SHORT COMMUNICATION

# First Case of Allochronic Onset of Two Primary Dermatofibrosarcoma Protuberans Lesions Proved by COL1A1-PDGFB Fusion Gene Analysis

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Dermatofibrosarcoma protuberans (DFSP) is a relatively rare cutaneous malignancy that often develops on the trunk and extremities in young to middle-aged people (1). Although recurrence is common, both metastasis and multifocal lesions are extremely rare in DFSP. We report here a new case of 2 primary DFSP proved by gene analysis.

## CASE REPORT

A 60-year-old woman was referred to Tottori university hospital with to a tumour on the left inguinal region. She had been aware of a slow-growing tumour for 5 years. Physical examination showed a 3.8×2.8 cm tumour on the left inguinal region (**Fig. 1**a). Histopathological examination of a biopsy specimen revealed a massive proliferation of mildly atypical spindle cells with a storiform pattern in the dermis to the subcutis (Fig. 1b). Immunohistochemistry showed that the spindle tumour cells were positive for CD34 (Fig. 1c). A diagnosis of DFSP was made. We resected the tumour with a 3-cm margin and confirmed margin clearance histopathologically. There was no sign of recurrence for 1 year, but the patient noticed another nodule on the left side of her neck (Fig. 1d). Physical examination revealed a 1.5×1.5 cm tumour. The biopsied specimen showed a massive proliferation of atypical spindle cells (milder than the first DFSP) in the dermis to the subcutis (Fig. 1e). Immunohistochemistry showed CD34 positivity (not shown). Although we suspected metastasis of DFSP, computed tomography showed no evidence of anther lesion. The lesion was removed with a 1-cm margin. The fusion gene of the first lesion was compared with that of second lesion by reverse transcription-polymerase chain reaction (RT-PCR). Both lesions possessed a chimeric *COL1A1-PDGFB* fusion gene. The first lesion had a chimeric COL1A1 in exon 38-PDGFB in exon 2 fusion gene (Fig. 1f). On the other hand, the second lesion had a COL1A1 in exon 47-PDGFB in exon 2 fusion gene (Fig. 1g). Finally, we made a diagnosis of allochronic primary DFSP.

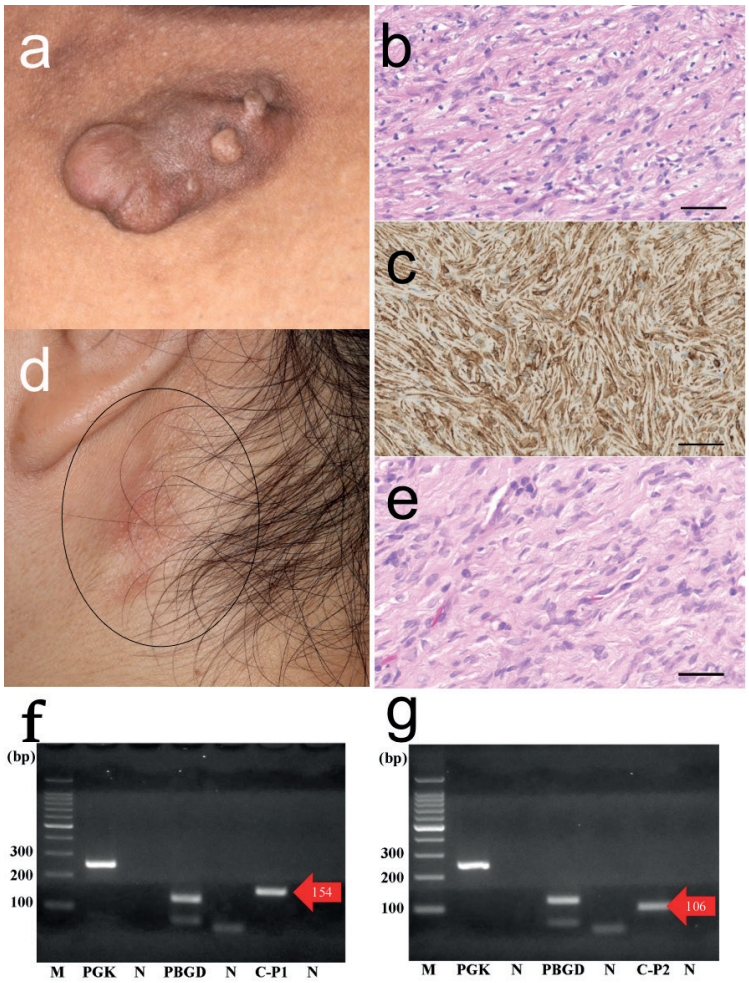


**Fig. 1.** (a) Clinical photograph of the tumour on the left inguinal region. Clinical photograph of the tumour, 3.8×2.8 cm, on the left inguinal region. (b) Haematoxylin-eosin (HE) staining of the first lesion showing massive proliferation of mildly atypical spindle cells with a storiform pattern. Scale bar: 50 μm. (c) The tumour cells were positive for CD34. Scale bar: 50 μm. (d) Clinical photograph of the tumour on the left side of the neck. (e) HE staining of the second lesion showing massive proliferation of atypical spindle cells (milder than the first dermatofibrosarcoma protuberans; DFSP). Scale bar: 50 μm. (f, g) COL1A1-PDGFB fusion genes of (f) the first and (g) second lesions by RT-PCR. M: molecular size marker; N: negative control (distilled water); PGK (phosphoglycerate kinase): 247 bp; PBGD (porphobilinogen deaminase): 127 bp. (f) C-P1 (COL1A1-PDGFB primer): 154 bp. (g) C-P2 (COL1A1-PDGFB primer): 106 bp.

## DISCUSSION

In the case of multiple DFSP lesions, as in the current case, differentiation of metastatic DFSP with a second primary DFSP is difficult. Several studies have shown that primary DFSP and metastatic DFSP have an identical chimeric *COL1A1-PDGFB* fusion gene (2, 3). However, in the current case, the chimeric fusion gene in the first lesion was different from that in the second lesion, indicating that the first and second lesions occurred independently. A few cases of allochronic primary DFSP have been reported (4–6), but there has been no case confirmed by examination of the *COL1A1-PDGFB* fusion gene. Therefore, in a case of multiple DFSP lesions, genetic analysis of the fusion gene is useful for differentiating metastatic DFSP from second primary DFSP.

*The authors have no conflicts of interest to declare.*

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