

SHORT COMMUNICATION

Pigmented Dermatofibrosarcoma Protuberans and Blue Naevi with Similar Dermoscopy: A Case Report

Yuko Ehara¹, Yuichi Yoshida¹, Tatsushi Shiomi² and Osamu Yamamoto¹

¹Division of Dermatology, Department of Medicine of Sensory and Motor Organs, and ²Organ Pathology, Faculty of Medicine, Tottori University, 86 Nishicho, Yonago-shi, Tottori 683-8503, Japan. E-mail: yukohara@med.tottori-u.ac.jp

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Pigmented dermatofibrosarcoma protuberans (Bednar tumour) is a rare neoplasm that accounts for 1–5% of all cases of dermatofibrosarcoma protuberans (DFSP) (1, 2). We report here a case of Bednar tumour and blue naevi with similar dermoscopy.

CASE REPORT

A 71-year-old Japanese woman was referred to our department for evaluation of 3 pigmented lesions on her left buttock. She had been aware of the lesions for a few months. Physical examination revealed 3 blue nodules, approximately 7, 8 and 4 mm in diameter, located on the left buttock (Fig. 1a). Dermoscopic examination revealed a homogeneous blue pigmentation with whitish-veil structures in each of the 3 lesions (Fig. 1b). However, the colour of the upper lesion was paler than that of the other lesions. Based on these findings, we suspected that these were blue naevi. The middle lesion was resected under local anaesthesia. Histopathological examination revealed diffuse proliferation of spindle cells containing melanin granules in the lower dermis (Fig. 1c). A diagnosis of cellular blue naevus was made. The remaining 2 lesions were

not resected, and a wait-and-see approach was taken.

After 2 years, the patient became concerned about the remaining 2 lesions, and returned to our department for treatment. The 2 blue nodules were slightly increased in size on the left upper and lower buttocks (Fig. 1a¹). There was no change in dermoscopic features compared with those seen 2 years previously (Fig. 1b¹). The 2 lesions were resected under local anaesthesia. The diagnosis of the lower small lesion was blue naevus. Histopathologically, the other lesion on the upper buttock showed massive proliferation of atypical spindle-shaped cells arranged in a storiform pattern from the lower dermis to the subcutaneous adipose tissue. In addition, there were scattered dendritic cells containing granular melanin pigments (Fig. 1c¹, 1d¹). Immunohistochemically, the spindle tumour cells were positive for CD34 (Fig. 1e¹), but negative for S-100 protein, while dendritic cells were only positive for S-100 protein (not shown). A diagnosis of Bednar tumour (pigmented dermatofibrosarcoma) was made. An additional extensive resection with a 3-cm tumour

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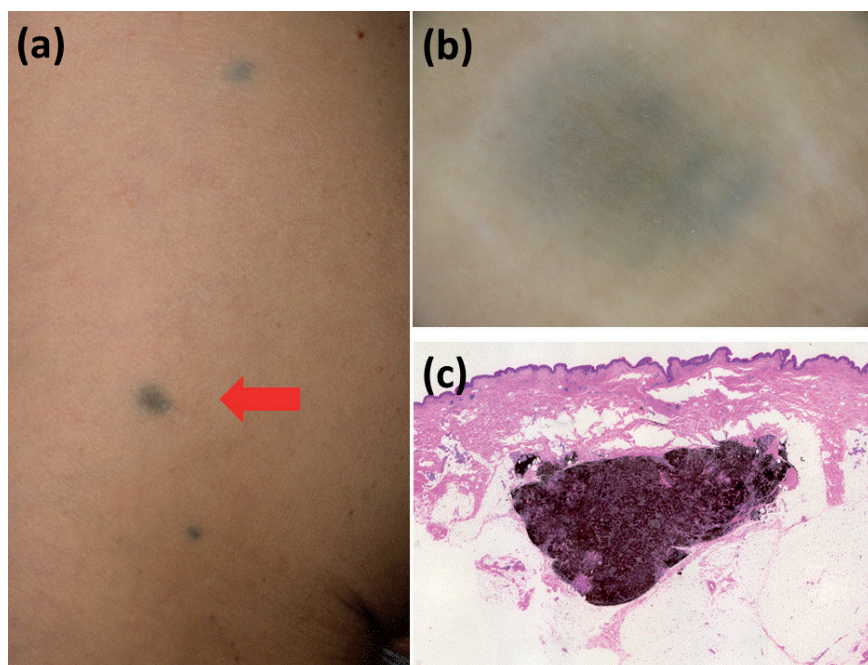


Fig. 1. (a) Three bluish pigmented nodules on the left buttock. (b) These lesions showed homogeneous black-bluish pigmentation and blue-whitish veil-like structures by dermoscopy (red arrow). (c) Diffuse spindle cell proliferation with melanin pigmentation was seen in the lower dermis. (haematoxylin and eosin; H&E $\times 20$).

margin was performed. After the operation, there has been no recurrence in 8 months of follow-up.

DISCUSSION

In 1957, Bednar (3) first described a peculiar tumour as “pigmented storiform neurofibroma” characterized histopathologically by a storiform pattern and the presence of melanin-containing cells. Further studies demonstrated that the non-pigmented portion of this tumour was identical to conventional DFSP, and it was proposed that this tumour be named Bednar tumour or pigmented DFSP (4). Goncharuk et al. (5) reported that Bednar tumour represented one part of a spectrum of neuroectodermal tumours including dermal melanocytosis, cellular blue naevus and conventional DFSP. However, the origin of these tumour cells has not yet been clarified and remains controversial.

There are various reports of Bednar tumour’s macroscopic colour: grey-white (6), brownish red (7), and blue or bluish-black (5, 8). However, there have been only a few reports of the detailed dermoscopic features of DFSP and Bednar tumours. Bernard et al. (9) reported 6 dermoscopic features: delicate pigment network (87%), vessels (80%), structureless light-brown areas (73%), shiny white streaks (67%), pink background coloration (67%) and structureless hypopigmented or depigmented areas (60%). These findings were often associated with a multicomponent pattern. Arborizing vessels were also seen in case of DFSP including one Bednar tumour (9). With regard to a blue naevus on hairy skin, structureless homogeneous pigmentation, which is often described as steel-blue coloration with possible white veils, is usually seen by dermoscopy (10).

In our case, dermoscopic observation showed homogeneous black-bluish pigmentation with whitish-veil structures and no vessels. We considered that the homogeneous pigmentation corresponded to a diffuse distribution of melanin-laden spindle cells in the lower dermis to the subcutaneous tissue. In addition, the whitish-veil structures were derived from the hyperplasia of collagen in the upper dermis. Since the tumour was located in deeper tissue, we only recognized paler pigmentation and no vessels in our Bednar tumour’s dermoscopy features.

To our knowledge, there has been no report of the concurrent occurrence of blue naevi and a Bednar tumour in same region.

Based on our findings, we consider that it is difficult to distinguish a Bednar tumour from a blue naevus by dermoscopy. We emphasize the importance of clinicians including Bednar tumours in the differential diagnosis when homogeneous black-bluish pigmentation with whitish-veil structures are observed by dermoscopy; a prompt biopsy should be performed for correct diagnosis.

The authors declare no conflicts of interest.

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