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Melanoma on Congenital Nevi: Case Report

Case Report

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Abstract

Introduction: Congenital nevi occur in about 1% of newborns. The most used classification is based on their size; small congenital naevus when the largest diameter is less than 1.5cm, intermediate naevi for those between 1.5 and 19.9cm, wide naevus when the diameter is greater than 20cm, and giant naevus when the size is greater than 40cm, with frequent satellite lesions. The risk of transformation into melanoma is estimated at 6.3% for giant nevi, it usually occurs before the age of 20, hence the indication of early excision. This risk is even lower as the size of the nevus is small, and the transformation is usually late. We report the case of a patient with melanoma occurring on a congenital melanocytic naevi of intermediate size.

Case Presentation: We report the case of a 54-year-old patient, with no pathological antecedents, who was born with a congenital nevus of the right shoulder with onset 7 months ago of a nodule within the old lesion, increasing rapidly of which the histological study had concluded to a melanoma.

Discussion: Melanoma can also occur in congenital nevi less than 10cm in size. They can be considered as precursors of melanoma. Because of their large number and frequency, prophylactic excision of all these nevi is not feasible. However, they should be removed as soon as possible when clinical or dermoscopic changes are observed.

Abbreviations: CMN: Congenital Melanocytic Nevus; PN: Proliferative Nodule.

Introduction

Congenital melanocytic naevi (CMN) are found in 1-2% of newborns. They are due to a proliferation of typical melanocytes in the dermis, epidermis or both. They are usually located at the proximal parts of the limbs, trunk, scalp and neck, but may be on any other surface of the skin. Their pigmentation can range from light brown to dark brown and depends on the melanin subtype and its concentration. Histological features may be heterogeneous within the same nevus [1]. Their classification usually depends on their size. Several classifications exist and the most used is the classification proposed by Krengel et al., in which the CMNs are listed in four groups according to the size of the nevus [2-5]. The risk of developing melanoma on a congenital nevus is correlated with the size of the nevus. The risk for small single CMN is very low, whereas where the CMN is > 40 cm projected adult size, and accompanied by multiple smaller CMN, the lifetime risk has been estimated at 10-15% [1, 6, 7]. In this case, we will present a patient with nodular melanoma discovered at the center of a CMN of intermediate size.

Case Presentation

Our patient is a 54-year-old woman who has been hypertensive for one year on treatment. She was born with a medium-sized congenital nevus located at the posterior surface of the right shoulder. For which, she never consulted. Until 7 months ago there was the appearance of a painful, bleeding, rapidly growing nodule on the congenital nevus, which prompted her consultation where a biopsy was performed and final pathological examination verified that it was a nodular melanoma with ulceration, Clark V, Breslow with 3.5mm thickness. The mitotic rate was 7mitosis/field and the presence of vascular embolus (Figure 1, 2). Subsequently, the patient presented herself in our training. The dermatological examination found an homogeneous pigmented, non-infiltrated, oval, well-defined, regularly contoured patch, measuring 9cm/3cm, centered by an heterochromous pigmented plaque, with a nippled surface, painless, non-bleeding at contact, making 5 cm of long axis, with presence within the plaque of a slightly hypertrophic linear scar (Figure 3). The dermoscopy had objectified the presence of a peripheral network with points and globules within the patch and a perifollicular hyperpigmentation (Figure 4), and

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on the plaque, the presence of a papillomatous appearance, with an heterogeneous pigmentation, a polymorphic vascularization made of comma, linear, arborizing and hairpin vessels, the presence of pseudo-comedones and the presence of terminal hair. (Figure 5). The rest of somatic examination was normal. During her hospitalization, an extension assessment was made of a cerebro-cervico-thoraco-abdominopelvic scan and an ultrasound of ganglion areas returning without abnormalities. Then she was referred to the department of plastic surgery where she received a resection of the entire nevus with margins of 2cm, and was put under guided healing. Final pathological evaluation confirmed free margins; no other melanoma sites were confirmed.

During the follow-up, ultrasonography of the axillary lymph node area showed the appearance of suspected lymphadenopathy, resulting in lymph node dissection that confirmed the metastatic nature of the lymph nodes. The patient was subsequently referred to oncology and radiotherapy department for further treatment.

Discussion

Congenital melanocytic nevi (CMN) are found in 1% to 6% of newborns [1].

They are presumed to result from errors in migration and proliferation of melanocytic progenitor cells during embryogenesis [1]. Although most CMN are present at birth, a subset termed "tardive nevi" may appear after birth [8]. Clinically, CMN are characterized by a smooth to mammillated surface, light- to dark-brown color, and well-demarcated borders. Over time, they may darken or lighten and acquire coarse hairs. Thus some CMN may show variations in color, borders, and surface [9]. CMN are usually classified according to size into small (<1.5cm), medium (1.5-19.9 cm), and large (>20cm) [10]. The estimated prevalence of CMN ranges from 0.5% to 31.7% 8. Small CMN have an estimated incidence of 1 in 100 live births, whereas large CMN is 1 in 20,000 live births [11-13]. Recent data suggest that patients with very large CMN and many satellite nevi are at greatest risk of developing cutaneous and extracutaneous melanoma and neurocutaneous melanocytosis [13, 14]. Knowledge of the dermoscopic structures and patterns found in CMN can assist in the diagnosis and follow-up of these lesions. Dermoscopic evaluation of a CMN begins with the assessment of specific dermoscopic structures, in particular the pigment network and aggregated globules [15].

Figure 1. HES staining G x 50, Melanocytic proliferation ulcerated with massive dermal invasion.

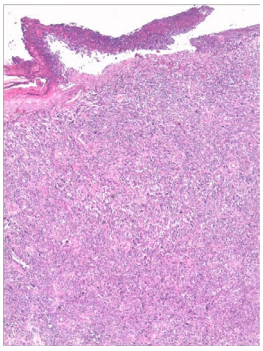


Figure 2. HES staining G x 200, Invasion of the hypodermis.

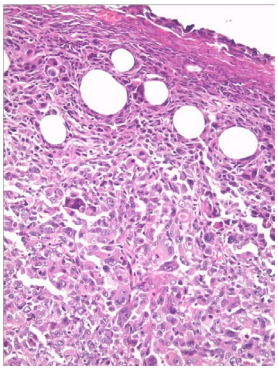


Figure 3. Clinical image.

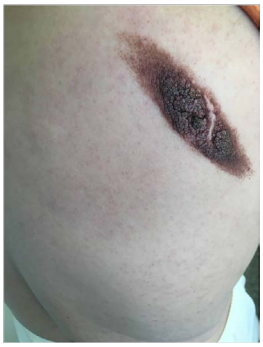


Figure 4. Dermoscopy of the patch.

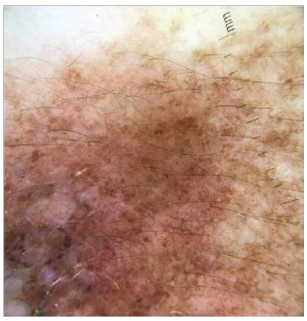
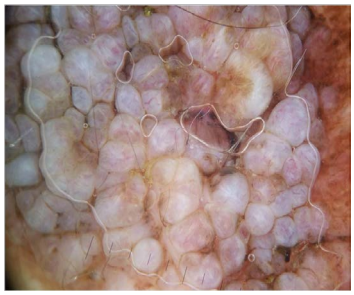


Figure 5. Dermoscopy of the plaque.



The presence of one or more of these structures will confirm that the lesion is a melanocytic lesion [16]. In CMN, the pigment network can be fine, thick, or both. It can be homogeneously distributed throughout the lesion, focal (patchy), or present only at the periphery [16]. Globules can be distributed diffusely throughout the lesion, distributed sparsely or densely, or clustered centrally and surrounded by reticulation. The globules can form a cobblestone-like arrangement or target-like structures in which a halo or a network appears to surround the globules [16]. Other dermoscopic structures frequently observed in CMN include milia-like cysts, perifollicular pigment changes, and hypertrichosis. In addition, almost 70% of CMN will reveal vascular structures under dermoscopy, in particular comma vessels, dotted vessels, serpentine vessels, and target network with vessels [17, 18]. If the dermoscopic of the lesion reveals any of the melanoma-specific structures that are atypical pigment network, streaks, negative pigment network, crystalline structures, atypical dots and globules, off-center blotch, blue-white structures over raised areas, regression structures, peripheral brown structureless areas or atypical vascular structures as it was seen in our case, then a biopsy should be contemplated [15]. It is important to acknowledge that most small and medium CMN are fairly homogeneous clinically and dermoscopically, and melanomas developing in such lesions tend to develop at the dermoepidermal junction. Thus dermoscopy can be relied upon to help in the detection of these melanomas [15]. In contrast, large CMN are often heterogeneous, displaying multiple islands of color and irregular topography, although each "island" within the large CMN tends to be fairly organized and homogeneous in its dermoscopic appearance [17]. That being said, melanom as developing in large CMN often develop below the dermoepidermal junction, and thus dermoscopy cannot be relied upon to help identify these melanomas [15].

The most difficult differential diagnosis of a melanoma arising in a CMN would be a proliferative nodule (PN) developing from the CMN, which is a more frequent occurrence than melanoma [19]. Several studies suggest that the incidence of PNs arising from CMNs ranges from 2.9% to 19% [20-22]. PNs have a stronger tendency to develop from giant CMNs than do melanomas, but they rarely develop from small and medium CMNs [19]. PNs usually develop from CMNs during childhood, and only a few cases have ever been described in adults [23]. Another feature than can help distinguish between PNs and melanomas is the location of the lesion. PNs occur in the dermis of the CMNs. Melanomas tend to arise in the more superficial part of the CMN, such as the epidermis and dermis, in small and medium-sized CMNs. However, in giant CMNs, it is found in deeper tissues [19]. Therefore, the appearance of a nodule in the superficial part of a giant CMN in a child, first suggests a PN. In contrast, the appearance of a nodule in the superficial part of a small or intermediate size CMN in an adult, as in our case, should first rule out the diagnosis of melanoma.

Congenital melanocytic nevi are considered giant or large when greater than 20cm in diameter in an adult or when they cover 2-5% of total body surface. Their malignant potential is well established; they have a high risk of developing melanoma [24]. Moreover, nearly 60% of melanomas in this group occur within the first decade of life [25]. Therefore, early preventive excision of giant nevi is recommended, though this may be surgically difficult depending on size and location [25-33]. On the other hand, only little attention has been given to malignant melanoma on congenital melanocytic nevi less than 10cm in diameter [34]. Rhodes and his collaborators have for the first time systematically investigated the development of melanomas on small CMNs in a large population [35, 36]. In two groups of 234 and 134 patients, respectively, tumor associated CMN less than 4.5cm in diameter were ascertained in 8.1% by histology study and in 14.9% by history. These authors estimated a cumulative melanoma risk of 2.6% to 4.9% for persons with small CMN who live to age 60 years. In view of the fact that small CMN less than 10cm are much more frequent than giant nevi as it was said before [11-13], the malignant potential of the small CMN might be of greater practical importance than that of the large ones [37]. As none

of the CMN-associated melanomas in the Rhodes and associates studies occurred before puberty and because such an event was only rarely reported in the literature, these authors suggest that preventive excision may be delayed until the beginning of puberty. Considering that even CMNs less than 10cm in diameter are potential precursors of melanoma, their risk apparently does not depend on the size of the nevus. Since the risk of developing melanoma will continue throughout life, however, we believe that excision could be the prophylactic method of choice for CMNs less than 10cm, too; but this can be postponed until the beginning of puberty. If not, the patient should be fully aware of the risk and typical aspects of melanoma associated with CMN and clinical and dermoscopic monitoring will be considered.

Authors' Contribution

All the authors contributed for the acquisition, analysis, interpretation of data for the work; and drafting the work or revising it critically for important intellectual content; and the final approval of the version to be published; and the agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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