

Case Letters

Lethal melanoma in a two-year-old child with multiple congenital melanocytic nevi

Sir,

Pediatric melanoma is rare, accounting for only 1–4% of all cases of melanoma and 1–3% of all pediatric malignancies.¹ Here we describe a rare case of malignant melanoma in a two-year-old boy with multiple congenital melanocytic nevi. This case of melanoma showed rapid progression, metastasis and eventual death.

A 2-year-old boy presented with multiple brown and black lesions on the back noticed since birth. It was noted that all lesions were flat and did not show changes until 10 months of age. This child was born out of a non-consanguineous marriage by normal vaginal delivery at term. There was no history of trauma or infection. There was no family history of melanoma or other malignancies. Examination revealed multiple hyperpigmented plaques of size varying from 0.5 cm to 3 cm. The plaques were mainly distributed on the back but some smaller ones were also found on the subcostal region of right flank. In total, lesions and satellites covered approximately 6% of body surface area. Face, chest, abdomen and limbs were spared. One of the plaques on the back was studded with a nodule of 5 mm × 5 mm size [Figure 1a].

Dermoscopy of the nodule on the back revealed incomplete pigmentation, blue-and-white veil [Figure 2a, c and e], atypical dots and globules [Figure 2b] and atypical reticular formations and striae [Figure 2d and f], showing features of malignant melanoma.

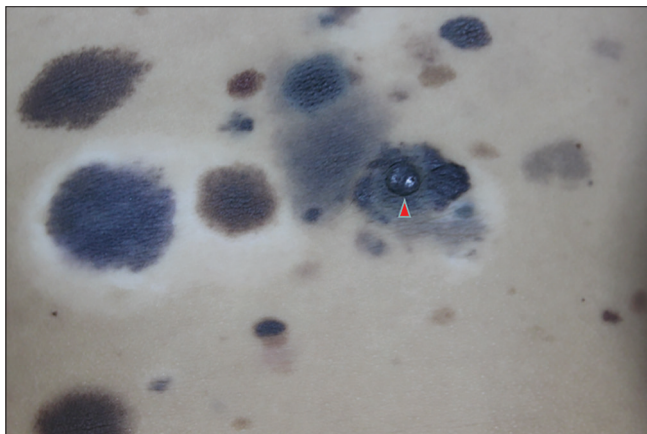


Figure 1a: Multiple brown and black plaques and a nodule of 5mm × 5 mm size on the back (the red triangle)

Parents did not consent for a skin biopsy. Follow-up visits were advised every 3 months so that the lesions could be monitored through dermoscopy.

Nine months later, parents agreed for a skin biopsy. Biopsy was done from the nodule arising from a hyperpigmented plaque on the back. Histopathologically, the tumor was composed of atypical melanocytes infiltrating the entire dermis extensively without any epidermal involvement [Figure 3a]. It was shown that the melanocytes had nuclear hyperchromasia. Mitotic figures could also be observed [Figure 3b]. Two mitotic figures were observed per 10 high power regions. The thickness of the tumor (Breslow) was 7 mm, the Clark level was IV and the tumor stage was evaluated as IIIB. The immunohistochemical examination revealed that atypical melanocytes were positive for S-100 [Figure 3c] and HMB-45 [Figure 3d]. The Ki-67 labelling index in dermal atypical melanocytes was 20–30% [Figure 3e]. Both HMB45 and Ki-67 positivity of melanocytes in deep dermis were highly suspicious of malignant transformation.

After 4 months the child presented with a 3 cm × 3 cm tumor on the right axilla. Lymph node metastasis was clinically suspected [Figure 1b]. Histopathological examination revealed atypical metastatic melanocytes with deep staining of the nucleus and melanin extensively infiltrating lymph node parenchyma [Figure 4a and b]. Immunohistochemical examination showed positive staining for S-100 and



Figure 1b: The enlarged lymph node of 3 cm × 3 cm size in the right axilla



Figure 2a: Uneven pigmentation (*blue circle*) and blue-white veil (*red star*) on dermoscopy



Figure 2b: Atypical dots and globules (*white arrows*) on dermoscopy

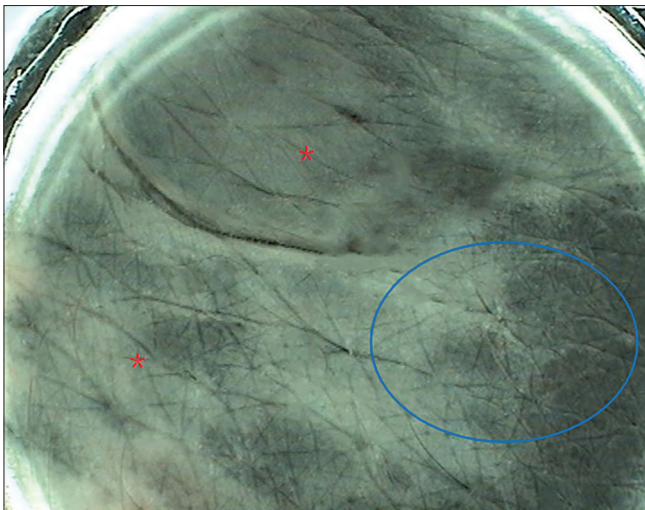


Figure 2c: Uneven pigmentation (*blue circle*) and blue-white veil (*red star*) on dermoscopy

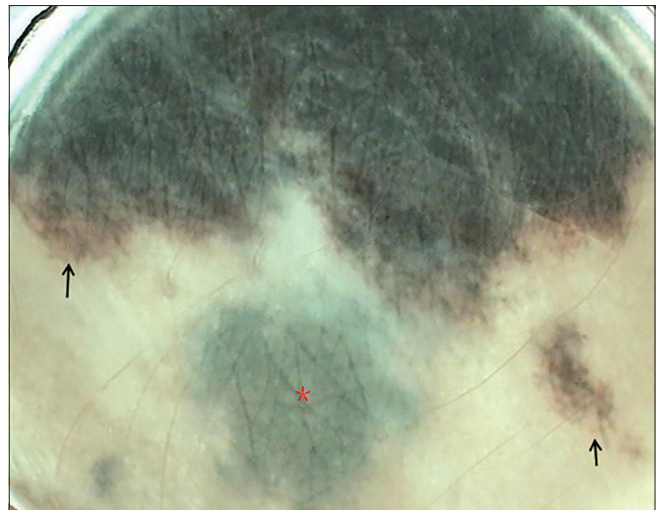


Figure 2d: Atypical network and streaks (*black arrows*) and blue-white veil (*red star*) on dermoscopy



Figure 2e: Uneven pigmentation (*blue circle*) and blue-white veil (*red star*) on dermoscopy

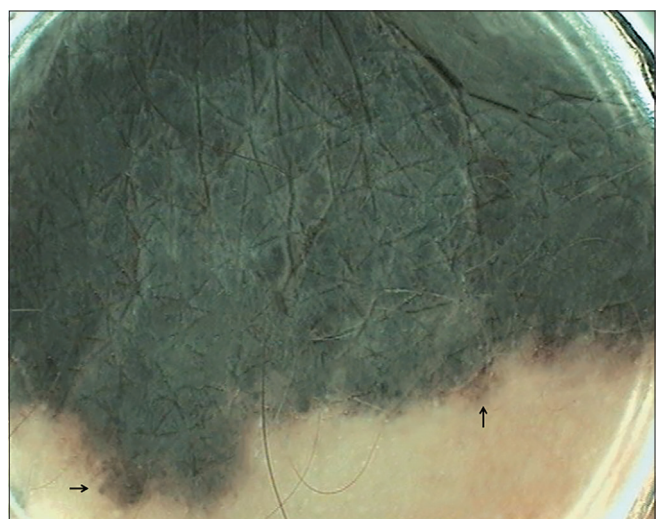


Figure 2f: Atypical network and streaks (*black arrows*) on dermoscopy

HMB45 [Figure 4c and d]. This case was diagnosed as malignant melanoma arising from a congenital melanocytic nevus with lymph node metastasis.

The child did not receive any computed tomography scans or treatment due to financial constraint. The child died a year after the initial diagnosis. It was informed that the boy was in a coma during the last months of his life. Therefore, it is speculated that he might have developed

widespread metastases involving the brain as well. Brain metastasis often occurs when a tumor is located near the spine.

The biggest challenge in diagnosis is to distinguish between melanomas arising from congenital melanocytic nevus and proliferating nodules that develop in congenital melanocytic nevus. There are several features that distinguish between proliferating nodule and melanoma: (1) lack of highly uniform cell atypicality; (2) lack of necrosis within the nodule; (3) rarity of mitosis; (4) evidence of maturation of cells in a mixed or transitional form; (5) lack of spread into the epidermis and (6) absence of destructive bloated growth.¹

It is generally accepted that a complete excision in early childhood decreases the risk of malignancy². Another option is a close monitoring by physical examination, including lymph node palpation and dermoscopy, with periodic follow-up and prompt excision if the lesion presents variations.

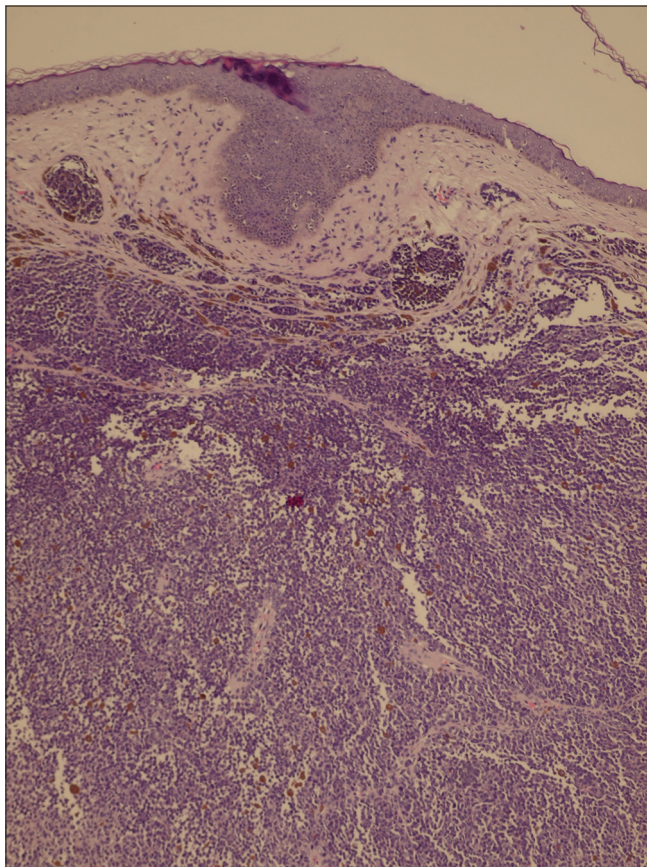


Figure 3a: Nodule on the back composed of atypical melanocytes widely infiltrating the entire dermis with no epidermal involvement (hematoxylin and eosin, ×400)

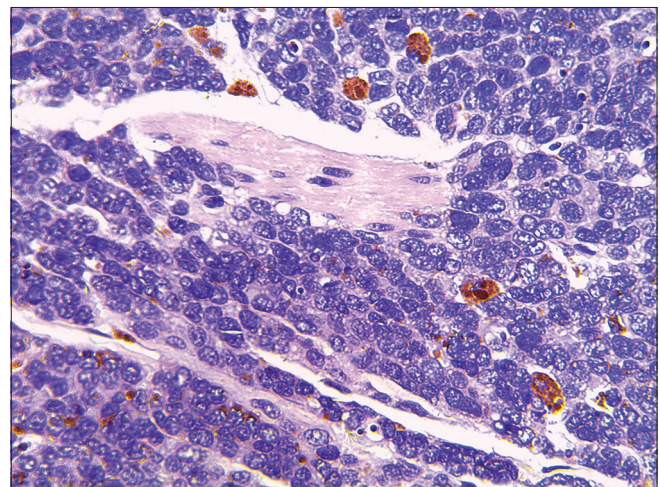


Figure 3b: Nodule on the back showing melanoma cells with nuclear hyperchromasia and mitotic figures (hematoxylin and eosin, ×400)

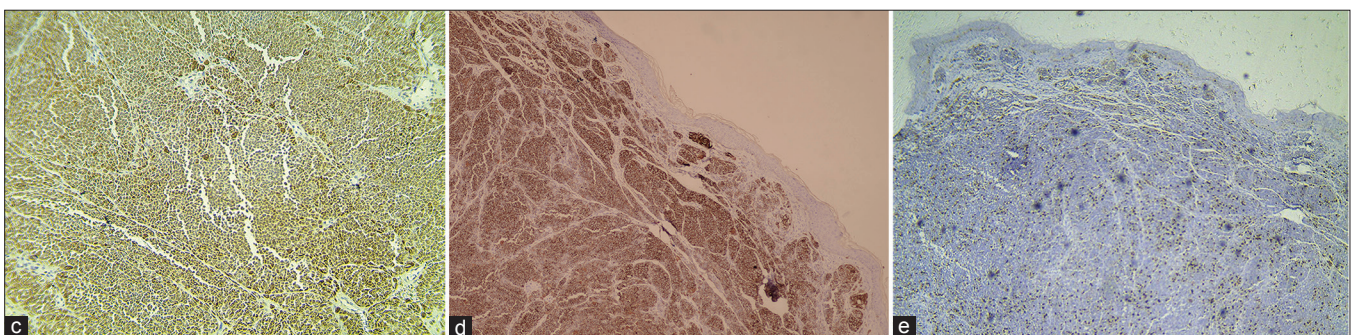


Figure 3 (c-e): Nodule on the back showing (c) S-100 stain positivity (S-100, ×100) and (d) HMB45 stain positivity (HMB-45, ×100). (e) Ki-67 labelling index in dermal atypical melanocytes was 20–30% (Ki-67, ×400).

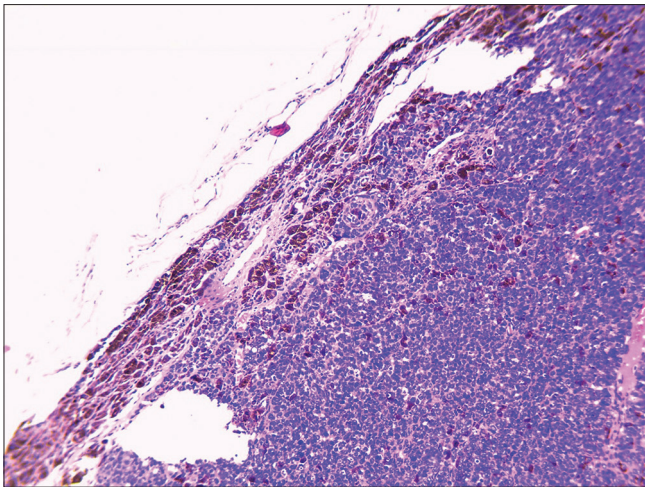


Figure 4a: Atypical metastatic melanocytes widely infiltrating in the parenchyma of lymph node (hematoxylin and eosin, $\times 100$)

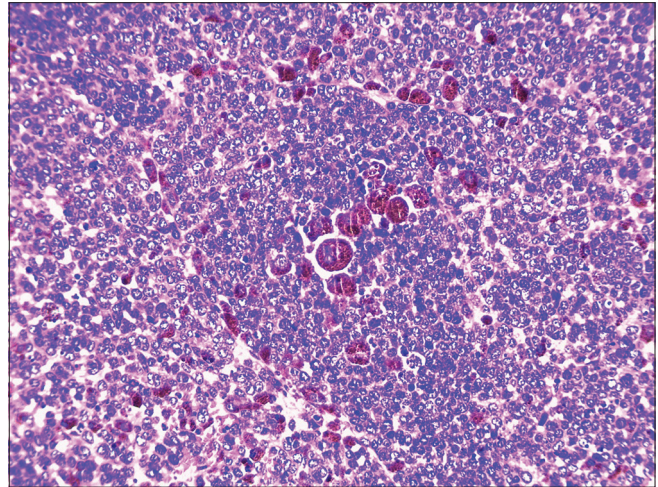


Figure 4b: Lymph node with irregularly sized and shaped atypical melanocytes with large deeply stained nuclei (hematoxylin and eosin, $\times 200$)

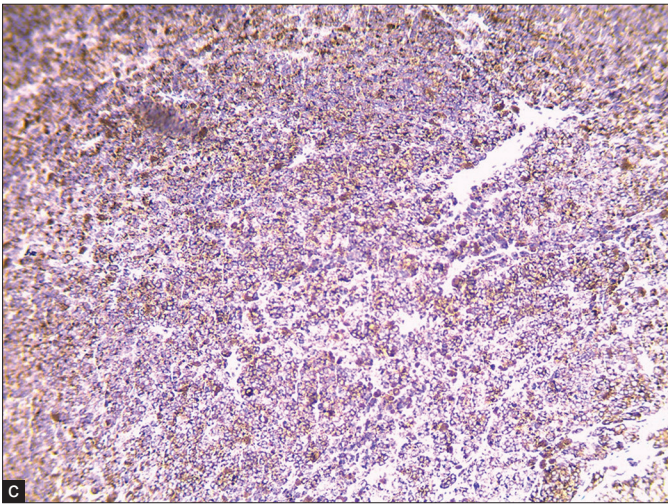
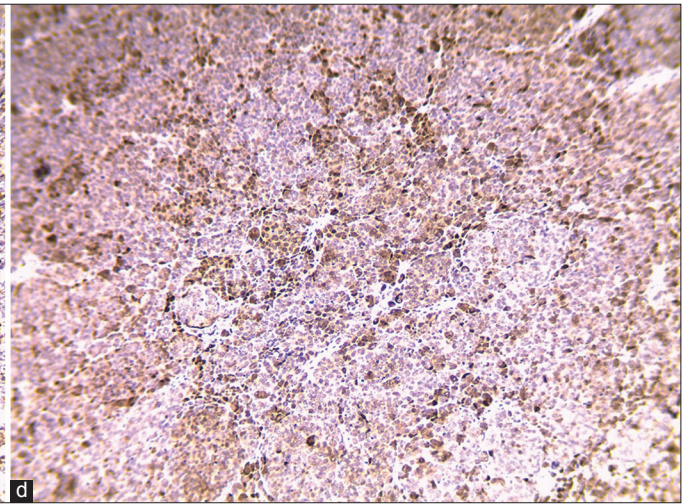


Figure 4 (c-d): (c) Lymph node showing S-100 stain positivity (S-100, $\times 100$).



(d) Lymph node showing HMB45 stain positivity (HMB-45, $\times 100$)

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the parents have given their consent for images and other clinical information to be reported in the journal. The parents understand that names and initials will not be published and due efforts will be made to conceal patient identity, but anonymity cannot be guaranteed.

We have obtained all appropriate patient consent forms.

Financial support and sponsorship

Shandong Province Science and Technology Development Project (2013GSF11835).

Conflicts of interest

There are no conflicts of interest.

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Access this article online	
Quick Response Code:	Website: www.ijdv1.com
	DOI: 10.4103/ijdv1.IJDVL_938_17

How to cite this article: Gao F, Niu G, Zhang B, Xin L. Lethal melanoma in a two-year-old child with multiple congenital melanocytic nevi. *Indian J Dermatol Venereol Leprol* 2019;85:89-93.

Received: February, 2018. **Accepted:** August, 2018.

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