Congenital melanocytic naevi (CMNs) are divided into giant, medium-sized, and small types. Giant CMN measures more than 20 cm in maximal diameter, whereas small CMN measures less than 1.5 cm (1). The lifetime incidence of melanoma in giant CMN is estimated to be between 6.3% (2) and 12% (1). Since the prognosis is poor (3), it is generally agreed that it would be desirable for giant CMN to be excised whenever possible. Melanomas arising from giant CMN are apt to occur before puberty and are occasionally of dermal origin (4).

Although the incidence of melanoma in medium-sized CMN is probably greater than that in a comparable area of normal skin, it is supposed that melanomas rarely arise from small CMN (4, 5). Some authors insist that the risk is related to lesion size (6), while other investigators disagree (4). The excision of small and medium-sized CMNs when feasible is advised by many authors (7, 8), although not all (1, 5). Melanomas arising from small and medium-sized CMNs are apt to occur after puberty, to be located at the margin of the naevi, and to be exclusively of epidermal origin (4).

CASE REPORT

A 17-year-old woman presented with a dome-shaped, brown-grey nodule measuring 3 × 3 mm around the centre of an oval black plaque measuring 13 × 10 mm on the left abdomen (Fig. 1). A black macule had been present at that site since birth. The nodule had developed approximately one year before presentation. Our initial diagnosis was melanoma arising from a small CMN.

Histopathologically, a scanning view demonstrated a dome-shaped nodule around the centre of the plaque (Fig. 2A). The plaque was composed of epithelioid and spindle cell nests in the lower epidermis, accompanied by regular elongation of rete ridges. The nodule was composed of epithelioid cells in the upper dermis and spindle cells in the deeper dermis without junctional components (Fig. 2B). There was apparent maturation. The spindle cells extended between collagen bundles singly or in double rows. Above these cells, there were reticular elongation of thin rete ridges and a subepidermal clear zone. Throughout the lesion, single melanocytes with melanin granules were increased in the basal layer. There was no nuclear atypia in either epithelioid or spindle cells of either the plaque or nodule. Since these cells were considered naevus cells, the above findings corresponded to the so-called “naevus in naevus.”

Immunohistochemically, naevus cells of both the plaque and nodule were positive for S-100 protein.
and negative for HMB45. Some (>30%) of the naevus cells in the nodule were positive for proliferating cell nuclear antigen, whereas a majority of naevus cells in the plaque were negative.

DISCUSSION

Several types of secondary melanocytic neoplasms can develop in CMNs, many of which are thought to be distinct from melanomas and are termed “proliferative nodules” (PN) (9–12). Although PN may be clinically worrisome, occasionally showing rapid growth and ulceration, these lesions tend to stabilize and regress (9–12). The benign nature of PNs has been demonstrated by techniques such as molecular analyses (13–15). PNs arise exclusively in relatively large CMNs during infancy (10–15). Even in the one exceptional case report describing the development of PN in a small CMN, the patient was a 3-month-old infant (15). To our knowledge, there have not been any previous case reports of PNs involving small CMNs after childhood. Although PNs have exclusively been reported in large CMNs during infancy, similar nodules can develop in small CMNs after childhood. While the former bears a close resemblance to melanoma, the latter may not do so.

REFERENCES