

〈Case Report〉

## **Fibrosarcomatous variant of dermatofibrosarcoma protuberans on the right cheek: A case report**

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**ABSTRACT** A 52-year-old man presented with a subcutaneous mass on his right cheek. The tumor was resected. Histopathological examination of the resected tissue revealed fibrosarcomatous deromatofibrosarcoma protuberans (FS-DFSP). Since the resection resulted in a large skin defect, his cheek was reconstructed using a deep inferior epigastric artery perforator flap (DIEP). As the pathological findings showed positivity for tumor cells at the excised end, radiation therapy was applied to his right cheek.

FS-DFSPs are found in about 10% of all DFSP cases, and are more malignant than other types of DFSP. Because there is a risk of local recurrence or distant metastasis, the patient should undergo close, long-term observation.

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### **INTRODUCTION**

Dermatofibrosarcoma protuberans (DFSP) is one of the intermediate fibrohistiocytic tumors, which may recur in a local area, although remote metastasis is rare. DFSP frequently develops in the limbs and trunk. The incidence of DFSP of the head and neck is low<sup>1)</sup>. On the other hand, Mentzel *et al.*<sup>2)</sup> reported that DFSP with a fibrosarcomatous lesion involving 5% or more of the tumor area (FS-DFSP) accounted for 9.85% of patients with DFSP. They indicated that the local recurrence and remote metastasis rates were 58 and 14.7%, respectively,

suggesting that the recurrence rate is higher than in patients with non-lesional DFSP.

In this article, we report a patient with FS-DFSP in the right buccal region, and review the literature.

### **CASE REPORT**

The patient was a 52-year-old male. He had a 2- or 3-year history of a subcutaneous mass in the right buccal region. As the mass had rapidly increased in size, he consulted a local clinic in December 2012. He had undergone an imaging study and biopsy in a local hospital. Based on the pathological findings, he

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Fig. 1. Photograph on the initial consultation  
A subcutaneous mass was noted (red round mark).

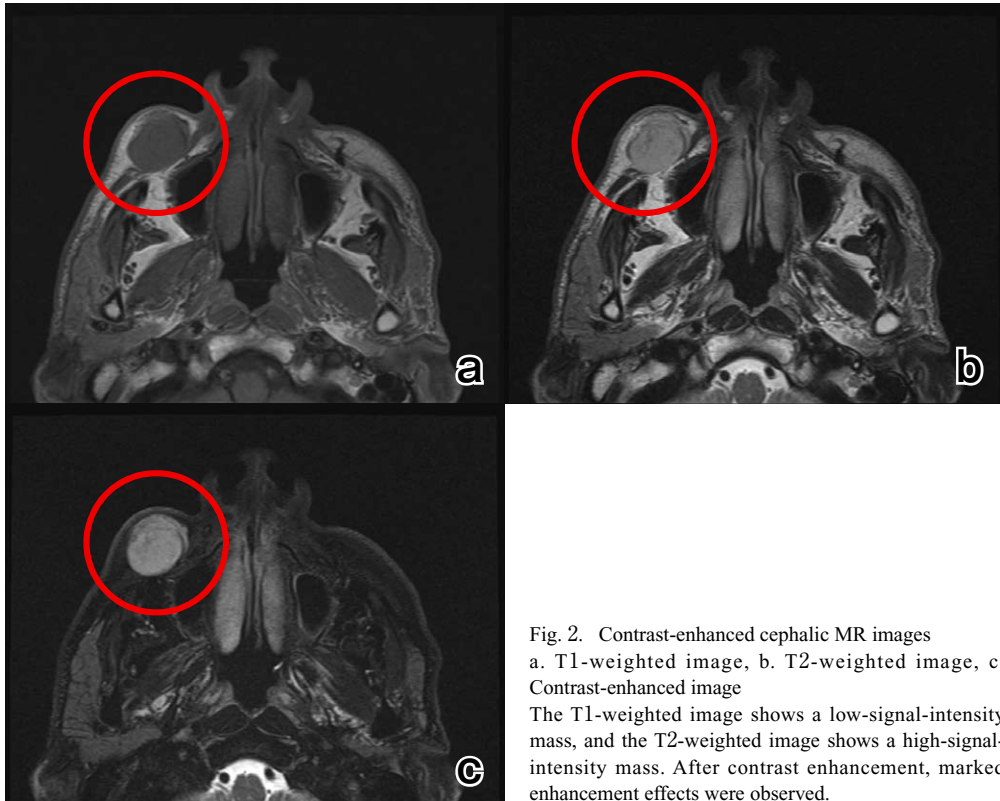


Fig. 2. Contrast-enhanced cephalic MR images  
a. T1-weighted image, b. T2-weighted image, c. Contrast-enhanced image

The T1-weighted image shows a low-signal-intensity mass, and the T2-weighted image shows a high-signal-intensity mass. After contrast enhancement, marked enhancement effects were observed.

was referred to our clinic for a suspected malignant soft tissue tumor, and the patient was referred to our department. On the initial consultation, an elastic-hard subcutaneous mass measuring approximately 5 cm was noted in the right buccal region (Fig. 1).

There was no assimilation between the mass and skin. There were no dermal changes. The mobility of the mass was favorable. There was no assimilation with the inferior base. Computed tomography (CT) and Magnetic resonance imaging

(MRI) were performed. T1-weighted images showed a low signal intensity in the right buccal region, and T2-weighted images showed a high signal intensity. After contrast enhancement, marked enhancement effects were observed (Fig. 2). Under a tentative diagnosis of a malignant soft tissue tumor, such as fibrosarcoma, resection was performed in January 2013.

Resection was conducted, with a 2-cm margin lateral to the tumor. At a site adjacent to the lower

eyelid, a 1-cm margin was established (Fig. 3a). Resection involving the cheekbone periosteum and large/small cheek muscles was performed, and the infraorbital nerve was amputated. It was possible to preserve the parotid duct (Fig. 3b). The site of the soft tissue defect was covered with artificial dermis. We planned to perform reconstruction after confirming the results of pathological examination.

As histopathological findings, the proliferation of spindle cells with oval nuclei was noted, showing

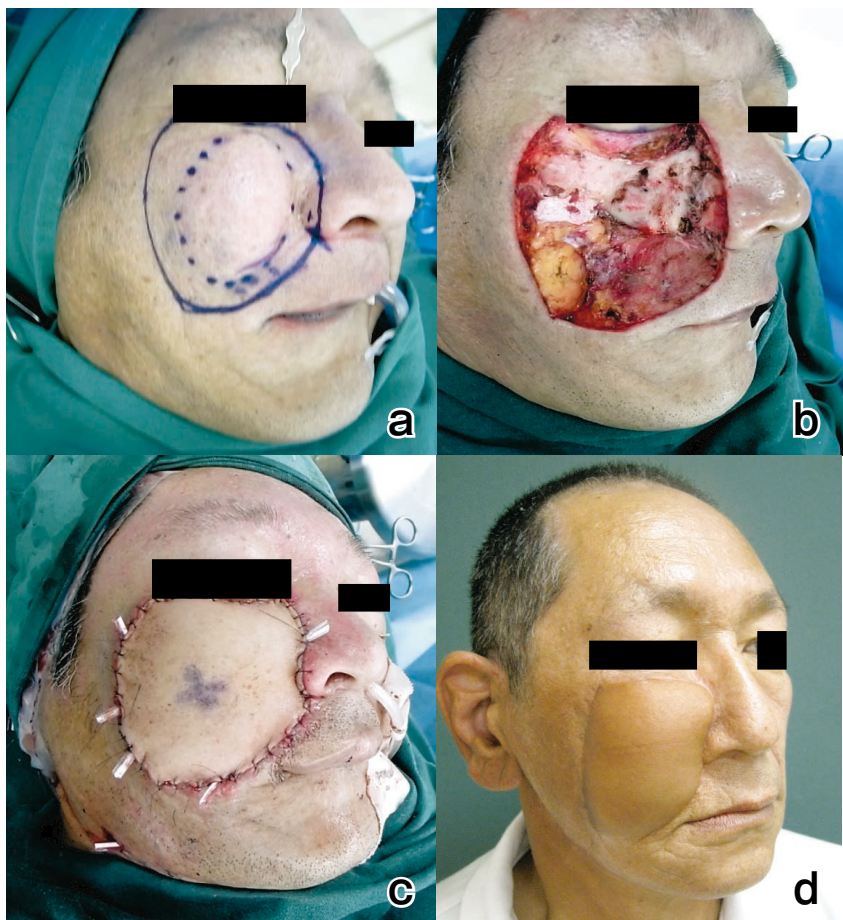


Fig. 3

a. Intraoperative photograph

The extent of resection was established, with lateral and lower eyelid margins of 2 and 1 cm, respectively.

b. After tumorectomy

Resection involving the cheekbone periosteum was performed.

c. After reconstruction of the right cheek with a flap

In the right buccal region, reconstruction was performed using a DIEP flap.

d. Condition 26 months after surgery

The buccal shape was favorable. There has been no relapse.

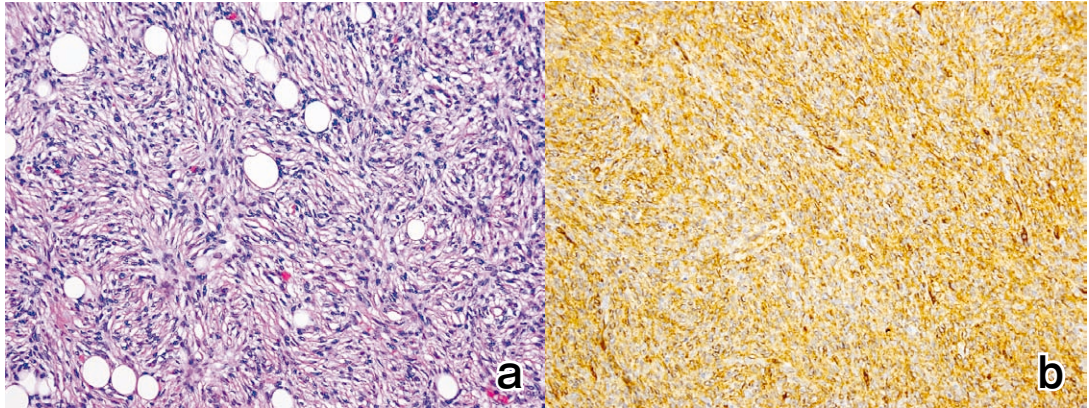


Fig. 4. Histopathological photograph (magnification  $\times 20$ )

a. HE staining

The proliferation of spindle cells was noted, showing a storiform pattern.

b. Immunostaining at the site of FS (anti-CD34 antibody)

At the site of FS, a diffusely positive reaction was also observed.

a storiform pattern. The border of the tumor was unclear, and adipose tissue infiltration was observed. Immunostaining showed a diffuse CD34-positive reaction, suggesting DFSP. However, the cellular density was high at the tumor center to margin. A herringbone pattern was partially noted. Based on these findings, the central lesion was considered to be a fibrosarcomatous change of DFSP (Fig. 4), leading to a diagnosis of FS-DFSP. The margin of the resected specimen at the inferior base was partially positive for tumor cells, whereas its lateral margin was negative.

In February 2013, reconstruction of the right cheek was performed. As the margin of the resected specimen at the inferior base was partially positive for tumor cells, the cheekbone surface was cut, and buccal reconstruction was conducted using a deep inferior epigastric artery perforator flap (DIEP flap). A flap was collected from the right abdomen, and anastomosed to the right facial artery/vein (Fig. 3c). The postoperative course was favorable. Complete graft survival was achieved. As postoperative adjuvant therapy, radiotherapy was applied to the right cheek at a total radiation dose of 60 Gy from April 2013.

There has been no relapse in the right buccal

region during the 26-month postoperative follow-up. As the flap was slightly over-sized, defatting was performed twice. Although irradiation-related pigmentation was observed in the right buccal region, apparent discomfort reduced. The results were cosmetically satisfactory (Fig. 3d). In the future, follow-up will be continued, considering local relapse and remote metastasis.

## DISCUSSION

DFSP was reported by Darier *et al.*<sup>3)</sup> in 1924 and by Hoffman<sup>4)</sup> in 1925. Remote metastasis is rare, but the tumor border is unclear, and local relapse may occur<sup>5)</sup>. As a rule, resection is performed. However, Goldblum *et al.*<sup>1)</sup> reported that the recurrence rate was 20% when establishing a surgical margin of 3 cm, whereas it was 41% when establishing that of 2 cm or less. DFSP frequently develops in the limbs and trunk. However, when it develops in the head and neck region, as demonstrated in the present patient, the surgical margin is insufficient in many cases. Especially in cases involving the face, the extent of resection is cosmetically and functionally limited, increasing the local recurrence rate<sup>6)</sup>. In the present case, the lateral space was sufficient, and a 2-cm margin was established, but,

on the cephalic side, a 1-cm margin was established to preserve the lower eyelid. Concerning inferior base margins, many studies have reported resection involving the fascia. In the present case, the tumor was adjacent to the cheekbone body, and resection involving the periosteum was performed. On pathological diagnosis, the peripheral margins were negative for tumor cells, but a portion of the inferior zygomatico-orbital margin was positive for them. Therefore, additional resection was conducted on buccal reconstruction. Thus, the tumor may have been completely resected. However, this patient was pathologically diagnosed with FS-DFSP. Mentzel *et al.*<sup>2)</sup> reported that the local recurrence and remote metastasis rates in FS-DFSP patients with FS changes involving 5% or more of the tumor area were higher than in those with non-lesional DFSP. Furthermore, Goldblum *et al.*<sup>7)</sup> performed extended resection (3 cm or larger) in 18 patients with FS-DFSP, and indicated that there was no remote metastasis. These results suggest the necessity of extended resection in patients with FS-DFSP. Postoperative adjuvant therapy may be necessary. William *et al.*<sup>8)</sup> reported that radiotherapy was useful for preventing relapse in patients whose surgical margin was positive for tumor cells or those in whom the cheekbone was adjacent to the tumor. We also selected adjuvant radiotherapy. Previous studies reported surgical margins of DFSP, but a consensus has not been reached. They should be decided in accordance with individual patients. In many patients with FS-DFSP, relapse is reportedly detected within 1 to 3 years after surgery. However, according to a study, relapse was detected after long-term follow-up<sup>2)</sup>. In the future, it may be important to continue strict follow-up over a long period, considering local relapse and remote metastasis.

In the present case, tumorectomy involving the buccal periosteum was performed, requiring reconstruction with a flap. Although reconstruction with skin grafting has also been selected for patients

in whom the periosteum is preserved, reconstruction with a flap may be cosmetically appropriate. Various studies have presented flap choices. However, we selected a DIEP flap. This is a perforating branch flap in which the deep inferior epigastric artery is used as a vascular pedicle, as reported by Koshima *et al.*<sup>9)</sup> in 1989. It has been applied in various fields<sup>10)</sup>. The reasons why this flap was selected included: (1) it is possible to harvest a flap in a supine position, and it is unnecessary to change the position during surgery, (2) the abdominal rectus muscle can be preserved, and a thin flap can be collected, (3) the site of flap harvest is discreet, and complications, such as abdominal wall hernia, may not occur, and (4) the anastomosed blood vessel length can be sufficiently maintained, and the vascular diameter is appropriate for vascular anastomosis. As a limitation, color matching with the face is unfavorable due to its abdominal origin. However, radiotherapy is performed after surgery, and it may be impossible to avoid flap pigmentation. As a result, pigmentation remained in the right buccal region, but the results were morphologically and cosmetically favorable.

### Funding

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### Conflict of interest

None

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