Rhabdomyosarcoma everywhere: Differential diagnosis on Imaging

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Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma of childhood. In this study, we aimed to stress the distinguishing properties of RMS from other small round cell, germ cell, peripheral nerve sheath, and vascular tumors, Langerhans cell histiocytosis (LHH), and other tumors (of bone, fibrous tissue origin) on the basis of imaging findings. Histopathological features of these lesions will also be presented for radiopathologic correlation of given neoplastic tissue types.
Background

Medical records and imaging data of patients diagnosed with RMS were retrospectively evaluated. Based on their localization, RMSs were grouped under the general groups of orbital, parameningeal, genitourinary, and extremity RMSs.
Findings and procedure details

RMS is a malignant tumor of childhood, which belongs to the broader small round cell tumor family. Cross-sectional imaging modalities are used to assess lesion localization and extension, lesion characterization, and differential diagnosis. The histological subtypes of RMS are embryonal (with botryoid and spindle cell variants), alveolar, and pleomorphic or undifferentiated. Embryonal subtype is the most common type of RMS and has a better prognosis. It commonly arises in the mucosal-lined structures of the nasopharynx, auditory canal, genito-urinary and gastro-intestinal tracts. Botryoid RMS usually occurs in the vagina, bladder and biliary tract. Spindle cell RMS arises from the head and neck region, the paratesticular soft tissue, and the abdomen. Alveolar RMS, which has the worst prognosis, commonly occurs on the trunk and lower extremities.

Orbital RMS tends to involve more the superior portion of the orbit. It is usually ill defined and an aggressive, invading frequently orbital walls and the adjacent soft tissues. CT is better for demonstration of bone destruction. At MRI, its imaging features are nonspecific. On postcontrast MRI, the tumor shows usually moderate to marked enhancement (Figure 1). Orbital RMS will be discussed along with cases of LHH, plexiform neurofibroma, hemangioma, and aneurysmal bone cyst.

LHH usually presents as unifocal bone disease with possible orbital involvement but also as multifocal or systemic disease. CT or MRI shows an osteolytic lesions commonly superior or superotemporal orbital region (Figure 2). There may be extension of the soft tissue into the epidural space as well as in the temporal fossa. On postcontrast CT and MRI, lesion show moderate to marked enhancement.

Presence of a plexiform neurofibroma (PNF) is diagnostic of NF1. PNF can involve any peripheral nerve but usually involve sensory nerve in the orbit. It appears as an ill defined irregular mass that cross multiple tissue planes. It can cause dysplasia of the greater sphenoid wing and expansion of the orbital bony rim (Figure 3).

Hemangiomas are very vascular masses. The tumor most commonly occurs in the superior nasal quadrant and most are extraconal. MRI may demonstrate peripheral and internal flow voids, findings that are characteristic of the hemangioma. Dark fibrous septa may also be demonstrated between the hyperintense lobules on T2WI. These vessels are an important distinguishing feature of hemangioma. An intense enhancement after contrast material injection always present (Figure 4 and 5).
Aneurysmal bone cyst is a solitary, expansile lesion. On MRI, it shows the various stages of blood products which appear as layers of different signal intensity on both T1WI and T2WI and multiple cysts (Figure 6).

Parameningeal RMS may originate from the masticator space, the parapharyngeal space, the nasopharynx, the nasal cavity and paranasal sinuses, the mastoid or the middle ear. Parameningeal RMS may show earlier spread to the intracranial compartment (Figure 7,8). Parameningeal RMSs will be presented in comparison with the images from Ewing sarcoma, osteosarcoma, lymphoma, fibroosseous lesion, neuroblastoma, infantile hemangiopericytoma, and granulocytic sarcoma cases.

Ewing sarcoma occurs in the head and neck most commonly the mandible. It presents as a very aggressive mass lesion with permeative destruction of bone and invasion into surrounding tissues. On T1WI, it is isointense to mildly hypointense; on T2W or STIR images it is hyperintense and shows variable contrast enhancement (Figure 9).

Osteosarcoma is a rare entity regarding the head and neck. The vast majority of lesions arise in either the mandible or maxilla. They present with a permeative lytic lesion. There is an associated soft tissue component, and an osteoid tumor matrix. The presence of tumor bone formation within osteosarcoma can aid in the diagnosis of this lesion (Figure 10).

Lymphoma in head and neck most commonly involves the lymph nodes. It may involve the masticator space, the paranasal sinuses and the orbits. The enlargement of the lymphoid ring of Waldeyer in combination with enlarged neck nodes may be seen in lymphoma. Lymphoma has a smooth appearance on imaging and shows moderate enhancement after IV contrast administration. Primary lymphoma of bone is rare and may occur in the mandibula and maxilla. It is seen as ill-defined lytic destructive lesion, most commonly with associated soft tissue mass. In the head and neck, Burkitt lymphoma presents most frequently in the orbit, jaw and meninges (Figure 11).

Fibrous dysplasia is a benign and slowly progressing pathological disorder of bone in which normal bone marrow is replaced by fibro-osseous tissue with expansion of the medullary cavity. CT shows bony expansion with ground glass appearance. MRI appearance is variable. The tumor shows usually heterogenous low signal intensity on T1WI and heterogenous low, intermediate or high signal intensity on T2WI and variable contrast enhancement (Figure 12).

Neuroblastoma is a common tumor of childhood, which involves the CNS either by direct extension or metastases. Neuroblastoma metastases commonly involve the orbit and calvarium. On MRI, the tumor masses have intensity similar to soft tissues and...
enhancement is marked. Bony spicules from the calvarium and splitting the coronal and sagittal sutures are characteristic finding in neuroblastoma (Figure 13).

Hemangiopericytoma has a nonspecific MRI appearance. Prominent serpentine vessels are typically present. It is usually infiltrative and can demonstrate areas of high signal intensity due to hemorrhage (Figure 14).

Granulocytic sarcoma may present prior or after the diagnosis of acute myeloid leukemia and is the commonest form of leukemic involvement of the orbit. It is irregular homogenous mass and has a nonspecific MRI appearance. It has homogenous enhancement on postcontrast sequences (Figure 15).

Genitourinary RMS most commonly involves the bladder (Figure 16), the prostate, the vagina and the uterus (Figure 17). MRI shows a heterogenous mass, which may invade adjacent structures. Genitourinary RMS will be discussed along with cases of Ewing sarcoma, PNET, desmoplastic round cell tumor, undifferentiated sarcoma, neuroblastoma, osteoblastoma, and hemangioma.

PNET may present as a well defined soft tissue mass. There are no characteristic imaging findings to differentiate this tumor from RMS. It should be considered in the differential diagnosis of small round cell tumors of any site (Figure 18).

Desmoplastic round cell tumor is an aggressive primary peritoneal malignancy. It is most commonly seen in the pelvic peritoneal cavity. MRI appearance of the lesion is often indistinguishable from RMS. It should be considered in the differential diagnosis of small round cell tumors of any site (Figure 19).

Undifferentiated sarcomas can be homogeneous but is more often heterogeneous due to areas of necrosis. At MRI, they have heterogeneous signal intensity and enhance heterogeneously after intravenous administration of contrast material (Figure 20).

Osteoblastoma appears as a lytic lesion with expansion. In the posterior elements of the spine, it appears as an expansile mass with associated soft tissue mass. The central portion of osteoblastoma reveal moderate to marked enhancement (Figure 21).

Ewing sarcoma presents as a very aggressive mass lesion with permeative destruction of bone and invasion into surrounding tissues. On T1WI, it is isointense to mildly hypointense; on T2W or STIR images it is hyperintense and shows variable contrast enhancement (Figure 22).
Hemangiomas are very vascular masses. These vessels are an important distinguishing feature of hemangioma. MRI may demonstrate peripheral and internal flow voids, findings that are characteristic of the hemangioma. An intense enhancement after contrast material injection always present (Figure 23).

The extremities represent the third most common site of origin of RMS (Figure 24). RMS of the extremities is almost of alveolar histology. The tumor has a propensity to metastasize to unusual sites (Figure 25). RMSs in extremities will be compared with Ewing sarcoma, synovial sarcoma, osteosarcoma, and desmoid tumor cases.

Ewing sarcoma of bone presents a very aggressive mass lesion with permeative destruction of bone and invasion into surrounding tissues. Metadiaphyses of the long bones are the most common site for Ewing's sarcoma (Figure 26).

Osteosarcoma is the most common pediatric malignant bone tumor and predominantly involve the metaphyses. It manifests as bone destruction with an associated soft-tissue mass. MRI usually show a large mass that is typically heterogeneous in appearance because of necrosis, hemorrhage, and ossification. MRI shows accurate intramedullary extent of the tumor (Figure 27).

Synovial sarcoma is well defined, lobulated in outline. At MR imaging, the lesion shows heterogeneous signal intensity on T2WI and may have fluid-fluid levels due to hemorrhage and necrosis within cystic components of the tumor (Figure 28).

Desmoid tumor is a locally aggressive and poorly circumscribed, with infiltration of the surrounding soft tissues. The signal intensity of the lesions depends on the pathologic nature of the lesion. Signal intensity of desmoid tumor is variable on both T1WI and T2WI. The amount of collagen and myxoid material within lesion affect signal intensity. Lesions usually enhance intensely following intravenous administration of contrast material (Figure 29).

Neuroblastomas can be found anywhere along the sympathetic chain. Less common sites are the pelvis and the neck. On CT, a neuroblastoma is seen as a large, heterogeneous, lobulated soft-tissue mass that shows mild heterogeneous enhancement. Coarse or curvilinear calcifications are usually seen. On MRI, the tumor is typically heterogeneous, with low signal on T1- and high signal intensity on T2-weighted images, and shows enhancement (Figure 30).
**Fig. 2:** Langerhans cell histiocytosis: coronal T2WI (a), coronal T1WI (b), contrast enhanced coronal T1WI (c), axial T2WI (d), axial T1WI (e), and contrast enhanced axial T1WI (f) show destructive lesion involving a right superior orbital rim (arrows). After intravenous Gd administration inhomogenous enhancement of a lesion is seen on the right side.

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Fig. 3: Plexiform neurofibroma; axial T2WI (a), axial contrast enhanced T1WI (b), coronal T2WI (c) and coronal contrast enhanced T1WI (d) show a irregular defined lesion which extension from intra/extraconal space to cavernous sinus. Contrast enhanced T1WI demonstrate heterogenous enhancement of the plexiform neurofibroma. On T1WI (b) is seen dysplasia of the left greater sphenoid wing and dural enhancement in the middle cranial fossa.

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Fig. 4: Capillary hemangioma; poorly marginated lesions is seen in the superomedial quadrant of the left orbit. The lesion show high signal intensity on axial (a), coronal (b) T2WI and low signal intensity on axial (c) T1WI. Contrast enhanced axial (d) T1WI shows enhancement of the lesion. T2WI demonstrate peripheral and internal flow voids regions (arrow).

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Fig. 5: Axial (a) T2WI, axial (b) T1WI, contrast enhanced axial (c) T1WI, axial (d,e) CT and reformat CT image (f) demonstrate a hemangioma in the right orbital lateral wall (arrow). MRI shows lobulated soft tissue lesions and marked enhancement. CT shows mildly expansile lytic lesion in in the right orbital lateral wall with thinning of cortex.

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Fig. 6: Axial (a) T2WI, axial (b) T1WI, axial (c) contrast enhanced T1WI, sagittal (d) T1WI, coronal (e) and axial (f) contrast enhanced T1WI demonstrate a aneurysmal bone cyst in the right orbita. T1 and T2WI show well defined multiloculated expansile lesion with fluid-fluid/hemorrhage levels.

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Fig. 7: Masticator RMS; axial (a) T2WI, axial (b) T1WI, axial (c) contrast enhanced T1WI, coronal (d) T2WI, coronal (e) and sagittal (f) contrast enhanced T1WI show a large heterogenous lesion in the left masticator space. Coronal T2WI (d) and contrast enhanced T1WI (e) show extension of lesion into the bilaterally cavernous sinus and dural enhancement. After intravenous contrast material administration considerable enhancement of the tumor and with strands of non-enhancing tissue within the tumor mass. No pathological enhancement of the adjacent brain.

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**Fig. 8:** Maxillary RMS: axial (a) T1WI, axial (b) T2WI and axial (c) contrast enhanced T1WI demonstrate a large left maxiller mass. The tumor shows low signal intensity on T1WI and heterogenous high signal intensity on T2WI and heterogenous contrast enhancement. The lesion extends into the left orbit, retroantral space, and adjacent nasal cavity.

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**Fig. 9:** Axial (a) T2WI, axial (b) T1WI, axial (c) and coronal (d) contrast enhanced T1WI demonstrate a Ewing sarcoma in the right maxillary sinus and nasal space.

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Fig. 10: Nasal osteosarcoma; an right intranasal midline lesion appearing intermediate signal intensity (arrow) on T1WI (a,b), high signal intensity (arrow) on T2WI (c,d). After intravenous Gd administration considerable enhancement of the lesion (arrow) and mucosal thickening in the right maxillary sinus is seen (e,f).

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Fig. 11: Right maxillary lymphoma; homogenous poorly defined mass with signal intensity similar to gray matter on both T1WI (a,b,c) and T2WI (d) and uniform contrast enhancement (e,f). Direct extension of enhanced mass into the masticator space and premaxillary region.

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Fig. 12: Left maxillary fibroosseous lesion; expansile sclerotic/lytic lesion involving the maxilla with mixed intermediate to high signal on T1WI (a) and variable mixed low, high signal on T2WI (b). Contrast enhanced axial (c) T1WI shows heterogenous enhancement in portions of the lesion. Axial (d,e) and sagittal (f) CT show expansion and ground glass matrix of left maxillary bone.

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**Fig. 16:** Intravesical RMS; ultrasound images (a,b) show a heterogeneous solid and cystic mass in the bladder, with Doppler US (c) demonstrating vascularization in the solid component of the mass. Sagittal (d) and axial (e) T2WI, axial (f) T1WI show the multiloculated cyst mass and solid elements. Contrast enhanced axial (g) T1WI shows enhancement of the lesion.

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**Fig. 17:** Vaginal RMS; sagittal T2WI (a), T1WI (b), contrast enhanced sagittal (c) and axial (d) T1WI show vaginal rhabdomyosarcoma. The tumor appears well defined and low signal intensity on T1WI and heterogenous high signal intensity on T2WI (arrow). After intravenous contrast material administration the lesion enhances inhomogenously (arrow).

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Fig. 18: PNET; axial (a) T1WI, axial (b) T2WI, axial (c) and coronal (d) contrast enhanced T1WI show a large heterogeneous presacral mass with intermediate to high signal intensity at T2WI and heterogeneous enhancement.

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**Fig. 19:** Desmoplastic round cell tumor; axial (a) T1WI, axial (b) and coronal (c) T2WI, sagittal (d) contrast enhanced T1WI show a large presacral mass with increased signal intensity at T2WI, decreased at T1WI and intense contrast enhancement.

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Fig. 20: Undifferentiated sarcoma; coronal (a) T2WI and coronal (b) T1WI demonstrate a well-circumscribed mass arising from the left abdominal wall. The lesion shows heterogeneously hyperintense on T2WI and hypointens on T1WI. Coronal contrast-enhanced T1 fat-suppressed MR image (c) shows enhancement of the mass.

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**Fig. 21:** Osteoblastoma; expansile vertebral lesion in corpus and posterior elements with epidural extension is seen sagittal (a), axial (b) T2WI and axial (c) T1WI and axial (d) contrast enhanced T1WI.

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**Fig. 22:** Sacral Ewing sarcoma; sagittal T2WI (a), T1WI (b) and contrast enhanced T1WI (c) show a large heterogeneous sacral mass with intermediate to high signal intensity at T2WI and heterogeneous enhancement. On MR images the mass lesion with destruction of bone and extension into the spinal canal is seen.

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**Fig. 23:** Hemangioma; a heterogenous lesion is seen in the right gluteus muscles consisting of both cystic and solid parts. On ultrasound images (a,b,c), the lesion is very vascular. Coronal (c) fat suppressed T2WI, coronal (d) T1WI show a heterogeneous mass.

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Fig. 24: Forearm RMS; coronal (a) T1WI, coronal (b) and axial (c) contrast enhanced T1WI show a well defined homogenous lesion of low signal intensity on T1WI and inhomogenous contrast enhancement.

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Fig. 25: Metastatic RMS; sagittal T2WI (a), T1WI (b), contrast enhanced sagittal (c) and coronal (d) T1WI show a large heterogeneous forearm mass with intermediate to high signal intensity at T2WI, heterogeneous enhancement. There is no extension of lesion into the elbow.

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Fig. 26: Ewing sarcoma. Sagittal T2WI (a), T1WI (b), contrast enhanced T1WI (c) of a case of Ewing's sarcoma. The tumor is hypointense on T1WI and heterogeneously hyperintense on T2WI with diffuse marrow infiltration and extraosseous spread.

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Fig. 27: Plain radiograph of the right leg shows permeative lytic destruction, periosteal reaction and Codman triangle. Coronal fat suppressed T2WI (b), T1WI (c) and contrast enhanced coronal (d) and sagittal (e) T1WI of a case of osteosarcoma. The tumor is hypointense on T1WI and hyperintense on T2WI. The tumor has an extra compartmental extension into the muscle plane.

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Fig. 28: Synovial sarcoma. Coronal T2WI (a), T1WI (b) and contrast enhanced T1WI (c) show a large heterogeneous thigh mass. The tumor is hypointense on T1W and heterogenous hyperintense on T2W sequences. The tumor has an extra compartmental extension into the muscle plane.

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Fig. 29: Left fibula desmoid tumor; expansile lytic lesion involving the fibula with mixed low to intermediate signal on T1WI (a) and variable mixed low, high signal on T2WI (b). Contrast enhanced T1WI (c) shows heterogenous enhancement in portions of the lesion.

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Fig. 30: Neuroblastoma; sagittal T1WI (a), T2WI (b), contrast enhanced T1WI (c) show a large heterogeneous presacral mass with intermediate to high signal intensity at T2WI, heterogeneous enhancement, and extension of the spinal canal.

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Fig. 14: Hemangiopericytoma; a well marginated heterogenous lesion in the left masticator space extends into the parapharyngeal space. Axial (a) and coronal (b) T1WI, axial (c) T2WI, contrast enhanced axial (d), coronal (e) and sagittal (f) T1WI shows heterogenous signal intensity on both T1WI and T2WI (a) and intense contrast enhancement. It demonstrates areas of high signal intensity on T1WI due to hemorrhage.

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**Fig. 15:** Axial (a) and sagittal (b) T1WI, axial (c) T2WI, axial (d) contrast enhanced T1WI show a chloroma (arrows). The lesion is seen originating paratymppanic region of the left temporal bone with intermediate signal intensity on T1WI and low signal intensity on T2WI. After intravenous contrast material administration the lesion enhances considerably.

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Fig. 13: Neuroblastoma; coronal (a) fat suppressed T2WI, sagittal (b) and axial (c) fat suppressed contrast enhanced T1WI, axial (d) fat suppressed T2WI show a heterogeneous paravertebral mass with intermediate to high signal intensity at T2WI and heterogeneous enhancement. On axial MR images, the extension of lesion into the spinal canal is seen.

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Conclusion

Although imaging findings are not specific to RMS, many other neoplastic masses can be ruled out by a careful analysis of location, internal structure, presence of vascular structures, calcifications, enhancement pattern and diffusivity of the masses using multimodal imaging.
References


