Synovial Sarcomas of the Head and Neck: MR Findings

OBJECTIVE. MR images of six synovial sarcomas of the head and neck were evaluated to determine their characteristic sites of origin, size, extent, intensity, and contrast enhancement. It was hoped that specific MR characteristics could be defined to suggest this entity.

CONCLUSION. A nonmucosal head and neck mass that is isointense to gray matter on T1-weighted images and is well defined yet heterogeneous, with septations, hemorrhage, cysts, calcification, or multilocularity, should raise suspicion of a synovial sarcoma. Because the appearance of synovial sarcomas varies and other masses may appear similar, no specific imaging characteristics define the entity.

Synovial sarcomas are aggressive, malignant soft-tissue tumors that occur primarily in young adults. Synovial sarcomas represent 8–10% of all soft-tissue malignancies [1], with 85% occurring in the extremities [2]. Additional sites, although rare, include the head and neck, lower back, chest, and abdominal wall. Because of the rarity of synovial sarcomas (soft-tissue tumors arising in the head and neck represent <10% of malignant tumors in these areas), they tend to be misdiagnosed, delaying treatment. Therefore, MR diagnostic criteria would likely be of immediate value. MR imaging is considered the procedure of choice for the detection and staging of these tumors [3]. We reviewed data on six patients at our medical center with histopathologically confirmed synovial sarcomas of the head and neck to define useful MR diagnostic criteria. Although studies of synovial sarcomas elsewhere in the body have been published, the reports of imaging characteristics of head and neck cases are, to our knowledge, limited to only two small series [4, 5].

Materials and Methods

The MR images of all patients who presented during the past 5 years with histopathologically proven synovial sarcomas were retrospectively reviewed by two neuroradiologists. The study population consisted of six patients (five women and one man) who were 34–69 years old. The lesions ranged in maximal dimension from 8 mm to 8.8 cm. They arose from the masticator space (n = 2), parapharyngeal space (n = 2), sinonasal cavity (n = 1), and pharynx (n = 1). All images were obtained at 1.5 T on a Signa scanner (General Electric Medical Systems, Milwaukee, WI). Sagittal and axial T1-weighted images were obtained at 500–733/11–17 (TR range/TE range). Axial fast spin-echo (with fat suppression) T2-weighted images were obtained at 2700–3500/90–100. Axial enhanced-fat-suppressed T1-weighted images (533–800/17–35) were obtained after gadopentetate dimeglumine was administered IV at a dose of 0.1 mmol/kg.

Images were evaluated for signal-intensity and contrast enhancement characteristics. The signal intensity of the synovial sarcoma was compared with that of muscle, fat, gray matter, and CSF on the T1- and T2-weighted images. The enhancement of each lesion was graded on a scale from none to marked (none = no definite enhancement, minimal = less enhancement than for glandular tissue (e.g., submandibular gland), moderate = enhancement similar to that of glandular tissue, and marked = enhancement similar to that of mucosa). In cases of nonuniform enhancement, the most intense area of enhancement was used for final tabulation.

Two neuroradiologists independently reviewed the scans to assess intensity characteristics, cystic areas, hemorrhage, intratumoral calcification, and sites of invasion. We distinguished calcification from intratumoral hemorrhage on the basis of the assumption that calcification would cause low signal intensity on all pulse sequences and, unlike hemorrhage, would not show evolution or resolu-
tion. We searched for tumoral extension in three domains: the skull base, the intracranial compartment, and the vascular structures.

Therefore, 20 observations were made for each of the six cases reviewed: (1) T1-weighted signal intensity of mass compared with muscle, (2) T1-weighted signal intensity of mass compared with fat, (3) T1-weighted signal intensity of mass compared with glandular tissue, (4) T1-weighted signal intensity of mass compared with CSF, (5) T1-weighted signal intensity of mass compared with gray matter, (6) T2-weighted signal intensity of mass compared with muscle, (7) T2-weighted signal intensity of mass compared with fat, (8) T2-weighted signal intensity of mass compared with glandular tissue, (9) T2-weighted signal intensity of mass compared with CSF, (10) T2-weighted signal intensity of mass compared with gray matter, (11) degree of enhancement (mild, moderate, or marked), (12) type of enhancement, (13) presence of locularity, (14) presence of heterogeneity, (15) presence of extension to the skull base, (16) presence of extension to the meninges, (17) presence of extension to the vascular structures, (18) presence of calcification, (19) presence of intratumoral hemorrhage, and (20) presence of cyst formation. The seven neuroradiologists' interpretations were concordant in 113 (94%) of 120 observations related to tumor intensity and extension. The seven discrepancies in interpretation were decided by consensus after joint review. The final consensus readings are reported in the results.

Results

On T1-weighted imaging, all six synovial sarcomas appeared hyperintense to muscle and CSF and hypointense to fat (Fig. 1). Five of six lesions were nearly isointense to gray matter. Well-defined margins were visualized for all six lesions.

On T2-weighted imaging, all lesions appeared hyperintense to muscle and hypointense to CSF. Two were hypointense to fat, and three were isointense to fat; the sixth was of mixed intensity. Two were isointense to glandular tissue, three were isointense to fat, and the sixth was heterogeneous, with portions isointense to fat, CSF, and glandular tissue (Fig. 2). Two lesions exhibited marked contrast enhancement, and four exhibited moderate contrast enhancement.

Half the tumors appeared unilocular, and half appeared multilocular with septations. All lesions were predominantly solid. The multilocular lesions appeared heterogeneous.

Half the lesions, with clear areas of necrosis and a second with clear evidence of hemorrhage. Although MR imaging is not the diagnostic gold standard for detecting calcifications, MR scans suggested the presence of calcification in four of six patients.

Extension to the skull base was present in three of six patients. Extension intracranially was less common, present in two patients (Fig. 3). In our study, arterial invasion was not seen, but the cavernous sinus was infiltrated in one case.

Discussion

Synovial sarcomas typically present in the extremities of young men, especially around the knees. Men are more commonly affected than women by a ratio of 3:2 [5]. The typical patient with head and neck synovial sarcoma presents with a painless mass and associated symptoms of dysphagia, hoarseness, and headache depending on the plane of spread and site of origin [6]. Tumor extension relates both to the particular histologic subtype of the lesion and to the time elapsed between the onset of symptoms and the time of treatment. The aggressiveness of the growth of the synovial sarcoma is well known and is a key feature in its diagnosis. The sarcoma may exhibit a solid, cystic, or mixed pattern of growth, typically with areas of necrosis admixed with hemorrhage [4]. The most common site of occurrence in the head and neck is the hypopharynx [6].

The cause of synovial sarcomas is uncertain. In spite of their name, these tumors do not arise from synovial tissue [5]. They are so named because of their histologic similarity to synovium. The tumors arise from the mesenchyma and are composed of fibrous connective tissue and synovial-like cells; the former is represented by a spindle cell fibrosarcoma-like component, and the latter are represented by a pseudopithelial component [7]. Jern- strom [8] defined the synovial sarcoma as a biphasic tumor that is “histologically striking and characteristic... composed of two sharply contrasted types of tissue. One type faithfully reproduces caricatures of synovial structures, while the other consists of a fibromatous element that corresponds to an ordinary fibrosarcoma.” However, synovial sarcomas of only one cell type (monophasic tumors) have also been described [5]. Mackenzie [7] classified monophasic synovial sarcomas as either spindle cell sarcomas or epithelioid sarcomas. All cases in our study were classified as monophasic spindle cell-type synovial cell sarcomas.

The overall prognosis of patients with head and neck synovial sarcomas, although less than 40%, is better than the 20% 5-year survival rate of this same tumor presenting in the extremities. The relationship of the histologic findings to the patient’s prognosis is not surprising in that tumors that are well differentiated carry a better prognosis than poorly differentiated sarcomas.

CT and MR imaging may be used to determine the site of origin, delineate tumor extension, detect lymphadenopathy, identify calcification, and evaluate for possible airway compromise. MR imaging, because of

Fig. 1—36-year-old woman with synovial sarcoma of parapharyngeal space.
A, Axial T1-weighted MR image reveals well-defined mass (asterisk) deep in relation to parotid gland, anteromedial to carotid sheath vessels, minimally displacing parapharyngeal fat (arrow). Signal intensity of mass is brighter than that of muscle.
B, Coronal T1-weighted MR image shows cap of parapharyngeal fat (arrow) along superior margin of synovial sarcoma (asterisk).
C, Mass (asterisk) enhances intensely on enhanced T1-weighted scans, similarly to right parotid gland but less than sinonasal mucosa (seen on more superior sections). Enhancement of mass was graded moderate.
MR Imaging of Head and Neck Synovial Sarcomas

Fig. 2.—54-year-old man with synovial sarcoma of parapharyngeal space.
A. T1-weighted scan shows that signal intensity of parapharyngeal synovial sarcoma (asterisk) is homogeneous. Mass abuts right pterygoid muscle (P) and obliterates parapharyngeal fat.
B. Signal intensity on T2-weighted scan is heterogeneous with lower intensity components centrally and posteriorly (white arrows) and higher intensity components (black arrows) anteriorly.
C. Gadolinium enhancement on T1-weighted imaging is also heterogeneous, with nodular areas of enhancement (arrows) intermixed with nonenhancing zones. On the basis of absence of enhancement, pterygoid (P) muscle does not appear to be infiltrated.

Fig. 3.—46-year-old woman with aggressive growth of synovial sarcoma of masticator space.
A. Synovial sarcoma (asterisk) had grown into posterior aspect of left maxillary sinus from masticator space and pterygopalatine fossa.
B. Signal intensity on T2-weighted scans was graded similar to that of fat.
C. Coronal gadolinium-enhanced fat-suppressed T1-weighted MR image shows growth of mass through skull base into inferior margin of left cavernous sinus (arrows). Mass had also grown into inferior left orbit through inferior orbital fissure from pterygopalatine fossa.
its improved soft-tissue resolution, is considered the procedure of choice for detecting and staging soft-tissue tumors; however, its usefulness in suggesting a particular histologic diagnosis remains limited [1].

Synovial sarcomas are frequently misclassified on imaging as benign masses because of their smooth margins, cystic components, and lack of aggressive infiltration [3]. Within the differential diagnosis of nonmucosal head and neck masses in these locations, one should consider neurogenic tumors, other sarcomas, and tumors of ectopic minor salivary glands. Schwannomas and neurofibromas often present like synovial sarcomas as neck masses. They are of intermediate signal intensity on T1-weighted MR images. The T2-weighted signal intensity varies according to whether the lesion is highly cellular (intermediate intensity) or cystic or stromal (nonhomogeneously high intensity) and whether the content of the tissue is Antoni A or Antoni B [9].

In the malignant category one must include rhabdomyosarcomas, which account for 84% of all soft-tissue sarcomas and 35–45% of those that occur in the head and neck [10]. On MR imaging, rhabdomyosarcomas are remarkably homogenous and have intermediate signal intensities on all imaging sequences. Fibrosarcomas account for 12–19% of all soft-tissue sarcomas; only 15% appear in the head and neck [10]. On MR imaging they tend to have low to intermediate signal intensities on all imaging sequences. Liposarcomas compose 15–18% of all malignant soft-tissue sarcomas. MR imaging shows a heterogeneous high T1-weighted signal intensity and an intermediate T2-weighted signal intensity. Leiomyosarcomas represent only 5–6% of all soft-tissue sarcomas, of which only 3–10% occur in the head and neck [10]. Hemangiopericytomas are uncommon vascular lesions that arise primarily in the lower extremities. However, 15% occur in the head and neck. On MR imaging, they enhance and have a low to intermediate T1-weighted signal intensity and a higher T2-weighted signal intensity. Angiosarcomas of soft tissue account for only 2–3% of soft-tissue sarcomas. On MR imaging, these tumors are aggressive bone-destroying lesions that enhance greatly. Synovial sarcomas do not seem to enhance as markedly as tumors of vascular origin, and, owing to the presence of cystic, hemorrhagic, or calcified regions of the tumor, the enhancement may be heterogeneous.

Minor salivary gland pleomorphic adenomas are one of the most common primary parapharyngeal lesions, presumably because of ectopic rests of glandular tissue in this area, and approximately 10% of all head and neck tumors are of glandular origin [11]. Pleomorphic adenomas are bright on T2-weighted scans and generally enhance homogeneously, although they may appear pleomorphic. The malignant versions of these tumors may be of variable cellularity. These tumors tend to have an intermediate signal intensity on T1-weighted MR images. The T2-weighted signal intensity depends on the cellularity of the neoplasm: highly cellular types usually are of intermediate signal intensity on T2-weighted images, and the stromal or less cellular variety has a high T2-weighted signal intensity.

Calcification, present in approximately 30% of synovial sarcomas [4], may distinguish this lesion from others. Characteristically, these appear as round, calcified concretions best visualized with CT scanning. Discrete solitary or multiple calcifications may, however, signify a chronic inflammatory process. On the other hand, diffuse calcifications in a lesion with poorly defined margins suggest a sarcoma (e.g., osteosarcoma).

Our results differed from some of the previously collected data. Sigal et al. [5] described the MR and CT characteristics in three cases of head and neck synovial sarcoma. Their first case revealed low signal intensity on T1- and T2-weighted images and minimal enhancement after the administration of gadolinium. The second case showed low signal intensity on T1-weighted images, high signal intensity on T2-weighted images, and heterogeneous enhancement. The third case showed mixed signal intensity on both T1- and T2-weighted images and heterogeneous enhancement. Because each had a different appearance on MR imaging, Sigal et al. concluded that the MR findings were nonspecific and that no characteristic factors existed to differentiate synovial sarcomas from other soft-tissue tumors arising from the head and neck.

We agree with other reports that on T1-weighted images the masses were mainly hypointense relative to fat. One group reported that no notable evidence was evident between the MR imaging characteristics of the monophasic and the biphasic pathologic subtypes [12] but found that the following combination of MR imaging characteristics suggested synovial sarcoma: hemorrhagic areas within the tumor, signal heterogeneity, fluid-fluid levels, and well-defined margins [12]. We have corroborated these findings.

On MR images, three of our six synovial sarcomas appeared unilocular and half appeared multilocular. All the multilocular lesions were noted to be heterogeneous, and one was cystic. One of the unilocular lesions was noted to have homogeneous signal intensity. No edema was seen, and the tumors were remarkable for their well-defined margins, unusual for such a malignant tumor. The predominant feature on the MR images in these six cases was the heterogeneous configuration of the mass.

We have found a wide spectrum of MR imaging findings in synovial sarcomas of the head and neck. The tumor may mimic the benign neoplasms that we have described. In general, however, a head and neck synovial sarcoma on T1-weighted images is most frequently isointense to gray matter and has a signal intensity similar to that of fat or glandular tissue on T2-weighted images. When one encounters these signal-intensity characteristics in a well-defined yet heterogeneous mucosal or nonmucosal lesion that also contains septations, hemorrhage, cyst formation, calcification, or multilocularity, one should consider the diagnosis of synovial sarcoma. Histopathologic sampling is required because no pathognomonic features define this lesion.

References