Malignant Melanoma on Congenital Melanocytic Nevus

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Congenital melanocytic nevi are considered to be precursors of malignant melanoma. Although the risk of malignant melanoma with medium and small congenital melanocytic nevi is uncertain, it is important to notice the possibility of malignant transformation in those lesions. We describe a 62-year-old woman who had had a brown soft verrucous tumor on her right lower back since birth. She first noticed a black nodule in the center of the tumor 5 years before which had ulcerated 3 months prior to presentation without healing. A biopsy specimen revealed malignant melanoma arising from a congenital melanocytic nevus.

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Key Words : Malignant melanoma, Congenital melanocytic nevus

Congenital melanocytic nevi have been regarded as precursors of malignant melanoma. The risk of malignant melanoma associated with melanocytic nevi lies between 19% and 85%. The lifetime risk of malignant melanoma with medium and small congenital melanocytic nevi is uncertain[1-3]. Therefore it is controversial to remove medium and small congenital nevi prophylactically.

Herein we report a patient with malignant melanoma arising from a congenital melanocytic nevus.

CASE REPORT

A 62-year-old woman had had a verrucous tumor on her right lower back since birth. The lesion had slowly enlarged in size and elevated with age. She first noticed a black nodule at the center of the lesion five years before, and this had become ulcerated with exudation three months prior to presentation and had not healed. She had no significant personal or family history of illness.

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A physical examination revealed a 17 × 9 cm brown, verrucous, soft tumor with multiple black papules on its surface on the right lower back. There was a 3 × 3 cm black, ulcerated, oozing mass at the center of the tumor (Fig. 1). Lymphadenopathy was absent.

A biopsy specimen from the brown verrucous tumor demonstrated well-circumscribed nests and cords of nevus cells in the dermis without junctional activity on the dermal-epidermal junction (Fig. 2). Nevus cells were present around and within the skin appendages, and were also extended between collagen bundles singly or in double rows. Nevus cells contained a moderate amount of melanin. Numerous ovoid or round cavities were present, which represented spaces occupied by lipids produced by degeneration of the nevus. A biopsy specimen from a central black nodule showed considerable junctional activity with downward streaming of tumor cells, which showed great variation in size and shape, and possessed anaplastic nuclei from the epidermis into the dermis (Fig. 3). Tumor cells also extended upward into the overlying epidermis. A considerable amount of melanin pigment was found within the tumor cells and melanophages. It was consistent with malignant melanoma of the invasive type and the maximum depth of invasion was 3.8 mm.

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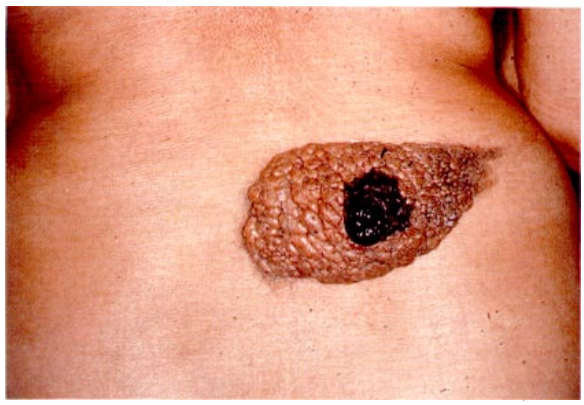

Fig. 1. A 17×9 cm-sized, brown, verrucous, soft tumor was noted on the right lower back. Multiple small black papules were seen on the surface of the tumor. At the center of the verrucous tumor, a 3×3 cm sized, black, ulcerated, oozing mass was found.

On computed tomography, the mass was localized outside of the muscles. Abnormallyenlarged lymph node or intra-abdominal pathology was not demonstrated. A whole body bone scan did not show any evidence of metastasis of the tumor.

The whole lesion was excised. Microscopic examination revealed that all resection margins were free from malignant cells.

DISCUSSION

Malignant melanoma may arise de novo, but they can develop at the site of pre-existing skin lesions such as congenital melanocytic nevi, acquired melanocytic nevi, and dysplastic nevi[4]. The theory that melanocytic nevi are precursors as well as markers of an increased risk of cutaneous malignant melanoma is supported by the spatial association observed clinically and histologically between nevi and malignant melanoma[1-6].

Congenital melanocytic nevi have been divided into three groups according to their size, such as small (less than 1.5 cm in diameter), medium (1.5 to 20 cm in diameter), and large or giant (greater than 20 cm in diameter)[4]. The lifetime risk of malignant melanoma associated with giant congenital nevi may be between 5% and 15%[7]. Usually, the greatest risk of malignant degeneration of giant congenital melanocytic nevi exists before the age of 10 years, whereas the risk of malignancy associated with smaller congenital melanocytic nevi is greatest after puberty[8,9]. It is unclear to what extent malignant melanoma develops in smaller congenital nevi[8,10]. According to the previous report, the patients with small congenital melanocytic nevi who live up to the age of 60 years are estimated to have a

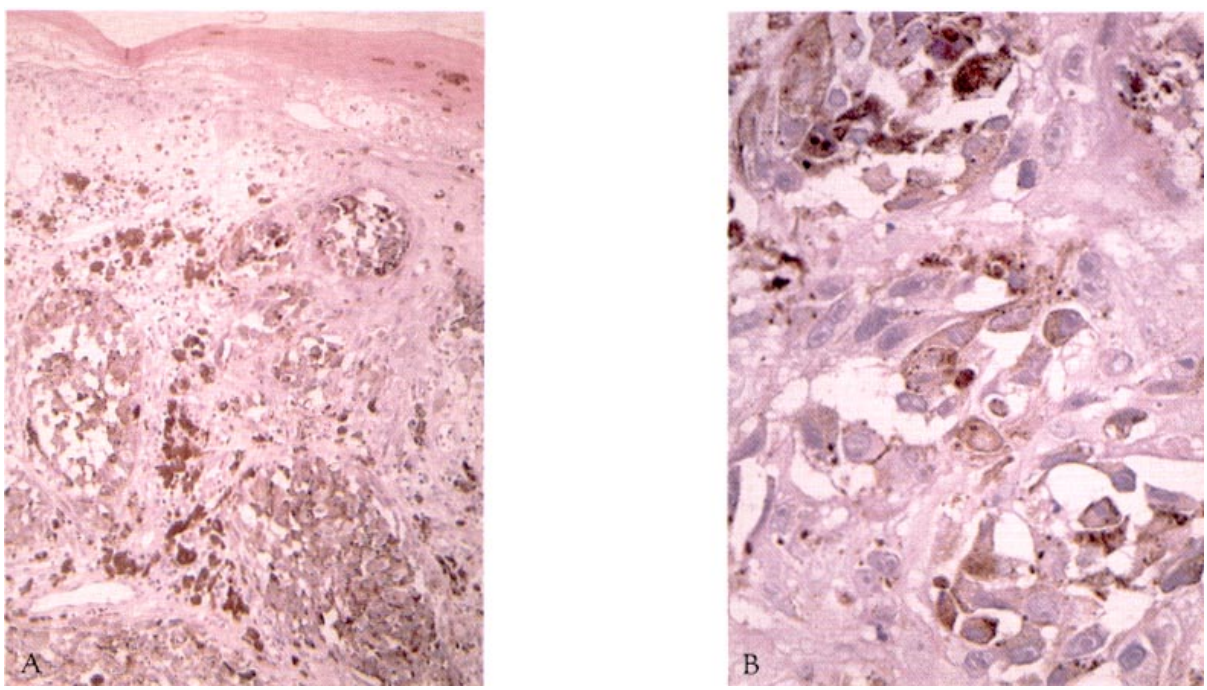

Fig. 2. A biopsy specimen from the central black ulcerated mass revealed anaplastic cells with variable sizes and shapes from the epidermis to the deep dermis. (H&E, A, ×100, B, ×400).

cummulative risk for malignant melanoma of 0.8% to 4.9%[9]. The nevus seen in our patient was 17 cm at its greatest diameter, a medium-sized congenital melanocytic nevus. Because of the size of the nevus and the patient's age, the risk of malignant transformation was estimated to be high. However, malignant melanomas arising in a small and medium-sized congenital melanocytic nevi have not been reported in Korea.

The process of malignant transformation from benign melanocytic nevi has not yet been clarified. Excessive UV radiation has been regarded as the major cause of malignant melanoma. Predilection sites for malignant melanoma are those receiving intermittent and intense sun exposure rather than those receiving the greatest cumulative sun exposure[11]. Therefore the trunk was reported as the most common site for a malignant melanoma associated with a melanocytic nevus followed by the limbs, whereas melanomas on the head and neck were least commonly associated with a melanocytic nevus[15]. A great number of melanocytes, genetic factors or immunosuppression have been regarded as causative factors for an increased malignant potential of nevus cells[12-14]. In our patient, the nevus was located on the lower back and this location is compatible with the previously reported predilection site for malignant transformation. Because the location of the nevus in this case was an area that was irritated easily, we thought that irritation could be a factor for malignant transformation of melanocytic nevi.

Histologically, benign melanocytic nevus cells have been found in association with a malignant melanoma in 4% to 72% of malignant melanoma patients[5,6]. If a malignant melanoma arises from a congenital melanocytic nevus, it usually originates at the epidermal-dermal junction[15]. Occasionally, however, the malignant melanoma in a giant congenital melanocytic nevus has arisen in the dermal population of the nevus cells[16,17].

The management of congenital melanocytic nevi depends primarily on their size and the perceived risk of development of malignant melanoma. Although the most important preventive measures for the development of malignant melanoma later in life are those taken during childhood, there is insufficient data to recommend prophylactic excision of all congenital melanocytic nevi. Therefore, patients with congenital melanocytic nevi should be examined periodically, and if an alteration in the nevus is detected, it should be evaluated by a physician and examinations including a skin biopsy should be done for the exact diagnosis and proper treatment.

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