Rhabdomyosarcomas in the Head and Neck: MR Imaging Evaluation

To determine the typical magnetic resonance (MR) signal intensity characteristics of rhabdomyosarcomas, short repetition time (TR)/short echo time (TE) (T1-weighted) and long TR/proton density and T2-weighted images of 13 patients with rhabdomyosarcomas of the head and neck were retrospectively reviewed. Seven patients received gadopentetate dimeglumine injections. The most common MR appearance was that of a homogeneous mass, isointense to both muscle and fat on long TR/short TE images and isointense or minimally hyperintense to muscle on short TR/long TE images. All lesions of the patients who received gadopentetate dimeglumine enhanced markedly. Two lesions had intratumoral hemorrhage, and six were markedly heterogeneous in signal intensity. Similar MR signal intensity patterns have been described for lymphomas and nasopharyngeal carcinomas. The forte of MR imaging lies in its ability to delineate precisely the extent of the rhabdomyosarcoma.

Rhabdomyosarcomas of the head and neck may occur in a variety of locations, including the nasopharynx, middle ear cavity, orbits, parapharyngeal soft tissues, and nasal/paranasal cavities. They often grow insidiously and may invade the intracranial space through the numerous foramina leading to the brain. These characteristics of the tumor make the multiplanar imaging capabilities of magnetic resonance (MR) imaging very useful for evaluating the full extent of the lesion. To our knowledge, no study to date has described the MR imaging features of head and neck rhabdomyosarcomas.

MATERIALS AND METHODS

Thirteen patients with histopathologically proved rhabdomyosarcomas of the head and neck underwent MR imaging at either the Hospital of the University of Pennsylvania or the Children's Hospital of Philadelphia over a 4-year period. There were six female and seven male subjects, with an age range of 5 months to 30 years (average age, 9 years). The tumors originated in the soft palate (n = 1), nasopharynx (n = 3), nasal cavity (n = 1), nasolabial fold (n = 1), infratemporal fossa (n = 1), tongue (n = 1), middle ear (n = 1), base of skull (n = 3), and submandibular soft tissue (n = 1). Ten of the rhabdomyosarcomas were of the embryonal type and three were alveolar.

The MR images were obtained with use of a GE Medical Systems 1.5-T unit (Milwaukee) and/or a Siemens 1.5-T unit (Iselin, NJ) and were evaluated independently by three neuroradiologists. Axial spin-echo (SE) images were obtained with the following parameters: 600–800/15–30 (repetition time [TR] msec/echo time [TE] msec), 2,500–3,000/30–45, 80–90. Coronal images were also obtained to evaluate intracranial and intraorbital extension with short TR/short TE SE sequences. Seven patients received an injection of gadopentetate dimeglumine (Magnevist; Berlex, Wayne, NJ) (dose = 0.1 mmol/kg) and were immediately imaged with SE 600–800/20–30 sequences with gradient moment nulling (flow compensation) applied. No delayed post-gadopentetate dimeglumine images were obtained.

The signal intensities of the tumors were compared to those of fat and muscle on the short TR/short TE, long TR/short TE, and long TR/long TE images. The lesions were assessed for signal intensity homogeneity and the presence of hemorrhage on the long TR/long TE images. The presence of enhancement following injection of gadopentetate dimeglumine was noted. Each tumor was assessed for the presence of intraorbital, intracranial, or lymph node extension. Intraorbital and intracranial extension was inferred by contiguous spread of similar intensity tissue from the main tumor bulk, which was sampled for biopsy. Because the tumors were treated with chemotherapy and/or radiation therapy, the tumor margins were never surgically confirmed and no patient had a complete excision of the tumor (biopsy or aspiration cytology specimens of readily accessible portions of the tumors were obtained for pathologic verification). Round lymph nodes that had abnormal signal intensity on long TR/long TE images and that were greater than 1.5 cm in size were suggestive of lymph node extension. In one case, biopsy of a lymph node was performed, and the results confirmed the presence of tumor. In all other cases, the lymph nodes were believed to be involved clinically. Because hyperplastic lymph nodes may approach 1.5 cm in size in this age group, the presence of neoplasm is not assured, since nodal dissections and biopsies were not performed.

RESULTS

See the Table for a summary of the MR findings. On the short TR/short TE images, the rhabdomyosarcomas were isointense or very minimally hyperintense in relation to muscle in 12 of the 13 cases (Fig 1) and were mixed in intensity in one case (Fig 2). This case demonstrated hyperintense regions, which were believed to rep-
Figure 1. Alveolar rhabdomyosarcoma of the tongue. (a) Sagittal 700/20 image through the tongue fails to reveal the lesion. It is isointense to the normal tongue musculature. (b) Sagittal 3,000/30 image demonstrates an inhomogeneous mass infiltrating the superior aspect of the tongue. (c) The 3,000/90 image depicts the inhomogeneity of the mass to a greater degree.

<table>
<thead>
<tr>
<th>Tumor Origin</th>
<th>Histologic Subtype</th>
<th>Patient Age</th>
<th>Signal Intensity in Relation to Muscle</th>
<th>Signal Intensity in Relation to Fat</th>
<th>Enhancement with Use of Gadopentetate Dimeglumine</th>
<th>Homo- or Heterogenous Intratumoral Hemorrhage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nasopharynx</td>
<td>Embryonal</td>
<td>2 y</td>
<td>+</td>
<td>-</td>
<td>Not given</td>
<td>Hetero</td>
</tr>
<tr>
<td>Nasopharynx</td>
<td>Embryonal</td>
<td>7 y</td>
<td>+</td>
<td>+</td>
<td>Not given</td>
<td>Homo</td>
</tr>
<tr>
<td>Nasopharynx</td>
<td>Alveolar</td>
<td>15 y</td>
<td>+</td>
<td>+</td>
<td>Not given</td>
<td>Homo</td>
</tr>
<tr>
<td>Soft palate</td>
<td>Embryonal</td>
<td>15 y</td>
<td>+</td>
<td>+</td>
<td>Not given</td>
<td>Homo</td>
</tr>
<tr>
<td>Nasal cavity</td>
<td>Embryonal</td>
<td>4 y</td>
<td>+</td>
<td>+</td>
<td>Not given</td>
<td>Homo</td>
</tr>
<tr>
<td>Submandibular</td>
<td>Embryonal</td>
<td>5 m</td>
<td>+</td>
<td>+</td>
<td>Not given</td>
<td>Homo</td>
</tr>
<tr>
<td>Tongue</td>
<td>Alveolar</td>
<td>9 y</td>
<td>+</td>
<td>+</td>
<td>Not given</td>
<td>Homo</td>
</tr>
<tr>
<td>Middle ear</td>
<td>Embryonal</td>
<td>3 y</td>
<td>+</td>
<td>+</td>
<td>Not given</td>
<td>Homo</td>
</tr>
<tr>
<td>Skull base</td>
<td>Embryonal</td>
<td>30 y</td>
<td>+</td>
<td>+</td>
<td>Not given</td>
<td>Homo</td>
</tr>
<tr>
<td>Infratemporal fossa</td>
<td>Embryonal</td>
<td>13 y mixed</td>
<td>+</td>
<td>+</td>
<td>Not given</td>
<td>Homo</td>
</tr>
<tr>
<td>Skull base</td>
<td>Embryonal</td>
<td>8 y</td>
<td>+</td>
<td>+</td>
<td>Not given</td>
<td>Homo</td>
</tr>
<tr>
<td>Nasolabial fold</td>
<td>Alveolar</td>
<td>2 y</td>
<td>+</td>
<td>+</td>
<td>Not given</td>
<td>Homo</td>
</tr>
<tr>
<td>Skull base</td>
<td>Embryonal</td>
<td>6 y</td>
<td>+</td>
<td>+</td>
<td>Not given</td>
<td>Homo</td>
</tr>
</tbody>
</table>

Note.—T1W = short TR/short TE SE, PD = long TR/short TE SE, T2W = long TR/long TE SE, = denotes isointense, + = hyperintense, - = hypointense.

In this category, isointensity also includes characteristics that one observer believed represented very minimal hyperintensity.

|                     |                  |              | Tumor had hemorrhage, but signal intensity of nonhemorrhagic portion is as listed. |

The long TR/short TE sequence revealed tumors to be of increased signal intensity compared to that of muscle in all cases, isointense with fat in two (15%) cases, slightly hypointense to fat in nine (69%) cases, slightly hyperintense to fat in one (8%) case, and with areas of intensity above and below that of fat in one case with hemorrhage (its nonhemorrhagic portions were isointense to fat). In five (38%) of the 13 cases, the signal intensity was significantly heterogeneous on long TR images yet remained either hypointense or hyperintense to fat. In the sixth case in which the tumor was heterogeneous in intensity, the tumor contained hemorrhage (Fig 2).

The long TR/long TE sequence revealed all lesions to be hyperintense to both muscle and fat. The signal intensity was inhomogeneous in six (46%) cases, with hemorrhagic products in two (15%). In all seven cases in which gadopentetate was used, postcontrast images demonstrated enhancement of the tumor.

No differences in signal intensity characteristics or invasiveness were noted when the embryonal and alveolar subtypes of head and neck rhabdomyosarcomas were compared.

DISCUSSION

Rhabdomyosarcoma is the most common soft-tissue sarcoma in children, occurring most frequently in children under 12 years of age (1–3). Forty percent of rhabdomyosarcomas occur in the head and neck (4). In this region, the orbits, nasopharynx, and middle ear are the most commonly affected sites (2–5). Histologically, the most common forms found in the head and neck are the embryonal and botryoid type, which rarely arise in skeletal muscle (as opposed to the alveolar type, which of-
Figure 2. Embryonal rhabdomyosarcoma of the infratemporal fossa with intratumoral hemorrhage. (a) Axial 600/20 image demonstrates a 5 x 6-cm mass (arrows), which originated from the infratemporal fossa and extended into the maxillary sinus. The mass shows mixed signal intensity with areas representing presumed subacute hemorrhage centrally (H). The mastoid air cells on the right side are opacified, probably due to obstruction of the right eustachian tube. (b) At a higher section, this axial 600/20 image demonstrates intraorbital extension of tumor (arrowheads) as well as subtle invasion of the middle cranial fossa (arrows). The intracranial involvement was not detected with CT. (c) Axial 3,000/35 image demonstrates inhomogeneous signal with evidence of high-intensity subacute hemorrhage encircled by an area of low intensity, probably representing hemosiderin. (d) Axial 3,000/90 image shows more susceptibility effect from the hemosiderin and high-intensity tumor compared with muscle and fat. Obstructed mastoid and maxillary sinus secretions are of higher intensity than tumor. (e) Post-gadopentetate dimeglumine image shows enhancement predominantly of the posterior and medial portion of the tumor (arrowheads). (f) Coronal post-gadopentetate dimeglumine image demonstrates enhancement on the surface of the inferior right temporal lobe (arrows) where the tumor has extended across the base of the skull to the middle cranial fossa.

ten occurs in the extremities of children (3,5,6). The term botryoid refers to the grapelike growth of the tumor from mucosal surfaces. The pleomorphic type of rhabdomyosarcoma tends to predominate in adults (3,6). The tumors often have a loose stromal network, which accounts for the high overall water content reflected in the high signal intensity on long TR/TE images. No signal intensity differences were noted between embryonal or alveolar rhabdomyosarcomas.

Rhabdomyosarcomas in parameningeal regions (nasopharynx, paranasal sinuses, nasal cavity, and middle ear) may invade the meninges in 35%-55% of these patients and may cause death in 90% of those patients with meningeal invasion (7,8). Bone erosion occurs in 18% of all parameningeal rhabdomyosarcomas and in up to 67% of middle ear tumors (2,7).

Coronal MR imaging is the best method for demonstrating intracranial extension. We detected intracranial spread of the tumors in 23% of the 13 patients with head and neck rhabdomyosarcomas and in 33% of patients with tumors in parameningeal sites. From the information provided by imaging, the radiation therapist will include the full margins of the tumor plus a 2-3-cm boundary. In some protocols, intracranial spread necessitates whole head or craniospinal irradiation (Goldwein J, oral communication, 1990). Spread of the tumor to lymph nodes in the neck is reported in 12%-50% of cases (3,4). Hematogenous spread may be to the lungs, bones, or brain (2,3).

Anecdotal descriptions of the MR characteristics of single rhabdomyosarcomas have been published (4,9-11). To our knowledge, this article is the first to describe the signal intensity findings in a group of patients with head and neck rhabdomyosarcomas. The findings that the tumors are isointense or nearly isointense to muscle on short TR/short TE (T1-weighted) images, variable in signal
intensity on long TR/short TE (proton density) images, and high in intensity on long TR/long TE (T2-weighted) images may be of some use in the differential diagnosis of head and neck malignancies. Som et al have reported that high-grade malignancies of the paranasal sinuses and salivary glands may have lower signal intensity on long TR/long TE images (12,13). Liposarcomas will usually have a component of fat intensity, high on short TR images and lower on long TR/TE images. The presence of areas of calcification, seen as signal voids with all pulse sequences, may suggest the diagnosis of chordoma, chondrosarcoma, chondroma, or osseosarcoma, tumors that may occur in young adults in locations similar to those of rhabdomyosarcomas. An intratumoral hemorrhage demonstrated at MR imaging has been reported as an atypical feature of rhabdomyosarcomas; however, in histopathologic studies, the tumors are known to bleed and have central necrosis (2,5).

Rhabdomyosarcomas should be easily differentiated from benign lesions in the head and neck in children (eg, branchial cleft cysts and thyroglossal duct cysts), since these lesions usually are of lower intensity than muscle on short TR images and high in intensity on both echoes of the long TR images. Lymphangiomas also tend to have the same "cystic" signal intensity pattern and demonstrate intratumoral hemorrhage more commonly.

Unfortunately, other relatively common tumors of the head and neck—lymphoma and nasopharyngeal carcinoma—usually have the same signal intensity characteristics as rhabdomyosarcomas (9,10,14). Both of these lesions also have a propensity for intracranial invasion through the skull base foramina. Lymphomas are usually homogeneous in signal intensity; heterogeneous signal intensity would suggest nasopharyngeal carcinoma or rhabdomyosarcoma (9). Patients with acquired immunodeficiency syndrome (AIDS) may develop head and neck Kaposi sarcomas, which may simulate rhabdomyosarcomas. The presence of generalized lymphadenopathy may suggest AIDS-related Kaposi sarcoma (14). Although the younger age of the patients with rhabdomyosarcomas will suggest the diagnosis, there may be overlap in the age group affected by lymphoma, AIDS, and nasopharyngeal squamous cell carcinoma, particularly in the Chinese population (15). The nonspecificity of the MR signal intensity characteristics underscores the role of aspiration cytology or surgical biopsy with use of clinical, computed tomographic, ultrasound, or MR guidance for definitive histologic diagnosis of head and neck masses.

References