

Junctional Anomalies in Congenital and Acquired Nevi in the First Years of Life

7.1 Introduction

Although the vast majority of prepubertal nevi has no special histological findings, a few of them show important architectural and cytological anomalies at the dermoepidermal junction. These lesions are entirely benign in their clinical evolution, but their histological features often resemble melanoma in situ and an undue diagnosis of melanoma is frequently made.

Similar features can also be found in nevi of adolescents, chiefly nevi found on special sites such as the scalp or genitalia.

7.2 Clinical Features

There are two situations in which these unusual junctional features can be found: congenital nevi and acquired “early onset” nevi of childhood.

In congenital nevi the lesion appears after birth as a flat and blackish spot large up to 1 cm in diameter, in the context of a medium-sized or giant congenital nevus; in time, lesions can rise above the skin surface. Occasionally, these dark macules or papules occur in combination with large neoplastic nodules or plaques deeply situated in the intradermal portion of a congenital nevus. The acquired “early onset” nevi are very dark small lesions which appear in the first years of life and progressively enlarge.

The congenital or acquired nevi excised during the first years of life with junctional pseudomelanomatous features are about 7% (Barnhill 1998) of all pigmented lesions, in this age range, that end up being examined histopathologically.

7.3 Histological Features

Both in congenital and acquired lesions, features are similar to a melanoma in situ (Fig. 7.1) or superficially invasive melanoma (Fig. 7.2), either de novo or in combination with a preexistent compound nevus, namely:

- The junction is colonized by large irregularly shaped me-

lanocytic nests, occasionally confluent with each other; the nests vary largely in size and shape; this involvement also affects the adnexal structures.

- Nests are large and occupy large parts of the epidermis, frequently reaching the granular or even spanning the entire epidermis to reach the cornified layer.
- Occasionally an obvious “dyshesive” pattern is present, with melanocytes inside the nests separated from each other.
- Pagetoid spread of single melanocytes above the junction is a frequent and occasionally striking finding (Fig. 7.3).
- At times a lentiginous pattern is prevalent.
- Cells can be atypical or even bizarre (Fig. 7.4); melanocytes with elongated dendrites and hyperchromatic nuclei are frequent in acquired lesions, also epithelioid cells with large dusty pigmented cytoplasm can be found; mitoses, although rare, are also possible.
- In the papillary dermis, a few melanophages and reactive granulation tissue are an almost constant finding in the acquired lesions.

The above-mentioned atypical findings (Mooi and Krausz 1990; Farmer and Hood 1990; Le Boit 1994) are, as a rule, restricted to melanocytes in the epidermis. In the acquired lesions, however, architectural and cytological atypia are occasionally present also in the papillary dermis, a detail which accentuates the similarity of these lesions with melanoma (Mihm and Googe 1990).

7.4 Differential Diagnosis

From a morphological point of view, the lesions described herein strongly resemble melanoma in situ, either de novo or one that developed in a congenital or early acquired nevus.

The correct diagnosis of a benign lesion is based in part on the young age of the patient and on the clinical history. These considerations notwithstanding, there are some objective histopathologic differences from melanoma arising in congenital nevi:

- In these nevi with junctional alterations, despite striking

architectural disorder, the nuclei of the cells at the junction are small and round and monomorphous. They tend to have very small nucleoli. The pagetoid spread of single melanocytes occurs mostly above (or combined with) well-formed junctional nests. The cells have a characteristic clear cytoplasm with dusty granules of melanin similar to that seen in the balloon cells found in balloon cell nevus of adults.

- In these lesions a dense inflammatory infiltrate in the papillary dermis is usually absent.
- The deep portion of the nevus shows an entirely innocent-looking benign morphology. As a rule, a gradual transition from atypical cells with superficially abundant pale cytoplasm to small round melanocytes with scant cytoplasm (characteristic of congenital nevus at deeper levels) is present; in general, the two populations are not separated by lymphocytes or by fibrosis.
- Mitoses are possible within the larger melanocytes but are rare.
- In benign lesions the junctional atypical component does not involve the epidermis beyond the dermal component of similar cells, let alone extend beyond the boundaries of the overall dermal component; on the contrary, in melanoma arising in a nevus, a junctional component and pagetoid spread are found also to the side of the nevus and in the perilesional epidermis; moreover, benign lesions are sharply circumscribed, whereas melanomas are not.
- From a clinical point of view, simulants of melanoma arising in congenital nevi are dark areas within a larger lesion; the atypical component does not overly a normal dermis.
- De novo melanomas of infancy are much deeper than their benign simulators, and a large intradermal component is usually present (almost all the metastasizing de novo melanomas reported in literature in this age range reached the deep dermis or subcutis); on the contrary, in benign lesions the atypical melanocytes or irregular nests never go beyond the papillary dermis.
- As said above, de novo melanomas in prepubertal patients are exceedingly rare and are utterly exceptional before 10 years of age.

In sum, when facing an infantile melanocytic lesion with a junctional “melanomatous proliferation,” a diagnosis of nevus should be made if:

- The lesion is small or the phenomena occur in a congenital nevus.
- The atypical features involve only the junction and the papillary dermis.
- Nuclei are small and monomorphous.

7.5 Further Reading

- Barnhill RL (1998) Childhood melanoma. *Semin Diagn Pathol* 15(3):189–194
- Farmer ER, Hood AF (1990) *Pathology of the skin*. Appleton & Lange, London, p 730
- LeBoit PE (ed) (1994) *Malignant melanoma and melanocytic neoplasms*. Hanley & Belfus, Philadelphia, p 217–219
- Mihm MC, Googe PB (1990) *Problematic pigmented lesions*. Lea & Febiger, Philadelphia, p 140–149
- Mooi WJ, Krausz T (1990) *Biopsy pathology of melanocytic disorders*. Chapman & Hall, London, p 139–142

7.6 Summary

- Atypical junctional melanocytic proliferation can occur in congenital nevi and in nevi that appear in the first years of life.
- Although such features of these lesions can be strikingly similar to those of melanoma, they do not imply the malignant nature of the lesion nor its proclivity to evolve in melanoma.
- Histologically, hints of the true benign nature of the lesions are the absence of pleomorphism and cellular necrosis and the presence of an utterly benign melanocytic nevus deeper in the dermis. The atypical component is restricted into the epidermis and the papillary dermis.

Fig. 7.1
Congenital nevus with atypical junctional proliferation

This large congenital nevus has been illustrated in Chap. 8 as well (Fig. 8.2) because of its large nodule situated in the deep part of the lesion (a).

A striking pagetoid spread of melanocytes above the junction is present in the epidermis (b). This spread is mostly made up of nests, but single melanocytes are also present (c).

Adequate clinical information is mandatory in cases like this, but a few details indicate that this phenomenon is not a sign of melanoma.

First of all, the very presence of large nests above the junction, without many single cells, is typical of the pagetoid spread we see in nevi (like the Spitz nevus) and is very rarely found in melanoma; moreover, melanocytes are monomorphous, nuclei are small and typical, and finally, necrotic melanocytes are not found

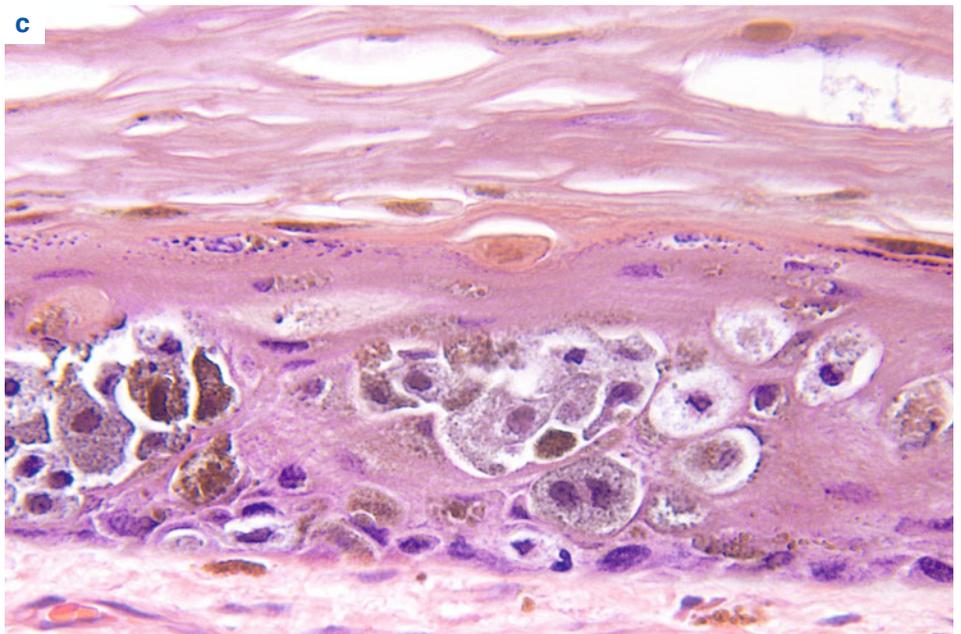
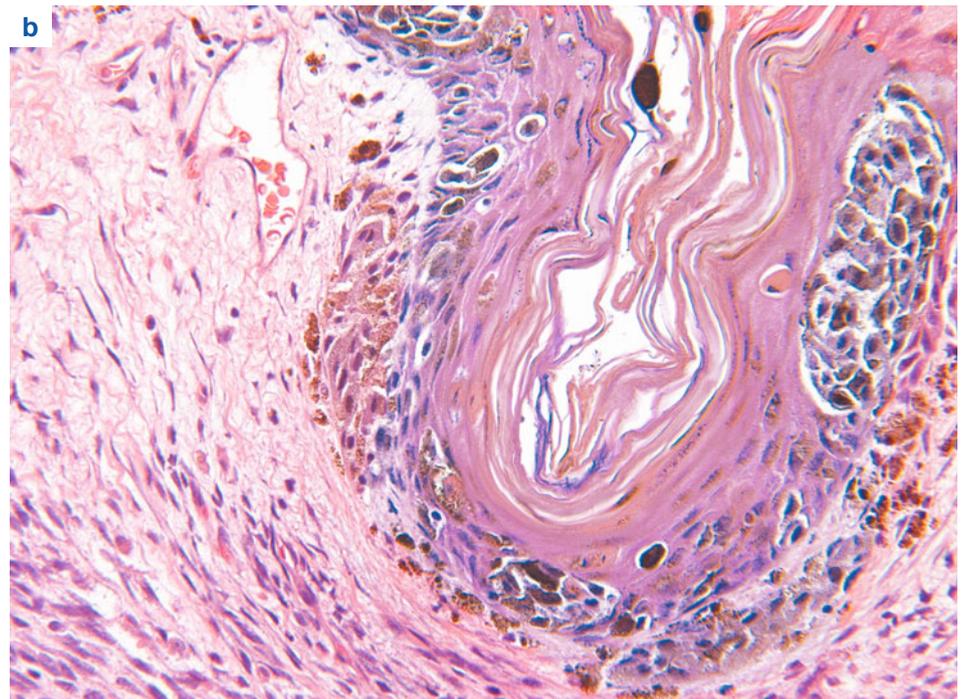
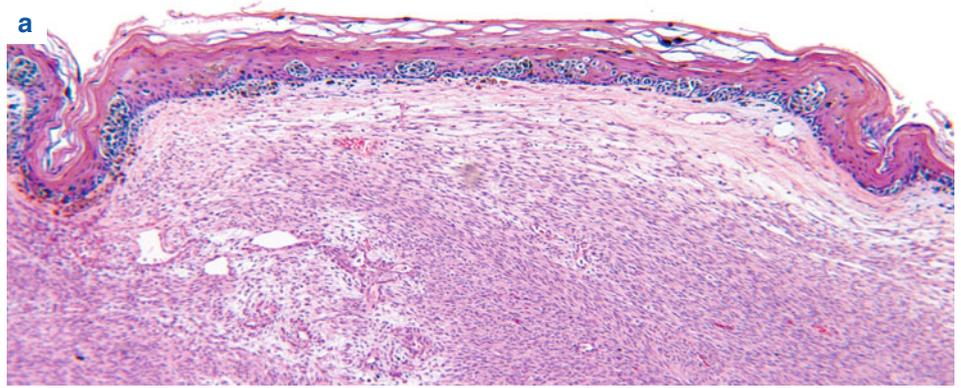


Fig. 7.2
Congenital nevus with atypical junctional proliferation

This lesion is a congenital nevus as can be inferred by its very depth. In fact, melanocytes occupy the entire span of the dermis (a) and the subcutaneous fibrous septa.

A junctional proliferation of melanocytes at the dermoepidermal border and in the adnexa is evident in this congenital lesion and a diagnosis of an evolving melanoma in situ must be taken into consideration (b).

The nuclei are small and atypical appearing (c) but monomorphic.

A strong piece of evidence that the lesion is benign is that the large, pale melanocytes in the papillary dermis merge with smaller ones more typical of a congenital nevus deeper in the lesion. A pagetoid pattern of melanoma is rare in infants and children even among the rare melanomas of that age group

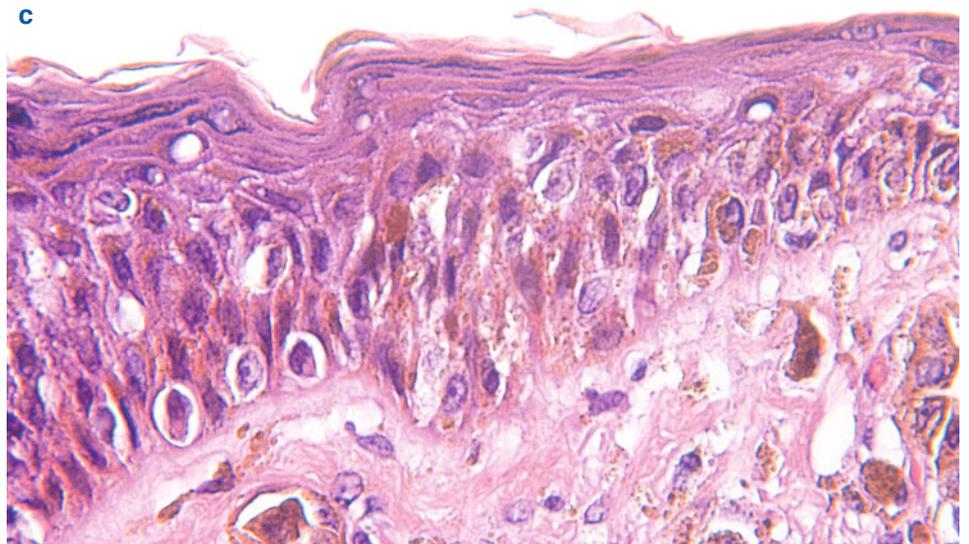
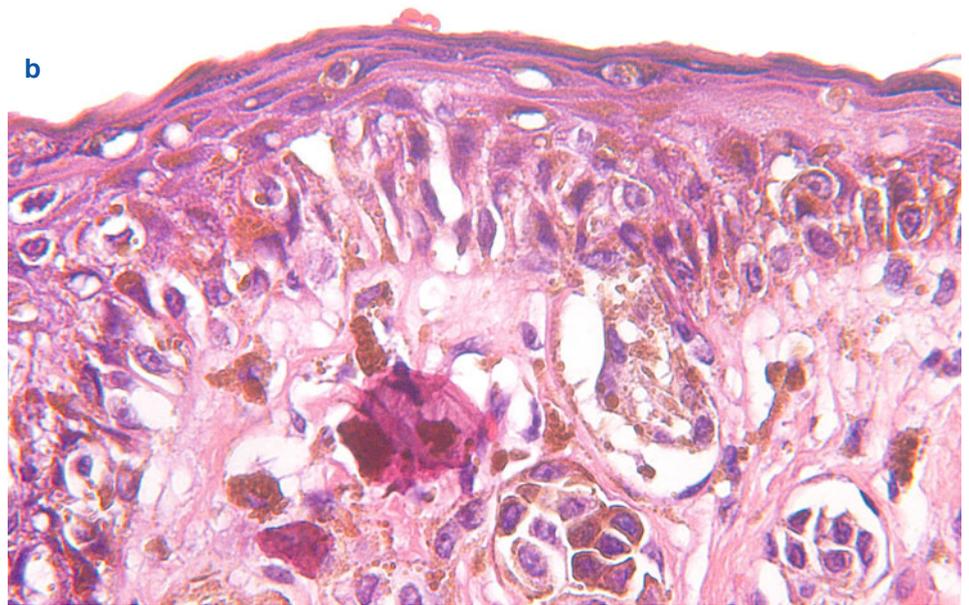
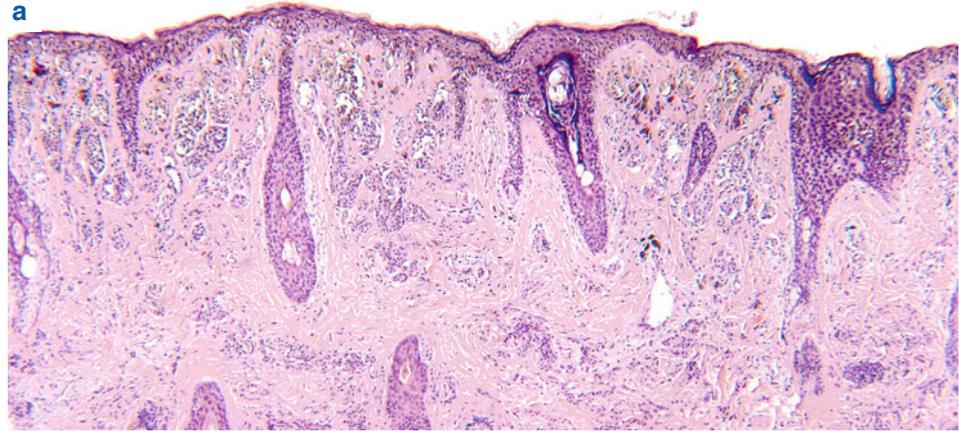


Fig. 7.3
Congenital nevus with
atypical junctional proliferation

The patient was a neonate, but the congenital nature of the lesion can be inferred also from the adnexotropism of melanocytes as can be seen in the last picture.

At the junction melanocytes are disorderly scattered in single units or in irregular confluent nests (a, b).

The pagetoid spread is mostly in the lower part of the epidermis, principally in rete ridges and does not go above the line connecting the tops of dermal papillae.

Figure (c) illustrates the adnexotropism

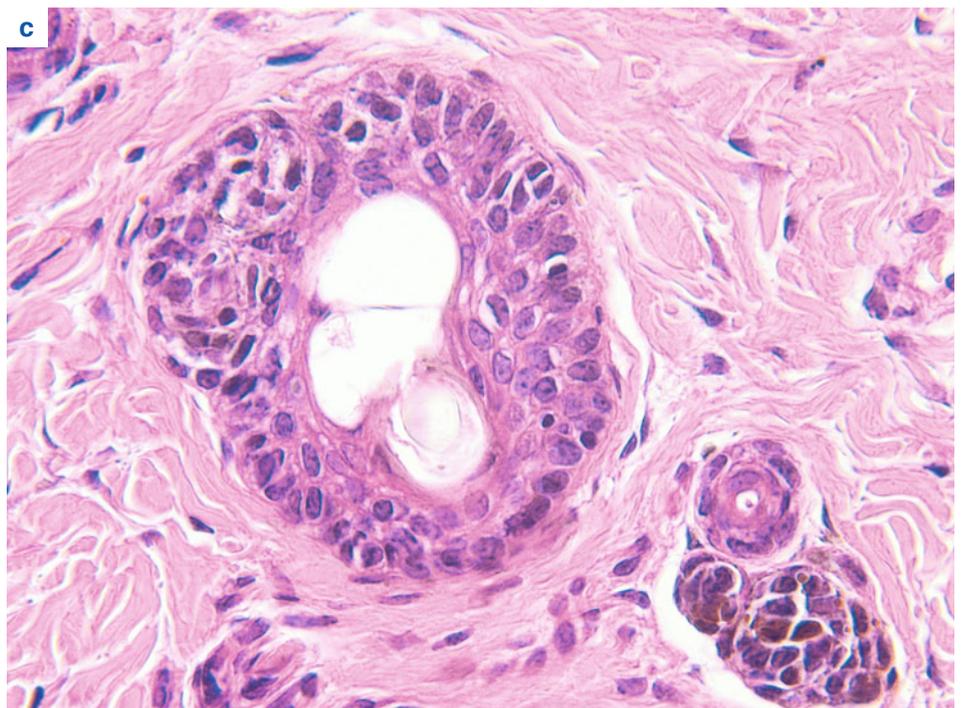
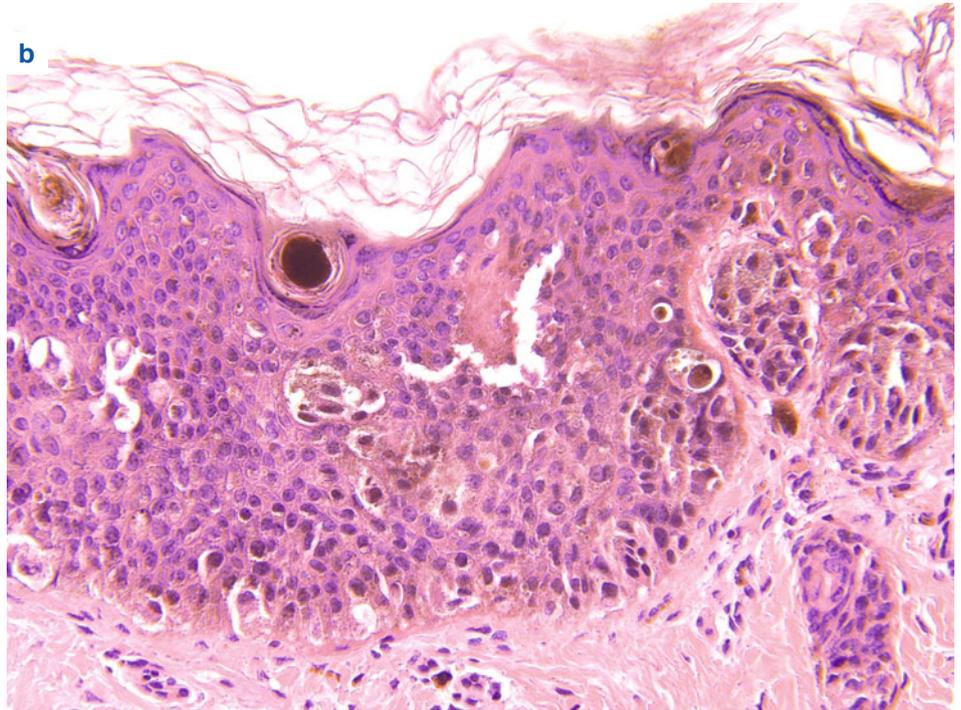
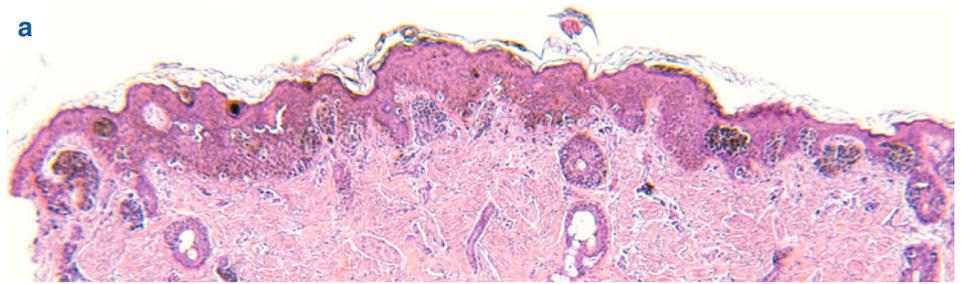


Fig. 7.4
Congenital nevus
with atypical junctional
proliferation (vs. melanoma
in situ in congenital nevus)

This lesion was excised from a 21-year-old male. The features in the dermis are those of a congenital nevus or of an acquired nevus with congenital features (a). The lesion is deep. Cells at the bottom are aligned in rows between collagen fibers and a perifollicular growth pattern is evident.

In this nevus however the junctional proliferation is striking (b) and a form of melanoma developed in a nevus must be carefully considered (c). Details indicating a benign nature of this finding are:

- Cells at the junction are most gathered in nests.
- Cytology is bland and cells are similar to the innocent intradermal component.
- Rete ridges are not effaced.

In cases like this, in postpubertal youngsters, the lesion must be entirely excised and a note of caution should be inserted in the pathological report

