CASE REPORT

Congenital melanocytic nevus mimicking a turban tumour in an 18-year-old Filipino male

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SUMMARY

Congenital melanocytic nevi (CMN) are nevi that are present at birth or arise within the first few weeks of life. They are often found on the trunk, head and neck and extremities. We report herein an unusual presentation of a CMN as a cerebriform tumour presenting as secondary cutis verticis gyrata on the scalp of an 18-year-old Filipino male.

BACKGROUND

The presentation of a congenital melanocytic nevus (CMN) as a secondary cutis verticis gyrata (CVG) on the scalp is uncommon. In this case, it is important to perform laboratory workup to establish the diagnosis and rule out disease entities associated with these conditions so that management can be individualised.

CASE PRESENTATION

An 18-year-old male consulted due to an enlarging tumour on the left parieto-occipital scalp. At

Figure 2 Clinical appearance of the tumour in the parieto-occipital scalp showing numerous folds and thickenings resembling the surface of a brain (posterior view).

Figure 1 Clinical appearance of the tumour in the parieto-occipital scalp showing numerous folds and thickenings resembling the surface of a brain (anterior view).

birth, there was a solitary round black patch with hair growth on the left occipital scalp measuring approximately 7.6 cm×7.6 cm. Lesion was stable until 6 years prior to consult when patient noted gradual enlargement of the lesion. It developed into a gyrated, doughy, hairless tumour on the left occiput extending to the left parietal scalp. This eventually prompted consult at our institution.

His medical history revealed no known comorbidities. His family history was unremarkable.

His sun exposure was limited to 3–4 times a week whenever he plays sports. He does not use sunscreen or any form of sun protection.

Focusing on the dermatological examination, the patient had a well-defined 20 cm×20 cm×8 cm round tumour on the left parieto-occipital scalp. The tumour is doughy and cerebriform in appearance. There are tufts of hairs growing in between the folds (*figures 1–3*). There are no other lesions on the rest of the body, mucosal surfaces and nails. Dermoscopy findings showed a globular pattern with sharply circumscribed aggregates of brown pigment that are diffusely scattered (*figure 4*).

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Figure 3 Clinical appearance of the tumour in the parieto-occipital scalp showing numerous folds and thickenings resembling the surface of a brain (lateral view).

INVESTIGATIONS

Biopsy of the tumour on the left occipital scalp showed nests of melanocytes maturing with descent in strands and cords in the reticular dermis with some increased single cells along the dermal–epidermal junction. Histopathological diagnosis is consistent with CMN, cerebriform type (figures 5 and 6). Cranial CT scan plain and with contrast showed normal findings. Fasting blood sugar was normal. Lipid profile showed

Figure 4 Dermoscopy findings showing sharply circumscribed round to oval aggregates of brown pigment.

Figure 5 Biopsy of the tumour on the left occipital scalp showing nests of melanocytes with maturation with descent into the reticular dermis with some increased single cells along the dermal–epidermal junction (low-power view).

elevated triglycerides and very low-density lipoprotein and low high-density lipoprotein and low-density lipoprotein.

DIFFERENTIAL DIAGNOSIS

Connective tissue hamartoma is a differential diagnosis that we can think of. It can present with hypertrophy and folding of the scalp. It is also present at birth or during early childhood with a history of increase in size like in our patient. However, without the biopsy findings, we cannot be certain if there is an increased amount of collagen or elastic fibres within the papillary or reticular dermis. Ectopic meningoepithelial hamartoma is another differential diagnosis. Like in our patient, it usually presents at birth or early childhood on the posterior aspect of the scalp. However, the biopsy findings would show involvement of the dermis and subcutaneous tissue with ‘proliferation of connective tissue elements and vessels admixed with meningoepithelial elements, without any sign of malignant transformation’.⁴ Cylindroma can also be considered because it commonly involves the head and neck area, including the scalp. The tumours can coalesce in the scalp, forming a confluent mass called ‘turban tumour’. However, based on the history, cylindroma is more common in females and it typically presents in the third decade of life. Furthermore, cylindroma is a proliferation derived from sweat glands. Amyloidosis is another differential diagnosis. But based on the patient’s history there is no weakness, paresthesias,

Figure 6 Biopsy of the tumour on the left occipital scalp showing nests of melanocytes extending to the level of the reticular dermis (high-power view).

Figure 7 Postoperative appearance of the patient after the tumour was excised (anterior view).

Figure 9 Postoperative appearance of the patient after the tumour was excised (lateral view).

numbness, easy fatigability, joint pains and weight loss. Histopathology would show amyloid deposits which are absent in our patient's case. Pachydermoperiostosis can also be considered but this can be ruled out because the patient does not have involvement of the face, oily skin, acne, thickening of the skin on the hands and feet, hyperhidrosis and clubbing of the digits.

TREATMENT

Warranted excision of CMN depends on the following factors: melanoma risk (large CMN), age of the patient (older age), anatomical location (proximity to vital structures), the presence of neurocutaneous melanosis (found in 2.5% to 45% of CMNs), cosmesis and complexity of removal.²

Figure 8 Postoperative appearance of the patient after the tumour was excised (posterior view).

For our patient, there was a history of progressive enlargement of the tumour so a biopsy is already warranted to establish the histological diagnosis of the tumour. We also referred him to the Division of Plastic Surgery.

OUTCOME AND FOLLOW-UP

Patient underwent a series of operations including excision of the tumour, insertion of the expander, advancement flap and reinsertion of the expander (figures 7–9).

DISCUSSION

CMN are nevi that are present at birth or arise within the first few weeks of life. These are often larger than benign acquired nevi and can reach more than 20 cm in size. Lesions may have a smooth, pebbly, rugose, verrucous, cerebriform or grossly lobular surface. CMNs with a cerebriform appearance may present as CVG.³

CVG is characterised by excessive growth of the skin on the scalp and folding of the skin into a cerebriform pattern. It can be classified as either primary or secondary. Primary CVG is further divided into essential (no neurological or ophthalmological abnormalities) or non-essential, which is associated with mental retardation, cerebral palsy, epilepsy or ophthalmological findings. Meanwhile, secondary CVG is due to underlying conditions such as inflammatory skin disorders (eg, eczema, psoriasis, folliculitis, impetigo, erysipelas, dissecting cellulitis), local neoplasms (eg, CMN, neurofibroma, fibroma, hamartoma) and systemic illnesses (eg, myxedema, acromegaly, amyloidosis, syphilis, leukaemia, acanthosis nigricans, metabolic syndrome). Clues for secondary CVG include early onset of the disease. Other clues are asymmetry and disordered furrows that are not oriented anterior to posterior.⁴

Cerebriform intradermal nevus (CIN) is a secondary form of CVG that is usually congenital but can also be acquired. The first case was reported in 1937 by Hammond and Ransom. The first manifestations appear at birth or early in life. At 5–10 years of age, these lesions are observed to grow rapidly. In the patient's

case, the tumour rapidly grew in size when he was around 11 years old. The most common locations of CIN are on the parietal and occipital regions of the scalp. It is not known how the cerebriform pattern of the tumour develops.⁵ CIN are thought to be related to giant CMN, but several differences were observed. CIN is usually a slightly or non-pigmented lesion (vs intense pigmentation in giant CMN). Second, there are absent or sparse hair follicles in CIN (vs increased number of hair follicles and nevus cells at the dermoepidermal junction in giant CMN) and lastly there is uncommon junctional activity in CIN compared with giant CMN.⁶

The patient's tumour measures 20 cm×20 cm×8 cm, which makes it a large CMN associated with risk for melanoma. Neurocutaneous melanosis can also be found ranging from 2.5% to 45% of cases for large CMN. For CIN, the risk of neurocutaneous melanosis is not yet established.⁷

Learning points

- ▶ As dermatologists, we should not be limited in performing skin biopsies when we come across with this kind of case.
- ▶ We must not forget the importance of a multidisciplinary approach in the management of our patients.
- ▶ We should keep in mind that we can actively manage these cases by ordering diagnostic exams to rule in/rule out diseases associated with this condition and refer to other specialties when the need arises.

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Contributors MEGS is the attending physician of the patient and was the one who requested for the workup and diagnostic tests to establish the patient's diagnosis. She wrote most of the sections of the manuscript as well. EC is the dermatopathologist who read the slide of the patient's biopsy and contributed to the writing of the manuscript. BUT is one of the surgeons of the patient and contributed to the writing of the manuscript.

Competing interests None declared.

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