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Case Report

Cerebriform congenital melanocytic nevus of scalp and its management using tissue expansion

Shruti Patel[1], Naveen Kumar[1]

[1]Department of Plastic Surgery, Lady Hardinge Medical College and Associated SSK Hospital, New Delhi, Delhi, India.

***Corresponding author:**
Naveen Kumar,
Department of Plastic Surgery,
Lady Hardinge Medical College and Associated SSK Hospital,
New Delhi, Delhi, India.

drnaveenvmmc@gmail.com

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ABSTRACT

Congenital melanocytic nevi are benign proliferations of cutaneous nevomelanocytes. Usually, they manifest at birth or become apparent within the first few years of life. The nevi show variable surface morphology (papular, rugose, verrucous, or cerebriform). Congenital melanocytic nevus showing cerebriform morphology is a rarity. Early diagnosis and surgical excision are usually recommended in congenital melanocytic nevus to prevent the future risk of malignant transformation which is higher in larger lesions, especially in giant forms (>20 cm in size). An excision of the lesion also helps to avoid the social and psychological consequences arising out of significant cosmetic deformity. We report a 21-year-old patient who presented with a cerebriform congenital melanocytic nevus measuring 10 cm × 7 cm × 2 cm in the right parietal region. Early-onset, pigmented lesion with a cerebriform surface, and the histopathology features of congenital melanocytic nevus were the points that favored the diagnosis of cerebriform congenital melanocytic nevus in our patient. He was treated with excision of the lesion and defect coverage with tissue expansion in two stages. Two rectangular tissue expanders were placed beneath the galea aponeurotica (one with a capacity of 300 cc in the left parietal region and another with 500 cc in the occipital region). Both the expanders were inflated twice to their capacity. Second stage surgery was performed after about 3 months in which the tissue expanders were removed and the pre-expanded scalp skin was used to drape the scalp defect that resulted from the excision of the lesion. An excision and a two staged reconstruction of the scalp using tissue expanders, may ensure a good aesthetic outcome in the management of intermediate to large sized congenital melanocytic nevus.

Keywords: Scalp, Nevi, Tissue expander, Melanocytic, Congenital, Cerebriform

INTRODUCTION

The congenital melanocytic nevi are seen in approximately 1% of new borns.[1] The congenital melanocytic nevi vary considerably with respect to their size, location, and morphology.[2] Cerebriform congenital melanocytic nevus is one of the rare forms, where the surface of the nevus is thrown into gyri and sulci giving a cerebriform appearance.[3-6] As there is a potential risk for the development of malignant melanoma in congenital melanocytic nevi, which is more in larger lesions, early diagnosis, and surgical excision are usually recommended. Moreover, patients seek excision of the lesion to avoid social and psychological consequences arising out of significant cosmetic deformity.[7]

We report a 21-year-old patient who presented with cerebriform congenital melanocytic nevus, who was treated with excision of the lesion and defect coverage with tissue expansion in two stages.

CASE REPORT

A 21-year-old man attended the plastic surgery department with multiple, pigmented nodules of variable sizes on the scalp, that were present since birth. The patient was born with a nodular swelling on the right parietal region of the scalp. The skin and hair covering the lesion were normal. The lesion had enlarged gradually over a period of years, and the overlying skin became thickened and dark in color with no other complications. During the past 3 years, the lesion showed a rapid increase in the size to cover most of the parietal region. The patient had normal intelligence without any past history of trauma, chronic skin disorder, or history suggestive of malignancy. He did not give a family history of similar illness.

Clinical examination revealed a painless, pigmented swelling (10 cm × 7 cm × 2 cm) with multiple nodularity (that imparted a cerebriform appearance) and normal scalp hairs, in the right parietal region [Figure 1]. There was no lymphadenopathy. His systemic examination was essentially normal. Radiography of the head ruled out any involvement of calvarium. Intra-cranial pathology was ruled out by computed tomography and magnetic resonance imaging of the head. The differential diagnoses considered were cerebriform congenital melanocytic nevus and diffuse neurofibroma. Because of aesthetic concern, the patient and the family opted for excision of the lesion.

Patient was operated in two stages. In the first stage, two tissue expanders (rectangular in shape), were inserted in the scalp beneath the galea aponeurotica. One with a capacity of 300 cc was placed in the left parietal region and another with 500 cc was placed in the occipital region with the incisions made at the junction of the lesions and the normal scalp [Figure 2]. Both the expanders were inflated twice to their capacity and a second stage surgery was performed after about 3 months in which the lesion was dissected, and completely removed. The periosteum was cauterized using bipolar coagulation. Tissue expanders were removed and the pre-expanded scalp skin was used to drape the resultant scalp defect. The wound healed completely without any complication [Figure 3]. Histopathological examination confirmed the diagnosis as congenital melanocytic nevus.

Figure 1: Pre-operative image of the patient with cerebriform congenital melanocytic nevus.

DISCUSSION

Cerebriform congenital melanocytic nevus is a rare morphological type of congenital melanocytic nevus.[3,5] The presence of sulci and gyri over the lesion gives it a cerebriform appearance. Histopathology features that distinguish congenital nevi from the acquired nevi include, the presence of nevomelanocytes within the lower dermis and the

Figure 2: Tissue expander *in situ* following first stage surgery in a patient with cerebriform congenital melanocytic nevus.

Figure 3: Post-operative image after the excision of the cerebriform congenital melanocytic nevus and the two staged reconstruction of scalp (using tissue expander).

extension of the same between collagen bundles, around hair follicles, eccrine apparatus, and sebaceous glands.[8] The risk of neoplastic transformation and cosmetic concern (as in our patient) necessitate the excision of the lesions.[7]

Early-onset, pigmented lesion with a cerebriform surface, and the histopathology features of congenital melanocytic nevus were the points that favored the diagnosis of cerebriform congenital melanocytic nevus in our patient.[1,8]

Reconstruction of the scalp following the excision of large lesions poses a challenge since it may not be possible to attain a good aesthetic outcome, through excision and skin graft. A staged repair using tissue expanders is one of the options that may provide closure with satisfactory results in the hair-bearing scalp.[9] Small lesions can be managed with partial scalp excision with primary closure or grafting.[9] However, in the case of primary cutis verticis gyrata, a "butterfly"-shaped scalp skin excision design along with sub-galeal scalp relaxation incisions was proposed by some of the researchers to effectively improve the appearance of the scalp.[10] Moehrle *et al*. have reported two patients with cerebriform giant melanocytic nevus of the scalp. One was treated with serial excisions under subcutaneous infusion anesthesia. The second patient underwent excision of the nevus (after implanting a tissue expander) and defect closure using rotation flap.[5]

Our patient sought a plastic surgery consultation mainly for the cosmetic reason. The challenge in our case happened to be the complete removal of the lesion while maintaining a satisfactory aesthetic outcome (preserving the normal hair pattern with minimal scarring). We opted for closure of the defect using tissue expansion since it tends to serve the aforementioned purpose.

In order to plan the tissue expansion using two rectangular-shaped tissue expanders, the lesion was roughly divided into two halves. Two expanders of appropriate volume and length (about 1.5 times the length of each half of the lesion with base width roughly equal to the width of either of the lesions) were selected to resurface the defect during the second stage. The total surface area gain provided by a rectangular-shaped tissue expander is about 40%, which is the highest that can be provided by expanders of any shape.[11] They could easily resurface the resultant scalp defect following complete excision of lesion.

CONCLUSION

An excision and a two staged reconstruction of the scalp using tissue expanders, may ensure a good aesthetic outcome in the management of intermediate to large sized congenital melanocytic nevus.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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