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Giant Cerebriform Nevus Cell Nevus of the Scalp: A Case Report

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Abstract

Giant cerebriform nevus cell nevus of the scalp is an extremely rare form of congenital melanocytic nevus. Giant cerebriform nevus of the scalp has a major psychosocial impact because of its unsightly appearance with fetid maceration. We report the case of a 35-year-old woman who had a painless, malodorous swelling of the cerebriform scalp measuring 20 × 17 cm in diameter with a wide base of insertion at the occipital level adhering to the deep planes. The excision associated with a skin plasty was carried out. Histology concluded that there was a giant cerebriform naevo-cellular nevus of the scalp.

Keywords

Giant Nevus, Tumor of the Scalp, Resection

1. Introduction

Giant cerebriform nevus cell nevus of the scalp is an extremely rare form of congenital melanocytic nevus [1]. The congenital melanocytic nevus is a benign proliferation of melanocytes of the epidermis, dermis or other tissue and is present at birth [2]. However, malignant transformation into melanoma is possible and this risk increases when it comes to giant nevus [1]. According to Wu [3], the lesion is defined as giant when its size is at least 20 cm. Giant cerebriform nevus of the scalp has a major psychosocial impact because of its unsightly appearance with fetid maceration. This disfiguration couple with the risk of malignant transformation often requires surgical therapy. If surgical treatment is unanimous; the surgical strategy varies between: total excision associated with a skin graft,

excision and rotation flap and implanted expander before excision.

The aim of our study is to report a case of a giant cerebriform nevus cell nevus of the scalp which was operated in our department.

2. Case

A 35-year-old female, without any remarkable past medical history, was hospitalized for a painless swelling of the scalp in the occipital region evolving since birth. It started with a slowly small painless enlarging nodule in the occipital region, gradually increasing over the years motivating some "traditional" treatments.

Physical examination found a cerebriform, solid and painless of 20 × 17 cm of diameter with a large base of insertion swelling of the scalp adherent to the deep layer in the occipital region. A fetid secretion was seeping out of the folds of the cerebriform (Figure 1).

The CT scan showed a large heterogenous occipital tumor with heterogeneous contrast enhancement. There was no associated bony lesion neither intracranial extension (Figure 2). The surgery was performed by a double team (neurosurgeons and plastic surgeons).

During the resection, a highly vascularized and well limited cutaneous tumor with sparing of the galea. Skin grafting was done and the skin graft was harvested

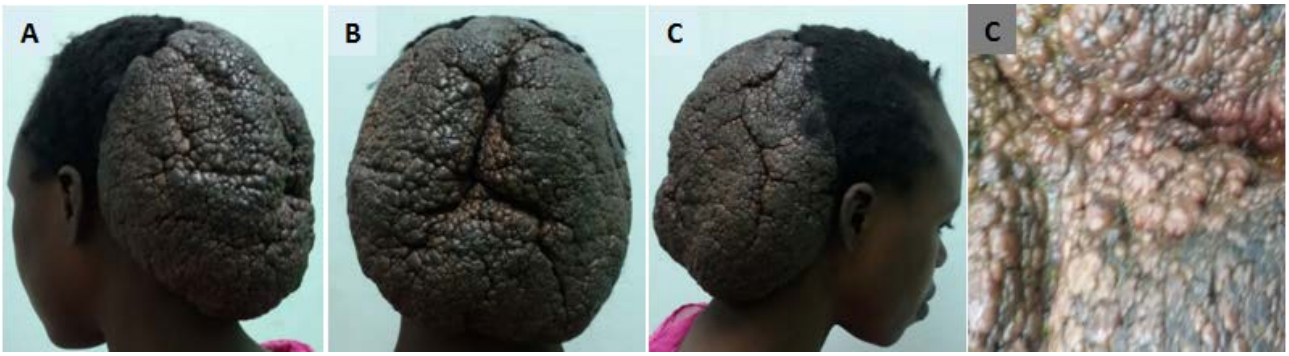

Figure 1. Appearance of the tumor on physical examination: Profile view of the left side (A), Back view (B) and Right side profile view (C). Purulent maceration between the cerebriform folds (D).

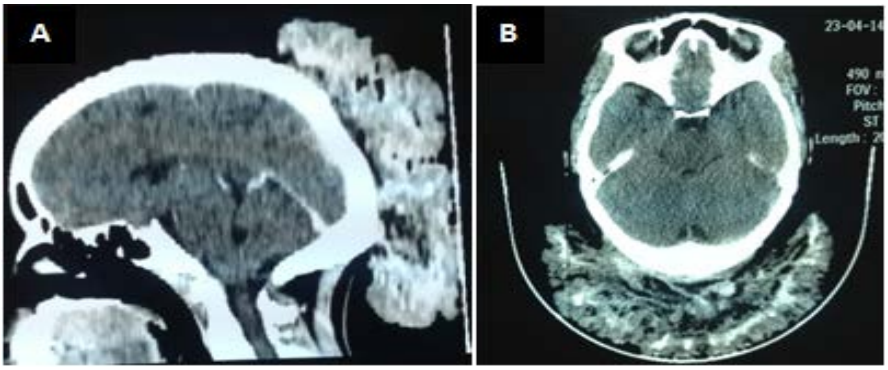

Figure 2. Brain CT showing the heterogenous occipital tumor, Sagittal reconstruction (A) and Axial section (B).

from the posterior aspect of the right thigh.Vaseline gauze and compressive (occlusive) dressing were used (Figure 3 and Figure 4).

Postoperative course was uneventful with good skin graft healing at postoperative Day 7 and Month 1 respectively (See Figure 5).

Histopathology examination of the surgical specimen found a benign naevopleomorphic cell nevus which was presented in the form of layer proliferation of rounded naevic cells loaded with melanin pigment extending into the dermis without cellular atypia.

The evolution at 6 months postoperatively was characterized by good healing

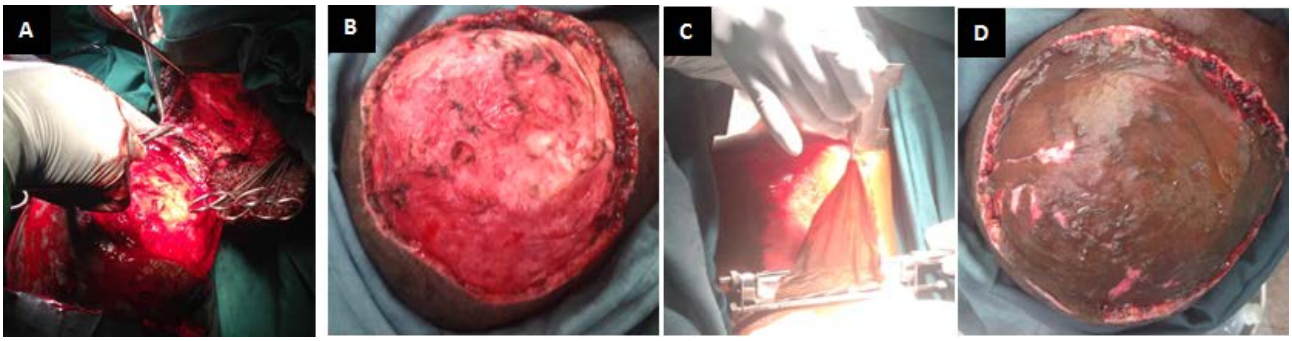

Figure 3. Hemorrhagic excision (A), Total tumor resection sparing the galea (B), Skin graft harvesting from the anterior aspect of the right thigh (C), Graft placement (D).

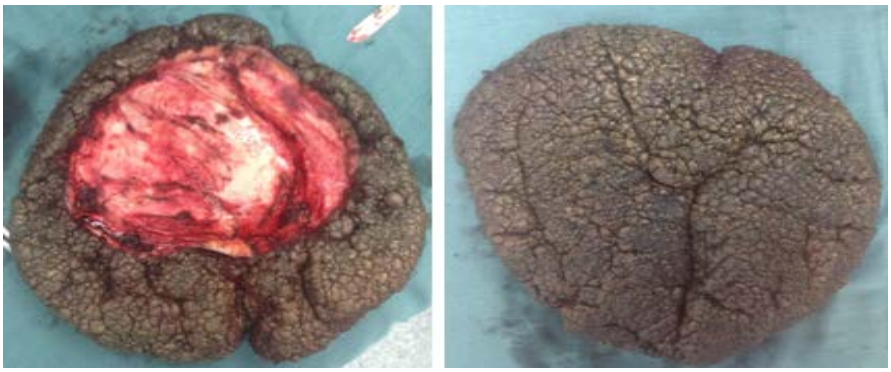

Figure 4. Tumor mass after complete monobloc resection.

Figure 5. Good skin graft healing with budding at postoperative day 7 (A) et (B). Complete healing at postoperative month 1.

of the graft, the absence of tumor recurrence and the psychological satisfaction of the patient. Due to the absence of hair on the graft, a capillary expander implant was considered. It wasn't carried out because of the inaccessible cost.

3. Discussion

Giant congenital melanocytic nevi are rare with estimated incidence less than 1/20,000 new borns [4]. Giant cerebriform nevus of the scalp is an extremely rare form. In our case it is a young adult (36 years old) like those of Lischner [5] and Moehrle [1]; however, two cases have been reported in patients over 50 years [6] [7]. The slight female predominance reported for giant congenital melanocytic nevus [8] [9] is not confirm in its scalp giant cerebriform.

The long consultation delay (36 years) observed for our patient is partly due to some multiple attempts of traditional treatment. This delay in consultation is also found in some European authors [1] [5]. This observation leads us to say that the delay in consultation is mainly linked to the very slow evolution of the tumor.

The reason for consultation is almost always aesthetic. The consultation is motivated by a paroxysmal social rejection at an advanced stage of the lesion due to the unsightful nature and fetid maceration. Our patient consulted after being abandoned by her husband because of her lesion. The congenital nature as well as the hyperchromic cerebriform aspect with maceration and fetor was unanimously reported [1] [5] [6] [7].

The occipital location in our case (occiput) is also reported by three other authors [1] [5] [7]. The large diameter of the lesion in our patient (20 cm) is wider than Gross [6], Hayachi [7] and Moehrle [1] which were 18 cm, 10 cm and 18 cm respectively. The giant lesion criterion still poses a problem of consensus. Hence, Wu [3] and Pai [10] retained a diameter of at least 20 cm while Illig *et al.* [11] retained a diameter > 10 cm.

Medical imaging studies did not find any bony lesion or intracranial extension in our case on CT as well as in those of Moehrle [1] and Hayachi [7] on MRI.

Surgical management was recommended due to cosmesis, psychosocial impact and malignant transformation [12]. Total resection was the surgical option chosen in our case like that of Moehrle [1] and Hayachi [7]. After the resection, we performed an autologous skin grafting while Moehrle [1] opted for a rotation flap. Gross [6] did only a biopsy because he discovered the lesion at the stage of metastatic melanoma.

In our study, similarly to Moehrle [1], the pathological examination concluded to a benign nevus. In contrast, the studies of Gross [6] and Hayachi [7], pointed in favor of malignant melanoma.

The postoperative course was uneventful for our patient with good management of the skin graft and a proper healing. We are considering capillary expansion in our patient. It wasn't carried out because of the inaccessible cost.

4. Conclusion

This is a rare pathology which poses a significant cosmetic and psychological

problem. Surgical resection with subsequent plastic surgery is the only means of correcting this esthetic problem while preventing an eventual malignant transformation.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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