

Congenital Giant Nevus Tumor: Proliferative Nodules or Melanoma?Dehhaze A^{1*}, Baali N², Dhaidah O¹, Quaboul M¹, Ejjiyar M¹, Bhihi A¹, Sahibi M¹, Elgouatri M¹, El Amrani MD¹, Benchamkha Y¹, Ettalbi S¹¹Plastic, reconstructive, aesthetic surgery and burn unit Mohammed VI's university hospital center Marrakesh, Morocco²Dermatologic department Mohammed VI's university hospital center Marrakesh, Morocco**Case Report*****Corresponding author**

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Abstract: The giant congenital nevus affects a considerable number of children who may be accompanied by proliferating nodules that in some cases become melanoma. We illustrate through a medical observation a case of giant congenital nevi with nodules of histologically confirmed proliferations with a clinical course inclining towards a melanoma demonstrating the difficulty histological diagnosis to differentiate between the two. The literature review confirms the problem of clinical differentiation, histological and evolutionary between these two diagnoses we push to look for other alternatives to decide and have a positive diagnosis and therefore adequate and especially early care of these cases.

Keywords: Congenital nevi, proliferative nodule, melanoma.

INTRODUCTION

Congenital nevi is a condition affecting a considerable number of children that may be stable over time or transforming into a malignant tumor, ie cutaneous melanoma, which is an extremely exceptional tumor in children, its frequency increases with age, representing 1 and 3% of pediatric cancers occurring mainly on a site at risk of xeroderma pigmentosum or congenital nevi. Its incidence is estimated at 2,10000 births with a risk of malignant transformation approaching 2.5.

We illustrate, through this observation, a case of congenital giant tumor nevus with proliferation nodules of atypical appearance, which posed the problem of differential diagnosis with melanoma given the unfavorable evolution with the result of the assessment of extension which was very favor of a malignant melanoma on a giant congenital nevus.

The objective of this observation is to demonstrate the difficulty of anatomopathological diagnosis in the case of proliferation nodules that may be clinically malignant melanoma.

MATERIALS AND METHODS

It's about a case report of giant congenital nevi whose evolution was poorly prognostic

RESULTS

Male child of 02 years and 5 months, from a non-consanguineous wedding, with a giant congenital nevus sitting in the left lower limb, part of the right lower limb, buttocks and almost the entire back. One month before hospitalization, two nodular lesions appeared; inguinal right and to the posterior aspect of the painful left thigh which gradually increased in size.

The clinical examination found a conscious child, in good general condition, with an invasive afebric lesion having a giant blackish lesion with a plumper surface, spreading all over the left lower limb,

partly the right lower limb, the buttocks, and almost all the back sparing only the top 1/3.

It is surmounted by two nodular lesions, the first erythematous 6 cm long axis located at the root of the left thigh, soft and painful on palpation, the second sitting on the posterior aspect of the left thigh, nodular rounded d about 8 cm in the long axis, of soft consistency and hard and painless center. Multiple rounded noses were scattered, of variable size.

The anatomopathological examination of the various subtotal exereses carried out shows, in total, a morphological aspect compatible with congenital giant nevi, with no sign of malignant transformation or incomplete resection.

A complete extension assessment, including an ultrasound showed a left inguinal swelling (8.7 * 5.9 cm) fleshy double component, echogenic irregular contours budding in the cystic portion with anechoic content, seat area bubbles and fine echoes mobile.

- Long popliteal dimpled (6.4* 3.5 cm) fleshy, echogenic, double-component, with irregular contours budding in the cystic portion with anechoic content.
- Important infiltration of fat and soft parts of neighborhoods.

The extension assessment was completed by thoraco-abdominopelvic CT, which showed a large mass of the locally infiltrating left inguinal region associated with bilateral pulmonary micronodules of secondary origin most likely.

A biological assessment, including a blood count, showed anemia at 9.2 g / dl hypochromic microcyte, hyperleucocytosis at 15 300 / mm³, renal function, hepatic function and hemostasis balance were without abnormalities. The surgical procedure consisted of an excision of the two lesions with a direct closure. The external resection limit appeared healthy, unlike the others that were tumorous.

The anatomic-pathological study of the operative specimen showed tumor proliferation nodules made of relatively larger sized melanoma cell layers than those of the adjacent nevus. These cells were globular or spindle-shaped, with abundant cytoplasm, often pigmented, and with a bulky, vesicular and nucleolus nucleus.

The mitotic activity is variable from one nodule to another, reaching in places 20-25 / 10 large fields. The stroma is fibromyxoid, covered with fibrinohematologic raptus and punctuated by small lymphocyte clusters. This proliferation infiltrates the deep dermis and hypodermis, reaching 20 mm thick.

On the surface, the cutaneous fragment is the seat of an intradermal and hypodermic naevic tumoral lesion producing some pseudoepithelial thecae in the subepidermal zone and especially deep diffuses layers. These plies are made of naevic elements showing signs of fragmentary neuroid maturation.

In conclusion, these were cell proliferations, some of which were atypical in appearance with high mitotic activity on giant congenital nevi. The immunohistochemical study was not done by default means.

Close monitoring of the child for a period of 1 month revealed no recurrence, however, the evolution was unfavorable and the prognosis was bleak, marked by the very early death of the child.

DISCUSSION

Congenital nevi (NC) are by definition present at birth [1], however, they can still appear in the first months of life up to 2 years [2]. Depending on their size, there are small size NCs (size <1.5 cm), medium

and large NCs (size between 1.5 and 20 cm) and giant NCs (> 20 cm) [3].

Clinically, giant NCs are often highly pigmented macules, progressively overlapping with hair (75% of cases) and are seated with warty, papular or nodular lesions more or less voluminous and very disturbing [4]. Many scattered satellite nevi are often associated [5].

Localization is ubiquitous, of segmental or metamerical disposition, neuromeningée melanosis should be sought, if the seat is axial or para-vertebral (neuroradiological exploration by medullary and / or cerebral MRI) [6,7]. The risk of transformation into melanoma is all the more frequent as the lesion is large.

Proliferative nodules may exist within the lesion, clinically achromic or highly pigmented, requiring systematic histological confirmation [8].

Also called cellular nodules or atypical nodules sitting in the superficial or reticular dermis or the superficial part of the hypodermis, thus comprising epitheloid, spindle cells or small cells [10].

These proliferation nodules may present numerous cell atypias, irregular nuclear, and several mitoses may thus erroneously carry a histological diagnosis of malignancy.

Mitoses are quite rare with a percentage of cells marked by Mi B1 is <15%, a rather coarse pigmentation and not finely dusty as in melanomas and nuclei are fairly uniform [9].

However, the differential diagnosis between malignant melanoma and atypical proliferation nodules (NP) occurring in giant congenital nevi remains very difficult, and very few studies have been done in this context, including the presence of a homozygous CDKN2A mutation. (P16, 9p21) described in family MM cases of uncertain significance but requiring close surveillance [11].

In the S.Fraitag study, a 4-color FISH technique was reported to aid in the "benign versus malignant" differential diagnosis in classical melanoma tumors, having shown that both NP and melanoma developed on NCG during in childhood are accompanied by chromosomal number abnormalities that do not allow the benign versus malignant differential diagnosis at this age unlike the melanoma occurring on NCG in adulthood [12]. No clinical, evolutionary, histological, immunohistochemical or molecular biology features have demonstrated its ability to clearly differentiate them.

In our observation, the adverse development marked by the occurrence of pulmonary metastasis with

the very early death of the child is very much in favor of a malignant melanoma.

CONCLUSION

The provision of new diagnostic methods would be of great interest to advance the knowledge of congenital giant tumor nevi and thus improve their prognosis.

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