

A Case of Desmoplastic Malignant Melanoma

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Desmoplastic malignant melanoma(DMM) is an uncommon variant of malignant melanoma featuring a proliferation of spindle cells with pronounced desmoplasia. DMM commonly occurs on the sun-damaged skin such as head and neck in older persons, and it can be associated with lentigo maligna. We report an unusual case of desmoplastic malignant melanoma that developed de novo on the trunk in a young patient.

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Key Words : Desmoplastic malignant melanoma

In 1971, Conley et al¹ described an invasive melanoma composed of spindle cells and abundant collagen, which they called "desmoplastic malignant melanoma(DMM)". The concept of non-pigmented, collagenizing spindle cell tumor associated with overlying, inconspicuous, intraepidermal lentiginous hyperplasia has been extended by Reed and Leonard², who described neural invasion or neuroid differentiation, namely neurotropism in DMM. DMM is an especially difficult subtype to recognize because a dermal fibroblastic component is predominant and a melanocytic proliferation at dermal-epidermal junction may be minimal or even absent. Consequently, DMM can be confused with a large number of other spindle-cell lesions. On reviewing Korean documentation only one case of DMM associated with acral lentiginous melanoma has been described³. We report an additional case of DMM developing de novo at a

young age on the back, which is a relatively rare location.

CASE REPORT

A 20-year-old man was admitted to our clinic for evaluation of an enlarging nodule on the back. It had been present for 5 years and had undergone recent change. The skin lesion showed a $1.3 \times 1.0 \times 0.4$ cm flesh colored soft, firm, nontender nodule on the right upper back area.(Fig. 1) The family and past medical history were not significant. On physical examination, specific findings were not revealed except the skin lesion. There was no palpable lymph node on both axillae. Laboratory tests including complete blood count, liver function test, urinalysis, and chest X-ray were within normal range. Histological examination of the biopsy specimen showed elongated and fusiform spindle cells in the dermis and extending to the subcutaneous tissue.(Fig 2) The constituent cells were separated by abundant collagens and form a nodular mass. The cells had enlarged and atypical nuclei, at least focally, resembling neoplastic melanocytes. There was no atypical melanocytic proliferation at the dermal-epidermal junction. Immunohistochemical staining was performed on paraffin embedded sections with S100 protein, smooth muscle actin(SMA), HMB45, desmin and CD68 antibody. The stainings with S100 protein and SMA were focally positive, and those with

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HMB, desmin, and CD68 antibody were not positive. (Fig 3) The diagnosis was made as malignant melanoma with a desmoplastic amelanotic spindle cell component. Breslow's thickness was 15mm. Clark's level was V. He was treated with an en-

bloc resection with 1cm margin of the mass, and there has been no recurrence for 1 year.

DISCUSSION

DMM is a rare variant of malignant melanoma accounting for approximately 1% of all cases in a population-based study⁴. DMM continues to cause difficulty in diagnosis for both clinicians and pathologists. The lack of sinister clinical pigmentation often results in delay in diagnosis, which may be further compounded by an incorrect histological diagnosis and failure to appreciate the invasive nature of this tumor⁵. DMM may develop as a complication of any of Clark's four major biological types of malignant melanoma, but has been reported most frequently in association with lentigo maligna^{1,2,6} and acral lentiginous melanoma⁷. In this case the tumor appeared to be arising in the dermis and there was no evidence of a preexisting nevus or other melanocytic lesion.

Fig. 1. Single 1.3 × 1.0 × 0.4cm flesh colored, soft non-tender nodule on the right upper back.

Fig. 2. A. Nodular focus composed of elongated amelanotic spindle cells with hyperchromatic nuclei in the dermis. Junctional activity is absent in overlying epidermis. Note that the immediate subepidermal zone is largely free from tumor. (Hematoxylin-eosin, × 40)
B. Atypical spindle cells arranged in loose fascicles extend into the deeper dermis and subcutis. (Hematoxylin-eosin, × 100)
C. Lt. High power view of lesion shows dense fibrosis with scattered spindle cells with large hyperchromatic nuclei; several mitoses are present. (Hematoxylin-eosin, × 200) **Rt.** Immunohistochemical stains of paraffin section shows positive reaction to S-100 protein. (× 200)

The clinical presentation of DMM differs from that of other melanomas. Male are more likely to develop DMM. The mean age of patients with DMM calculated from major clinicopathological series is 60 years, with the majority in the sixth or seventh decade⁸. However, DMM should not be considered as occurring exclusively in the elderly, as a small number of much younger patients^{2,5} have been reported including our case. DMM most commonly occurs on the head and neck, but has also been reported as a common feature of acral lentiginous melanoma. In the current series^{9,10,11}, the head and neck was the most common sites for both genders whereas the most common site for other cutaneous melanoma was the back in males and the lower limbs in females. Thus as it has previously been suggested^{2,4}, DMM tends to occur in sun-exposed areas. An increased association with lentigo maligna melanoma, which requires solar degeneration for diagnosis, supports this link with sun exposure. Others, less frequently involved sites include the upper limb, lower limb, and trunk⁶. There were also reports of primary DMMs arising from the mucous membranes, including the lip², gingiva¹¹, and conjunctiva¹², maxillary alveolus¹³, anus¹⁴, and vulva¹⁵. Our present case showed a slow growing flesh colored nodule on the back in young age. These clinical features belonged to the less common clinical feature of the disease. Most cases have feature of a poorly circumscribed downward dermal proliferation of elongated spindle cells that, at least focally, contain enlarged atypical and hyperchromatic nuclei⁹. Mitotic figures are usually present in the atypical cells but may require careful searching to identify them⁵. Neurotropic melanomas are considered as a variant of desmoplastic melanoma demonstrating prominent perineural growth and invasion and sometimes featuring neuroid differentiation. DMM usually presents as an area of dermal thickening or scarring. When a clinically obvious nodule is present, most are amelanotic. The absence of pigmentation within the spindle cells is probably the major cause for histological failure to recognize DMM as melanoma. And up to 40 to 50% of cases may not have a junctional or lentiginous component¹⁰. Immunohistologic features of DMM differ from those of conventional melanoma. If a problematic spindle-cell skin lesion is a suspected melanocytic process, HMB45 is unlikely to provide confirmatory evi-

dence of the diagnosis of DMM. Similarly, because of the variability in S100 expression in this neoplasm, the absence of S100 staining should not be relied on too heavily to exclude DMM if the clinical and histologic features favor that diagnosis¹⁶.

As long as the cause of desmoplastic transformation of melanoma remains unknown, definition must be empirical, and may also be inaccurate. Benign mesenchymal proliferations figure prominently in the differential diagnosis of DMM. These include schwannoma, neurofibroma, leiomyoma, and especially the dermal fibrous histiocytomas⁵. Careful attention to the presence of an atypical junctional melanocytic component and to cells with enlarged atypical nuclei and mitotic figures are the most helpful features in excluding benign mesenchymal lesions. The large size and asymmetrical silhouette of many of these lesions differ from those of most benign lesions. One subtle clue to the diagnosis at low magnification is the frequent presence of a lymphocytic infiltrate, distributed as nodular aggregates of infiltrating lymphocytes throughout the tumor. A search for enlarged and hypercellular nerves will also help in recognizing the neurotropic type of DMM. S-100 protein is not expressed by dermatofibroma cells, although it may be found in the cells of some smooth muscle neoplasms. Malignant soft tissue tumors that should be distinguished from DMM include superficial malignant epitheloid schwannoma, malignant fibrous histiocytoma, storiform fibrous histiocytoma, leiomyosarcoma, fibrosarcoma, and nerve sheath sarcoma. Additional malignant lesions within the differential diagnosis are spindle-cell/squamous cell carcinoma, spindle-cell malignant melanoma and metastases, particularly from a renal cell carcinoma². The presence of an atypical junctional melanocytic component is the most helpful feature in distinguishing the nonmelanocytic lesions from DMM.

The desmoplastic nevus, a variant of Spitz nevus, must also be distinguished from DMM¹⁷. Desmoplastic nevus typically occurs on the extremities of young patients. The lesion is wedge-shaped and may have very pleomorphic cells, but mitotic figures are extremely rare. Abundant collagen separates the cells. The junctional component is sparse and does not show the features of lentigo maligna.

As with other types of malignant melanoma, adequate, prompt surgical excision is the initial treatment

of choice because multiple local recurrence is the major therapeutic problem and appears to antecede metastatic disease. In our case, we performed local excision and there is no signs of metastasis until now.

We report herein a rare and unusual case of primary DMM in 20-year-old man, and it has not been described in Korean literature as yet.

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