Synovial sarcoma of the temporomandibular joint area: report of a case

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Synovial cell sarcoma is a relatively rare tumor of mesenchymal origin. It is a high-grade neoplasm that microscopically shows a monophasic or biphasic cellular pattern and includes epithelial features as well as supporting tissue features. Surgical excision is the primary mode of treatment. Postoperative radiotherapy and chemotherapy also is seen to be helpful. Between 3% and 10% of cases originate in the head and neck. A review of relevant literature shows less than 10 cases of synovial cell sarcoma of the temporomandibular joint area reported in the English literature. We report an additional case of biphasic synovial cell sarcoma arising in the temporomandibular joint area, which caused ear pain, tinnitus, and hearing loss, and we further discuss the clinical features, histopathology, differential diagnosis, and treatment modality. **(Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2007;104:e62-e65)**

Synovial cell sarcoma is a malignant neoplasm that arises not from synovial membrane, but rather from pluripotent mesenchymal cells near or even remote from the articular surface that is most commonly found in the extremities.[1] It comprises 8% to 10% of all soft tissue sarcoma, and only 3% to 10% of synovial cell sarcoma occurs in the head and neck.[2] The most common sites in the head and neck are the hypopharynx and retropharynx. It is an extremely rare tumor of the temporomandibular joint area. To our knowledge, there are only 7 cases of synovial cell sarcoma involving the temporomandibular joint area reported in the English literature.[2-4] We present an additional case and review the clinical features of this disease.

CASE REPORT

A 21-year-old woman appeared at the Oral and Maxillofacial Surgery Department of Mackay Memorial Hospital with the chief complaint of a tender mass over the right preauricular region for approximately 3 years. The discomfort over the right temporomandibular region was noted for a long time, and the patient paid no attention to it until the mass became progressively larger with increased tenderness. She had visited a local otolaryngologist's clinic several days before for the ear pain, tinnitus, hearing loss, and mouth opening limitation and was referred to our department. She denied any history of trauma, facial surgery, bruxism, or clenching habits and had not had occlusal appliance therapy or any other form of nonsurgical care. Her medical history was noncontributory except for systemic lupus erythematosus. Physical examination on admission revealed a tender, nonfluctuant mass in the right temporomandibular area about 3 cm in diameter. The skin overlying the swelling was intact without ulceration or induration. The range of mouth opening was limited to about 20 mm.

A computed tomographic scan revealed a focal soft-tissue enhancing tumor near and inferior to the right temporomandibular joint area posterior to the condylar neck and ramus of the right mandible. Significant ossification or calcification was seen in the periphery of this lesion. No destruction of the adjacent right mandible was seen (Fig. 1). Panoramic radiography revealed a well-circumscribed mixed radiolucent-radiopaque mass over the posterior border of the mandible, causing a depression in the bone (Fig. 2). There was no evidence of lymphadenopathy.

A fine needle biopsy was performed, which revealed a spindle cell tumor, favoring a benign stromal neoplasm with ossification or with reactive bony formation at the elevated periosteum.

Treatment, including surgical procedure, was discussed with the patient, and she elected to undergo operative excision. The local excision was performed through the preauricular incision. The histological examination revealed features of synovial sarcoma with marked osseous metaplasia at the periphery. Microscopically, a biphasic tumor was identified, with predominance of hypercellular fairly uniform hyperchromatic spindle cells with fibrosarcomalike features, which

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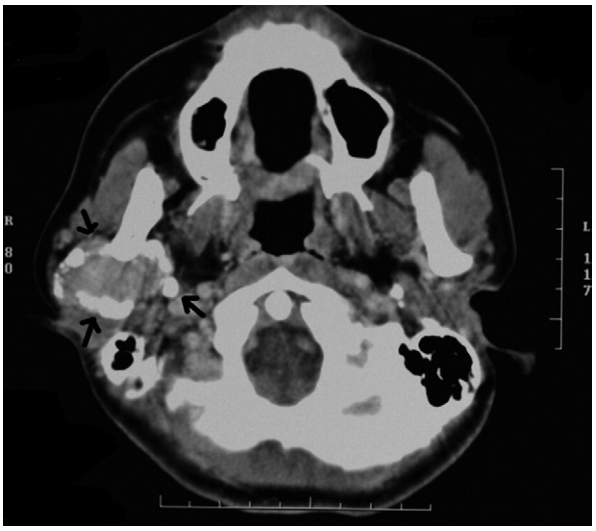

Fig. 1. Axial computed tomography reveals a focal soft-tissue enhancing tumor near and inferior to the right temporomandibular joint (*arrows*). Extensive ossification or calcification is seen in the periphery of this lesion.

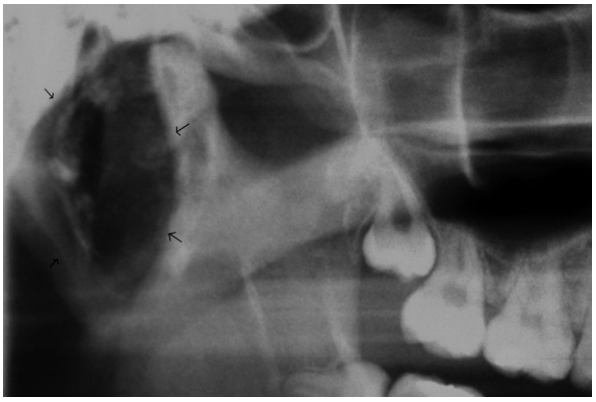

Fig. 2. Panoramic radiograph reveals a well-circumscribed, mixed radiolucent-radiopaque mass on the posterior border of the mandible, measuring about 3 × 4 cm (*arrows*).

occasionally contained epithelial-type tumor cells that formed round or slender to irregular glands (Fig. 3, *A*). Marked ossification forming delicate or compact bony trabeculae was seen in the hard and bony pieces (Fig. 3, *B*). The ossification was formed abruptly from the spindle cells without apparent osteoid formation like that seen in conventional osteosarcoma, and the term *osseous metaplasia* was used. The spindle cells were strongly stained for CD99 (Fig. 4, *A*), whereas the epithelial type of tumor cells and small numbers of spindle cells were positive for cytokeratin antigen 7 (Fig. 4, *B*) and epithelial membrane antigen (Fig. 4, *C*). The tumor cells were negative for CD34 or smooth muscle actin.

The postoperative recovery of the patient was uneventful, and all clinical signs subsided. She was discharged 10 days

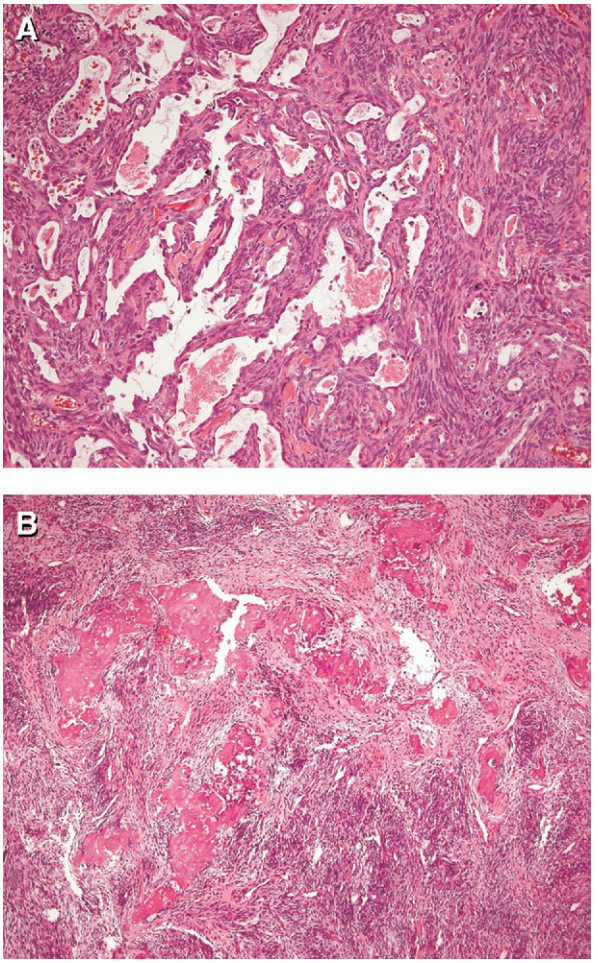

Fig. 3. Photomicrography. **A,** Plump spindle cells surround isolated glandlike epithelial component (hematoxylin-eosin, original magnification ×100). **B,** Marked ossification forming delicate or compact bony trabeculae is seen in the hard and bony pieces (hematoxylin-eosin, original magnification ×40).

after the surgical procedure. During this admission, the routine tumor workup was arranged and revealed no evidence of distant metastasis. Adjuvant radiotherapy for 64.5 Gy and chemotherapy with cisplatin, epirubicin, and ifosfamide for 4 courses was completed to prevent distant metastasis and tumor recurrence. The patient has remained free of recurrent tumor 5 years after discharge. A subsequent panoramic radiograph revealed that the previous depression of the mandible was remodeling.

DISCUSSION

Synovial cell sarcomas are found in younger patients between the ages of 15 and 40 years, 90% are younger than 50 years. Males appear to be slightly more susceptible than females; the average male to female ratio is 1.2:1. Synovial sarcomas usually occur in paraarticular locations of the lower extremities.[3-6] The most com-

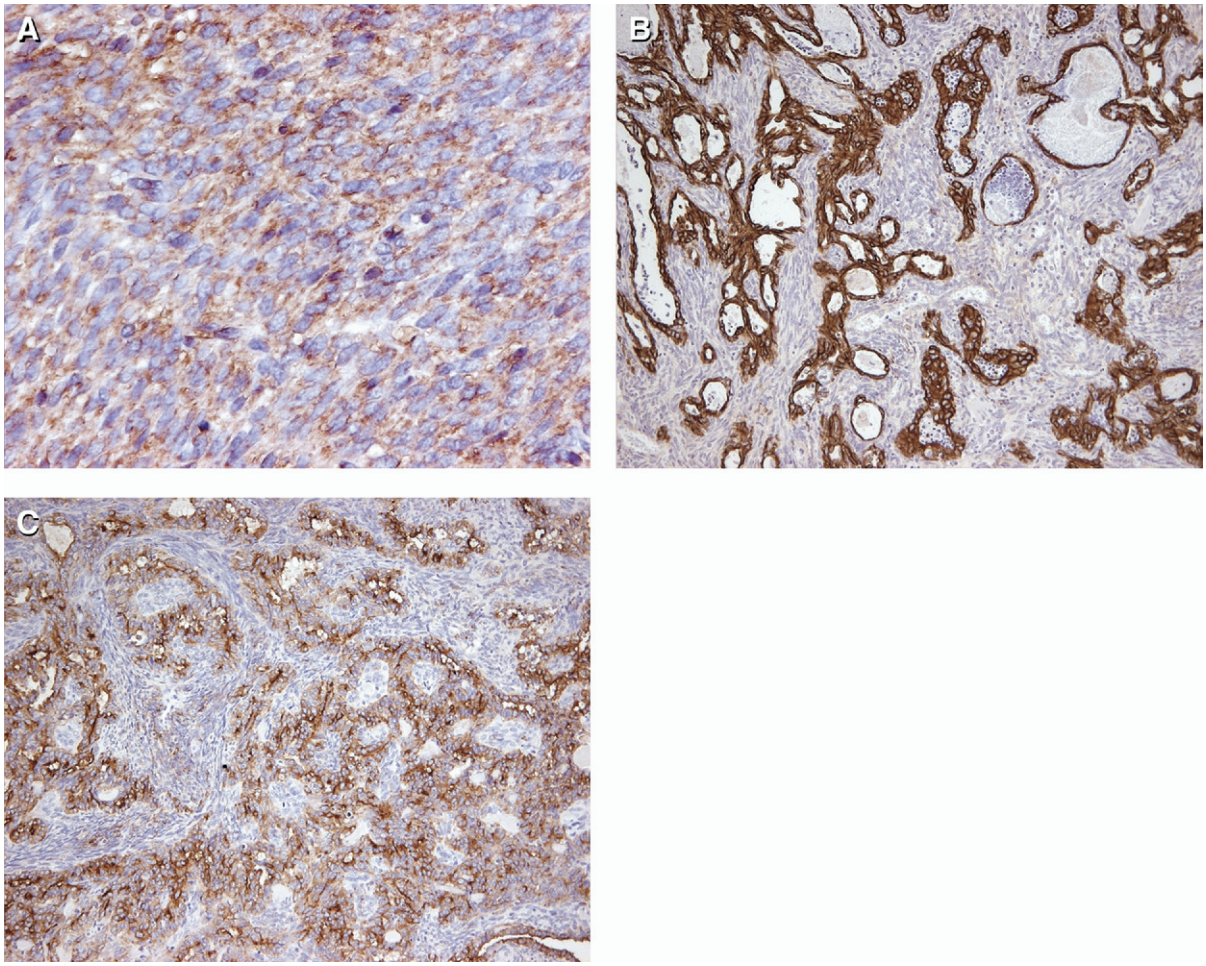

Fig. 4. Immunohistochemical stain. **A,** The spindle cells are strongly stained for CD99 immunoreactivity (original magnification ×100). **B,** The epithelial type of tumor cells and small numbers of spindle cells are positive for cytokeratin-7 immunoreactivity (original magnification ×100). **C,** Epithelial type of tumor cells is also positive for epithelial membrane antigen immunoreactivity (original magnification ×100).

mon sites in the head and neck are the hypopharynx and the retropharynx. It is an extremely rare tumor of the temporomandibular joint area. There have been less than 10 reported cases of synovial sarcoma arising in the temporomandibular joint area.[2,3]

The chief complaint of most patients is a progressively enlarging painless mass of several months' duration. In the head and neck region, other findings include a painful mass, dysphagia, hoarseness, and dyspnea, accompanied by some limited range of motion.[3-5] The preoperative duration of symptoms is highly variable and is most commonly 2 months to 4 years. A history of trauma to the involved area does not appear to be a predisposing factor.[3,4]

The computed tomographic imaging appearance of synovial sarcomas of the head and neck has been described as multilocular tumor with smooth margins and heterogeneous enhancement after injection of contrast medium. It may appear benign in some cases when a well-demarcated, homogeneous lesion with calcification is found in the head and neck.[7]

Findings at surgical removal yield well-circumscribed spherical multinodular or lobulated tumors covered by a thin fibrous membrane or pseudocapsule. Cystic and hemorrhagic foci are present.[5] The histopathologic findings are the same for synovial sarcomas arising in the head and neck and the extremities. Synovial sarcomas classically show a biphasic cellular pattern consisting of a stroma of fibroblast spindlelike cells in which are scattered pale epithelial-like cells arranged in glandular formations, nests, or cleftlike spaces. The epithelial-like cells may be cuboidal or columnar and form papillary projections. Synovial sarcoma consisting of only 1 cell type, either spindle or

epithelial, has been described (the monophasic variant).[1,3,5,8] Microcalcification has been found in some cases.[5] Pathological diagnosis is often difficult. It can be mistaken for epithelioid sarcoma, fibrosarcoma, malignant glandular schwannoma, malignant schwannoma, clear cell sarcoma, hemangiopericytoma, eccrine spiradenoma, mesothelioma, pigmented villonodular synovitis, as well as spindle cell carcinoma and metastatic carcinoma.[4-6] In addition to clinical information, a number of immunohistochemical stains are available to facilitate the diagnosis.[4]

Cytogenetic unity is also supported by the observation that a specific reciprocal chromosomal translocation t(X;18)(p11.2;q11.2) is found in each form of synovial sarcoma.[8,9] The presence of this translocation confirms the diagnosis of difficult cases that show unusual histological features or are located in unusual sites. Chromosome 18 contains the *SYT* gene, which fuses with *SSX1* or *SSX2* from chromosome X. The *SYT-SSX1* fusion is associated with biphasic variants; *SYT-SSX2* fusion is associated with monophasic variants.[10]

Wide excision with negative margins remains the mainstay of therapy. Most metastases are blood borne. Regional lymph node dissection is not believed to be necessary unless clinical lymphadenopathy or enlarged nodes on imaging studies are present.[1,3-5] With local excision as the only mode of treatment, recurrence rates as high as 60% to 90% have been reported in the head and neck region.[1,2,4] Postoperative radiotherapy is advocated for improved local control rates. Large doses in the range of 65 Gy or more is considered to be necessary, but radiotherapy alone is not recommended.[3-5,11,12] Chemotherapy has been used in an attempt to prevent the occurrence of distant metastases in advanced disease or in patients with residual gross tumor.[3,4] Metastases are developing in approximately 50% of patients and are most often found in the lung. Other favored sites of tumor metastasis are lymph nodes and bone marrow.[4] Although not normally a feature of sarcomas, metastasis of regional lymph nodes occurs in 12.5% of cases of the head and neck and 23% of cases in the extremities.[8]

The prognosis of head and neck synovial sarcoma is poor; 5-year survival rates range from 36% to 76% and 10-year survival rates range from 11% to 63%.[3,12] The prognosis of head and neck synovial sarcoma is poor; 5-year survival rates range from 36% to 76% and 10-year survival rates range from 11% to 63%[3,12]; overall survival rates for synovial sarcoma are 55% survival at 5 years and 38% survival at 10 years.[1] Favorable prognostic factors appear to be early diagnosis and the performance of wide surgical excision. The size of the primary tumor and the depth of invasion may be the most important prognostic indicators. Patients whose primary tumor is larger than 4 cm in diameter have a poorer outcome. Calcifying-type synovial sarcoma has been reported to have a better prognosis.[1,5]

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