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Desmoplastic Melanoma Arising in The Ankle

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ABSTRACT

Desmoplastic melanoma (DM) is an uncommon but potentially devastating malignancy that can be cured with early recognition and surgery. DM often occurs in elderly men and develops on sun-exposed areas with the head and neck being the most common site of origin followed by the extremities and trunk. We report a rare case of DM occurring in the ankle in an 85-year-old woman. Magnetic resonance imaging of the affected ankle showed a 4.3 x 4.3 x 2.5 cm sized mass which was located in the subcutaneous region, attached to lower leg muscles, the fibula, and the calcaneus. No metastasis to the lung, liver, bones or abdominal lymph nodes was found. In spite of below knee amputation, the patient died of lung metastases 10 months after surgery. Among several factors causing early death of the patient, a large size, a deep location and rapid growth of the tumor seem to be most important.

INTRODUCTION

Desmoplastic melanoma (DM) is an uncommon but potentially devastating malignancy that can be cured with early recognition and surgery. DM has clinical as well as histological features that may be subtle and overlooked, or misdiagnosed as other benign or malignant lesions that would require less aggressive therapy for cure [1]. The difficulty in its diagnosis lies in that most patients have amelanotic skin lesions [2]. DM often occurs in elderly men and develops on sun-exposed areas with the head and neck being the most common site of origin followed by the extremities and trunk [3,4,5,6]. Although the incidence of malignant melanoma is much lower

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in the Japanese than in Caucasians, the subungual and periungual sites are commonly found in Japanese. DM in the foot is, however, very rare [7]. We report a case of DM occurring in the ankle.

CASE REPORT

The patient was an 85-year-old woman. She noticed a non-symptomatic mass in her left ankle. Three month later, she was referred to us with the suspicion of malignancy. The colour of the skin over the mass looked normal. The mass was hard as a brick and fixed to the base but had mobility to the skin. No lymph nodes were palpable in the ipsilateral popliteal fossa and inguinal region. Bone metastasis was ruled out by bone scan. There was no evidence of metastasis to lung, liver or abdominal lymph nodes by computed tomography. Computed tomography of the affected ankle showed a iso-density mass, the periphery of which was irregularly

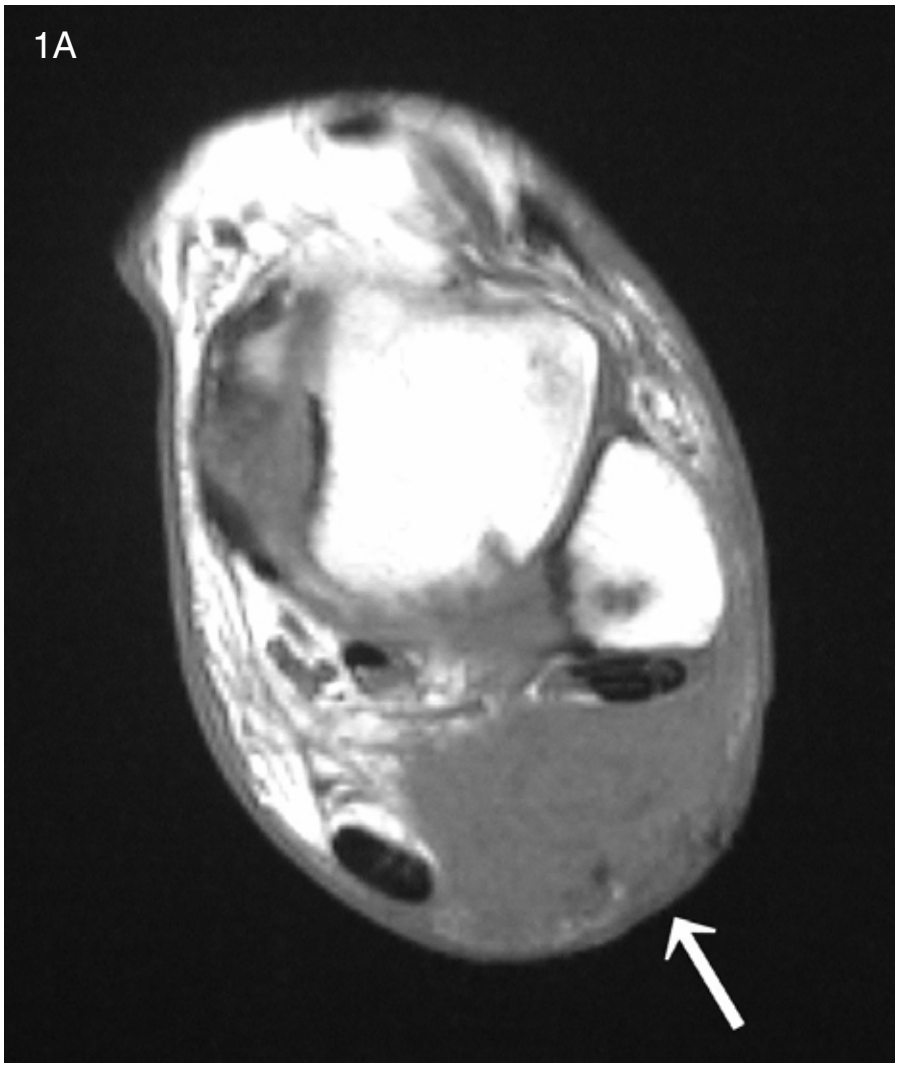
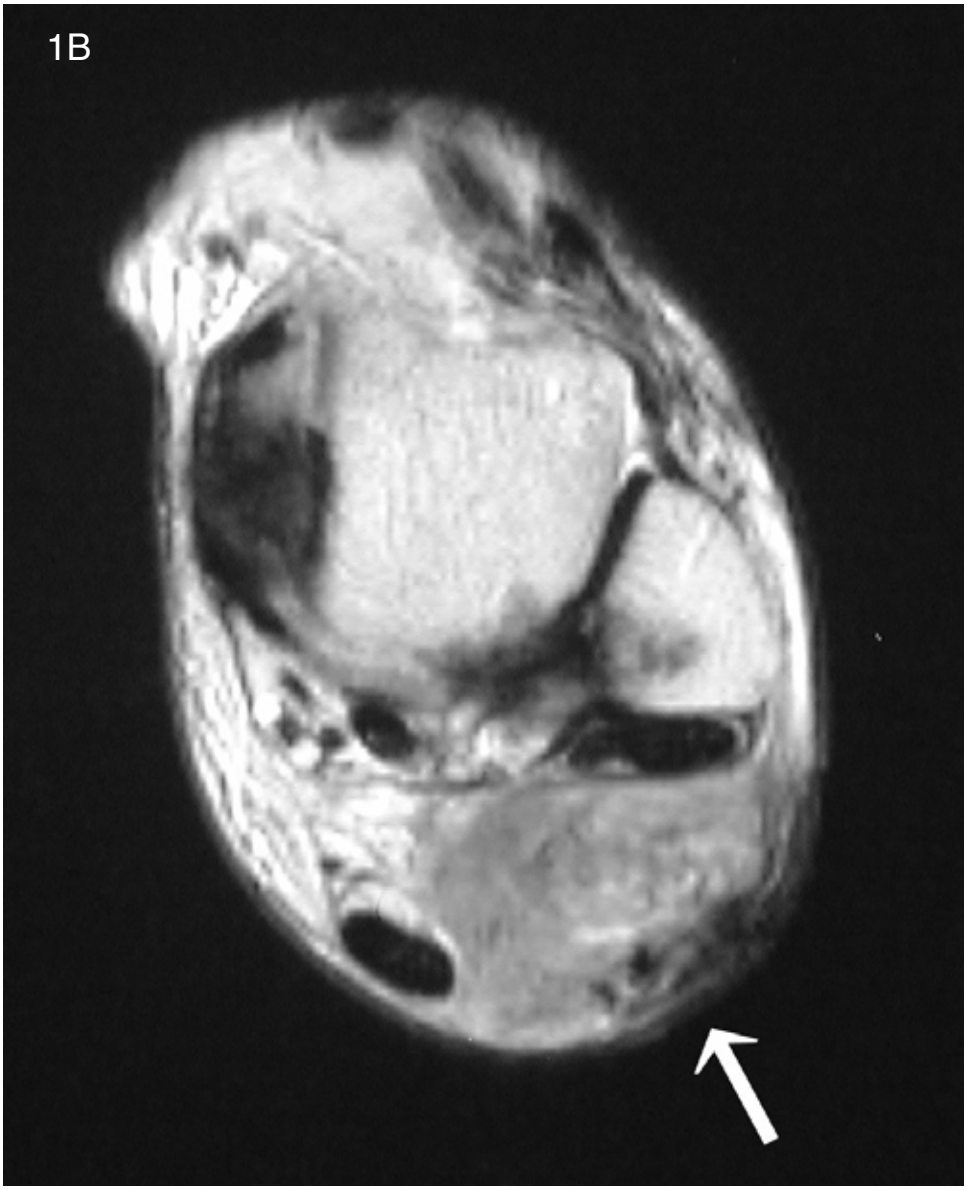


Fig. 1 Magnetic resonance imaging demonstrating a mass located in the subcutaneous region, attached to lower leg muscles, the fibula, the calcaneus and the Achilles tendon. The tumour had iso-signal intensities on T1 weighted images (A: axial image) and inhomogenously high signal intensities on T2 weighted images (B: axial image, C: sagittal image).



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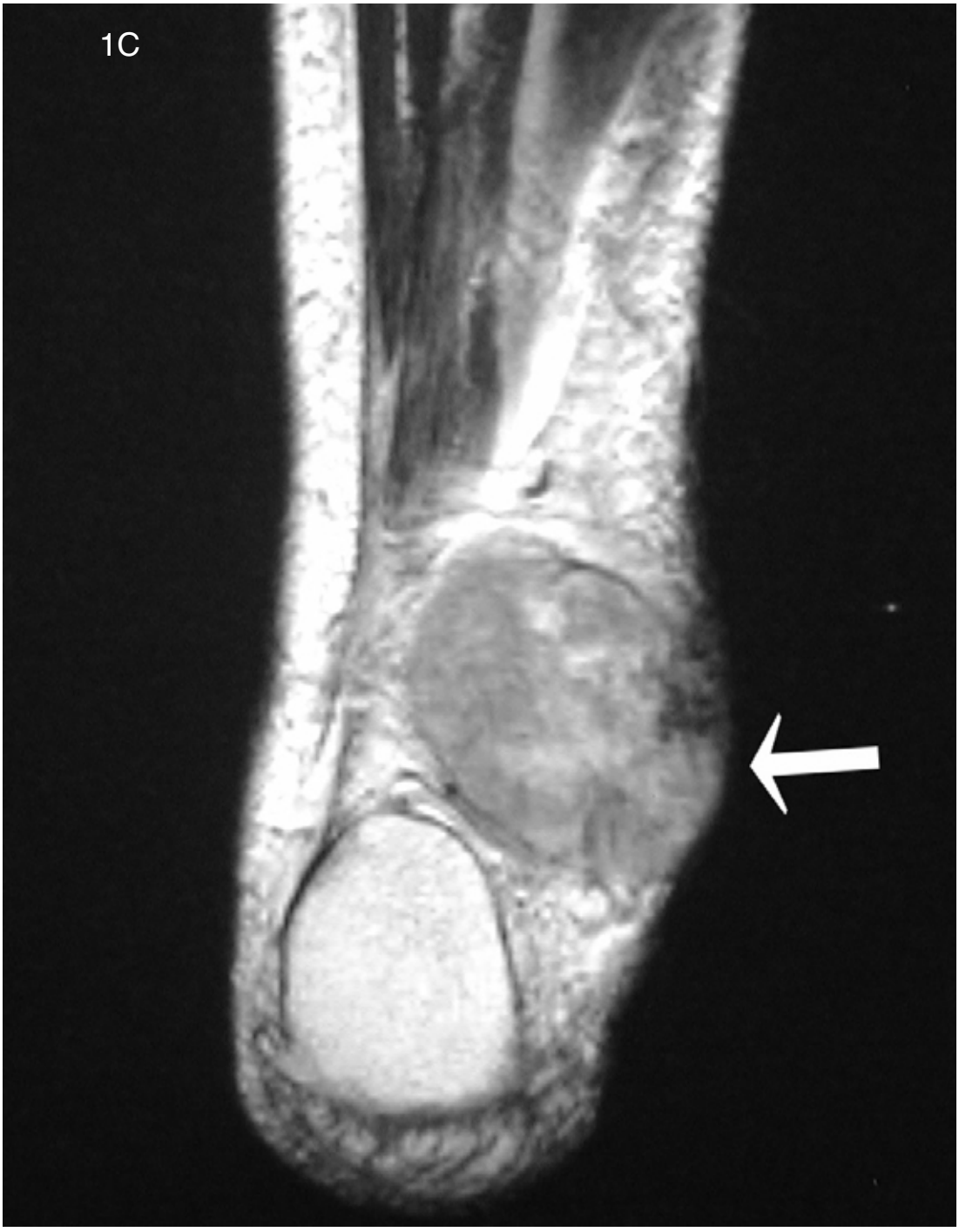
enhanced. Magnetic resonance imaging demonstrated a mass located mainly in the subcutaneous region, attached to lower leg muscles, the fibula, and the calcaneus, deviating the Achilles tendon medially. The mass was poorly circumscribed and 4.3 x 4.3 x 2.5 cm in size. It had iso-signal intensities on T1 weighted images and inhomogenously high and low signal intensities on T2 weighted images. The tumour was inhomogenously and highly enhanced after gadolinium injection (Fig 1).

Histological examination of open biopsy specimen demonstrated proliferation of short spindle to polyhedral cells in the dense collagenous stroma. These cells had



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small nuclei with moderately atypia showing little pleomorphism. They had narrow cytoplasm with slightly eosinophilic fine-granular appearance. Cellular density was moderate and mitotic figure was almost not detected. Focally there was a cluster of atypical cells containing brown granules in the cytoplasm, considered to be melanin

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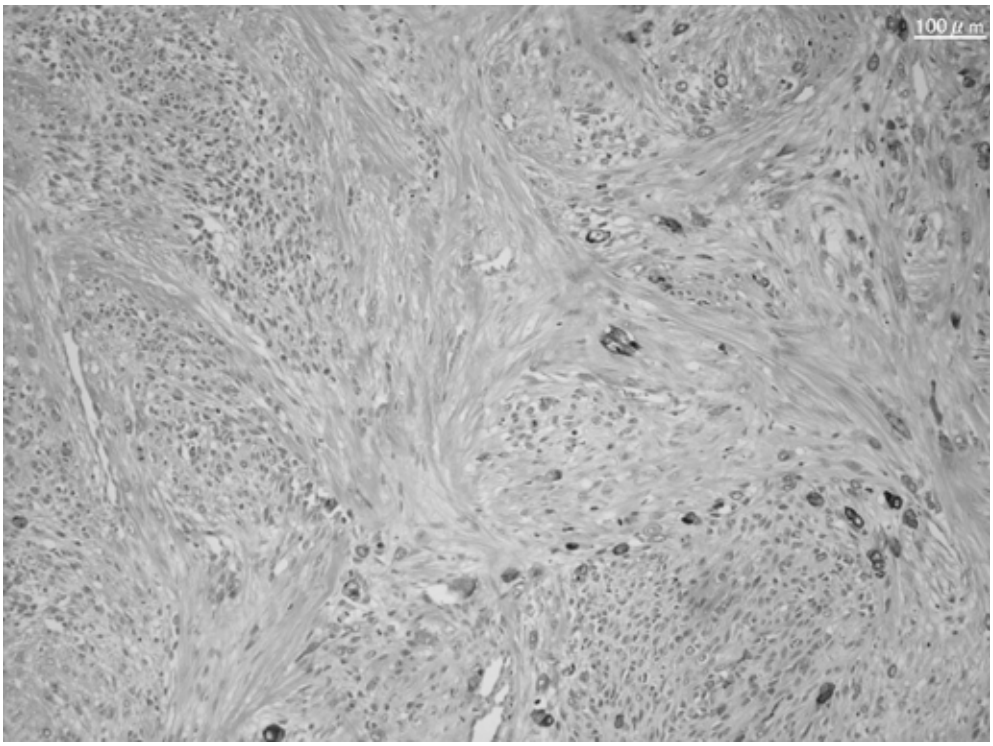

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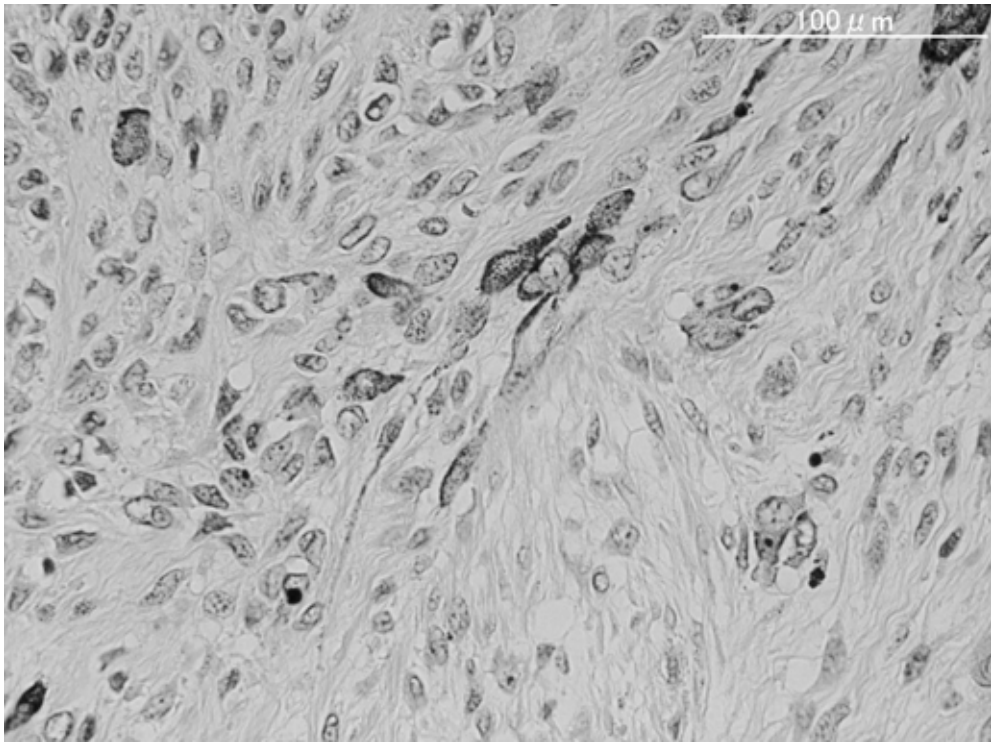

Fig 2: Microphotographs of the open biopsy specimen demonstrating proliferation of short spindle to polyhedral cells in the dense collagenous stroma. These cells had small nuclei with moderately atypia showing little pleomorphism. Focally there was a cluster of atypical cells containing brown granules in the cytoplasm, considered to be melanin pigments. (A: low power view, B: high power view).

pigments (Fig 2). Immunohistochemically positive staining for vimentin, S100 protein and αsmooth muscle actin in these cells was observed. Immunohistochemical staining after melanin removal showed that a few cells were reactive with HMB-45. Melanin A antigen, CD 34, CD 68 and desmin were negative. From these histological findings, desmoplastic melanoma (DM) was suspected.

Below knee amputation 7 cm apart from the tumour-involved skin was performed because of the location and extent of the tumour. Histological examination of sentinel lymph nodes revealed no signs of metastasis.

Macroscopically the tumour, 40 x 25 mm in size, had almost clear margin, and was opaque coloured with focally black-pigmented areas. No direct invasion into the neighbouring calcaneus was found. Microscopic examination of the removed specimen revealed similar histological findings as the biopsy specimen except for a higher cellular density and more atypical feature. There was a fascicular pattern of spindle cells between variably increased collagen fibers. Occasionally mitotic figures (9 to 10 cells per 10 HPF) were observed. In the black pigmented area, clusters of melanin-containing polyhedral cells were revealed. Histologically the tumour cells had invasive growth pattern along the septum of the fatty tissue in the dermis. There were no abnormal melanocytes observed in the epidermis covering the tumour. Immunohistochemically the tumour cells had the same results as the biopsy specimen. Ki 67 positivity was counted in 60 %. The histological diagnosis was desmoplastic malignant melanoma. Six months after surgery, lung metastases were found. The patient died of lung metastases 10 months after surgery.

DISCUSSION

DM is a rare and atypical form of melanoma. Establishing a correct diagnosis is difficult as DM is often clinically innocuous and unlike other subtypes of malignant melanoma [4]. This type of melanoma has usually invaded into the deep reticular dermis or into the subcutis by the time the biopsy is taken. In 2003, Parodi et al stated that histopathological identification of DM is confusing because of the intense fibrous reaction in the dermis and minimal, atypical melanocytic proliferation at the dermal-epidermal junction and often that DM is still misdiagnosed unfortunately as a variety of entities, including simple scar, fibrohistiocytic neoplasms, neural tumours, and superficial fibromatoses-with potentially devastating consequences[2]. Histologically, early lesions are characterized by superficial tumour fascicles, and randomly diffuse hypercellularity in the upper dermis identified as elongated hyperchromatic pleomorphic spindle cells with stromal myxoid feature. Neuroidal melanocytic structures, invasion of adventitial dermis, islands of inflammation, and epidermal lentiginous melanocytic hyperplasia are often present. The most reliable and characteristic features of an early lesion of DM are aggregates of lymphocytes, tumour cells showing cytological atypia, stromal myxoid appearance, and poor circumscription of the dermal infiltrate [1]. The present case also had these characteristic features except aggregates of lymphocytes.

In equivocal cases, the use of immunohistochemistry (in particular S-100 and neuron-specific enolase) may be helpful in establishing the diagnosis [2]. In the present case, immunohistochemical staining with S100 protein antibody and vimentin was positive. Focally the tumour cells were reactive with ÉøSMA and HHF-35 (muscle specific actin). A few cells were reactive with HMB-45. DM is almost always S-100 positive. Mature scars were readily differentiated from DM by light microscopy. In contrast, immature scar and DM had many features in common including hypercellularity, nodular lymphoid infiltrates, myxoid stroma, and atypical nuclei [8]. An S-100 study is often necessary to distinguish between tumour and early scar in margins of resection. S-100 staining has also proved to be a valuable adjunct in determining the extent of the tumour at the peripheral margins, particularly for hypocellular and amelanotic tumors [9]. DM is usually HMB-45 negative. All lesions of 128 cases evaluated by Skeleton et al. were negative for HMB-45, a marker for premelanosomes. Riccioni et al. reported three cases of desmoplastic melanoma (DM) rich in smooth muscle actin. They thought that actin-rich elements differentiate toward mesenchymal elements, paralleling the phenotypic changes seen in sarcomatoid carcinomas [10].

The clinical behaviour of DM is close to soft tissue sarcoma with high rates of local recurrence, low incidence of lymph node metastases, and propensity to develop lung metastases [4]. Skeleton et al. evaluated the relationship of histologic features to disease-free survival of 128 cases. Factors that correlated with survival included sex, tumour location, tumour depth, and the presence of stromal mucin. The 5-year disease-free survival rate was 68% for all cases and 61% for lesions more than 4 mm deep. Wide excision of the primary lesion with clear margins and close follow-up are necessary. It is important that the lesion will be radically extirpated at the time of initial surgery [1]. Recently Gyorki et al. described their experience with lymphatic mapping and sentinel lymph node biopsy (SLNB) in patients with DM to characterize the biological behaviour of these tumors. SLNB detected subclinical metastases of DM to regional lymph nodes [5]. SLNB at the time of resection can provide useful information to guide early treatment and, coupled to lymphadenectomy in positive patients, may limit tumour spread and prevent recurrence [6]. Because of the high local recurrence rate for DM located in the finger, amputation is recommended in an effort to gain effective tumor control [2]. In the present case, SLNB was negative. In spite of curative excision by below knee amputation the patient died of lung metastases 10 months after surgery. Among the several factors causing early death of the patient in our case are thought a large size of the tumour (40 x 25 mm) and its deep location.

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