

Case Report

Congenital cerebriform intradermal nevus presenting as cutis verticis gyrata: a case report

Ashish Jawarkar*, Bhumika J. Gharia, Amrish N. Shah

Department of Pathology, Parul Institute of Medical Sciences and research, Vadodara, Gujarat, India

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***Correspondence:**

Dr. Ashish Jawarkar,

E-mail: pathologybasics@gmail.com

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ABSTRACT

Cerebriform intradermal nevus is a rare disorder characterised by development of folds and furrows on the scalp, giving it a convoluted appearance resembling surface of the brain. There are two main forms of CVG, primary and secondary. Secondary form may appear at any age and is usually secondary to causes such as tumors, Cerebriform intradermal nevus (CIN), neurofibromas or amyloidosis. There is high risk of development of malignant melanoma in patients of CIN presenting as CVG. Hence early diagnosis and treatment is important.

Keywords: Bull dog scalp, Cerebriform intradermal nevus, Cutis verticis gyrata

INTRODUCTION

Cerebriform intradermal nevus is a rare disorder characterised by development of folds and furrows on the scalp, giving it a convoluted appearance resembling surface of the brain.¹ It is a secondary cause of Cutis Verticis Gyrata (CVG), varyingly described as Cutis sulcata, bulldog scalp and cutis capitis strata. CIN is an intradermal lesion histologically.¹ First case in literature of Cerebriform intradermal nevus resembling cutis verticis gyrata was described by Hammond et al in 1937.² Early diagnosis and treatment of CIN is important as there is high risk of development of malignant melanoma.³ We would like to present a case of congenital Cerebriform intradermal nevus presenting as CVG and treated by wide excision and grafting.

CASE REPORT

Six years male child presented to the out-patient department with occipital lesion, having loose folds of scalp in the area and absence of hair at the centre-clinically cutis verticis gyrata. (Figure 1) Patient did not

show any signs of mental retardation. The lesion was present since birth, did not increase in size but folds showed increasing thickness with age. We received wide local excision specimen of involved scalp with 1cm clear margins.



Figure 1: Scalp over the occipital area shows cerebriform folds, hair is sparse over the central part of the lesion.



Figure 2: The excised lesion as a whole.



Figure 3: Cut surface-the folds are thick with no grossly apparent tumor in the epidermis or the dermis. Few spots of hyperpigmentation noted in the superficial dermis.

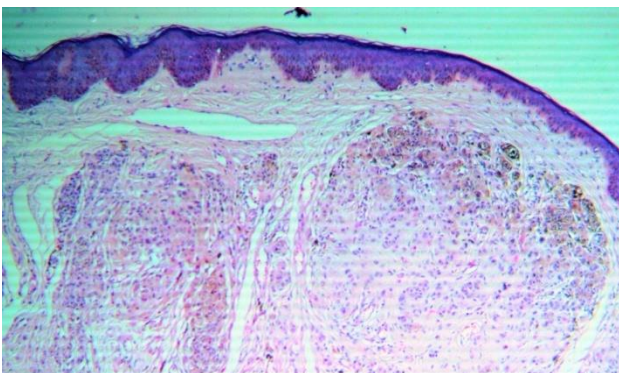


Figure 4: Tumor in the superficial dermis comprised of melanocytes in nests with well-defined cell boundaries.

The scalp folds were approximately 2.5cm thick (Figure 2, 3) multiple sections were taken for microscopic examination. Microscopy showed a tumor in the superficial dermis comprised of melanocytes in nests with well-defined cell boundaries, abundant eosinophilic to amphophilic cytoplasm containing coarse melanin granules. They have uniform round to oval nuclei, finely

dispersed chromatin, delicate nuclear membrane and small distinct eosinophilic nucleoli. Abundant eosinophilic to amphophilic cytoplasm containing coarse melanin granules was seen. There was no junctional activity and no evidence of atypia (Figure 4, 5, 6).

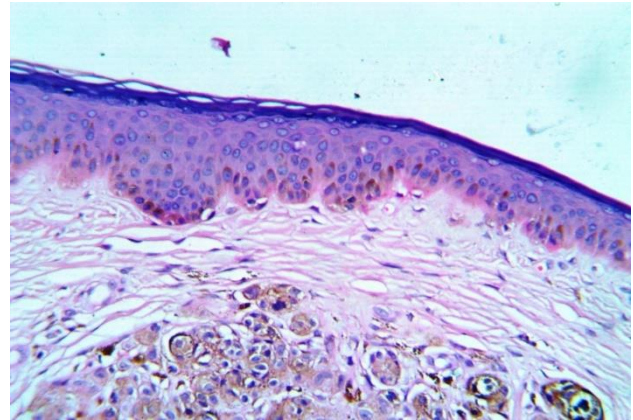


Figure 5: There was no junctional activity and no evidence of atypia.

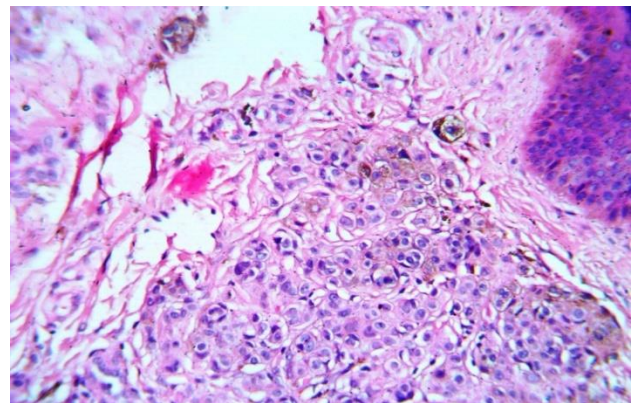


Figure 6: Individual cells were round to oval with uniform nuclei, finely dispersed chromatin, delicate nuclear membrane and small distinct eosinophilic nucleoli.

DISCUSSION

Cutis verticis gyrata is a morphological description for condition of the scalp and not a disease entity by itself. It refers to deep furrows and convolutions seen that represent outer surface of the cerebrum.⁴ The lesion is mainly localized to hairy scalp particularly in the occipital region. The condition is usually asymptomatic. There are two main forms of CVG, primary and secondary. The primary form is associated with mental retardation, epilepsy, cerebral palsy, cranial abnormalities, ophthalmological abnormalities like strabismus or a combination of these.⁵ Secondary form may appear at any age and is usually secondary to causes such as tumors, intradermal nevi (CIN), neurofibromas or amyloidosis. Patients with CIN have no mental retardation or systemic disease.

However, there is high risk of development of malignant melanoma to the tune of 6.3-12%.⁶ Hence early diagnosis and treatment is important. Treatment is complete surgical excision followed by grafting. In cases where complete excision is not possible, close follow-up of the lesion is advisable.⁷

CONCLUSION

Cerebriform intradermal nevus is a rare secondary cause of Cutis verticis gyrata. Early diagnosis and treatment is important as there is risk of development of malignant melanoma.

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