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This information is current as of August 6, 2025.

AJNR Am J Neuroradiol 2004, 25 (6) 1103-1105 http://www.ajnr.org/content/25/6/1103

Synovial Sarcoma of the Head and Neck: A Case of Predominantly Cystic Mass

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Summary: We report a case of predominantly cystic synovial sarcoma partly adherent to the hyoid bone in the submental area. The mass demonstrated posterior acoustic enhancement at sonography and a complex cystic mass with mural nodules and solid septa at CT.

Although rare, synovial sarcoma is one of the most common malignant soft-tissue sarcomas in children and adolescents (1). The head and neck is involved in 6.8% of all synovial sarcomas (1). In previous reports, the imaging features of synovial sarcoma in the head and neck included a well-defined, predominantly solid mass with a cystic or hemorrhagic component and calcification (2–4). We describe a case of predominantly cystic mass in the submental area, which was pathologically confirmed as a synovial sarcoma.

Case Report

An 11-year-old girl was admitted to hospital because of a palpable mass in the left submental area. The mass was palpable 3 months before admission. Initial fine needle aspiration revealed some follicular epithelial cells with mild atypia and columnar cells and was presumed to be a thyroglossal duct cyst. Sonography showed a predominantly cystic mass and internal solid septa (Fig 1A). Color Doppler sonography showed increased vascularity around the solid septa (Fig 1B). The relationship between the mass and hyoid bone was not clearly demonstrated sonographically. The mass was thought to be an atypical lesion associated with thyroglossal duct remnant. Nonenhanced axial CT showed a well-defined mass with a central low-attenuation area (Fig 2A). The central portion was 20.2 H. There was no remarkable fat infiltration adjacent to the mass. Contrast-enhanced axial CT disclosed peripheral, enhancing mural nodules and septation that showed moderate contrast enhancement. The lesion abutted the hyoid bone partly at its upper medial aspect (Fig 2B). Contrast-enhanced coronal CT showed the mylohyoid muscle was compressed, and the boundary between the upper part of the mass and the mouth floor was indistinct (Fig 2C). No lymphadenopathy larger than 1 cm in its maximal diameter was seen. The CT finding was thought to be a predominantly cystic mass with a malignant component

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such as a carcinoma arising from thyroglossal duct remnant. The lesion was removed en bloc. On surgical field, some adhesion between the mass and the adjacent muscles were seen. Microscopic findings were consistent with classic biphasic synovial sarcoma (Fig 3A). Immunohistochemical staining revealed positive immunoreactivity to CD99, BCL2, and CK (AE1/AE3) antibodies (Fig 3B).

Discussion

Synovial sarcoma can occur at all ages but most frequently affects young adults and adolescents (2). Synovial sarcomas often arise adjacent to joints, especially around the knees. These tumors do not originate from synovial tissue, but rather from pluripotential mesenchymal cells near or even remote from articular surfaces (2, 4). In the head and neck, the hypopharynx is the most commonly involved site perhaps because of the abundant synovial tissue at the hypopharynx. Other locations in the head and neck reported in the literature include the masticator space, parapharyngeal space, sinonasal region, and pharynx (4). In our case, the mass was located near the hyoid bone in the submental area. In this area, the synovial tissue is present at the retrohyoid bursa, which is between the thyrohyoid membrane and the hyoid bone (3-5).

Imaging findings of synovial sarcoma in the head and neck have been described. Usually, synovial sarcomas manifest as predominantly solid masses on CT scans or MR images with well-defined smooth margins; infiltration of the adjacent soft tissue is a less frequent finding (2-4). The lesions may appear as either a homogeneous or heterogeneous mass, according to degree of hemorrhage or necrosis. About half of the cases in the previous reports were those with homogeneous lesions (2, 4). Contrast enhancement pattern is variable, but a moderate degree of enhancement was frequently described in a previous report (2). Approximately 30% of synovial sarcomas contain calcifications that may be apparent on the imaging study (4). The presence of calcifications tends to be associated with better survival (3, 6). In our case, no visible calcification was seen.

Synovial sarcoma is a frequently misclassified imaging finding of a benign mass because of its smooth margin, cystic components, and lack of aggressive infiltration (2). In our case, the submental paramedian location, partly abutting the hyoid bone, and the predominantly cystic nature mimicked a lesion associated with thyroglossal duct remnant. Despite the youth of this patient, the lesion was regarded as a carcinoma arising from thyroglossal duct remnant. There was, however, no cal-

Received February 11, 2003; accepted after revision November 8.

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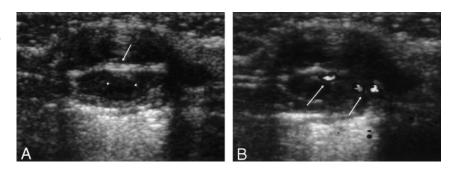
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Fig 1. Sonograms.

A, Gray-scale sonogram shows a well-defined 1.6×2 -cm mass with posterior acoustic enhancement. An echogenic central septum (white arrow) and slightly low echoic peripheral thick wall (white arrowheads) are seen.

B, Color Doppler sonogram shows increased vascularity at the septal solid portion of the lesion (*white arrows*).



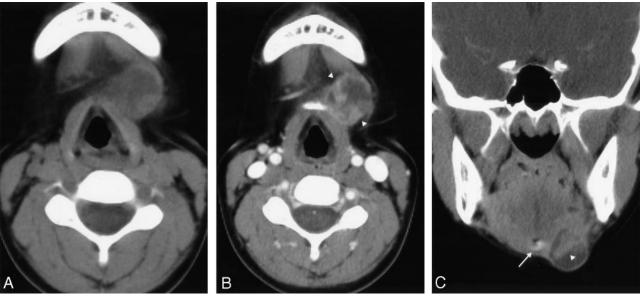


Fig 2. CT scans.

A, Noncontrast axial CT scan shows a well-defined submental mass with central, low attenuation. A solid mural component is present. CT attenuation number of central portion is 20.2 H. There is no remarkable fat infiltration adjacent to the mass.

B, Contrast-enhanced axial CT scan shows moderately enhanced peripheral wall and nodular mural components (white arrowheads). Mass abuts partly to the hyoid bone, at its medial aspect.

C, Contrast-enhanced coronal CT scan shows a thick central solid septa (*white arrowhead*). The hyoid bone (*white arrow*) is located next to the medial aspect of the lesion. The boundary between the upper part of the mass and mouth floor is indistinct.

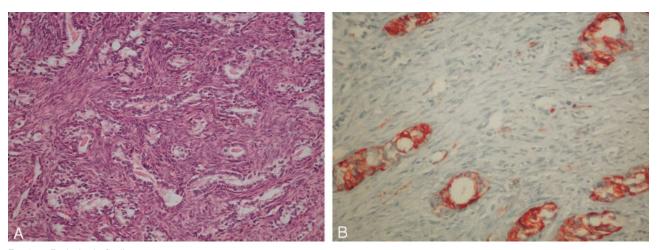


Fig 3. Pathologic findings.

A, Microphotograph of the solid portion shows classic biphasic synovial sarcoma with glandular structures surrounded by a spindle cell component. The glandular structures have intraluminal eosinophilic secretions (Hematoxylin and eosin, ×200).

B, Immunohistochemistry for cytokeratin highlights the epithelial elements. The spindle cell elements reveal focal cytokeratin immunoreactivity (cytokeratin [AE1/AE3], ABC method, ×200).

AJNR: 25, June/July 2004

cification, which is frequently seen in the carcinoma of thyroglossal duct remnant (7, 8). The predominantly cystic appearance of our case is not similar to other previously reported imaging findings. Schwannoma and neurofibroma, with a cystic change, and carcinoma of a thyroglossal duct cyst may be considered in the differential diagnosis (2).

Conclusions

A pure cyst or complex cysts in the anterior midline or paramedian neck are usually benign cystic lesions such as a thyroglossal duct cyst in the pediatric age group (9); however, neoplasm such as a carcinoma of thyroglossal duct cyst or cystic neurogenic tumor should be considered when any solid mural nodules or thick septa coexist in the cystic lesion. Rarely, synovial sarcoma with a prominent cystic change should also be included in the differential diagnosis. In addition, the region near the hyoid is a potential site of origin for synovial sarcoma in the head and neck.

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