

## Dermatofibrosarcoma Protuberans of the Parotid Gland -A Case Report -

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Dermatofibrosarcoma protuberans (DFSP) typically presents during the early or mid-adult life, and the most common site of origin is the skin on the trunk and proximal extremities. DFSP of the parotid gland is extremely rare and only one case has been reported in the literature. We present here a case of a 30-year-old woman with DFSP occurring in the parotid gland, and we discuss the differential diagnosis. The patient is alive and doing well one year after her operation.

**Key Words :** Dermatofibrosarcoma Protuberans-Parotid Gland

Epithelial tumors make up the majority of salivary gland neoplasms, while mesenchymal tumors of this organ are uncommon. Dermatofibrosarcoma protuberans (DFSP) of the salivary gland is extremely rare and only one case has been reported in the parotid gland.<sup>1</sup>

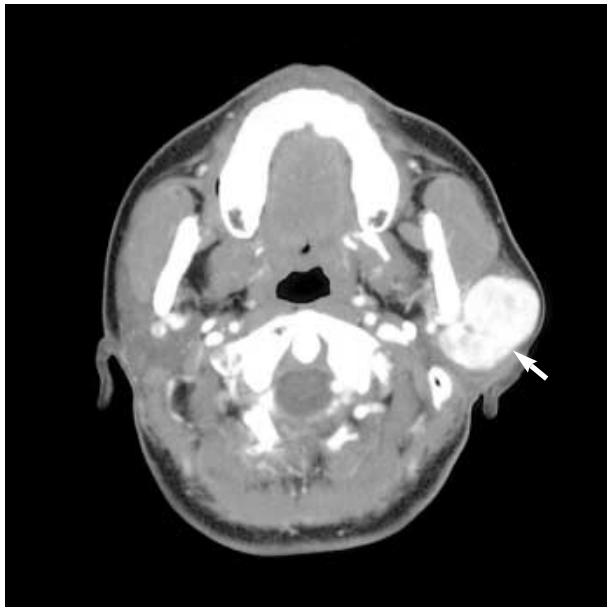
DFSP is a low-grade sarcoma with an infiltrative growth pattern and a high rate of local recurrence. It typically presents during early or mid-adult life as a nodular cutaneous mass.<sup>2-4</sup> Males are affected more frequently than females. Although these tumors may occur at almost any site, they most frequently arise in the soft tissue of the trunk (50 to 60%), followed by the proximal extremities (20 to 30%) and the head and neck (10 to 15%).<sup>5</sup> The ordinary DFSP is composed of uniform, bland spindle cells that are positive for CD34 and arranged in a distinct storiform or cartwheel pattern. Because of the rarity of this tumor in the salivary gland and the potential for erroneous diagnosis as another neoplasm, we present a case of DFSP of the parotid gland with a review of the relevant literature.

### CASE REPORT

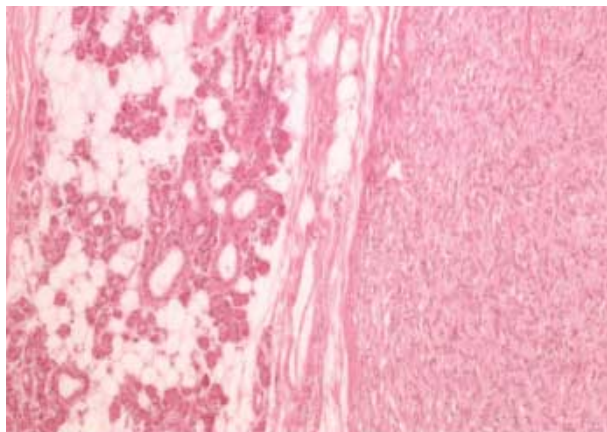
A 30-year-old woman came to the Otolaryngology Clinic at the Asan Medical Center with a 2-year history of a slowly enlarging mass inferior to the left angle of the mandible. On physical examination, the mass was located in the parotid gland and it was firm, mobile and nontender. The rest of the head and neck examination were normal. The chest X-ray and laboratory examination were unremarkable with no evidence of metastatic disease. A computed tomography scan showed a 4 cm sized, well-circumscribed mass in the parotid with no extraparotid extension being noted (Fig. 1). The fine needle aspiration biopsy revealed several clusters of spindle cells, suggesting a spindle cell neoplasm with uncertain malignant potential. The patient was treated with total parotidectomy. The postoperative course was uneventful and she is alive and doing well one year after the surgery.

A well-demarcated, round, firm mass was eccentrically present in the parotid gland, and it measured 4.3 cm in diameter. The mass was completely enclosed within a fibroadipose capsule of

the parotid gland and there was no evidence of extraparenchymal extension. The cut surface of the mass was whitish tan and slightly myxoid without any necrosis. It showed multifocal areas of hemorrhage, and there was no abnormality in the remaining salivary gland. Microscopically, the tumor was relatively well circumscribed from the surrounding parotid gland (Fig. 2). The tumor was uniformly cellular and consisted of short spindle cells arranged in a distinct storiform pattern with inconspicuous vasculature (Fig. 3). The tumor focally showed an infiltrative growth, but there was no necrosis. The tumor cells had slightly coarse nuclear chromatin with inconspicuous nucleoli (Fig. 4). There was mild nuclear pleomorphism present with low mitotic activity

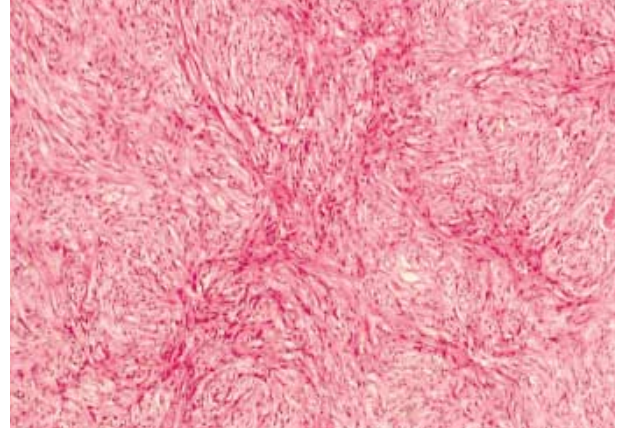


**Fig. 1.** A computed tomography scan shows a 4 cm sized, well-circumscribed mass in the parotid with no extraparotid extension (arrow).

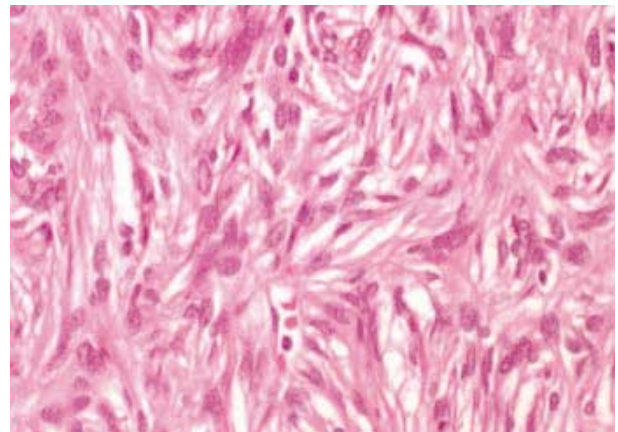


**Fig. 2.** The tumor in the parotid gland is well-demarcated from the surrounding parotid gland.

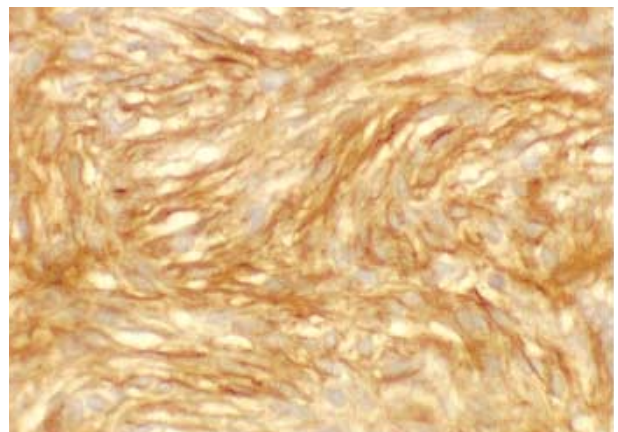
(1/10HPF). In certain areas, thick collagen bundles were scattered between the tumor cells. On immunohistochemical staining, the tumor cells exhibited diffuse and strong immunoreactivity for



**Fig. 3.** Spindle cells are arranged in a distinct storiform pattern.



**Fig. 4.** The tumor cells on the high power examination show short spindle cells with slightly coarse nuclear chromatin, no nucleolus, minimal pleomorphism and low mitotic activity.



**Fig. 5.** The tumor cells are intensely immunoreactive for CD34.

CD34 (Fig. 5), Bcl-2 and CD99. The cells were not immunoreactive for CD56, S-100 protein, smooth muscle actin and cytokeratin.

## DISCUSSION

Dermatofibrosarcoma protuberans (DFSP) was first described in 1924 by Darier and Ferrand<sup>6</sup> as a “progressive and recurring dermatofibroma”, and it is a low-grade malignant tumor that typically presents on the trunk and proximal extremities as a slow-growing, painless, firm, cutaneous plaque. However, unusual sites for DFSP including the vulva and parotid gland have been reported.<sup>6</sup> Histologically, DFSP is composed of a uniform population of slender fibroblasts arranged in a distinct storiform or cartwheel pattern around an inconspicuous vasculature, and this tumor is characterized by the nearly constant expression of CD34. This is an extremely rare case of DFSP arising in the parotid gland, because the tumor was completely enclosed within a fibroadipose capsule of the parotid gland, and it displayed not only the typical histologic features of DFSP, but also CD34 expression. Although the parotid gland is an unusual site for DFSP, careful histologic and immunohistochemical examinations can lead to the diagnosis of DFSP.

In the differential diagnosis, solitary fibrous tumor, benign fibrous histiocyoma, schwannoma, myoepithelioma, and other spindle cell sarcomas have to be considered. Solitary fibrous tumor is the most difficult entity to distinguish from this case because of the somewhat similar histologic findings and overlapping immunohistochemical staining results. Solitary fibrous tumor is characterized by a variety of growth patterns. There is frequently a mixture of cellular spindle cell areas with a distinct hemangiopericytic vasculature and foci of sclerosis or myxoid changes. DFSP, unlike solitary fibrous tumor, shows remarkable uniformity, a lack of the hemangiopericytic pattern and a distinct storiform pattern around an inconspicuous vasculature. Because of overlapping immunohistochemical results for DFSP and solitary fibrous tumor, caution is required in the differential diagnosis.

Benign fibrous histiocyoma may resemble DFSP, but it is usually negative for CD34 and Bcl-2. Schwannoma contains Antoni A and B areas, and it is S-100 protein positive which are not seen in DFSP. Other spindle cell tumors of the parotid gland in the differential diagnosis include spindle cell myoepithelioma, malignant peripheral nerve sheath tumor, fibrosarcoma and malignant fibrous histiocyoma. A proper use of immunohistochemical markers in conjunction with the characteristic light microscop-

ic features helps to differentiate these tumors from DFSP.

The histogenesis of this tumor is still controversial. Cytogenetic studies of DFSP have recently shown two consistent features: a reciprocal translocation,  $t(17;22)(q22;q13)$ , and supernumerary ring chromosome derived from the translocation,  $r(17;22)$ .<sup>7,8</sup> Both  $t(17;22)$  and  $r(17;22)$  result in a fusion of the collagen type I  $\alpha 1$  gene (COL1A1) on chromosome 17 and the platelet derived growth factor B-chain gene (PDGFB) on chromosome 22.<sup>9,10</sup> This gene rearrangement creates a chimeric COL1A1-PDGFB gene, which is believed to play an important role in the development of DFSP. As a consequence, the COL1A1-PDGFB molecular detection appears to be a powerful ancillary tool for the diagnosis of DFSP. In addition to the characteristic morphologic features and CD34 immunoreactivity, molecular studies will serve us in the future to enhance our understanding of DFSP.

In conclusion, DFSP is a rare mesenchymal tumor that is not restricted to the soft tissue. We report here on a case of DFSP arising in the parotid gland. Although the characteristic histologic and immunohistochemical features assisted us to conclude the diagnosis of DFSP, other spindle cell tumors occurring in the salivary gland should be excluded before making a diagnosis of DFSP.

## REFERENCES

1. Junaid TA, Ani AN, Ejeckam GC. Dermatofibrosarcoma protuberans in the parotid gland: a case report. *Br J Oral Surg* 1975; 12: 298-301.
2. Taylor HB, Helwig FB. Dermatofibrosarcoma protuberans: a study of 115 cases. *Cancer* 1962; 15: 717-25.
3. Burkhardt BR, Soule EH, Winkelmann RK, Ivans JC. Dermatofibrosarcoma protuberans: study of 56 cases. *Am J Surg* 1966; 111: 638-44.
4. McPeak CJ, Crux T, Nicastri AD. Dermatofibrosarcoma protuberans: an analysis of 86 cases-five with metastases. *Ann Surg* 1967; 166: 803-16.
5. Gloster HM. Dermatofibrosarcoma protuberans. *J Am Acad Dermatol* 1996; 35: 355-74.
6. Weiss SW, Goldblum JR. *Enzinger and Weiss's soft tissue tumors*. 4rd ed. St. Louis: Mosby, 2001; 491-507.
7. Naeem R, Lux ML, Huang SF, Naber SP, Corson JM, Fletcher JA. Ring chromosomes in dermatofibrosarcoma protuberans are composed of interspersed sequences from chromosome 17 and 22. *Am J Pathol* 1995; 147: 1553-8.
8. Pedeutour F, Simon MP, Minoletti F, *et al.* Translocation  $t(17;22)(q22;q13)$  in dermatofibrosarcoma protuberans. A new tumor-associated chromosome rearrangement. *Cytogenet Cell Genet* 1996; 72: 171-4.

9. O'Brien KP, Seroussi E, Dal Cin P, *et al.* Various regions within the alpha-helical domain of the COL1A1 gene are fused to the second exon of the PDGFB gene in dermatofibrosarcomas and giant-cell fibroblastoma. *Genes Chromosomes Cancer* 1998; 23: 187-93.
10. Simon MP, Pedeutour F, Sirvent N, *et al.* Dereglulation of the platelet-derived growth factor B-chain gene via fusion with collagen gene COL1A1 in dermatosarcoma protuberans and giant cell fibroblastoma. *Nature Genet* 1997; 15: 95-8.