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Clinical Image

Giant Congenital Melanocytic Nevus (GCMN) - A New Hope for Targeted Therapy?

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Abstract

We present a 6-month-old male patient, who was consulted with dermatologist by his parents, because of a pigmented lesion, present since birth, covering almost the all skin of the back and buttocks. A sharply bordered, unequally coloured congenital pigmented nevus, measuring approximately 21 cm in diameter was observed in the whole body skin examination. The lesion was affecting the lower 2/3 of the skin of the back and the top half of the gluteus area, extending to the lateral part of the tors, forward the abdomen and the upper lateral part of the hips, composed by multiple darker-pigmented nests and several lighter areas, with single depigmented zones, hairy surface, irregularly infiltrated on palpation. Congenital melanocytic nevi are presented in approximately 1% of newborns, while giant congenital melanocytic nevi (GCMN) are the most uncommon subtype of them; with occurrence rate 1 in 50,000 births. They affect 2% of a total body surface or presenting in a diameter larger than 20 cm in older children. Although not common, the possible malignant transformation remains one of the most important considerations related to them, as the related lifetime risk of melanoma is 4% to 10%. Treatment recommendations include non-surgical methods as dermabrasion only within the first two weeks of life, for prevention the possible melanocytic deeper migration, while serial surgical excisions or tissue expanders could be useful treatment tool even in later stages. Nevertheless, cosmetic result is not always satisfactory, and the risk of malignant changes remains, in cases of previous melanocytic migration in deeper layer. Recent article suggests the potential role in the treatment of GCMN with NRAS inhibitor trametinib, approved for treatment of advanced melanoma, associated with underlying NRAS mutations. Although promising, the drug could be useful in paediatric patients, only with associated NRAS gene mutation. It is still unclear whether it could be helpful, independent of the NRAS status.

We present a 6-month-old male patient, who was consulted with dermatologist by his parents, because of a pigmented lesion, present since birth, covering almost the all skin of the back and buttocks. A sharply bordered, unequally coloured pigmented nevus, measuring approximately 21 cm in diameter was observed in the whole body skin examination. The lesion was affecting the lower 2/3 of the skin of the back and the top half of the gluteus area, extending to the lateral part of the tors, forward the abdomen and the upper lateral part of the hips, composed by multiple darker-pigmented nests and several lighter areas, with single depigmented zones, hairy surface, irregularly infiltrated on palpation (Fig. 1a, 1b, 1c, 1d). Several satellite pigmented lesions were also observed on the upper and lower extremities, measuring from one to 3-4 cm in diameter, most of them- hairy surfaced. Atopic dermatitis was also seen as an additional abnormality. The performed screening procedures, including MRI and CT, did not reveal any abnormalities, neither data for neurologic involvement.

Congenital melanocytic nevi are presented in approximately 1% of newborns, while giant congenital melanocytic nevi (GCMN) are the most uncommon subtype of them; with occurrence rate 1 in 50,000 births [1]. They affect 2% of a total body surface or presenting in a diameter larger than 20 cm in older children [1]. Although not common, the possible malignant transformation remains one of the most important considerations related to them, as the related lifetime risk of melanoma is 4% to 10% [2]. Treatment recommendations include non-surgical methods as dermabrasion only within the first two weeks of life, for prevention the possible melanocytic deeper migration, while serial surgical excisions or tissue expanders could be useful treatment tool even in later stages [1]. Nevertheless, cosmetic result is not always satisfactory, and the risk of malignant changes remains, in cases of previous melanocytic migration in deeper layer [3].

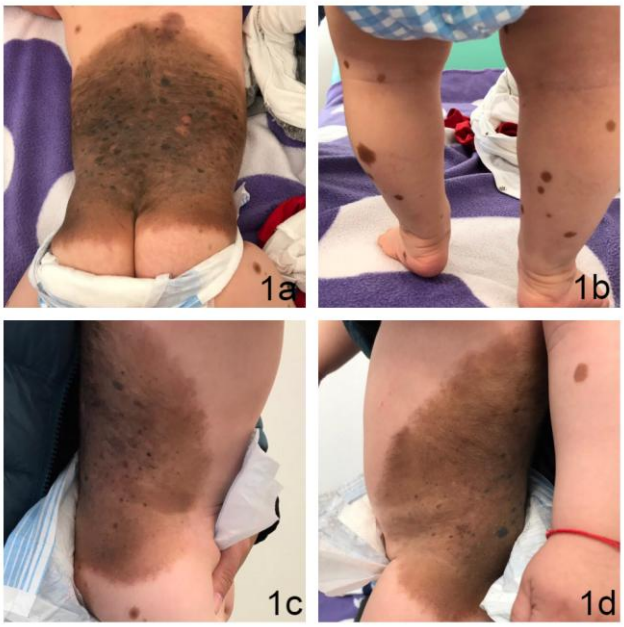

Figure 1: – Clinical manifestation of a giant congenital melanocytic nevus in 6-month-old child, affecting the lower 2/3 of the skin of the back and the top half of the gluteus area (1a), extending to the lateral part of the tors, forward the abdomen and the upper lateral part of the hips (1c, 1d), composed by multiple darker-pigmented nests and several lighter areas, with single depigmented zones, hairy surface, irregularly infiltrated on palpation and several satellite lesions (1b)

Recent article suggests the potential role in the treatment of GCMN with NRAS inhibitor trametinib, approved for treatment of advanced melanoma, associated with underlying NRAS mutations [3]. Although promising, the drug could be useful in paediatric patients, only with associated NRAS gene mutation. It is still unclear whether it could be helpful, independent of the NRAS status. Meanwhile, the future following up of these patients is mandatory [1].

Despite some of the patient could undergo screening by dermatoscopy, this tool is not useful in cases with full-size infiltration and equally dark-brown to black colour, as the remarkable dermoscopic finding could be noted only in the hypopigmented areas, as in the presented cases [4]. The clinical examination could note sigh of infiltration, while PET-scan and reflect confocal microscopy should be added as helpful tools in the following up period. Excision biopsy could be performed in infiltrated areas, but it is not sure and safety enough method for observation of malignant transformation. Therefore, total surgical excisions, by serial operations or tissue expanders remain the most effective treatment option, with minimal risk of malignant transformation and maximal therapeutic effectiveness.

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