

■ Case Report

Synovial sarcoma of the neck

Boyunda görülen sinoviyal sarkom olgusu

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ABSTRACT

Synovial sarcomas are rare malignant neoplasms commonly arising from articular tendons and joint capsules. Despite being termed synovial sarcomas due to their histologic similarity to the synovium, they rarely involve a synovial structure. Although they mostly occur in lower extremities, rare cases originating from the thorax, abdomen, head, and neck have also been reported. A 60-year-old male patient was admitted to the hospital with a complaint of swelling in left side of the neck. CT revealed a mass of approximately 6.5x5 cm, occupying the left parapharyngeal space and left submandibular fossa and protruding into the pharynx lumen. The lesion was non-infiltrative, well circumscribed, and uniformly ovoid. The patient underwent surgery, and subsequent pathological examination confirmed the diagnosis of synovial sarcoma. Despite their rarity, synovial sarcomas should be considered, along with Ewing's sarcoma, rhabdomyosarcoma, and other sarcomas, in the differential diagnosis of a large juxtaarticular mass containing calcifications.

Keywords: synovial sarcoma, computed tomography, jugulodigastric space, neck

ÖZ

Sinoviyal sarkomlar artiküler tendonlardan ve eklem kapsüllerinden köken alan nadir malign tümörlerdir. Histolojik olarak sinoviyuma benzerlikleri nedeniyle sinoviyal sarkom olarak isimlendirilmelerine rağmen, nadiren sinoviyal yapı içerirler. Genellikle alt ekstremitelerde görülmekle birlikte nadir olarak toraks, abdomen, baş-boyun bölgesinde sinoviyal sarkom tespit edilen olgular bildirilmiştir. 60 yaşında erkek hasta boyun sol yarısında şişlik şikayeti ile hastanemize başvurdu. Çekilen bilgisayarlı tomografide sol parafaringeal boşluğu ve sol submandibular fossayı doldurup, farenk lümenine protrude yaklaşık 6,5x5 cm boyutunda kitle tespit edildi. Kitle düzgün sınırlı, ovoid şekilli ve çevre dokulara infiltrasyon göstermemekteydi. Cerrahi olarak total çıkarılan kitlenin histopatolojik tanısı sinovyal sarkom olarak belirlendi. Nadir görülmelerine rağmen sinoviyal sarkomlar Ewing sarkomu, rabdomiyosarkom ve diğer sarkomlarla birlikte kalsifikasyon içeren jukstaartiküler kitlelerin ayırıcı tanısında düşünülmelidir.

Anahtar kelimeler: sinoviyal sarkom, bilgisayarlı tomografi, jugulodigastric boşluk, boyun

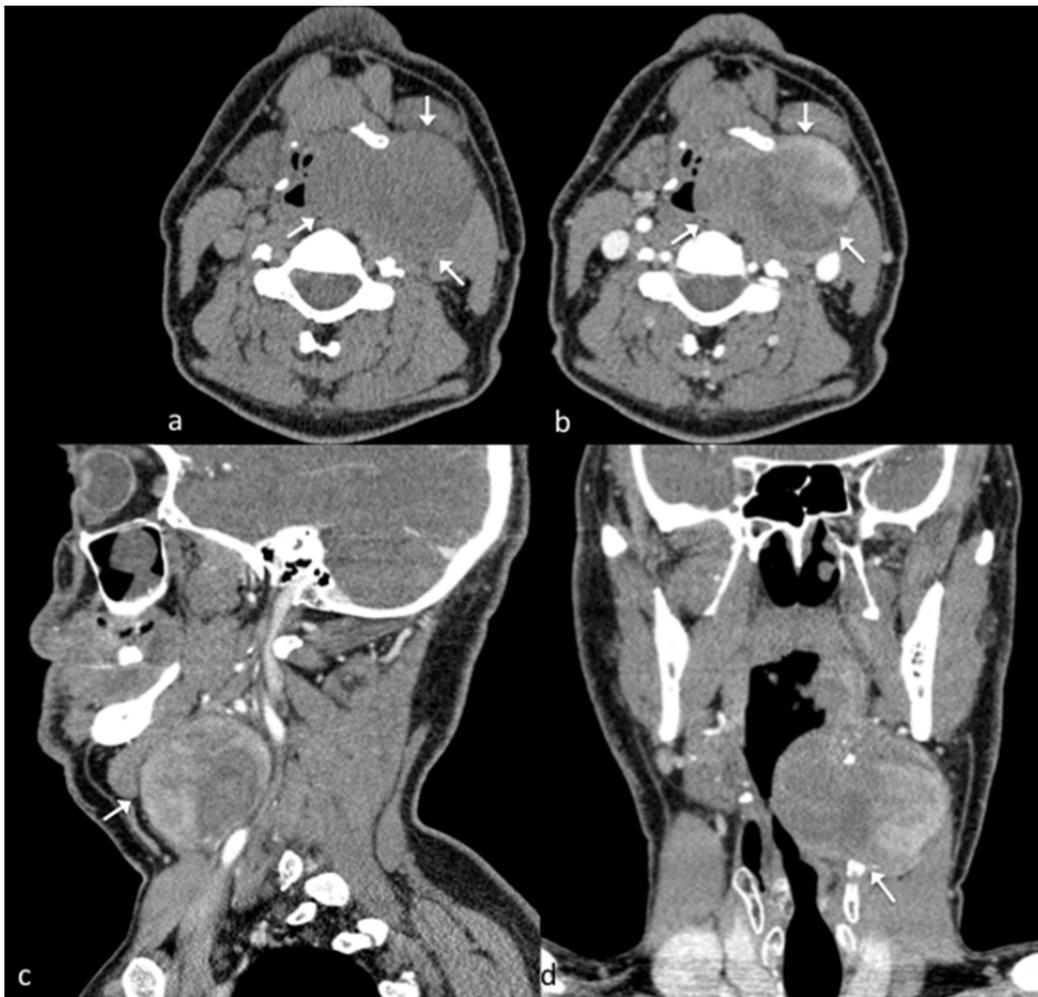


Figure 1. Synovial sarcoma of the jugulodigastric region visualized on (a) unenhanced and (b) enhanced axial, sagittal and coronal CT slices. The lesion, involving the left jugulodigastric region, is protruding into the larynx. It is located beneath the submandibular gland and above the thyroid cartilage. Some parts of the lesion show better contrast enhancing

INTRODUCTION

Synovial sarcoma is a rare malignant neoplasm that commonly arises from articular tendons and joint capsules; however, the synovial relationship is not always clear. Synovial sarcoma is the third most common soft tissue sarcoma in adults and accounts for approximately 10% of soft tissue sarcomas [1,2]. Despite being termed synovial sarcomas due to their histologic similarity to the synovium, they rarely involve a synovial structure and are usually seen near the joints [3], mostly affecting the extremities. Although they generally occur in the lower extremities, rare cases originating from the thorax, abdomen, head, and neck have also been reported. About 9% of synovial sarcomas are located in the head and neck region. Synovial sarcomas are usually seen in adolescents and young adults. Most studies have shown that the median age of synovial sarcomas is in the third decade and approximately 66% of the patients are male. Synovial sarcoma in the head and neck region is mostly located in the hypopharynx, the parapharyngeal space, and postpharyngeal region [4].

This case is presented because it is a rare lesion that is usually overlooked in the differential diagnosis of neck solid lesions.

CASE

A 60-year-old male patient was admitted to the hospital with a complaint of swelling in the left side of the neck. On physical examination, a mass of approximately 3x3 cm was palpated in the left jugulodigastric area. Ultrasonography revealed a heterogeneous echogenic mass of about 5 cm in diameter on the left side of the neck. The patient was then evaluated with neck CT.

There was a mass approximately 6.5x5 cm in size, occupying the left parapharyngeal space and left submandibular fossa and protruding into the pharynx lumen. The defined mass was observed as hypodense from the surrounding muscle tissue in the unenhanced CT examination. The lesion was non-infiltrative, well circumscribed, and uniformly ovoid. On contrast-enhanced CT, the anterolateral segment showed more contrast enhancement than the posterior (**Figure 1**).

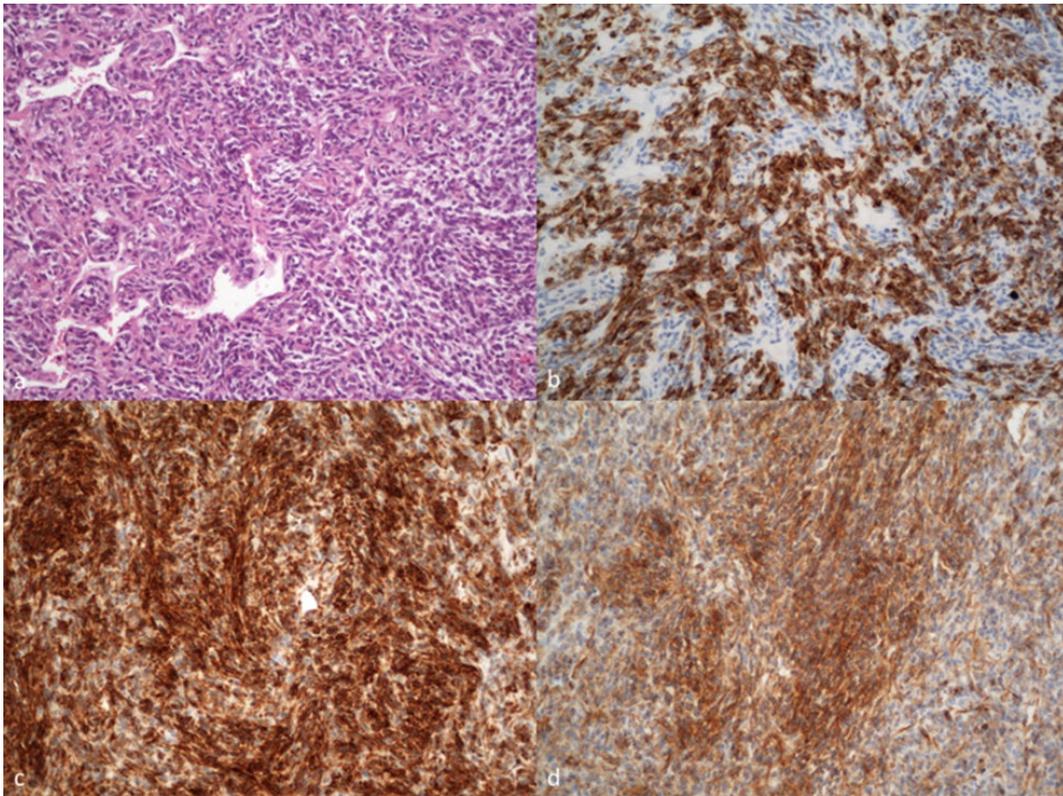


Figure 2. (a) Uniform spindle cells and hemangiopericytomatous vasculature (H&Ex200), (b) Cytokeratin AE1/AE3 positivity in tumor cells (x200), (c) Strong immunoreactivity for bcl2 protein in tumor cells (x200), and (d) CD99 positivity in tumor cells (x200)

The lesion was totally resected. The pathological outcome was reported as synovial sarcoma (**Figure 2**).

DISCUSSION

In the differential diagnosis of tumors in the digastric area, cystic lesions, such as brachial cleft cysts and lymphangioma, metastatic lymph nodes, and salivary gland tumors need to be considered [5]. In the present case, there was no cystic component, and the mass was purely solid. Therefore, branchial cleft cysts and lymphangioma were not considered in the differential diagnosis.

Salivary gland tumors that usually originate from the parotid gland tail should also be considered in the differential diagnosis of lesions located in the jugulodigastric area. Pleomorphic adenomas are the most common salivary gland tumors. Pleomorphic adenomas are usually visualized as spherically shaped, hypodense masses with lobulated contours. Necrotic areas and calcification may also be detected. The contrast agent enhances smaller tumors to a greater extent than larger tumors [6]. Another common benign salivary gland tumor is Warthin's tumor, which is usually divided into multifocal and bilateral tumors. Warthin's tumors are seen bilaterally and multifocally and differ from other lesions with their well-defined, homogeneous and generally lobulated structure. They may

be solid or may contain cystic components. These tumors have moderate enhancing on CT [7]. However, in the present case, the contour feature, shape and contrast pattern of the lesion were different.

Adenoid cystic carcinoma and mucoepidermoid carcinoma are the two most common malignant salivary gland tumors, with the other examples of this group being acinar cell cancer and squamous cell cancer. Malign lesions tend to be smaller and are seen more rarely. On contrast-enhanced CT, low-grade lesions are visualized as masses with cystic and solid components. Aggressive tumors are mostly solid masses with irregular borders. High-grade lesions may invade the surrounding tissues and involve the external jugular vein or cause the narrowing of the carotid artery [8]. In the present case, the lesion was purely solid, and there was no evidence of the invasion of the surrounding tissue.

Another group of tumors that should be considered in differential diagnosis are metastatic lymph nodes. The jugulodigastric node is the first lymph node to receive lymphatic drainage from the tonsils, pharynx, mouth, and face. Pathological lymph nodes can be seen in the jugulodigastric area, especially in the tonsils, pharynx mouth, and facial tumors [9]. Vascular lesions, lymphoproliferative diseases, lymphoma, and sarcomas may

also be present in this region. However, neurogenic tumors and sarcomas are rarely observed in this area [5].

Although synovial sarcomas took their name from their similarity to the synovium, they are considered to originate from primitive mesenchymal cells that undergo differentiation to synovial cells. The main histologic variants of these sarcomas are: a biphasic type consisting of spindle and epithelial cells that usually form glandular structures, a monophasic type consisting of only spindle cells, and a poorly differentiated type consisting of cells that resemble small, round, blue cells. The monophasic variant is the most common among these histopathological types. Synovial sarcoma is associated with a specific t(X;18) (p11; q11) translocation, involving SS18 (SYT), SSX1, SSX2, or SSX4. The characteristic pathologic features of synovial sarcoma include tumoral calcification, cystic changes, and necrosis [10].

The most common CT appearance of synovial sarcoma is that of a heterogeneous deep-seated soft-tissue mass. The CT attenuation of synovial sarcomas are similar to or slightly lower than that of muscle and lower attenuation areas, and necrosis and hemorrhage are also common [12]. A contrast-enhanced CT scan shows heterogeneous enhancement in 89%–100% of cases, but smaller lesions may be more homogeneous [11]. CT is also useful for detecting calcification and bone involvement in synovial sarcoma. This technique typically reveals a non-infiltrative, well-circumscribed soft tissue mass. Although punctate, peripheral calcifications are other entities identified in most CT images [13], we did not observe them in our case; the lesion was non-infiltrative, well-circumscribed, and uniformly ovoid and showed heterogeneous enhancement.

MRI is the modality of choice for the diagnosis and initial staging of synovial sarcoma because of the information provided by intrinsic signal characteristics and superior soft-tissue contrast [14]. On T1-weighted MR images, synovial sarcoma appears as a prominently heterogeneous multilobulated soft tissue mass with signal intensity similar to or slightly higher than that of muscle. Prominent heterogeneity with predominant high signal intensity is also a feature of these lesions on T2-weighted MR images. In addition, a triple signal pattern, representing the hypointense, isointense, and hyperintense areas on T2-weighted images, is typically seen. This heterogeneity and triple sign on T2-weighted MR images is the result of a mixture of solid cellular elements, hemorrhage, necrosis, and calcified or fibrotic collagenized regions [11]. In the present case, an MRI scan was not performed since the

lesion was well-circumscribed and the CT images were considered adequate for surgery.

CONCLUSION

Despite being rare tumors, synovial sarcomas should be considered, along with Ewing's sarcoma, rhabdomyosarcoma, and other sarcomas, in the care of young adults and adolescents with a large juxtaarticular mass containing calcifications. CT and MRI each have their own advantages in evaluation, and the combination of the two approaches can improve the accuracy of the diagnosis. Considering the slowly progressing nature of synovial sarcomas, in the early stage when clinical findings are not sufficient, imaging modalities are the only way of achieving early diagnosis. However, the final diagnosis relies on the results of a pathological investigation.

DECLARATION OF CONFLICT OF INTEREST

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