only 0.08% occurred in children under the age of 20 years, with a mortality rate of 0.01[13,14].

The prognosis of small and medium-sized single CMNs is usually excellent, and their lifetime risk of developing melanoma is estimated to be around 1%, being extremely rare before puberty. On the other hand, in patients with large or giant congenital melanocytic nevi, the risk is

Figure 2 | A: An epidermis with a junctional and deep intradermal component as usually seen in congenital nevi that form nests, with the presence of non-pigmented cells in the deeper region, with a central sector with altered architecture and distribution in melanocytes' thecas and nests surrounded by melanophages. B: With higher resolution there are irregular nests of atypical melanocytes with anisokaryosis and anisonucleosis, showing intracytoplasmic finely granular melanin. These cells have an abnormal intraepidermal distribution and the nests extend into the papillary dermis, where pigmented melanophages are found as well



estimated to be around 5%, being greater as the size increases[1-4,15].

Clinical awareness of possible malignant transformation is crucial. Any suspicious change in a previously stable nevus should alert to the possibility of malignancy and a biopsy should be performed. The gold standard for diagnosis of malignant melanomas is histopathology. Melanomas arising on small and medium CMN predominantly develop from the epidermis, as opposed to the melanomas emerging from giant CMNs, which originate from dermal melanocytes[1,6].

Dermoscopy is crucial in their evaluation, as it is an inexpensive, quick, non-invasive method that can provide the physician with very useful diagnostic information of early malignant transformation. The physician should know the dermoscopic features of CMN to avoid unnecessary excisions, and to recognize a potential melanoma when it begins to develop[5,6,15].

It is important to remember that giant CMNs can compromise the central nervous system, by presenting neurocutaneous melanosis. Individuals with three or more CMN or with multiple satellite melanocytic nevi, or a large CMN in a paravertebral or axial location such as the back, head, or neck, have higher risk and imaging (ideally with a brain MRI with contrast) is mandatory[2, 6, 15].

The treatment of CMNs is one of the most complicated areas of surgical and dermatologic oncology, and there are no world-wide accepted guidelines to treat this type of lesion, especially because it is not entirely clear whether removing de nevus reduces the risk of melanoma. Some surgical options described include serial resection, skin grafts, and the use of tissue expanders, as it was done in our patient[12].

We report a case of a melanoma in a 9-year-old female child, arising on a medium-sized congenital nevus on the nose. The present case highlights the usefulness of dermoscopy in the early detection of significant changes in a melanocytic lesion under follow-up and alerts us to the importance of strict control in children with congenital melanocytic nevi, even if they are low risk. Also, it shows the excellent results of the oncological surgery and posterior aesthetic reconstruction of a complex lesion of the face.

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Conflict of interest: None to declare

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