CASE REPORT

Basal cell carcinoma arising in a congenital melanocytic naevus in an adult

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SUMMARY

Congenital melanocytic naevi (CMN) are common skin lesions. They harbour a risk of malignant transformation, and various lesions have been described as developing within them. A basal cell cancer occurring within a CMN has never previously been described. A case is described of a woman aged 52 years presenting with a slow-growing, symptomatic 3 cm lesion in the centre of a 10×5 cm CMN on her right upper back. Diagnostic core biopsy revealed an ulcerated, infiltrative basal cell carcinoma which was then further excised. The scar has healed with no evidence of local recurrence at 1-year follow-up.

BACKGROUND

Congenital melanocytic naevi (CMN) are neural crest-derived, benign proliferations of melanocytes, determined in utero, and present on the skin within the first year of life.^{1–2} The maximal size that a CMN is projected to attain by adulthood has traditionally been used to categorise CMN, and to determine the risk of adverse outcomes,¹ the so-called 'naevus burden'.² More recent works examine histological criteria and involved anatomical regions to create a more sophisticated classification system for this heterogeneous group of lesions.^{1–3}

Nearly 1 in 20 children is born with a CMN, one of the most common skin lesions in newborn infants.² In addition to their surgical challenges and psychosocial sequelae, they can be associated with rare syndromes or transform into melanoma.² They are also rarely associated with other malignancies, including rhabdomyosarcoma, liposarcoma, peripheral nerve sheath tumour and squamous cell carcinoma. We present a case of a basal cell carcinoma (BCC) occurring within a CMN, which has never previously been reported to the best of the authors' knowledge.

CASE PRESENTATION

A woman aged 52 years was referred to our unit with symptomatic ulceration in the centre of a CMN on her right upper back. She was otherwise fit and well, and a non-smoker. She had noticed that it had become ulcerated. Examination revealed a 10×5 cm pigmented lesion. This was a low profile papule, only slightly raised from the skin unlike a raised, sebaceous naevus, in which BCCs are more common.⁴ There was a 3 cm central area of depigmentation, with poorly demarcated edges, firmer in texture than the naevus and displaying surface telangiectasia and ulceration (figures 1 and 2). A core

biopsy performed by the referring team identified it as an infiltrative BCC.

INVESTIGATIONS

See histology results for core and excision biopsy.

TREATMENT

An excision biopsy was performed under a local anaesthetic with primary closure. The histology confirmed a 30 mm nodular BCC within a benign compound naevus, limited to the dermis, with no perineural or lymphovascular invasion (figures 3 and 4). The nearest peripheral margin was 11 mm and deep 8.5 mm. There was an uninvolved lymph node in the subcutaneous fat (figure 5). The wound healed without complication postoperatively.

OUTCOME AND FOLLOW-UP

At 6-month follow-up, there was no evidence of local recurrence and the patient was discharged.

DISCUSSION

CMN patients live with the risk of malignant transformation. Classically, a melanoma,² though other mesenchymal or epithelial-derived malignancies occur, reflecting the cell lines found within CMN.⁵ With a 'medium' sized CMN such as this,² our patient's cited risk of melanoma as she grew up would have been ~4%.² It is surprising then that the malignancy that she developed was a BCC, never previously described within a CMN.

It is known that BCCs and melanocytic naevi are positive predictive factors for each others' incidence,⁶ and that they may occur close to, or 'collide' with each other.⁷ Cases of BCC arising in

Figure 1 Basal cell carcinoma within congenital melanocytic naevi: a 10×5 cm longstanding pigmented low profile papule with a 3 cm central, poorly demarcated area of depigmentation, surface telangiectasia and ulceration.

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Figure 2 Basal cell carcinoma within congenital melanocytic naevi: a 10×5 cm longstanding pigmented low profile papule with a 3 cm central, poorly demarcated area of depigmentation, surface telangiectasia and ulceration.

Figure 3 The histopathology slides, stained with H&E, of the basal cell carcinoma at ×200 magnification.

Figure 4 The histopathology slides, stained with H&E, of the precursor lesion at ×100 magnification.

association with other naevi, such as atypical, sebaceous, basal cell, dysplastic and epidermal naevi of various kinds have failed to reach a consensus on whether these represent two distinct tumours or whether they are inter-related.^{4 8 9} They discuss the potential for ‘crosstalk’ between melanocytes and basal keratinocytes, which are both located in the basal layer of the epidermis.⁸ It is likely that rather than a transformation, as is described in other malignancies such as melanoma, this was a concurrent histopathological process.

How does this relate to clinical practice? We know that within a CMN, there are different cell lines—epidermal and

Figure 5 The histopathology slides, stained with H&E, of the excised lymph node at ×40 magnification.

mesenchymal—of which various malignant and benign transformations have been described. These are usually easy to differentiate histologically, but can be difficult to tease apart clinically.¹⁰ In recognition of this, of the different excision margins required for BCC compared with other secondary skin malignancies in a CMN, and of the size and complexity of reconstruction involved in excising an entire CMN, our recommendation echoes others; that lesions should be excised if they show suspicious features on clinical examination and dermoscopy. Each case should be managed individually, taking into account patient/parent preference and multidisciplinary. Patient/parent education is important and long-term follow-up by plastic surgeons/dermatologists is recommended as per specific lesion guidelines.

Learning points

- ▶ Congenital melanocytic naevi (CMN) are common, and carry a burden of disease, the ‘naevus burden’.
- ▶ The risk of malignant transformation most commonly to melanoma is well known with CMN.
- ▶ Basal cell carcinomas occurring in association with naevi has been documented, but never within a CMN.
- ▶ It is important to manage lesions discretely within the CMN according to histology, in recognition of the surgical challenges in excising the entire CMN.

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Competing interests None declared.

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