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Imaging of Rhabdomyosarcomas of the Head and Neck

Joseph T. Latack¹ Raymond J. Hutchinson² Ruth M. Heyn²

Rhabdomyosarcoma (RMS) is the most common childhood malignancy of the head and neck. The Intergroup Rhabdomyosarcoma Study now divides head and neck RMS into three categories by site of origin: (1) orbital, (2) parameningeal (middle ear, paranasal sinuses, and nasopharynx), and (3) all other head and neck sites. CT is clinically applicable in the diagnosis of RMS of the head and neck, in treatment planning, and in the follow-up of patients with these tumors. Specific areas of applicability include (1) determination of the presence/absence of intracranial and meningeal involvement, (2) definition of tumor extent to guide radiation therapy planning, and (3) demonstration of tumor regression or recurrence during and after treatment. CT has played an important role in the dramatically improved prognosis seen in RMS over the last 10 years. The role of MR in evaluating these patients is not yet defined, but it has promise because of the ease of obtaining multiple projections and the avoidance of ionizing radiation.

Rhabdomyosarcomas (RMSs) are the most common soft-tissue sarcomas of childhood and account for 4–8% of all malignancies in patients under the age of 15 years [1, 2]. Forty percent of all RMSs arise in the head and neck [3]. In this article we review the imaging findings of RMSs of the head and neck and discuss their role in the diagnosis, staging, and follow-up of these neoplasms.

Materials and Methods

In the last 6 years, 17 patients have presented at the University of Michigan Medical Center with RMS of the head and neck. All 17 patients were imaged with CT after IV infusion of contrast material. Contiguous 5-mm-thick axial images of the head and neck were obtained in all patients. Contiguous 5-mm-thick coronal images of the head and neck were also obtained in eight patients. Contiguous 1.5-mm-thick axial bone-algorithm images of the base of the skull and temporal bone were obtained in 13 patients. After initiation of treatment the patients received CT follow-up, initially at 3-month and later at 6- to 12-month intervals. MR images of the head and neck were obtained in axial, sagittal, and coronal projections in three patients. A superconducting 0.35-T magnet and spin-echo sequence were used with a repetition time (TR) of 0.5–2.0 sec and echo time (TE) of 28 and 56 msec. Chest films, chest CT scans, and radionuclide bone scans were the only other routine imaging procedures. The radiographic findings were correlated with the clinical data, surgical findings, and pathologic specimens.

Results

The RMSs in our series arose in the orbit, middle ear, nasopharynx, nasal cavity, paranasal sinuses, and soft tissues of the neck. Imaging findings were variable, depending on the site of origin of the RMS. The site of origin, age of patient at the time of diagnosis, cell type, clinical group, presenting symptoms, and clinical course of RMS patients are found in Table 1.

Of the five orbital RMSs, two presented as eyelid masses and CT demonstrated

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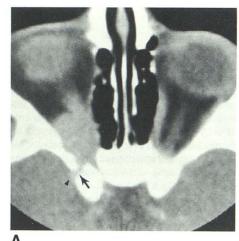
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TABLE 1: Site of Origin, Age, Cell Type, Clinical Grouping, Presenting Symptoms, and Clinical Course in Head and Neck Rhabdomyosarcomas

Site: Case No.	Age at Diagnosis	Cell Type	Clinical Group	Presenting Symptoms	Clinical Course
Orbit:					
1	10	Embryonal	II	Mass in upper lid	Alive; NED >3 yr
2	1	Embryonal	II	Mass in upper lid/brow	Alive; NED >3 yr
3	5	Embryonal	П	Double vision, slight proptosis, third nerve palsy	Alive; NED >3 yr
4	6	Embryonal	Ш	Rapidly progressive proptosis	Alive, but with intracrania extension of disease
5	6	Embryonal	11	Rapidly progressive proptosis	Alive; NED >3 yr
Parameningeal, mide	dle ear:	,		, , , ,	
6	7	Embryonal	III	Diminished hearing at school examination and bleeding from canal	Alive; NED >3 yr
7	3	Embryonal	III	Bleeding from canal	Alive; NED 3 yr
8	5	Embryonal	III	Otorrhea, facial palsy, mass	Died of progressive RMS
9	10	Embryonal	III	Ear pain and mass	Alive; NED 2 yr
10	3	Embryonal	III	Facial palsy and facial swelling	Alive; NED >3 yr
Parameningeal, naso	opharynx:	,		, , , , , , , , , , , , , , , , , , , ,	,
11	4	Embryonal	Ш	Nasal obstruction, weight loss, strabis- mus, ptosis of lid	Died of progressive RMS
Parameningeal, nasa	al cavity:			,	
12	46	Alveolar	Ш	Nasal obstruction, epistaxis, serous otitis media	Alive but with disease
13	18	Alveolar	Ш	Nasal obstruction, epistaxis, serous otitis media	Died of progressive RMS
Parameningeal, ethn	noid sinus:				
14	20	Embryonal	Ш	Nasal obstruction, epistaxis, serous otitis media	Alive; NED >3 yr
Parameningeal, max					
15	12	Alveolar	III	Nasal obstruction, mass in medial orbit and upper cheek	Alive; NED >3 yr
Other sites (soft tiss	ues of head and	neck):			
16	1	Embryonal	IV	Neck mass	Died of progressive RMS
17	16	Embryonal	IV	Neck mass	Alive; NED >3 yr

Note.—NED = no evident disease; RMS = rhabdomyosarcoma.



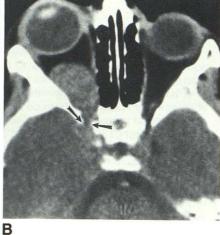


Fig. 1.—Case 3, 5-year-old girl. Rhabdomyosarcoma of orbit. Axial, 5-mm-thick orbital CT images after IV contrast enhancement.

A, Relatively well-demarcated, intraconal, homogeneous mass of posterior and lateral aspect of right orbit. Optic nerve displaced medially. Destruction of superior orbital fissure (arrow) with tumor extension intracranially (arrowhead).

B, 3 weeks later. Mass has enlarged. Destruction of superior orbital fissure more marked (arrows).

only minimal intraorbital extension (cases 1 and 2). The other three orbital RMSs were relatively well-defined, homogeneous masses located in the posterior lateral aspect of the orbit and occupying about one-third of the volume of the orbit (cases 3–5) (Fig. 1A). Two lesions were intraconal and one was both

intra- and extraconal. After IV contrast enhancement all lesions enhanced to the same degree as surrounding muscle. Two of the RMSs eroded the superior orbital fissure in the process of extending into the cavernous sinus (cases 3 and 4) (Fig. 1). This intracranial extension was also verified by MR

in case 4. Repeat pretreatment CT 4 weeks after the original study showed a significant increase in the size of the orbital mass in cases 3 and 5 (Fig. 1B).

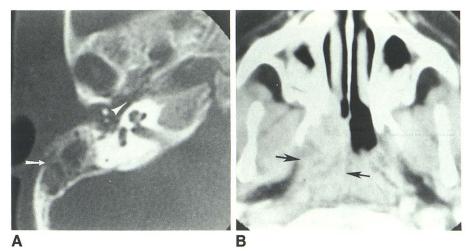
Five RMSs arose in the middle ear. One was confined entirely to the middle ear and on CT was seen as a soft-tissue mass without bone destruction (case 6). A second dumbbell-shaped RMS arose in the middle ear, extended by a narrow strand of tumor through the eustachian tube, and enlarged again in the nasopharynx (case 7) (Fig. 2). The other three middle-ear RMSs were seen as contrast-enhancing masses centered in the middle ear and involved the middle cranial fossa, the posterior cranial fossa, and the surrounding soft tissues of the scalp (cases 8–10) (Fig. 3A). Marked destruction of the temporal bone was present (Fig. 3B). MR in case 9

yielded a high-signal abnormality on T2-weighted images (Fig. 4).

Five RMSs arose in the nasopharynx, nasal cavity, or the paranasal sinuses. One of these arose in the posterior nasopharynx, extended medially across the midline, anteriorly into the nasal cavity (Fig. 5A), and anterosuperiorly into the apex of the orbit and the floor of the middle cranial fossa (case 11) (Fig. 5B). Two RMSs originated in the nasal cavity, one in the anterior ethmoid sinus, and one in the maxillary sinus; all four had very similar CT appearances. A soft-tissue mass involved the nasal cavity, maxillary sinus, anterior ethmoid air cells, and orbit with destruction of the bone separating these various cavities (Fig. 6). MR of the RMS arising in the ethmoid sinus (case 14) showed a signal abnormality similar in size

Fig. 2.—Case 7, 3-year-old girl. Rhabdomyosarcoma of middle ear. Axial CT of temporal bone and nasopharynx after IV contrast enhancement.

- A, 1.5-mm-thick bone-algorithm image of right temporal bone. Mass causing opacification of right tympanic cavity, mastoid, and eustachian tube. Surrounding bone of mastoid (arrow) and eustachian tube (arrowhead) destroyed.
- B, 5-mm-thick image of nasopharynx. Rhabdomyosarcoma has extended from middle ear to nasopharynx (arrows) via eustachian tube.



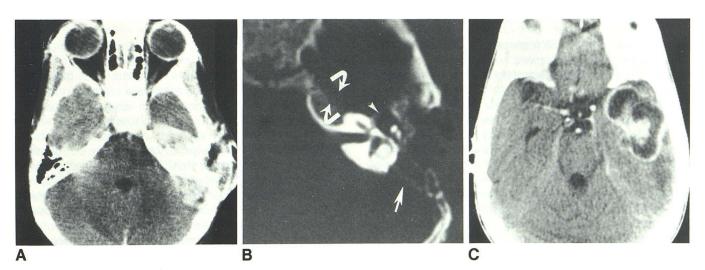


Fig. 3.—Case 8, 5-year-old boy. Rhabdomyosarcoma of left middle ear. Axial CT of temporal bone and brain.

A, 10-mm-thick image of brain after IV contrast enhancement. Enhancing mass centered in left temporal bone with extension into middle cranial fossa, posterior cranial fossa, and soft tissues lateral to temporal bone.

B, 1.5-mm-thick bone-algorithm image of left temporal bone. Soft-tissue mass of and adjacent to middle ear cavity. Destruction of bone of anterolateral petrous apex (curved arrows), tegmen (arrowhead), posterior mastoid (straight arrow), and external auditory canal.

C, 10 months after initial CT study. IV contrast-enhanced 10-mm-thick image of brain. Necrotic mass of right temporal lobe secondary to direct extension of rhabdomyosarcoma from temporal bone.



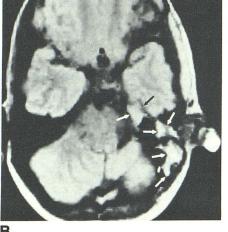
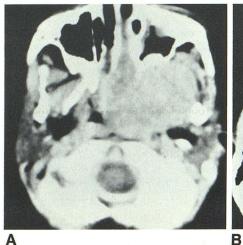


Fig. 4.—Case 9, 10-year-old boy. Rhabdo-myosarcoma of left middle ear.

A, Axial, 10-mm-thick brain CT image after IV contrast enhancement. Enhancing mass centered in region of middle ear with extension to middle cranial fossa, posterior cranial fossa, and soft tissues lateral to temporal bone.

B, Spin-echo pulse sequence, 2-sec TR, 28-msec TE. High-signal mass in left petrous apex, middle ear, mastoid, external auditory canal, and adjacent soft tissues (arrows). Absence of signal in uninvolved bony labyrinth and internal auditory canal.



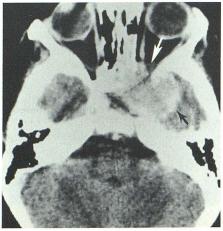


Fig. 5.—Case 11, 4-year-old girl. Rhabdomyosarcoma of posterior nasopharynx. Axial, 5mm-thick CT image after IV contrast enhancement.

A, Level of nasopharynx. Soft-tissue mass involves nasopharynx, posterior aspects of nasal cavity bilaterally, left maxillary sinus, and left infratemporal fossa. Destruction of left pterygoid plates and anterior displacement and destruction of posterolateral wall of left maxillary sinus.

B, Level of orbits and lower middle cranial fossa. Extension of mass into left middle cranial fossa and left orbit (arrows).

and configuration to the CT attenuation abnormality. The orbital involvement in this patient, however, was not as well seen on MR because of the lack of bone signal and the poorer spatial resolution.

Two RMSs arose in the soft tissues of the neck. On CT both of these lesions were large, relatively homogeneous soft-tissue masses located beneath the sternocleidomastoid muscle (Fig. 7). Some of the margins of the mass were distinct while other margins, demonstrating the aggressive nature of these lesions, blended into adjacent tissue planes. After contrast enhancement, these masses enhanced the same as or somewhat less than surrounding muscle. Although vertebral destruction and intraspinal extension was not appreciated on CT in either patient, myelography demonstrated a complete block to contrast media at the region of the mass in one patient (case 16).

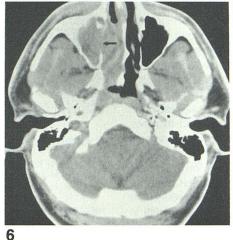
None of the 17 patients had positive chest films, chest CT scans, or radionuclide bone scans at the time of presentation, and only one (case 11) developed a chest metastasis later in the disease course. In two patients, CT demonstrated metastatic RMSs in neck lymph nodes; in a third patient with a

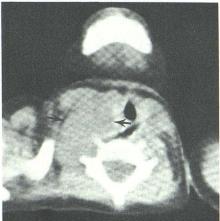
palpable abdominal mass, abdominal paraspinal adenopathy was confirmed by CT.

All patients were rescanned 3 months after diagnosis. Eleven patients who ultimately achieved a complete response were free of disease on the 3-month scan. Three patients with a diminished mass but persistent disease on the 3-month scan have all done poorly: one died with extensive intracranial extension from a middle ear primary (case 8) (Fig. 3C), another died of intracranial spread from a nasopharyngeal primary (case 11), and the third is alive but with significant tumor at the original site (case 12). Three patients were free of disease on the initial 3-month follow-up examination, but later developed a recurrence, two at the original tumor site and one at a distant site. One local recurrence occurred in a patient with an RMS that arose in the neck and initially compressed the cervical spinal cord (case 16). After a 1-year disease-free period, the tumor recurred at the original site and ultimately led to this patient's death. The second local recurrence, after a disease-free interval, was in a patient with frontal lobe extension from an adjacent orbital RMS (case 4). This patient is alive but with significant intracranial disease. The recurrence

Fig. 6.—Case 12, 46-year-old man. Rhabdomyosarcoma arising in nasal cavity. Axial, 5-mmthick CT image after IV contrast enhancement. Soft-tissue mass involves anterior nasopharynx, right maxillary sinus, and right nasal cavity. Bone destruction of medial wall of right maxillary sinus

Fig. 7.-Case 16, 1-year-old boy. Rhabdomyosarcoma of soft tissues of the neck. Axial. 5-mm-thick CT image of neck after IV contrast enhancement. Homogeneous mass (arrows), less dense than surrounding muscle, beneath right sternocleidomastoid. Borders of mass distinct laterally but other borders blend with surrounding tissue. Bone destruction or intraspinal extension not apparent on CT, but myelography showed block at site of soft-tissue mass.





away from the original site was seen in a patient (case 13) whose RMS originated in the nasal cavity and, after a 1-year disease-free interval, seeded the subarachnoid spaces with tumor, eventually leading to her death.

Discussion

The Intergroup Rhabdomyosarcoma Study divides head and neck RMSs by site of origin into three categories: (1) orbital, (2) parameningeal, and (3) all other head and neck sites [3, 4]. As previously reported and as seen in our patients, orbital RMS patients often present with proptosis and have a good prognosis, with a disease-free survival rate of 80-90% [5]. The good prognosis associated with orbital RMS may be related to early detection, lack of orbital lymphatics, and prevention of early spread by the rather complete bony wall of the orbit [2]. Secondary orbital involvement was also relatively common in our series, as four RMSs extended into the orbit from other primary sites. Parameningeal RMSs occur adjacent to the meninges of the skull base, and in our series consisted of tumors from three different regions: (1) middle ear, (2) posterior nasopharynx and posterior paranasal sinuses, and (3) nasal cavity and anterior paranasal sinuses. These lesions have the poorest prognosis of the head and neck RMSs because of their tendency to invade the base of the skull early and extend intracranially [6, 7]. Parameningeal RMSs often present with bloody discharge from the nose or ear [8, 9]. The disease-free survival rate of our parameningeal RMS patients again approached the 47% reported in the literature [4, 10-12]. The two RMSs of other head and neck sites originated in the soft tissues of the neck. Other reported sites of origin of RMS include oropharynx, oral cavity, hypopharynx, larynx, parotid gland region, and soft tissue of the scalp and face [13, 14]. The prognosis for these lesions of other sites is between that of orbital and parameningeal RMSs, with a survival rate of about 78% [15]. These RMSs either present as a palpable mass or with symptoms referable to that part of the respiratory or digestive tract that the lesion involves [16, 17].

RMSs develop from primitive pluripotential mesenchymal cells and thus may arise anywhere in the body [4, 18]. The Intergroup Rhabdomyosarcoma Study classifies these neoplasms into four cell types: (1) embryonal, (2) botryoid (variant of embryonal), (3) alveolar, and (4) pleomorphic. Although different histologically, extraosseous Ewing's sarcoma and undifferentiated small round-cell mesenchymal sarcoma, type indeterminate, are grouped with the RMSs by the Intergroup Rhabdomyosarcoma Study because of similar clinical behavior and response to treatment protocol [19-21]. There is not a good correlation between cell type and prognosis in head and neck RMSs [22, 23]. The prognosis better correlates with a clinical grouping system also defined by the Intergroup Rhabdomyosarcoma Study [24], which defines group I lesions as localized tumors that are completely resected. Group II patients have gross resection of their tumor but are left with microscopic residual. Group III patients have an incomplete resection of the RMS with gross residual disease remaining, while stage IV patients present with distant metastasis. This staging system, based on the amount of tumor remaining after surgery, is somewhat dependent on the skill, aggressiveness, and intent of the surgeon.

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The CT evaluation of our patients with head and neck RMS consisted of 5-mm axial and, when possible, coronal images of the appropriate region. CT screening of the head and neck in the region of the lymphatic drainage of the tumor is also used since unexpected lymphadenopathy was detected in two patients (cases 12 and 14). When parameningeal lesions are suspected, thin, 1.5-mm images, often with bone and soft-tissue algorithms and photography, may be necessary to image bone destruction and enhancement of involved meninges. On CT, all the RMSs in our series were seen as softtissue masses of either the orbit, temporal bone, nasopharynx-paranasal sinuses area, or soft tissues of the neck. Although a few lesions were relatively well demarcated (Figs. 1 and 7), most were poorly defined, inhomogeneous masses distorting soft-tissue planes and destroying bone. IV contrast material enhanced the RMS generally to the same degree as adjacent muscle.

The worst prognostic indicator in head and neck RMS is meningeal involvement [14]. Tumor cells in the CSF indicate meningeal involvement; however, CT imaging can confirm and define the extent of meningeal involvement [25, 26]. Orbital RMSs spread to the meninges by eroding the superior orbital fissure (Fig. 1). Middle-ear RMS extended through the tegmen tympani, eustachian tube, and semicanal for the tensor tympani to involve the middle cranial fossa meninges (Figs. 3 and 4); and through the posterior mastoid to reach the posterior cranial fossa (Fig. 3). Nasal cavity, paranasal sinus, and nasopharyngeal RMS reached the meninges by extending through basal foramina, destroying the thin roofs of the paranasal sinuses, and by breaking through the floor of the orbit with eventual extension intracranially through the superior orbital fissure (Fig. 5) [27, 28]. One RMS arising in the neck involved the meninges by direct extension into the adjacent cervical spinal canal.

The role of MR in RMS of the head and neck is not clear. The ability to easily obtain coronal and sagittal images is an obvious advantage as is the lack of ionizing radiation in the young population most often afflicted with this neoplasm. The absence of bone signal in MR makes it more difficult to determine bone destruction and the path of the RMS within bone. With the equipment used in these patients, the poorer resolution of MR outweighed its advantages and usually yielded a poorer depiction of the extent of the mass than CT did.

CT has three functions in the evaluation of head and neck RMS: (1) as a diagnostic tool, (2) as a means to define the extent of disease for treatment planning, and (3) in follow-up. In our series, some patients with RMSs of the head and neck had clinical signs and symptoms not conclusively indicating an aggressive process. The CT demonstration of a poorly defined soft-tissue mass with bone destruction was then an indication of the aggressive nature of the lesion and led to immediate biopsy and treatment. Other RMSs in our series were obviously aggressive tumors on clinical examination. CT, although not necessary for diagnosis in these cases, was still performed to define the extent of the lesion and provide information necessary in staging and treatment planning. Most lesions in the middle ear, nasopharynx, and paranasal sinuses do not lend themselves to good surgical removal. At present, these lesions are treated with chemotherapy and radiation after biopsy [29]. Imaging directly influences therapy. If intracranial extension is demonstrated, base-of-brain irradiation, whole-brain irradiation, and intrathecal chemotherapy may all be added to the standard therapeutic regimen [30]. Any CT-detected, clinically silent, distant metastasis also necessitates additional irradiation [31].

The first follow-up CT study 3 months after initial therapy was a good indicator of the patient's prognosis. In our series, complete regression of tumor was seen in 14 patients and only partial regression in three. Of the three patients with residual disease at 3 months, all have had subsequent tumor progression. Absence of tumor on the 3-month follow-up CT study, however, is an imperfect assessment of residual disease. Three of the 14 patients in whom the tumor disappeared by CT subsequently had recurrence. Differentiation of scarring

from residual disease was sometimes a problem and necessitated more frequent follow-up scanning.

The prognosis of head and neck RMS has improved markedly in the last 15 years. Survival rates of less than 20% have risen to over 70% [32, 33]. The increased success is related primarily to a multidisciplinary approach in the treatment of this neoplasm, aided by the use of CT in diagnosis, staging, treatment planning, and follow-up [33].

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