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## Aggressive giant dermatofibrosarcoma protuberance occurring in the parotid gland: Clinical case

**Relevance:** *Dermatofibrosarcoma protuberans (DFSP) is a cutaneous malignancy that arises from the dermis and invades deeper tissue. The cellular origin of DFSP is not clear. Evidence supports a fibroblastic, histiocytic, or neuroectodermal cellular origin.*

The article shares a clinical case of the treatment of aggressive dermatofibrosarcoma of the parotid salivary gland and describes the sequence of treatment and the treatment outcome.

**Results:** A 38 years-old woman with a slow-growing large right parotid mass underwent a total parotidectomy with auriculectomy and reconstruction using ALT flap. The surgery in the scope of a wide resection was followed by 4 cycles of chemotherapy and postoperative radiotherapy up to 40 Gy. The tumor recurrence after 10 months was treated by the second surgery followed by chemoradiotherapy. No new recurrence was observed in the following two years.

**Conclusion:** In the observed case, the giant tumor in the parotid salivary gland recurred after the initial surgery and adjuvant treatment. A tumor in the parotid region can recur despite aggressive initial treatment with wide resection and chemoradiotherapy.

**Keywords:** dermatofibrosarcoma protuberance, parotid salivary gland, CD 34, clinical case.

### Introduction

DFSP was first described in the literature in the early 1890s. In 1924, Darier and Ferran first described it as a progressive giant tumor disease, dermatofibroma.

Hoffman formally introduced the term "Dermatofibrosarcoma protuberance" (DFSP) in 1925 [1]. DFSP is a relatively rare low-grade soft tissue cancer. Large salivary gland sarcomas make up one-tenth of all mesenchymal cancers [2, 3]. Metastases are rare. DFSP is a locally aggressive tumor with a high recurrence rate [1]. The preferred method of treatment is surgical resection with negative margins [4, 5]. Several articles described adjuvant radiation therapy or chemotherapy performed in large incurable tumors [6]. Inhibitors such as the tyrosine kinase Imatinib or Sorafenib are more effective than other classical drugs [6]. We report a case of large DFSP with a rare localization in a parotid region. In parotitis, a salivary gland requires major resection; reconstruction is done with an external ALT patch. This case is reported in accordance with the SCARE criteria [7].

### Case report

A 38-year-old Russian woman complained of edema, slowly growing for eight years. Eight years ago, the patient underwent the parotid gland resection under local anesthesia at a local district hospital. Microscopic examination showed signs of angiofibrosis. A relapse happened after one year. The tumor histological examination revealed no homogeneous cells. Seven years later, the tumor status of the patient was rapidly growing. The general practitioner referred the patient to the Kazakh Institute of Oncology and Radiology. During the clinical examination, the tumor was positive, dense, slightly painful in the area of paroti-

tis, extending to the mastoid area and growing on a broad base of the atrial cartilage with a size of 18.0×8.0×9.0 cm. No paralysis of the face and growth of lymph nodes on the neck was registered (Fig. 1).



Figure 1 – The primary tumor in the patient's right parotid region

MRI showed a homogeneous mass on the right side with a positive margin, no soft tissue, the presence of non-neuronal areas of skin and subcutaneous fat. The features suggested sarcoma (Figure 2).

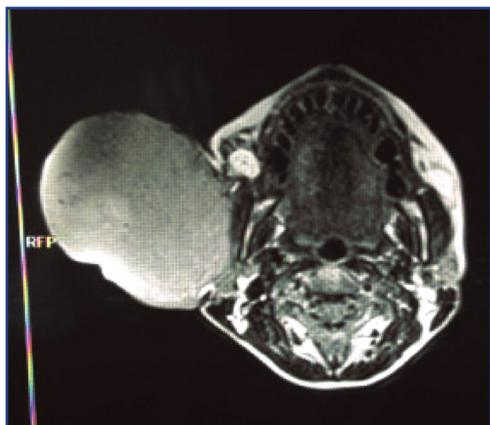


Figure 2 – DFSP formation on MRI image

A multidisciplinary group has decided on a right parotidectomy followed by the defect reconstruction by microvascular anastomosis of the pelvic flap.

Histopathological examination revealed short stem cells, hypercellular, moderate atypia, nuclear pleomorphism, and high mitotic activity with a pronounced storiform pattern (Figure 3). The tumor was CD34 positive on immunohistochemistry (Figure 4). The last histopathology evidenced a growing dermatofibrosarcoma. In the postoperative period, the patient decided to seek treatment at the regional oncological hospital where she received four cycles of dacarbazine 400 mg, doxorubicin 100 mg, and radiotherapy up to 40 Gy. The tumor recurred.

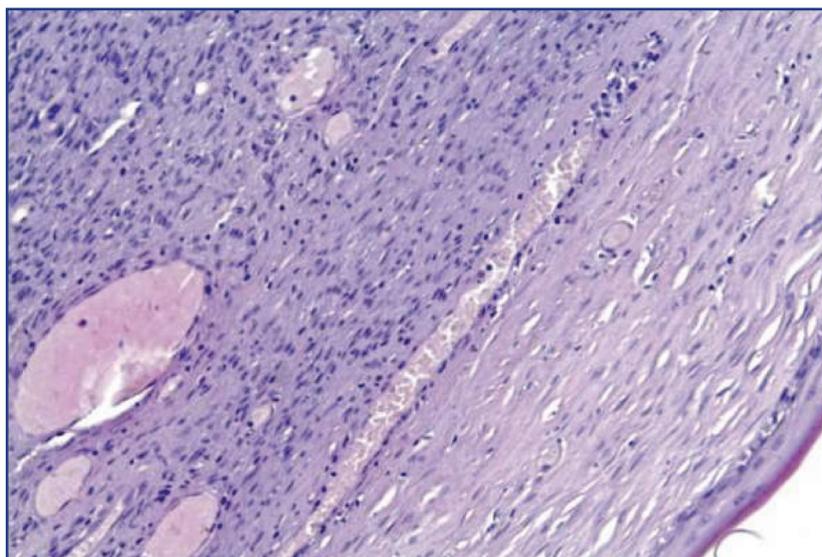


Figure 3 – Microscopic view of the tumor

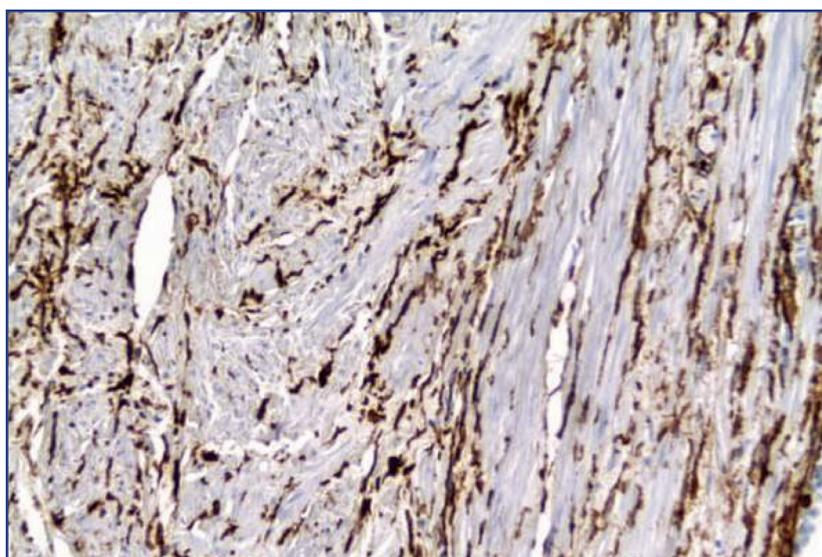


Figure 4 – Immunohistochemical analysis, CD200 x200, at ten months' follow-up

PET-CT revealed a 4 cm formation in the area of right mumps. The recurrent tumors were resected without complications (Figure 5). The microscopic image showed signs of dermatofibrosarcoma. After the second resection, the patient received adjuvant radiation therapy up to 80 Gy.

Tyrosine kinase inhibitors such as Imatinib or Sorafenib were not administered due to their absence at the regional cancer hospital. The patient refused to continue treatment in Almaty for private reasons. The patient was lost for follow-up for 28 months after the latest treatment.



Figure 5 – The tumor's view 10 months after surgery

### Discussion

DFSP is a tumor with fibrochimistosis, with high local recurrence, infiltrative margins, and rare metastases [8]. This tumor occurs mainly on the trunk and proximal parts of the legs and is prone to recurrence. The location on the parotid gland or mumps is very rare. The routine examination includes MRI (preferably) or CT and histological study of biopsy material. In DFSP, computed tomography showed a well-defined tumor mass, prone to hypotension, increased muscle tissue, and a uniform contrast enhancement. Homogeneous magnetic resonance imaging showed the tumor homogeneity and iso- or hypointensity in relation to muscle tissue on post-contrast enhanced T2 images [9]. Macroscopically, the tumor represents an improved and encapsulated mass. Histological signs that lead to cancer include a high mitosis rate (four or more mitoses in 10 high-capacity regions), hyperteliosis, mild atypia, and nuclear pleomorphism. DFSP is difficult to diagnose histologically because many tumors show a similar picture. DFSP differs from single fibrous tumors by homogeneity, no hemangioperetic pattern, and especially a fuzzy shape of the vessels around the vein. Therefore, immunohistochemistry should be used with caution when differentiating DFSP from fibroids. An effective fibrous histiocytoma may resemble DFSP but is usually negative. Schwannoma is positive for CD34, Bcl-2, and S-100 protein, which is not observed in DFSP [10]. DFSP is characterized by the actual translation t (17; 22) (q22; q13).

COL1A1-PDGFB is observed with the creation of a synthetic transcript [10].

The main treatment for DFSP is surgical resection with negative margins. In our case, due to the large size of the tumor, we decided to create an anastomosis to ensure gradual post-surgery restoration of the defect. Several publications reported the Mohs technique to reconstruct positive edges [11].

The 5-year survival rate for head and neck sarcomas is about 50%. Most authors that the degree, size, and depth

of sarcomas act as prognostic factors regardless of the tumor localization. The tumor causes many local recurrences in the head and neck area. In general, salivary gland sarcomas are aggressive tumors with a 40-64% recurrence rate, rare hematogenous metastases, and a mortality rate of 36-64%. Due to the small number of reported cases of DFSP in the parotid gland, the literature data is unclear. [2]. In our case, we observed a relapse after 11 months despite an aggressive postoperative treatment that included 4 courses of chemotherapy and radiation therapy up to 40 Gy. Secondly, a subsequent operation is performed only as adjuvant therapy [12]. Some studies also suggest adjuvant tyrosine kinase inhibitors [5, 6]. Relapses and metastases are possible over time.

### Conclusion

DFSP is a rare tumor with infiltrative margins, high local recurrence, and distant metastases. DFSP is rarely found in the parotid glands or parotid area and is more frequent on the trunk or extremities. Complete resection is an essential and basic treatment; other treatments' prognosis and effectiveness have not been confirmed. In large and recurring tumors, radiation therapy or chemotherapy can be administered. Frequent relapses require a long-term follow-up.

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