

9

Congenital melanocytic naevi

Congenital melanocytic naevi (CMN) are pigmented naevi which are present at birth. The term is usually restricted to naevocellular naevi and excludes other congenital melanocytic naevi such as blue naevi. The diagnosis of CMN is typically a clinical one but there are certain specific histological features seen in most but not all cases. The naevus cells in CMN characteristically involve the lower dermis, infiltrate in lines between collagen bundles and involve the skin appendages.¹ In some cases they extend deep into the subcutaneous fat and may involve fascia or even muscle.

CMN vary from small to giant-sized, covering over half the body surface. There are many and varied criteria for classification of the size of CMN including actual and predicted adult maximum diameter and percentage of body surface area involved by the lesion. The most universally accepted at present is that used by the New York University Registry.^{2,3}

Small: Greatest diameter less than 1.5cm or predicted to reach that size in adult life

Medium: Greatest diameter between 1.5 and 19.9cm or predicted to reach that size in adult life

Large: Greatest diameter 20cm or more, or predicted to reach that size in adult life

9.1 Risk of melanoma in patients with congenital melanocytic naevi

9.1.1 Small

An early histology-based study suggested that small CMN may be precursors for some cases of cutaneous melanoma developing in adult life,⁴ but there have been no satisfactory studies to confirm or quantify the risk of malignancy in these lesions.

9.1.2 Medium

The single prospective study of medium-sized (1.5–19.9cm in diameter) CMN⁵ showed no development of melanoma in 227 subjects with 230 CMN followed for a median of 5.8 years to a median age of 19.1 years. Similarly, in a study of CMN of all sizes, no melanomas were observed (0.18 expected) in those 232 patients with CMN < 20cm in diameter.⁶ However, individual case reports indicate that these lesions can, on rare occasions, be melanoma precursors.⁷ As the few reported cases of melanoma in medium-sized CMN have occurred in adult life, very long-term studies would be required to accurately quantify the risk.

9.1.3 Large

There are many studies which confirm the increased risk of melanoma in individuals with large CMN^{8–11} but the estimated magnitude of the risk varies widely between retrospective and prospective studies, those from academic and non-academic centres, those from small and large series, and those using the NYU and other classifications of size.¹² Other probable confounding factors limiting the veracity of results include the effects of surgical intervention and non-surgical treatments, the fate of cases lost to follow-up

and histological misdiagnosis. Well-conducted prospective studies suggest the risk is in the order of 2–5%,^{3,12,13} but an accurate answer will probably never be obtained.

The highest risk of developing melanoma in these patients is under the age of ten¹⁴ and in particular, under the age of five.¹⁵

Other factors associated with increased risk are giant size, axial location on the trunk, and the presence of multiple satellite naevi beyond the main lesion.¹⁴ These are also risk factors for neurocutaneous melanosis (NCN), the association of CMN with proliferation of melanocytes in the central nervous system (CNS),¹⁶ and all patients with these features should have magnetic resonance imaging investigation. Multiple large scattered lesions are also a risk factor for NCN.¹⁶

Melanoma development in satellite lesions is exceptional but has been reported.¹⁷

It is important to appreciate that melanoma in patients with large CMNs occurs in areas other than the cutaneous naevus in approximately half the cases.¹³ The commonest site of non-cutaneous melanoma in these patients is in the central nervous system. Patients with symptomatic NCN have more than a 50% risk of developing CNS melanoma.¹⁶

Evidence summary	Level	References
The risk of melanoma in patients with small and medium CMN has not been quantified with appropriate studies. Individual case reports, however, suggest that melanoma does occur in medium sized CMN, but rarely and only in adult life	IV	4–7, 12
Patients with large congenital melanocytic naevi (CMN) are at increased risk of development of melanoma both in the naevus and elsewhere. Well-conducted prospective studies suggest the risk is in the order of 2–5%, with most melanomas developing in the first decade of life	III-3	8–11

9.2 Approach to management of patients with congenital melanocytic naevi

Non-surgical techniques such as dermabrasion, laser therapy and chemical peeling may produce some cosmetic improvement but cannot be considered to reduce the risk of melanoma.¹⁸

In CMN, melanoma can occur in the deep dermis or below so in general, excision, if attempted, should be to the depth of the fascia. However even then, because of occasional even deeper involvement, recurrences can occur and the malignancy risk is not completely eradicated.

Because melanoma can occur in patients with large CMNs in sites away from the cutaneous naevus,¹² even complete surgical excision of the CMN cannot be said to have removed the risk of melanoma in patients with large lesions.

With giant lesions which have multiple satellites on the remaining skin, total removal is impracticable. Removal of the thickest and most infiltrated areas may be appropriate¹⁸ and biopsy or removal of any areas showing concerning alteration is mandatory.

Patients with symptomatic neurocutaneous melanosis in whom there is a high mortality rate and a high risk of CNS melanoma¹⁶ should be spared aggressive prophylactic surgery.¹⁸

The authors of a large systematic review⁸ concluded ‘...there is no conclusive evidence in the existing literature that the complete excision of large congenital melanocytic naevi decreases the prevalence of melanoma’.

Several papers have compared the rates of development of melanoma in treated and untreated patients with large CMN respectively.^{3,13,17} Melanoma development occurred more in untreated than treated patients. The findings are rendered very unreliable by the differences in patient selection and the lack of detail about patients subjected or not subjected to surgery respectively. It is likely that the treated lesions were smaller than the untreated lesions and therefore less likely to develop melanoma. Several of the untreated lesions in which melanoma developed were so large that treatment was not feasible.¹⁷

Evidence summary	Level	References
The evidence does not support routine prophylactic removal of small and medium CMNs	IV	3–6, 11, 13–17
In patients with large CMNs, prophylactic surgery may lessen the risk of melanoma in those individuals with more easily removable lesions, which will tend to be ones at the smaller end of the range. There is no evidence that surgery in patients with large (including very large and giant) CMNs reduces the risk of melanoma	IV	7–13, 17

Recommendations

Small and medium congenital melanocytic naevi	Grade
1. Prior to puberty, decisions regarding removal of these lesions be based on cosmetic considerations alone	C
2. Patients and parents be informed that the evidence regarding risk in adult life does not support routine prophylactic removal of these lesions	C
3. Patients report any suspicious changes in these lesions	C
4. Biopsy or removal of any lesions showing suspicious features be undertaken	C
Large congenital melanocytic naevi (more than 20cm in diameter)	
5. Lifetime surveillance be undertaken whether or not any surgery has been performed. This could include baseline photography and three-monthly evaluation for the first year of life, followed by six-monthly evaluation for the next three years, and then yearly evaluation	C
6. Patients and parents report immediately any concerning changes that occur between follow-up visits	C
7. Biopsies be undertaken immediately of any areas which show suspicious features	C

9.3 Good practice points

- All decisions regarding surgical management involve prolonged discussion with the parents, and later the patient, covering estimated risk of melanoma, what is involved in the surgery, the number and length of hospitalisations, possible morbidity of the procedures, and likely end cosmetic result
- MRI of the brain be undertaken in patients with large CMN in an axial distribution and those with multiple large scattered lesion, if the facilities are available. Some features of neurocutaneous melanosis, such as hydrocephalus, are amenable to treatment

References

1. Tannous ZS, Mihm MC, Jr., Sober AJ, Duncan LM. Congenital melanocytic nevi: clinical and histopathologic features, risk of melanoma, and clinical management. *J Am Acad Dermatol* 2005; 52(2):197–203.
2. Kopf AW, Bart RS, Hennessey P. Congenital nevocytic nevi and malignant melanomas. *J Am Acad Dermatol* 1979; 1(2):123–130.
3. Hale EK, Stein J, Ben Porat L, Panageas KS, Eichenbaum MS, Marghoob AA et al. Association of melanoma and neurocutaneous melanocytosis with large congenital melanocytic naevi—results from the NYU-LCMN registry. *Br J Dermatol* 2005; 152(3):512–517.
4. Rhodes AR, Sober AJ, Day CL, Melski JW, Harrist TJ, Mihm MC, Jr. et al. The malignant potential of small congenital nevocellular nevi. An estimate of association based on a histologic study of 234 primary cutaneous melanomas. *J Am Acad Dermatol* 1982; 6(2):230–241.
5. Sahin S, Levin L, Kopf AW, Rao BK, Triola M, Koenig K et al. Risk of melanoma in medium-sized congenital melanocytic nevi: a follow-up study. *J Am Acad Dermatol* 1998; 39(3):428–433.
6. Swerdlow AJ, English JS, Qiao Z. The risk of melanoma in patients with congenital nevi: a cohort study. *J Am Acad Dermatol* 1995; 32(4):595–599.
7. Illig L, Weidner F, Hundeiker M, Gartmann H, Biess B, Leyh F et al. Congenital nevi less than or equal to 10 cm as precursors to melanoma. 52 cases, a review, and a new conception. *Arch Dermatol* 1985; 121(10):1274–1281.
8. Watt AJ, Kotsis SV, Chung KC. Risk of melanoma arising in large congenital melanocytic nevi: a systematic review. *Plast Reconstr Surg* 2004; 113(7):1968–1974.
9. Krengel S, Hauschild A, Schafer T. Melanoma risk in congenital melanocytic naevi: a systematic review. *Br J Dermatol* 2006; 155(1):1–8.
10. Chan YC, Giam YC. A retrospective cohort study of Southeast Asian patients with large congenital melanocytic nevi and the risk of melanoma development. *J Am Acad Dermatol* 2006; 54(5):778–782.
11. Zaal LH, Mooi WJ, Klip H, van der Horst CM. Risk of malignant transformation of congenital melanocytic nevi: a retrospective nationwide study from The Netherlands. *Plast Reconstr Surg* 2005; 116(7):1902–1909.
12. Zaal LH, Mooi WJ, Sillevius Smitt JH, van der Horst CM. Classification of congenital melanocytic naevi and malignant transformation: a review of the literature. *Br J Plast Surg* 2004; 57(8):707–719.
13. Egan CL, Oliveria SA, Elenitsas R, Hanson J, Halpern AC. Cutaneous melanoma risk and phenotypic changes in large congenital nevi: a follow-up study of 46 patients. *J Am Acad Dermatol* 1998; 39(6):923–932.
14. Marghoob AA, Schoenbach SP, Kopf AW. Large congenital melanocytic nevi and the risk of developing malignant melanoma: a prospective study and review of the world literature. *J Invest Dermatol* 1995; 104:563.

15. DeDavid M, Orlow SJ, Provost N, Marghoob AA, Rao BK, Huang CL et al. A study of large congenital melanocytic nevi and associated malignant melanomas: review of cases in the New York University Registry and the world literature. *J Am Acad Dermatol* 1997; 36(3 Pt 1):409–416.
16. DeDavid M, Orlow SJ, Provost N, Marghoob AA, Rao BK, Wasti Q et al. Neurocutaneous melanosis: clinical features of large congenital melanocytic nevi in patients with manifest central nervous system melanosis. *J Am Acad Dermatol* 1996; 35(4):529–538.
17. Bett BJ. Large or multiple congenital melanocytic nevi: occurrence of cutaneous melanoma in 1008 persons. *J Am Acad Dermatol* 2005; 52(5):793–797.
18. Marghoob AA, Borrego JP, Halpern AC. Congenital melanocytic nevi: treatment modalities and management options. *Semin Cutan Med Surg* 2003; 22(1):21–32.