Synovial sarcoma of the pharynx: A case report

Levent Saydam, MD
Ahmet Kizilay, MD
M. Tayyar Kalcioglu, MD
Bulent Mizrak, MD
Fatih Bulut, MD

Abstract

Synovial sarcoma is a malignancy not usually encountered in the head and neck region. We describe a case of synovial sarcoma of the posterior pharyngeal wall in a 14-year-old girl. The mass was completely excised via the transoral route. Postoperatively, the patient received irradiation to 60 Gy. At 40 months of follow-up, the patient remains disease-free.

Introduction

A synovial sarcoma is a malignant tumor made up of primitive pluripotential mesenchymal cells. It is responsible for approximately 10% of all soft-tissue sarcomas.[1,2] These tumors are usually encountered in the periarticular areas of the lower extremities; only about 10% of cases occur in the head and neck region.[1]

The first case of synovial sarcoma of the head and neck was described by Jernstrom in 1954.[3] In this article, we report a new case of synovial sarcoma that originated in the posterior pharyngeal wall in a child.

Case report

A 14-year-old girl came to us with a 3-month history of progressive breathing difficulty and dysphagia. She had a muffled voice but was in no apparent respiratory distress. Physical examination revealed the presence of a midline submucosal mass in the posterior pharyngeal wall that extended from the nasopharynx down to the level of the epiglottis. Findings on examination of the neck were normal.

Contrast-enhanced computed tomography (CT) revealed the presence of a nonhomogenous mass that was primarily made up of cystic components; in addition, some solid components were scattered throughout the parenchyma (figure 1). The mass was clearly surrounded by a thin capsule at all levels. Despite the close proximity of the mass to the major neck vessels, no vessel involvement was observed. We did not perform a preoperative biopsy because the lesion's location might have led to airway problems.

After discussing various treatment options with the patient and her family, we decided on excision via a transoral approach with a possible midline mandibulotomy and tracheotomy. We were able to adequately expose the mass with a soft-palate retractor and a Jensen's tongue retractor. After establishing a submucosal dissection plane, we completely removed the mass and its entire capsule transorally.

The postoperative period was uneventful, and there was no need for the tracheotomy. The pathologist reported a synovial sarcoma. An extensive postoperative search for distant metastases was negative. Two representative sections of a hematoxylin-and-eosin-stained specimen revealed biphasic cellularity (figure 2). Immunohistochemistry was positive for cytokeratin (figure 3). The patient underwent postoperative irradiation to 60 Gy, and she was free of recurrent and distant metastatic disease at the 40-month follow-up (figure 4).

Discussion

Synovial sarcoma is a malignant soft-tissue neoplasm that arises from primitive pluripotential mesenchymal cells near to or remote from articular surfaces. Other terms used to describe this entity are *synovial cell sarcoma* and *malignant synovioma*.[2,4] Although the term *synovioma* implies a benign condition, further investigations have

From the Department of Otolaryngology, Bayindir Medical Center, Ankara (Dr. Saydam); the Department of Otolaryngology (Dr. Kizilay and Dr. Kalcioglu) and the Department of Pathology (Dr. Mizrak), Inonu University School of Medicine, Malatya; and the Section of Otolaryngology, SSK Hospital, Malatya (Dr. Bulut), Turkey.

Reprint requests: Levent Saydam, MD, Department of Otolaryngology, Bayindir Medical Center, 06520 Sogutozu, Ankara, Turkey. Phone: +90-312-428-0808; fax: +90-312-428-0690; e-mail: leventsaydam@usa.net

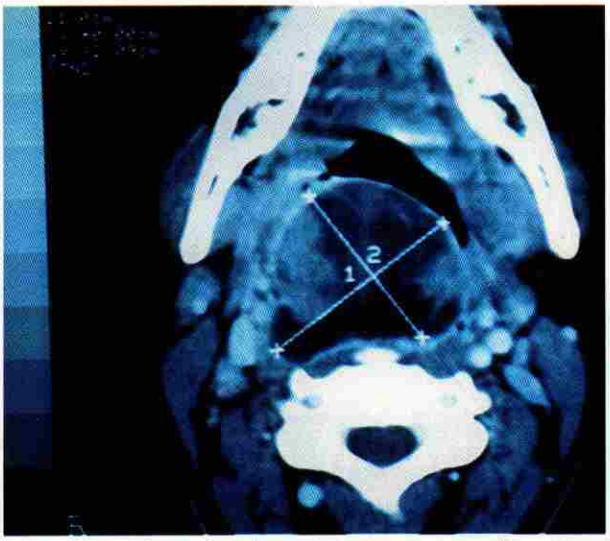

Figure 1. CT shows that the nonhomogenous mass fills a large portion of the oropharyngeal compartment.

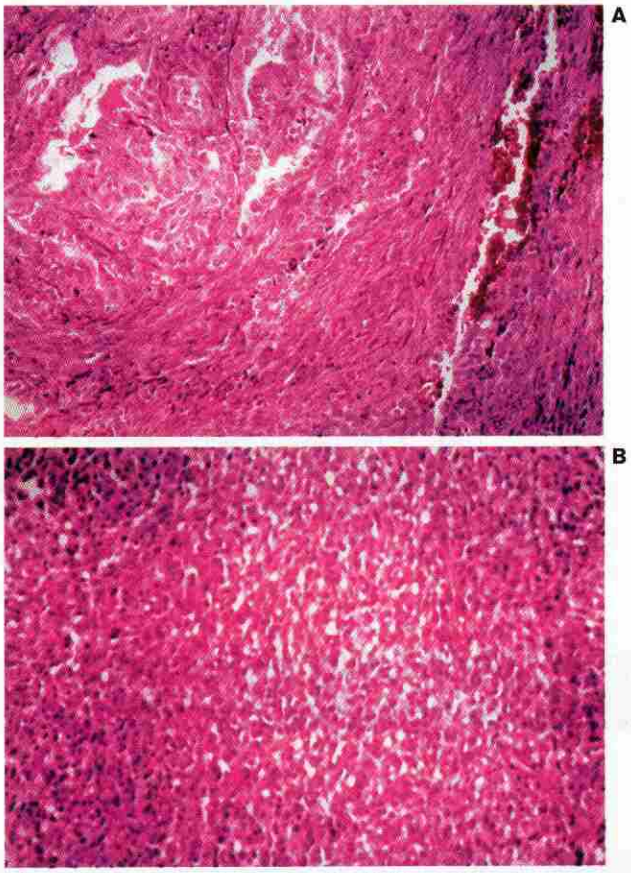

Figure 2. ***A:*** *The neoplastic growth exhibits a biphasic pattern. Glandular structures can be seen in the upper left and a sarcomatous area in the lower left.* ***B:*** *The spindle-cell area resembles a fibrosarcoma and other spindle-cell tumors (H&E, original magnification × 100).*

exposed the aggressive nature of this neoplasm and eventually led to its classification as a high-grade tumor by the American Joint Committee on Cancer.[5]

Incidence. Synovial sarcomas account for 8 to 10% of all sarcomas.[1] Although these tumors occur predominantly in the extremities, they are also known to arise in the head and neck region.[2] Since the first description of a head and neck synovial sarcoma by Jernstrom[3] in 1954, fewer than 100 cases have been reported in the literature, and they account for 3 to 10% of all reported cases.[2]

Clinical characteristics. The most common site of synovial sarcoma of the head and neck is the hypopharynx.[4,6] These tumors have been encountered in other areas of the head and neck as well, including prevertebral and parapharyngeal areas and maxillofacial, pharyngeal, and laryngeal sites.[7] Synovial sarcomas have a male preponderance (3:2) and a predilection for patients between the ages of 25 and 36 years.[8] Patients with synovial sarcoma of the head and neck tend to be younger than those who have such tumors in the extremities.[7,9]

The tumor usually appears as an asymptomatic mass until it attains sufficient volume to cause pressure effects on neighboring structures. This process is usually more rapid in patients with head and neck synovial sarcoma than in those whose tumor is in an extremity. Lockey reported that patients with head and neck tumors were diagnosed an average of 20 months earlier than those with extremity tumors.[9] This slower growth rate (and perhaps the resultant lower rate of lymph node metastasis) might explain why head and neck patients have a better prognosis.

Histologic characteristics. Although monophasic forms of synovial sarcoma have been reported, its primary histopathologic feature is biphasic cellularity (figure 2).[10] Microscopically, the tumor is made up of epithelial and sarcomatous components. The glandular appearance created by epithelial malignancy can mislead the pathologist to a diagnosis of a salivary gland tumor. Likewise, its sarcomatous structure can lead to an incorrect diagnosis of a malignant schwannoma or fibrosarcoma. Immunohistochemistry is key to a definitive diagnosis. The monophasic form exhibits primarily sarcomatous features; the monophasic epithelial form is rare.

Treatment. The relatively small number of patients with synovial sarcoma of the head and neck precludes us from drawing firm conclusions with respect to a precise treatment protocol. For now, the primary mode of treatment is wide surgical excision that includes a normal-appearing tissue cuff in order to obtain tumor-negative margins. However, this cannot always be accomplished in head and neck cases because of the proximity of the tumor to vital structures.

The results of multimodality treatment appear to be better than those seen with any single-modality treatment

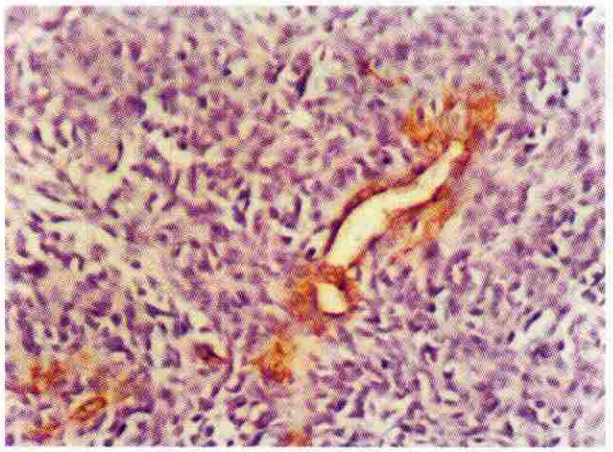

Figure 3. Cytokeratin positivity is seen in the tumor cells (streptavidin-biotin immunohistochemistry, DAB, original magnification × 100).

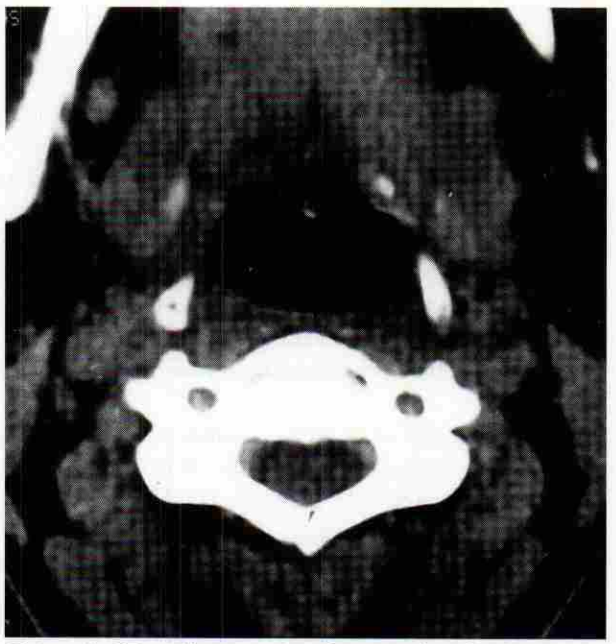

Figure 4. CT of the posterior pharyngeal wall obtained 31 months postoperatively detects no evidence of recurrent disease. The patient remains disease-free 40 months postoperatively.

or with any form of salvage therapy.[9,11] The role of radiotherapy is confined to reducing the risk of local recurrence following resection; radiation does not improve long-term survival. Chemotherapy has a similar role in that it can decrease the risk of distant metastasis. We did not administer chemotherapy in this case because the patient refused it.

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