Synovial Sarcoma of the Head and Neck

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- Synovial sarcoma, a malignant soft-tissue tumor that occurs primarily in the extremities, is seen rarely in the head and neck. Although known to behave aggressively in the extremities, recent reports suggest that the course of synovial sarcoma may be more indolent when it occurs in the head and neck. Five new cases of synovial sarcoma of the head and neck are described herein. Of four patients with adequate follow-up, three died of pulmonary metastases at 1, 2 1/2, and 8 years following initial treatment. The fourth patient is alive with disease at the primary site six years after diagnosis. The poor prognosis for patients with this disease suggests that aggressive treatment is necessary.

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Synovial sarcoma, a malignant soft-tissue tumor that occurs primarily in the extremities, is rarely seen in the head and neck. In 1927, Lawrence Weld Smith termed this lesion synovialoma, imparting a benign connotation. The truly aggressive nature of this malignancy was not recognized until many years later. Synovial sarcoma accounts for 8% to 10% of all sarcomas. It was first described in the head and neck in 1954, when Jernestrom reported a case of synovial sarcoma of the pharynx. Fewer than 50 head and neck synovial sarcomas have been reported since. The limited clinical experience with this lesion in the head and neck is in sharp contrast to a much greater experience in the extremities.

Several large series of synovial sarcoma of the extremities have been described. Cadman et al. reported 134 cases of synovial sarcoma at all sites with a 25% five-year survival. An additional 136 cases, with a five-year survival of 40%, were contributed by Hajdu et al. The two largest series of synovial sarcoma of the head and neck are collections of case reports from a number of different centers. Roth et al. cited a 47% five-year survival among 22 patients with synovial sarcoma of the neck. Eleven cases of orofacial synovial sarcoma were described by Shmookler et al., but a 66% survival in nine patients reflected a median follow-up of only 2.9 years. Reports of synovial sarcoma in the head and neck literature have been limited to isolated case studies. When discussing synovial sarcoma of the extremities, most authors recommend treating this lesion with aggressive surgical therapy followed by radiation therapy. However, recent reports suggest that synovial sarcoma may be indolent, and can be treated by simple excision without radiotherapy. Despite the controversy regarding treatment of synovial sarcoma of the head and neck, the clinical features of synovial sarcoma are similar regardless of site.

Patients present with synovial sarcoma usually between age 25 to 36 years, with men outnumbering women 3:2. Earlier discovery of synovial sarcoma in the head and neck results from symptoms of pain, dysphagia, hoarseness, and cosmetic deformity. Metastasis to regional lymph nodes occurs in 12.5% of cases in the head and neck and 23% of cases in the extremities. Pulmonary metastases caused death in ten of 22 cases in Roth's series of head and neck synovial sarcomas and occurs in 33% to 81% of cases in the extremities. The dismal prognosis for patients with synovial sarcoma of the extremities has led to several treatment options. Surgical procedures for primary sarcoma of the extremity include local excision, en bloc resection of the entire anatomic compartment, and amputation. Sarcomas of soft tissues tend to infiltrate along fascial and muscle planes far beyond the palpable limits of the tumor. Therefore, most authors conclude that surgery should aim for removal of all gross tumor and encompass a wide cuff of normal tissue. Although most sarcomas are radioresistant, adjunctive radiation therapy is advocated. The role of chemotherapy is not well defined. Recent attention has focused on the use of adjuvant chemotherapy before metastatic disease is detectable. The use of chemotherapy in children with sarcomas may improve the control of distant metastases and recurrences at the primary site. However, studies assessing the use of chemotherapy in adults with sarcomas have been disappointing in the demonstration of significantly improved survival.

One major factor that may lead to a misdiagnosis of synovial sarcoma of the head and neck is a divergence of opinion about its histopathologic features. Hajdu et al. stated that a primitive mesenchymal precursor cell differentiates into fascia, aponeurosis, tendon, bursae, and synovial membranes (Fig 1). The unicellular origin theory of Hajdu et al. postulates that malignant degeneration of this primitive mesenchymal cell causes synovial sarcoma. Contrary to earlier beliefs, synovial sarcoma seldom arises from...
formed synovial membrane, although mature synovium and synovial sarcoma exhibit a similar two-cell histology.\textsuperscript{22,23} Classically, biphasic synovial sarcoma manifests both spindle cells and epithelioid cells (Figs 2 and 3).\textsuperscript{3}

Synovial sarcomas consisting of only one cell type have been described, but not all pathologists agree that monophasic variants exist. Mackenzie\textsuperscript{24} classifies monophasic synovial sarcomas as either spindle-cell sarcomas or epithelioid sarcomas. Recognizing that most synovial sarcomas are poorly differentiated mesenchymal tumors, Russell\textsuperscript{25} recommends that staging of patients with synovial sarcoma be limited to stages 3 and 4. Despite differing opinions regarding classification of synovial sarcoma, the histopathology is the same for those arising in the head and neck and those arising in the extremities.

A child recently presenting with synovial sarcoma of the postauricular region and the limited experience with this tumor in the head and neck prompted a review of the cases treated at UCLA.

PATIENTS AND METHODS

The UCLA Central Tumor Registry was searched for records of cases of synovial sarcoma in the head and neck from 1974 to 1986. Five previously unreported cases are described (Table).

**CASE 1**—A 35-year-old woman presented for evaluation of a painless 2-cm left gingivobuccal lesion. There was no cervical adenopathy. A biopsy specimen of the lesion was consistent with synovial sarcoma. Results of a preoperative evaluation, including a chest roentgenogram were normal. She underwent a left composite mandibulectomy and suprahyoid neck dissection with Vitallium tray and iliac crest bone graft reconstruction. She received 5000 rad (50 Gy) of radiotherapy to the primary site and left neck. At one year, a 1-cm recurrence was treated with wide local excision along with removal of the infected Vitallium tray. Bilateral lung metastases were diagnosed seven years after initial therapy, in spite of control at the primary site. The patient underwent surgical removal of the lung nodules and died of progressive pulmonary failure eight years after initial diagnosis.

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**Table: Synovial Sarcoma of the Head and Neck, UCLA 1974-1985**

<table>
<thead>
<tr>
<th>Patient, No./ Age, y/Sex</th>
<th>Size, cm</th>
<th>Location</th>
<th>Treatment*</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/35/F</td>
<td>2</td>
<td>Left gingivobuccal sulcus</td>
<td>1975, composite resection hemimandible, suprahyoid neck dissection, 5000 rad (50 Gy); 1976, WLE recurrence; 1983, lung metastases</td>
<td>Dead of disease at 8 y</td>
</tr>
<tr>
<td>3/21/M</td>
<td>4</td>
<td>Hypopharynx</td>
<td>1983, WLE, 6490 rad (64.9 Gy), chemotherapy</td>
<td>1985, massive recurrence requiring tracheostomy; dead of disease at 2½ y</td>
</tr>
<tr>
<td>4/48/F</td>
<td>3</td>
<td>Anterior neck</td>
<td>1982, WLE, thyroidectomy, 6000 rad (60 Gy); 1983, neck dissection, lung metastases; chemotherapy</td>
<td>Dead of disease at 12 mo</td>
</tr>
<tr>
<td>5/8/M</td>
<td>4</td>
<td>Postauricular</td>
<td>1985, WLE, 5000 rad (50 Gy), chemotherapy</td>
<td>No evidence of disease at 6 mo</td>
</tr>
</tbody>
</table>

*WLE indicates wide local excision.
CASE 2.—A 27-year-old woman presented with intermittent right otalgia. Examination disclosed a 2-cm submucosal lesion in the right posterior nasopharynx. A biopsy specimen showed a synovial sarcoma. A chest roentgenogram was normal. She underwent 6600 rad (66 Gy) of radiotherapy to the primary tumor, and received eight courses of systemic combined chemotherapy consisting of dacarbazine, doxorubicin hydrochloride, and cyclophosphamide. A recurrence was noted in 1982, and she was referred to UCLA. A transpalatal, wide local excision was performed after the patient refused radical skull base surgery. Nasopharyngeal recurrences in 1983, 1985, and 1986 were excised locally, and the patient is currently alive with disease at the primary site, invading the clivus.

CASE 3.—A 21-year-old man presented with progressive dysphagia, odynophagia, and weight loss. Examination revealed a 4-cm submucosal mass in the pharynx extending from the fossa of Rosenmüller to the hypopharynx. A biopsy specimen showed synovial sarcoma. The patient underwent wide local excision through a lateral pharyngotomy approach. He received 6490 rad (64.9 Gy) of radiotherapy to the pharynx. The tumor recurred within three months and the patient was treated with cyclophosphamide, vincristine sulfate, daunomycin, and doxorubicin with partial response. The patient eventually developed massive recurrent tumor requiring tracheostomy for airway maintenance. He died of widespread pulmonary metastases 2 1/2 years after initial diagnosis.

CASE 4.—A 45-year-old woman discovered a painless, 1-cm mass in the lower right cervical region. Thyroid suppressive therapy was instituted for a suspected thyroid nodule. A thyroid scan and chest roentgenogram were normal. In December 1982, a total thyroidectomy was performed with excision of an adjacent paratracheal mass. Histopathology was consistent with synovial sarcoma. She received 6000 rad (60 Gy) of radiotherapy to the neck through opposed lateral fields. She was referred to UCLA six months later for a 4-cm recurrence. She underwent total laryngectomy with radical neck dissection and wide resection of the previous scar and overlying soft tissue. Despite treatment with systemic chemotherapy, she developed bilateral lung metastases and died one year following initial diagnosis.

CASE 5.—An 8-year-old boy presented with a 4-cm slowly enlarging mass in the left postauricular region in July 1985. There was no history of antecedent trauma or ear disease. An excisional biopsy specimen demonstrated a biphasic synovial sarcoma. Chest roentgenogram, bone scan, and bone marrow aspirate were normal. He then underwent wide local excision of the tumor bed with a 3- to 4-cm cuff of skin and normal soft tissue; reconstruction was performed with a cervical rotation advancement flap. He received 5000 rad (50 Gy) of radiotherapy to the postauricular and occipital regions. The patient recently completed a planned 12-month course of systemic chemotherapy with dactinomycin and vincristine and is without recurrent disease at 15 months.

COMMENT

In summary, five cases of synovial sarcoma of the head and neck are described. Patient 5 is newly diagnosed. Of the remaining four patients, three are dead of lung metastases at 1, 2 1/2 and 8 years following initial diagnosis. The fourth patient is alive with disease at the primary site six years after initial diagnosis. All four patients developed at least one recurrence. Three patients were treated with wide local excision and postoperative radiotherapy, while the fourth received radiotherapy as a primary modality. Adjunctive systemic chemotherapy was used in three patients.

This experience, though small, suggests that synovial sarcoma of the head and neck is equally as aggressive as synovial sarcoma of the extremities. A marked tendency for local recurrence and pulmonary metastases lowers survival to approximately 40% at five years, and 25% at ten years.1-3 We believe that until more experience with this lesion is obtained, treatment guidelines of synovial sarcoma of the extremities should be extrapolated to cases in the head and neck. Wide resection is indicated in all cases and should encompass a large cuff of normal tissue.1,14 Radiotherapy should be used postoperatively in all cases for the treatment of residual microscopic disease.18,19 The role of adjuvant chemotherapy in this disease is under investigation, but its greatest potential may lie in the prevention or delay of distant metastases.26

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References


