Malignancy of the Parotid Gland with Primary Symptoms of a Temporomandibular Disorder

Hizuru Miyamoto, DDS
Visiting Research Fellow
Oral and Maxillofacial Surgery Unit
The University of Adelaide
Adelaide, Australia

Staff Oral and Maxillofacial Surgeon
The First Department of Oral and Maxillofacial Surgery
Aichi-Gakuin University School of Dentistry
Nagoya, Japan

Hiroaki Matsuura, DDS
Visiting Research Fellow
Oral and Maxillofacial Surgery Unit
The University of Adelaide
Adelaide, Australia

Staff Oral and Maxillofacial Surgeon
The First Department of Oral and Maxillofacial Surgery
Aichi-Gakuin University School of Dentistry
Nagoya, Japan

David F. Wilson, MDS, FFOP (RCPA)
Associate Professor
Oral Pathology Unit
The University of Adelaide
Adelaide, Australia

Alastair N. Goss, DDSc, FRACDS (OMS), FICD
Professor and Director
Oral and Maxillofacial Surgery Unit
The University of Adelaide
Adelaide, Australia

Correspondence to:
Professor Alastair N. Goss
Oral and Maxillofacial Surgery Unit
Department of Dentistry
The University of Adelaide
Adelaide, South Australia
Australia, 5005
Fax: +61-8-8303-4402
E-mail: oral.surgery@adelaide.edu.au

The purpose of this case report is to highlight the possibility that malignant neoplasms of the deep lobe of the parotid gland may present as a temporomandibular disorder (TMD). Two patients who were eventually diagnosed with deep-lobe parotid tumors were retrospectively analyzed clinically, radiographically, and pathologically. For the first patient, there was a prolonged delay in establishing the correct diagnosis; for the second patient the delay was shorter. This was done primarily by computed tomographic examination. Despite aggressive surgical, radiotherapeutic, and chemotherapeutic treatment, both patients died from their malignant disease. All clinicians who treat TMD must be aware of the rare possibility of deep-lobe parotid tumors presenting as a TMD.
J OROFAC PAIN 2000;14:140–146.

Key words: temporomandibular joint disorders, parotid neoplasms

Parotid malignancies occur infrequently, accounting for only 1% to 3% of all head and neck cancers.[1] Because tumors of the parotid gland can sometimes show symptoms of a temporomandibular disorder (TMD), misdiagnosis is possible.[2] The specific reason for this is that the deep lobe of the parotid gland lies in a medial relationship to the mandibular ramus. Enlargement of the tumor places it directly next to the medial capsule of the temporomandibular joint (TMJ) and its innervation by the third division of the trigeminal nerve.

Sarcomas arising in the major salivary gland are exceedingly rare neoplasms, accounting for approximately 0.3% to 1.5% of all neoplasms of the salivary glands.[3–5] Malignant fibrous histiocytoma is the most frequently encountered sarcoma in the parotid gland. Other primary sarcomas of the parotid described in the literature are rhabdomyosarcoma, fibrosarcoma, neurosarcoma, and osteosarcoma.[4]

For a diagnosis of primary sarcoma of the major salivary glands to be made, 4 basic criteria should be met[4]:

1. The patient does not have, and has never had, a sarcoma elsewhere.
2. Metastasis to the gland from malignancies of the skin or mucosa of the upper respiratory/digestive tract should be excluded.
3. The gross and microscopic appearance should be consistent with an origin within the gland, rather than invasion of the gland by a sarcoma of the adjacent soft tissues.
4. Within the limits of microscopic study of multiple sections, a carcinosarcoma (so-called true malignant mixed tumor) must be excluded.

It is common for sarcomas in the parotid gland to present initially as an asymptomatic mass, followed by pain or tenderness.[6,7] A few patients may notice facial nerve paralysis if growth of their neoplasm is rapid.[8]

Undifferentiated carcinoma arising in the parotid gland is a rare type of neoplasm, accounting for approximately 3% of all neoplasms of the salivary glands.[9,10] Undifferentiated carcinoma arising in the parotid gland is a malignant epithelial tumor that consists of very poorly differentiated cells.[9] There is some difficulty in making a differential diagnosis of the tumor when encountering such poorly differentiated types as adenoid cystic carcinoma, epidermoid carcinoma, and adenocarcinoma. The undifferentiated carcinoma with a spindle-shaped figure may occasionally be mistaken for sarcoma of parotid gland origin.[9] Undifferentiated carcinoma of the salivary gland is associated with Epstein-Barr virus.[11] The prognosis of undifferentiated carcinoma of the salivary gland is poor.[12]

In this article, 1 case of sarcoma and 1 case of undifferentiated carcinoma originating in the deep lobe of the parotid gland in patients who presented with primary TMD symptoms are reported.

Case Reports

Patient 1

A 31-year-old woman who was medically fit initially consulted a specialist in private practice in Adelaide, Australia, in April 1993 with pain in the left TMJ. The patient had presented with a history of an acute sport injury to the left TMJ. The patient had not responded to non-surgical treatment over the previous 3 months, so the specialist performed a TMJ reconstruction with a fascial graft repair of the disc. Initially, the patient did well, with a good range of jaw movement, but some muscular pain remained. One year after the initial presentation, the patient reported ongoing pain and sought a second opinion. Examination of the patient at this time revealed tenderness, which was well-localized to the joint, and a limited range of jaw movement. There were no sensory or motor nerve deficits and no swelling. An arthrogram showed gross internal derangement and marked bony remodeling in the left TMJ. The joint was explored, and the gross derangement was confirmed. Discectomy was performed under anesthesia. A free-floating piece of tissue in the joint space was submitted to histopathology. This was thought both surgically and, subsequently, histopathologically to be a remnant of the fascial graft.

In the immediate postoperative period, the patient was pain-free and had a full range of jaw movement. Twenty-one months after the initial presentation and 9 months after the second surgical procedure, the patient presented again with the chief complaint of pain. The pain was diagnosed as consistent with muscle spasms. The patient also reported vague complaints of intermittent numbness of the lip.

Further non-surgical treatment of the masticatory muscles was performed for 3 additional months, at which point the area of numbness of the lip became more marked. Accordingly, computed tomography (CT) was performed, and it showed a large tumor in the left parapharyngeal space (Figs 1a and 1b). The tumor was reported as most likely representing an aggressive salivary gland neoplasm that arose in the deep lobe of the gland.

The patient's care was transferred to the University Teaching Hospital, where a full head and neck cancer workup was performed, followed by an extensive surgical procedure. The latter involved removal of the left mandibular ramus and TMJ, removal of the parotid gland with sacrifice of the facial nerve, and removal of an intracranial extension via the glenoid fossa. There was macroscopic surgical clearance. This was followed by 60 Gy of radiation.

Seven months after the surgery and radiotherapy, massive bilateral recurrence in the neck was clearly evident. The great vessels of the neck were completely surrounded by the tumor, which also extended down into the mediastinum. Palliative chemotherapy treatment with doxorubicin in 3-weekly cycles was performed. This slowed further development of the tumor, but it continued to grow. Five years after initial presentation, the patient died by exsanguination following erosion of the carotid vessels.

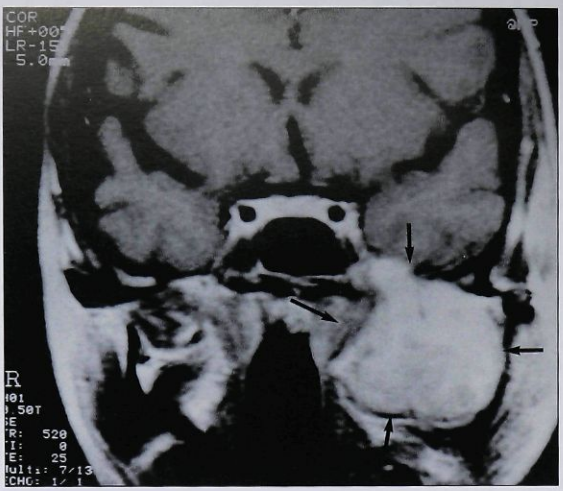


Fig 1a Patient 1. Coronal CT scan showing a large mass immediately medial to the left mandibular ramus (*arrows*). The base of the skull is eroded with an intracranial extension. The mandibular ramus is similarly eroded near the lingual. The parapharyngeal space is involved and the pharynx is deviated.

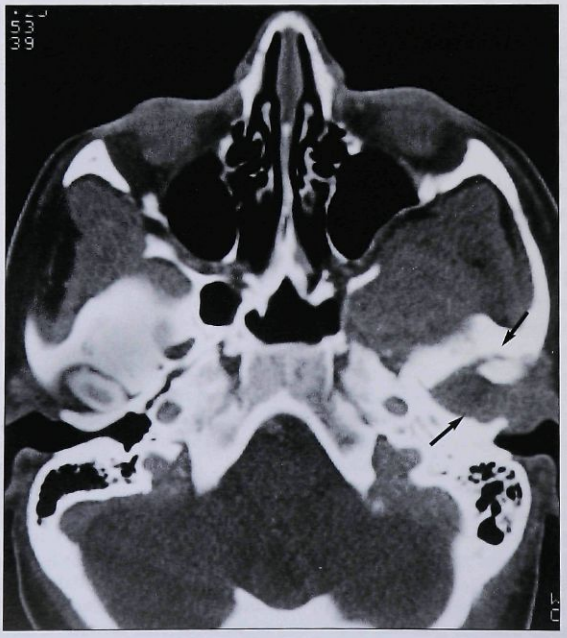


Fig 1b Axial CT scan showing the tumor extending into the posterior aspect of the left TMJ fossa (*arrows*).

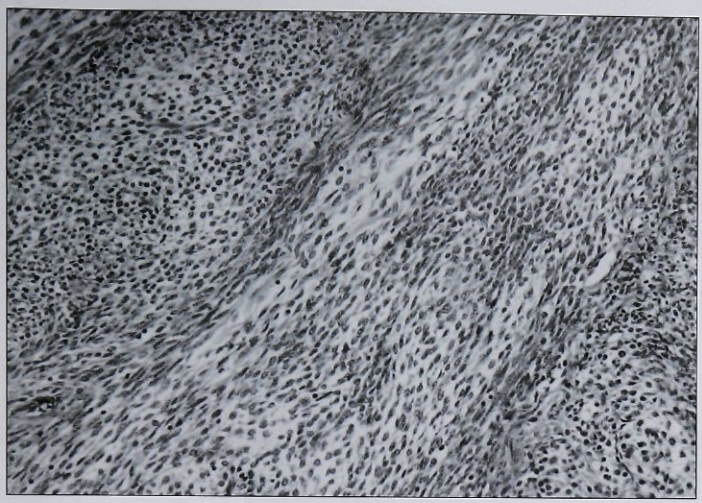


Fig 1c Histopathology of patient 1. This shows the cell-rich, fascicular pattern (hematoxylin-eosin stain; original magnification ×40).

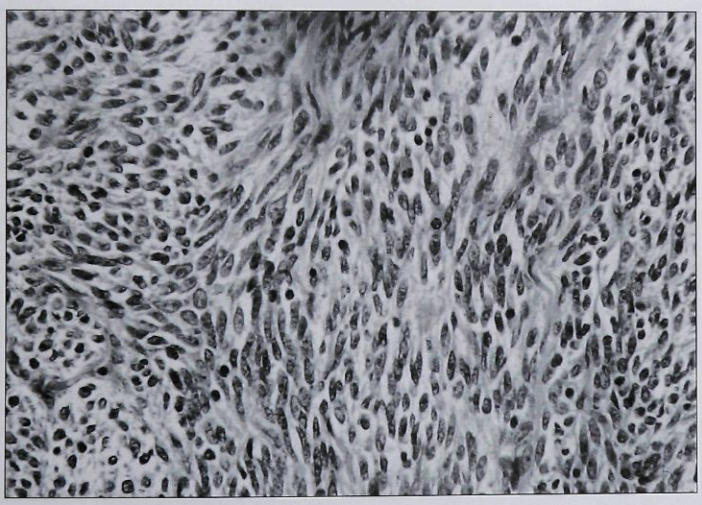


Fig 1d Histopathology showing the predominant spindle cell pattern of the tumor cells (hematoxylin-eosin stain; original magnification ×200).

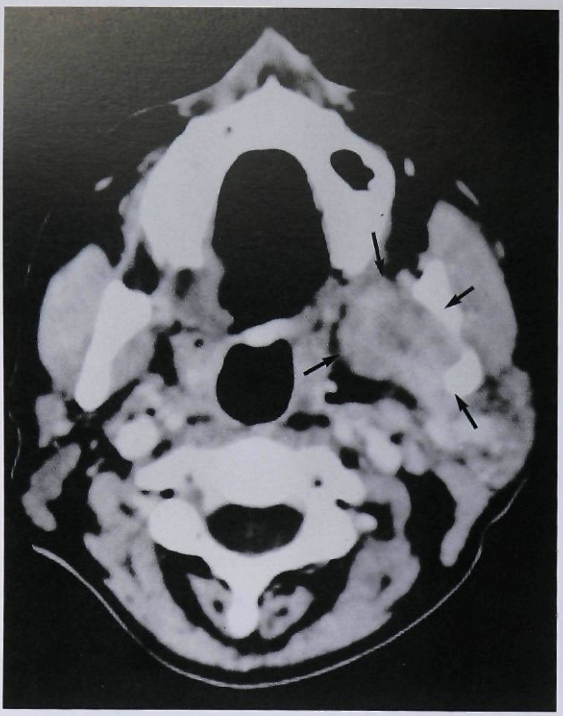
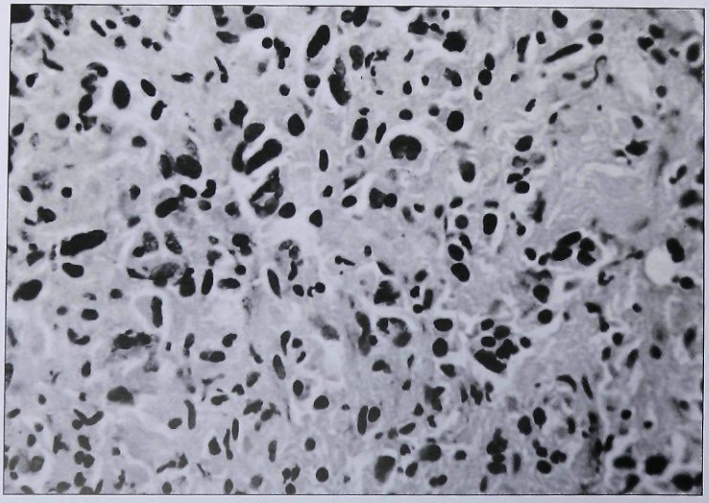


Fig 2a *(Left)* Patient 2. Axial CT scan showing a large mass immediately medial to the left mandibular ramus at the level of the sigmoid notch (*arrows*). The tumor extends behind the condyle. The parapharyngeal space is involved and the pharynx is deviated.

Fig 2b *(Below)* Histopathology of patient 2, showing a poorly differentiated malignancy with marked cellular nuclear pleomorphism (hematoxylin-eosin stain; original magnification ×400).



Pathologic Findings. Initial aspiration cytology done at the time of the CT scan showed fragments of highly cellular neoplastic tissue. Microscopic examination of tissue obtained from subsequent surgery and embedded in paraffin showed cell-rich, infiltrating fasciculi of plump, spindle-shaped cells (Fig 1c). The tumor cells appeared round to elongated and exhibited a predominance of nuclei, occasional nucleoli, and inconspicuous eosinophilic cytoplasm (Fig 1d). Mitoses were identified. Immunoperoxidase studies revealed positive staining for muscle-specific actin and alpha smooth muscle actin. However, the tumor cells tested negative for desmin, bovine keratin, and S100. Because of the equivocal immunoperoxidase results, the neoplasm was classified as a malignant spindle cell tumor, although the possibility of the lesion being of smooth muscle or of myofibroblastic or myoepithelial origin was considered.

Patient 2

A 60-year-old woman who was generally medically fit consulted a private prosthodontic practice in Adelaide in April 1997 with severe facial pain on the left side. The patient initially presented with an 8-week history of neck pain, which subsequently had become severe facial pain. Chiropractic, physiotherapeutic, and general medical treatment had not been beneficial. The patient had a previous history of osteoarthrosis of the neck and had mild TMJ problems.

Examination confirmed severe pain and tenderness in the left TMJ area. The patient could not open her mouth and had a slight facial weakness of the mandibular branch on the left side, but no swelling was present. A second opinion was sought and a CT arthrogram was performed. This showed an internally deranged disc with disc displacement. The CT scan was reported as showing no evidence of skull base pathology. A second CT scan of the head and neck was arranged but was delayed because of the presence of radiopaque contrast within the joint space from the CT arthrogram.

Shortly after this, the patient was admitted to a hospital due to severe pain, and arthrocentesis of the left TMJ was performed, with initial good results. A consultation was arranged with a specialist physician to address her progressive facial nerve weakness, and an orthopedic consultation was arranged to address her neck problem. Treatment as an idiopathic Bell's palsy was commenced, and the cervical spine was cleared for significant pathology. The delayed CT scan was then performed and showed a deep-lobe parotid tumor approximately 35 mm in diameter (Fig 2a).

The patient was transferred to the University Teaching Hospital, where a full head and neck workup was carried out. A left parotidectomy was then performed with sacrifice of the facial nerve. Access was gained by a mandibulotomy at the angle and fixed with a 4-hole plate. Less-than-optimal repositioning of the condyle in the fossa occurred. Subsequently, the patient experienced severe TMJ pain, and accordingly a discectomy was performed, along with a tarsorrhaphy to protect the left eye. A microscopic tumor was found that infiltrated the TMJ disc.

The patient was then scheduled for radiotherapy, but when this was commenced, it was noted that there was extensive mediastinal involvement. Palliative chemotherapy was arranged, but the patient died within 1 month from overwhelming head, neck, and chest involvement.

Pathologic Findings. Microscopic examination of the parotid gland and TMJ disc tissues revealed an infiltrative, poorly differentiated malignant tumor (Fig 2b). Cytologically, the tumor consisted predominantly of polyhedral cells exhibiting pronounced nuclear pleomorphism, hyperchromatism, and frequent mitoses. In some areas the lesion exhibited malignant spindle cells. The tumor tested positive for cytokeratins Cam 5.2 and bovine keratin but was negative for S100, smooth muscle actin, desmin, and HMB 45. A diagnosis of poorly differentiated carcinoma[13] of the parotid gland with invasion into the TMJ was made.

Discussion

Two rare cases of malignancy originating in the parotid gland are reported in this article. Both patients presented with primary TMD symptoms, the first being consistent with advanced internal derangement and the second with an acute, closed lock. Imaging studies directed at the joints confirmed the TMD, without evidence of more widespread pathology. Both patients were finally referred to one of the authors for diagnosis and management.

In the first case, the TMD was believed to be consistent with a previous temporofacial graft repair, and in the second case, it was associated with intense intra-articular pain and closed lock. In both cases, the tumors had microscopically invaded the intra-articular space.

When a sarcoma is located in the deep lobe of the parotid gland, a diagnosis of synovial sarcoma

should be considered. However, the characteristics of the tumor in patient 1 were not compatible with this diagnosis. In such cases, it is difficult to identify the origin of a tumor based solely on its anatomic location.[5] The basic 4 criteria for a diagnosis of primary sarcoma in the salivary glands were fulfilled in this case. Malignant spindle cells were found in the tumor. Thus, the overall findings confirmed a spindle cell sarcoma. Although immunohistochemistry may be of special value in excluding metastatic spindle carcinomas and melanomas,[4] further typing by immunoperoxidase studies was not possible in both cases. The cytologic features were not typical of either smooth muscle or nerve sheath origin in patient 1.

The first treatments performed for both patients were for TMD. In patient 1, non-surgical and surgical therapy appeared to be effective in the short term. This resulted in prolonged delay in the true diagnosis. In retrospect, this patient should have been diagnosed at the time of the second operation. The free-floating fragment of tissue did not show classical features of malignancy, and an altered fascial graft seemed a reasonable explanation to both the pathologist and the surgeon at the time. The patient had good symptomatic relief, presumably by the removal of the tumor's extension into the joint space. Arthrograms were abnormal but did not demonstrate the true nature of the pathology. Clinicians must be aware that conventional TMJ views, radioarthrotomograms, and CT arthrograms all cover a narrow field and do not clearly show the structures immediately medial to the joint. All the joint imaging for both patients was reviewed retrospectively by the treating clinicians, reporting radiologists, and independent experts, and even with the benefit of hindsight the medial tumor could not be positively identified. Magnetic resonance imaging (MRI) of the TMJ similarly does not show the medial structures, although the view to establish the angulation should alert the radiologist. There was a much faster progression of the tumor in patient 2, with prompt diagnosis. The precise pathology was recognized in both patients by head and neck CT scans and later by MRI.

Surgical treatment by a wide-field resection is required according to the clinical stage and histologic stage of the malignant tumor, usually with sacrifice of the facial nerve.[4] Postoperative adjuvant radiotherapy may be effective.[10] Prognosis in a reported series has generally been poor.[14,15] Both patients presented here received aggressive treatment, which slowly failed in the first patient and was rapidly shown to be ineffective in the second patient.

All practitioners who treat TMD must be aware of the occasional possibility of deep-lobe parotid tumors mimicking TMD. Reliance on imaging the TMJ alone may occasionally be a fatal mistake.

Acknowledgments

We would like to thank Dr A. Whyte, radiologist, for the imaging, and the Head and Neck Cancer Team for multidisciplinary management.

References

1. American Cancer Society. Cancer Facts and Figures. New York: American Cancer Society, 1979:62–66.
2. Sakashita H, Miyata M, Miyamoto H, Kurumaya H. Glomus tumor origination in the parotid region. J Oral Maxillofac Surg 1995;53:830–834.
3. Eneroth CM. Histological and clinical aspects of parotid tumors. Acta Otolaryngol Suppl (Stockh) 1964;191:5–99.
4. Luna MA, Tortoledo ME, Ordonez NG, Frankenthaler RA, Batsakis JG. Primary sarcoma of the major salivary glands. Arch Otolaryngol Head Neck Surg 1991;117:302–306.
5. Sakashita H, Miyata M, Miyamoto H, Minato H. Adenocarcinoma originating in the sublingual gland: Report of a case. J Oral Maxillofac Surg 1997;55:764–767.
6. Massarelli G, Tanda F, Vincenzo F, Oppia L. Hemangiopericytoma of the parotid gland. Virchows Arch [A] 1980;368:81–89.
7. Auclair PL, Langloss JM, Weiss SW, Corio RL. Sarcoma and sarcomatoid neoplasms of the major salivary gland region. Cancer 1986;58:1305–1315.
8. Sakashita H, Miyata M, Okabe K, Miyamoto H, Saito K, Tsuji M, et al. Clinical evaluation of treatment of space-occupying lesion in the parotid gland. Jpn J Oral Diagn Oral Med 1997;10:397–403.
9. Nagao K, Matsuzaki O, Saiga H, Sugana I, Shigematsu H, Kaneko T, et al. Histopathologic studies of undifferentiated carcinoma of the parotid gland. Cancer 1982; 50:1572–1579.
10. Kane WJ, McCaffrey TV, Oslen KD, Lewis JE. Primary parotid malignancies: A clinical and pathological review. Arch Otolaryngol Head Neck Surg 1991;117:307–315.
11. Sheen TS, Tsai CC, Ko JY, Chang YL, Hsu MM. Undifferentiated carcinoma of the major salivary glands. Cancer 1997;80:357–363.
12. Hui KK, Kuna MA, Batsakis JG, Ordonez NG, Weber R. Undifferentiated carcinomas of the major salivary glands. Oral Surg Oral Med Oral Pathol 1990;69:76–83.
13. Seifert G, Sobin LH, Thackray AC. Histological Typing of Salivary Gland Tumours. Berlin, New York: Springer-Verlag, 1991:17–31.
14. Stimson PG, Valenzuela-Espinoza A, Tortoledo ME, Luna MA, Ordonea NG. Primary osteosarcoma of the parotid gland. Oral Surg Oral Med Oral Pathol 1989;68:80–86.
15. Yeh CK, Fox PC, Fox CH, Travis WD, Lane HC, Baum BJ. Kaposi's sarcoma of the parotid gland in acquired immunodeficiency syndrome. Oral Surg Oral Med Oral Pathol 1989;67:308–312.