

## CASE REPORT

# A Giant Undifferentiated Sarcoma of Parotid Gland: A Case Report and Review of Literature

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## ABSTRACT

Malignant spindle cell tumors of the parotid gland are a diagnostic challenge. The clinical and histologic assessments were consistent with a primary sarcoma of the parotid gland. In this article, we present a case of 79-year female with huge right parotid swelling which came out to be an undifferentiated sarcoma on histological examination only. The huge size of the lesion, rarity and less number of reported cases on undifferentiated sarcoma prompted us to report the particular case. The major differential diagnostic considerations were malignant fibrous histiocytoma (MFH), fibrosarcoma, osteosarcoma, spindle cell carcinoma, carcinosarcoma and primary undifferentiated sarcoma of the parotid gland.

**Keywords:** Parotid swelling, Malignant spindle cell tumors, Primary sarcoma.

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## INTRODUCTION

Parotid malignancies represent 1 to 3% of all head and neck cancers,<sup>1</sup> while sarcomas comprise only 0.3 to 1.5% of all major salivary gland neoplasms.<sup>2,3</sup> The most frequently described sarcomas are rhabdomyosarcoma and malignant fibrous histiocytoma (MFH). However, less commonly a variety of other tumors can occur, including leiomyosarcoma, neurosarcoma, angiosarcoma, osteosarcoma, fibrosarcoma, liposarcoma, hemangiopericytoma, Kaposi sarcoma, synovial sarcoma and alveolar soft sarcoma.<sup>2</sup> We report an unusually giant parotid undifferentiated sarcoma presenting at our institute.

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## CASE REPORT

A 79-year-old female presented to our institute with one year history swelling involving right parotid region extending to involve almost whole of anterior triangle and posterior triangle in neck. The mass has gradually progressive in nature and reached to present state in due course of time. It was 25 × 20 cm, nontender, local temperature was normal, bosselated surface, ill-defined margin, variegated consistency, decreased mobility, skin was involved (Fig. 1). The findings from the patient's oropharyngeal examination were normal; there was no contralateral mass in her neck and there was no facial nerve dysfunction. Chest radiographs, complete hemogram and blood chemistry values were within normal limits. Computed tomographic scan revealed a right parotid mass around 25 × 20 cm involving sternocleidomastoid muscle (Figs 2 and 3). Fine needle aspiration performed elsewhere revealed it to be an pleomorphic adenoma.

A right total parotidectomy with modified radical neck dissection was performed sacrificing sternocleidomastoid, involved skin and preserving spinal accessory nerve, internal jugular nerve. Facial nerve was sacrificed (Figs 4 to 7). The specimen was sent for tumor evaluation and margins of resection. The specimen weighed 3 kg and measured 24.6 × 19 cm (Fig. 8). Microscopic examination showed that individual tumor cells were spindle to polygonal in shape with abundant eosinophilic cytoplasm. They had pleomorphic nuclei with prominent nucleoli. Mitotic activity was moderate, with up to four mitoses per 10 high power fields.



**Fig. 1:** Large parotid mass

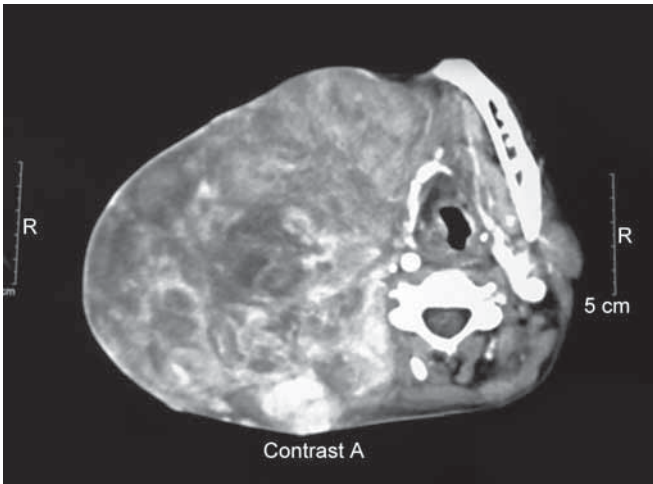


Fig. 2: Axial view

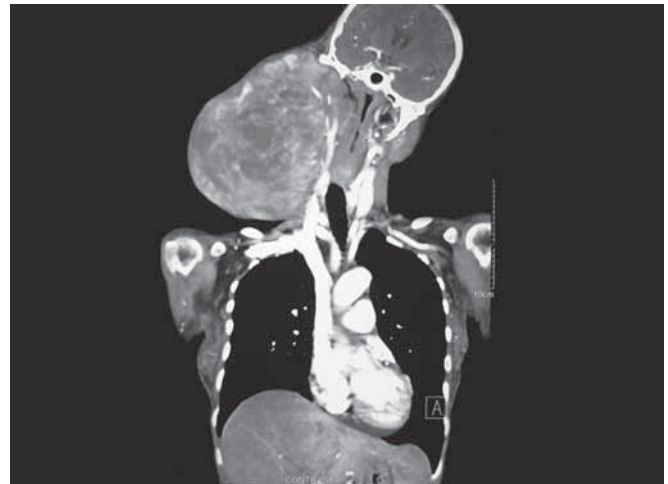


Fig. 3: Coronal view showing extension



Fig. 4: Incision placed



Fig. 5: Exposure of lesion

No definite necrosis was seen. Scattered patchy aggregates of lymphocytes were identified, and lymphoid aggregates with small germinal centers were present at the periphery of the tumor. The tumor consisted of solid sheets of cells completely effacing the normal parotid gland. The cells surrounded residual normal ducts. In most of the tumor, the cells were arranged in patternless sheets; however, in one area they formed a vague storiform pattern. The individual tumor cells were spindle to polygonal in shape with abundant eosinophilic cytoplasm. They had pleomorphic nuclei with prominent nucleoli. There was no vascular or lymphatic invasion.

The patient was subsequently treated with radiation and adjuvant chemotherapy with adriamycin (Fig. 9).

## DISCUSSION

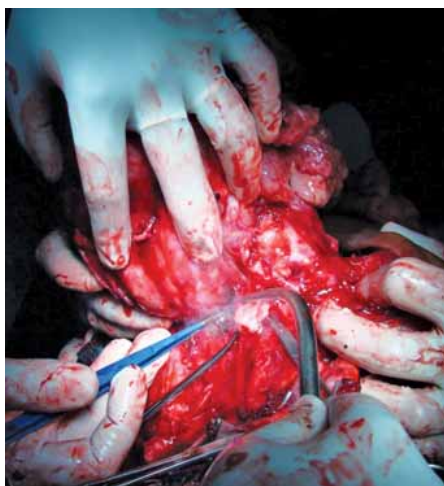
Malignant spindle cell tumors within salivary glands are a diagnostic challenge. The differential diagnosis is extensive, including carcinoma, carcinosarcoma, spindle cell carcinomas, melanoma, sarcoma of any origin and even lymphoma. These tumors may be primary or secondary.

Malignant fibrous histiocytoma can occur in deep soft tissues and in salivary glands.<sup>1</sup> Our tumor showed two microscopic features of MFH: cellular pleomorphism and a focal storiform pattern of growth. MFH, however, and in particular pleomorphic MFH, is a controversial entity.<sup>4</sup> Since, cellular pleomorphism was moderate and a storiform pattern was not prominent in our case, the overall features were felt to be inconsistent with MFH.

A fibrosarcoma was considered; however, the tumor lacked the typical histologic appearance of fibrosarcoma.

The possibility of an osteosarcoma was ruled out owing to the absence of bone formation and lacked the typical lace-like osteoid of osteosarcoma.<sup>7</sup> Carcinosarcoma, a biphasic neoplasm with malignant features in both the epithelial and stromal components, is also very rare in the parotid gland.<sup>5</sup> The absence of focal bone formation and lack of epithelial component ruled out the possibility of this diagnosis.

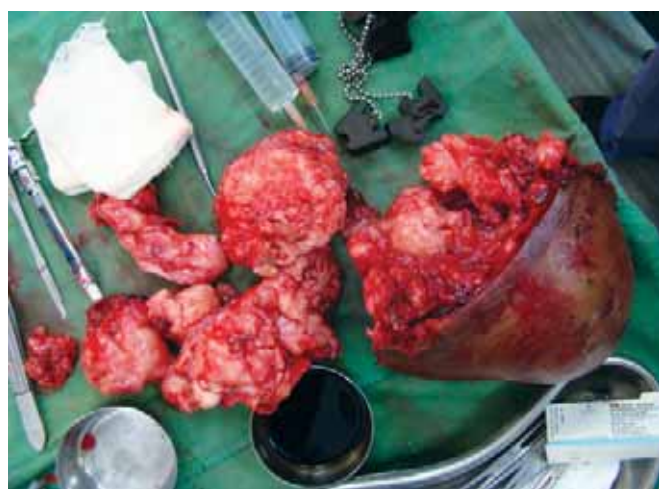
Spindle cell carcinomas are variants of squamous cell carcinomas with histologic features of both a squamous cell carcinoma and a spindle cell sarcomatoid tumor.<sup>6</sup> Histologically, there was no evidence of a squamous cell carcinoma



**Fig. 6:** Dissection in progress



**Fig. 7:** Delivery of specimen



**Fig. 8:** Excised lesion with involved skin



**Fig. 9:** Postoperative view

component in this case. This patient also had no history of a primary lesion elsewhere.

The criteria for a primary sarcoma in the salivary glands (as described by Luna et al) were met in this case.<sup>2</sup> Despite numerous imaging studies and investigations, a primary lesion elsewhere was not identified, therefore, to the best of our ability, we excluded a metastatic malignancy, either sarcoma or other tumor. The tumor originated in the parotid gland and was not an extension of a sarcoma from adjacent soft tissue.

Undifferentiated sarcomas involving major salivary glands are uncommon. In an early series by Auclair et al 367 sarcomas and sarcomatoid neoplasms of the major salivary gland regions were studied.<sup>3</sup> Ten of these cases were poorly differentiated sarcomas, unable to be further classified. In 1991, Luna et al reported a series of 11 primary sarcomas of the parotid gland: three MFHs, two neurosarcomas, two rhabdomyosarcomas, two fibrosarcomas and two osteosarcomas.<sup>2</sup> In their article, the authors summarized the reported literature in regard to primary sarcomas of the major salivary glands. Of the 74 identified cases,

only eight were sarcomas of undesignated type. Since this series, eight additional cases of major salivary gland sarcomas have been described in the literature: osteosarcoma<sup>7</sup> (4 cases), four leiomyosarcomas<sup>8,9</sup> (2 cases), rhabdomyosarcoma<sup>10</sup> (1 case) and chondrosarcoma<sup>11</sup> (1 case). In most instances, a sarcomatous lesion in a major salivary gland can be classified; however, a small number of cases defy precise classification.

In our case, the other problem was unusually giant size of neoplasm been harbored by the patient making surgical resection a challenge and at same time to preserve an uninjured facial nerve.

In summary, this case demonstrates an unusual giant undifferentiated parotid gland sarcoma. This case also illustrates the difficulty in evaluating malignant poorly differentiated neoplasms of salivary gland origin. The most important neoplasms to exclude are spindle cell carcinomas and carcinosarcomas. Careful assessment of the bone formation in such tumors is warranted, since the metaplastic vs neoplastic nature of the bone is important in ultimately deciding whether the tumor is an osteosarcoma.

## REFERENCES

1. Kane WJ, McGaffrey TV, Olsen KD, Lewis JE. Primary parotid malignancies. A clinical and pathologic review. *Arch Otolaryngol Head Neck Surg* 1991 Mar;117(3):307-315.
2. Luna MA, Tortoledo E, Ordonez NG, Frankenthaler RA, Batsakis JG. Primary sarcomas of the major salivary glands. *Arch Otolaryngol Head Neck Surg* 1991 Mar;117(3):302-306.
3. Auclair PL, Langloss JM, Weiss SW, Corio RL. Sarcomas and sarcomatoid neoplasms of the major salivary gland regions: a clinicopathologic and immunohistochemical study of 67 cases and review of the literature. *Cancer* 1986 Sep;58(6):1305-1315.
4. Fletcher CD. Pleomorphic malignant fibrous histiocytoma: fact or fiction? A critical reappraisal based on 159 tumors diagnosed as pleomorphic sarcoma. *Am J Surg Pathol* 1992 Mar;16(3):213-228.
5. Latkovich P, Johnson RL. Carcinosarcoma of the parotid gland: report of a case with cytohistologic and immunohistochemical findings. *Arch Pathol Lab Med* 1998 Aug;122(8):743-746.
6. Ishibashi T, Hojo H. Spindle cell carcinoma of the parotid gland. *J Laryngol Otol* 1995 Jul;109(7):683-686.
7. Stimson PG, Valenzuela-Espinoza A, Tortoledo ME, Luna MA, Ordonez NG. Primary osteosarcoma of the parotid gland. *Oral Surg Oral Med Oral Pathol* 1989 Jul;68(1):80-86.
8. Kang J, Levinson JA, Hitti I. Leiomyosarcoma of the parotid gland: case report and review of the literature. *Head Neck* 1999 Mar;21(2):168-171.
9. Oncel S, Daganay M, Ozer A, Arslanoglu S, Ermete M, Erdagan N. Leiomyosarcoma of the parotid gland. *J Laryngol Otol* 1996 Apr;110(4):401-403.
10. Gil-Benso R, Carda-Batalla C, Navarro-Fos S, Pellin-Perez A, Llombart-Bosch A. Cytogenetic study of a spindle cell rhabdomyosarcoma of the parotid gland. *Cancer Genet Cytogenet* 1999 Mar;109(2):150-153.
11. Bocklage T, Feddersen R. Unusual mesenchymal and mixed tumors of the salivary gland. *Arch Pathol Lab Med* 1995 Jan;119(1):69-74.