Nonprogressive size reduction of limbs involved in giant congenital nevi: A magnetic resonance imaging study

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Giant congenital nevi are hamartomas of neuroectodermal origin predominantly involving the trunk; two or more body segments are also commonly involved. A feature observed when giant congenital nevi are involved in a limb is a nonprogressive reduction in limb size. This feature has been evaluated, and demonstrates alterations only in the skin and subcutaneous tissue of the affected limb, with all other structures normal compared with the unaffected contralateral limb. The present magnetic resonance imaging study clearly demonstrates the substitution of subcutaneous tissue and fat cells by nevus cells, leading to a dramatic volume reduction in these tissues. The volumes of the deeper structures were not affected.

Key Words: *Giant congenital nevi; Limb size reduction; Nonprogressive*

Two patients presenting with giant congenital nevi (GCN) involving 30% and 50% of body surface area, respectively, had, in addition, involvement of the upper leg in one case and the lower leg in the other case (Figure 1).

Both patients presented with clearly evident asymmetry of the involved limb when compared with the contralateral side. It was clear that the function and length of the affected limb was comparable with the unaffected side.

In each case, magnetic resonance imaging (MRI) studies demonstrated the absence of normal subcutaneous structures in the region of the GCN. The deeper lying structures (bone and musculature) resembled the unaffected side.

DISCUSSION

A GCN is a pigmented nevomelanocytic nevus involving $\geq 20\%$ of the body surface area (1). GCN are associated with an estimated lifetime risk of malignant transformation of 6.3% (2).

These cosmetically unsightly lesions often cause emotional distress and have severe psychological sequelae. Nevus cells are of two different populations: one population of darker-staining larger cells being present superficially in the dermis; and a population of cells in the deeper dermis containing little pigment (3). Nevus cells may be found as deep as the fascia and muscle levels (1,2,4). The most aggressive form of therapy, therefore, does not remove all nevus cells (3). A nonprogressive reduction in the size of the limbs involved in GCN has been previously postulated and has been evaluated using electron myographic studies, which have indicated no conduction difference between the affected and unaffected limbs; limb length also does not differ when comparing the affected and unaffected sides (1).

Discrepancies between the affected and unaffected sides have previously been clinically and electron myographically evaluated. These studies have now been expanded by incorporating MRI evaluations that have clearly indicated that the difference in volume between a GCN-involved limb and the unaffected contralateral side is due to a lack of subcutaneous tissue in the affected region. This volume difference in the affected limb remains proportional as the child grows, which explains the term 'nonprogressive limb reduction'.

Réduction non progressive de la taille de membres atteints de naevus géant congénital une étude d'imagerie par resonance magnetique

Les naevus congénitaux céants sont des hamartomes d'origine neuroectodermique qui se retrouvent principalement au tronc. Plus de deux régions du corps sont généralement impliqués. Un fait observé quand le naevus congénitaux céants se retrouve sur un membre, est une diminution non progressive de la taille du membre. Ce fait a été évalué et indique seulement une atteinte de la peau et du tissu sous-cutané du membre affecté et les structures environnantes sont normales, lorsque comparé au membre contralatéral non atteint. L'imagerie médicale par résonance magnetique a clairement démontré que le tissu sous-cutané et les cellules adipeuses étaient substitués par les cellules naeviques expliquant le volume réduit significatif dans ces tissus. Le volume des tissus profonds n'est pas affecté.

Figure 1 A giant congenital nevus involving the lower leg. The discrepancy in the size in the calf area is clearly evident

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Figure 2) A giant congenital nevus involving the trunk and upper leg. The difference in size of the two thighs are clearly evident

Figure 3) An axial magnetic resonance imaging comparison between the two lower limbs. The left limb is affected by the giant congenital nevus, demonstrating the dramatic reduction in subcutaneous tissues when compared with the normal side (right)

CONCLUSION

A GCN is associated with severe cosmetic handicap and a well-known risk of malignant transformation. Even with treatment, patients often develop severe emotional distress syndromes. The clinical situation is further complicated if a limb is involved. A macroscopically evident,

Figure 4) A coronal magnetic resonance imaging view of the patient depicted in Figure 3, confirming the difference in subcutaneous tissue volume with no other structural differences

static or nonprogressive reduction in limb size and shape occurs. This limb volume reduction has also been evident at birth (Figure 2).

This condition does not affect the function of the limb in the cases studied. MRI studies have shown near total absence of the normal subcutaneous tissues in the areas of the overlying GCN (Figure 3). The nevus cells are clearly evident in the area, with a paucity of subcutaneous tissues, especially when compared with the normal contralateral limb. As a result, this leads to a dramatic decrease in bulk in the region of the lesion (Figure 4). The larger area covered by the GCN makes it easier to appreciate the phenomenon.

This phenomenon was also appreciated in other affected areas of the body and has been detected in areas of the scalp that were involved in other cases evaluated.

Thus, it is easy to understand why nevus cells can be found down to the fascia and muscle levels in the region of the GCN.

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