A Case of Pigmented Clear Cell Acanthoma

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Pigmented clear cell acanthoma (CCA) is considered to be a variant of CCA and a pigmented counterpart to CCA. Pigmented CCA is characterized by macroscopically visible brown pigmentation, easily observed dermal melanophages on low-power magnification and an increased number of melanocytes and melanin in the epidermis. We report a case of pigmented CCA which is a rare variant of CCA. (Ann Dermatol 16(3) 105~108, 2004)

Key Words: Pigmented clear cell acanthoma

Clear cell acanthoma (CCA) is a quite distinctive neoplasm first described in 1962[1]. Typically, CCA presents as a flat or dome-shaped, sharply demarcated papule or nodule, sometimes with keratotic scale and bright red or pink colour. Most of these lesions are macroscopically not pigmented[2,3]. The diagnosis of CCA can easily be made by distinct histopathological features which have been described in detail[2-5]. Pigmented clear cell acanthoma was first proposed in 1994. The term "pigmented clear cell acanthoma" was proposed for CCA with macroscopically visible brown pigmentation and an increased number of melanocytes and melanin at the light-microscopic level[6]. We report a case of pigmented CCA, which is macroscopically and histologically pigmented variant of CCA.

CASE REPORT

The patient is a 60-year-old Korean man, who has more than a 10-year history of dark brownish pruritic papules on the left upper chest, which bleed the intermittently (Fig. 1). The first clinical diagnosis was pigmented seborrheic keratosis versus pigmented basal cell carcinoma.

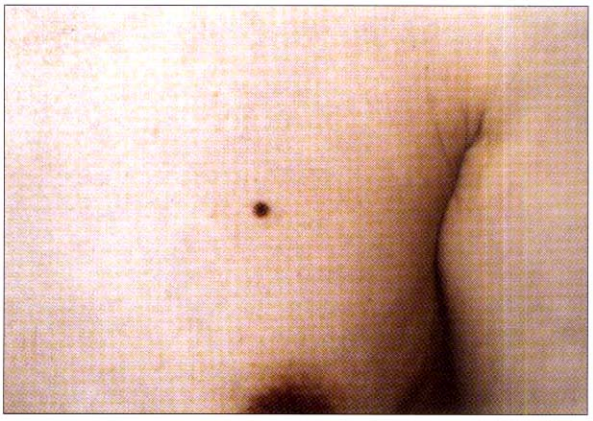

Fig. 1. Single dark brownish pigmented papule on the left upper chest.

A biopsy specimen of the papule from the patient's chest showed psoriasiform epidermal hyperplasia. The neoplastic keratinocytes had abundant pale- staining cytoplasm (Fig. 2A), which was periodic acid-Schiff (PAS)-positive, containing large amounts of glycogen (Fig. 3). Many melanophages were detected in the papillary and reticular dermis (Fig. 2A). Compared with the patient's normal epidermis in the tumour-free margin, which is shown in the right side of Fig. 4A, an increased number of melanocytes with long dendrites and melanin granules, interspersed among keratinocytes, can be seen on the left side of Fig. 4A.

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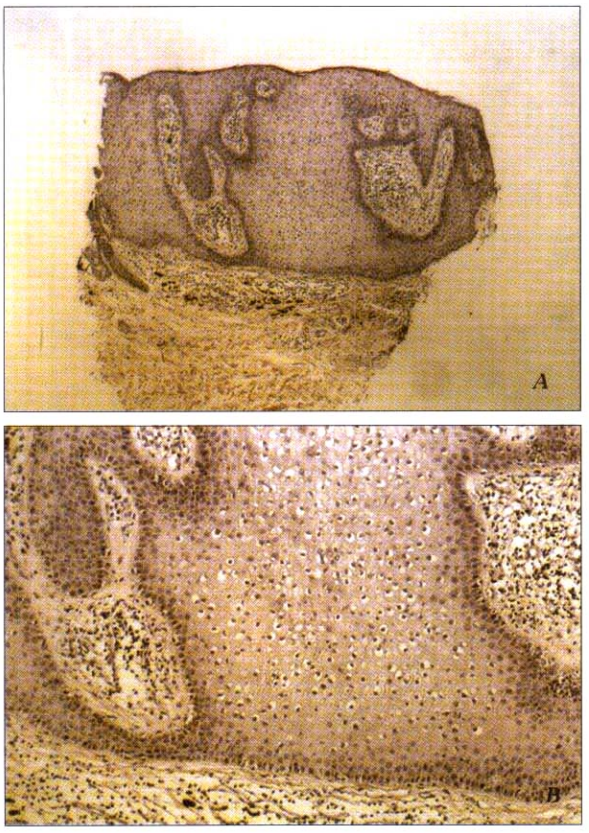

Fig. 2. (A) Psoriasiform hyperplasia is seen on the epidermis. Many melanophages, which are noticeable already on low-power magnification, are detected in the papillary and reticular dermis (H&E stain, ×40). (B) The cells within the thickened epidermis appear strikingly clear (H&E stain, ×200).

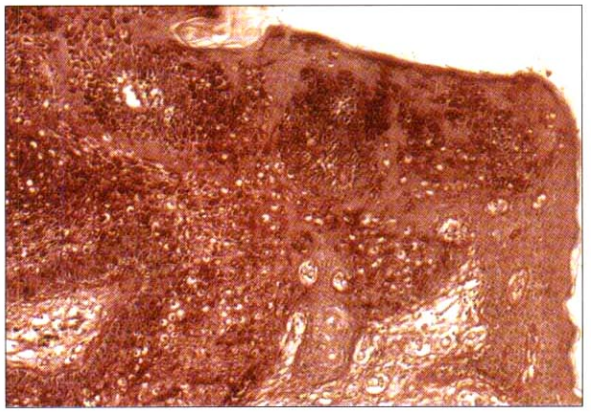

Fig. 3. Positive staining with periodic acid-Schiff (PAS) is seen in the clear cells (PAS stain, × 100).

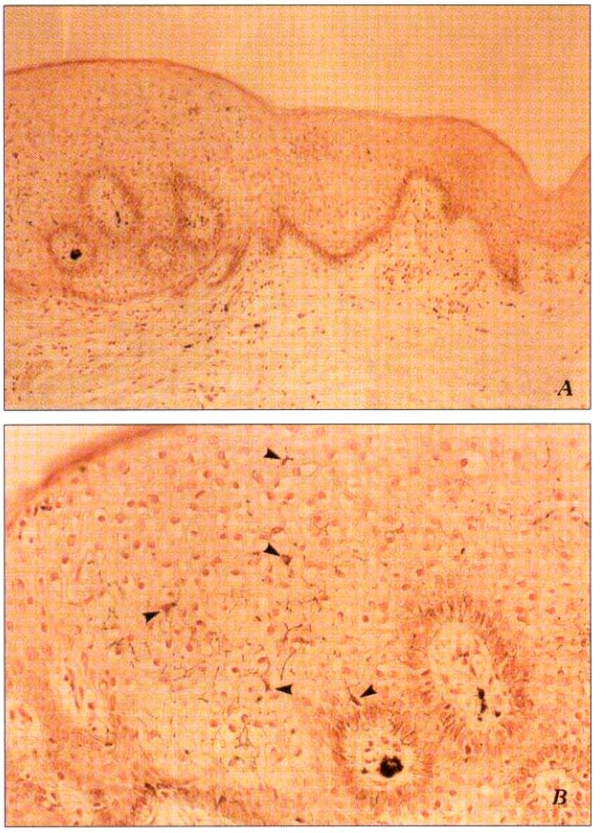

Fig. 4. (A) Compared with the normal epidermis (tumour-free margin) on the right side, an increased number of melanocytes with long dendrites and melanin granules, interspersed among the keratinocytes, can be seen on the left side of Fig. 4A (Masson-Fontana silver stain, ×100). (B) Melanocytes, located in the spinous layer, have several dendrites which contain large amounts of melanin (arrow head, Masson-Fontana silver stain, ×200).

Melanocytes, located in the spinous layer, have several dendrites containing large amounts of melanin (Fig. 4B, arrow head). In contrast, keratinocytes have no dendrite, with few or no melanin granules (Fig. 4). These clinical and histological features were consistent with the findings of pigmented clear cell acanthoma.

DISCUSSION

Clear cell acanthoma (CCA) is a relatively uncommon benign epidermal tumour. The nature of

CCA has not been clarified. However this tumour is considered to be a benign epidermal neoplasm in which metabolic disorder is caused by the accumulation of glycogen within the tumour cells[1]. Known pathogenesis of CCA is the deficiency of phosphorylase, except in the basal cell layer, which is normally present in the epidermis and is necessary for the degradation of glycogen[5].

The diagnosis of CCA can easily be made by distinct histopathological feature[5]. All epidermal cells within the thickened epidermis, with the exception of cells of the basal cell layer, appear strikingly clear and slightly enlarged. When staining is carried out with the PAS reaction, the presence of large amounts of glycogen is revealed within the cells. The rete ridges are elongated and may show intertwining. An absence or a marked reduction of melanocytes and melanin along with the impairment of melanosome transfer to keratinocytes have been described as a characteristic feature of CCA[3,4,6,7].

Pigmented CCA is a variant of CCA. Clinically, this tumour appears macroscopically to have a brown pigment. An important histological findings in these tumours are melanophages in the papillary dermis, which are already noticeable on low-power magnification and served to draw our attention to the presence of dendritic melanocytes in the epidermis[6]. Dermal melanophages in CCA therefore provide a clue to the diagnosis of a pigmented CCA[6]. Melanocytes with long dendrites, interspersed among the keratinocytes, are remarkable. Melanocytes and their dendrites contain large amounts of melanin. However, keratinocytes contain relatively few or no melanin granule[5]. This finding is confirmed by the Masson-Fontana stain. In our case, a large number of melanocytes, with melanin granules contained in their dendrites, are seen in the spinous layers compared with keratinocytes which have no dendrites and few or no melanin granules (Fig. 4). This feature is an identical histopathological finding to pigmented CCA reported by Langer et al[6]. This designation should be adopted to pay tribute to the existence of a pigmented counterpart to CCA, comparable to pigmented basal cell carcinoma as the pigmented counterpart to common basal cell carcinoma[6].

Pigmented CCA can be confused with melanoacanthoma. Melanoacanthoma is a rare variant of pigmented seborrheic keratosis. These two diseases have common clinical and histological features, such as pigmented papules and a striking proliferation of dendritic melanocytes. In pigmented clear cell acanthoma, electron microscopy reveals glycogen granules in the tumour cells, except cells of the basal cell layer. In the upper portion, the amount of glycogen is increased, and the granules are seen to infiltrate between the tonofilament[5]. In melanoacanthoma, the block in transfer of melanin from melanocytes to keratinocytes is often only partial[8] and the melanocytes may proliferate as nests, extending from the basal layer into the superficial layers of the epidermis[9]. However, in pigmented clear cell acanthoma, melanocytes are interspersed among the pale-stained keratinocytes[6].

In our patient, there are characteristic findings of pigmented CCA. First, the lesion is macroscopically pigmented. Second, melanophages which are already noticeable on low-power magnification, are detected in the papillary and reticular dermis. Third, an increased number of melanocytes and melanin is seen in the epidermis.

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