

# Rhabdomyosarcoma – site matters

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## Abstract

For the clinical radiologist imaging a soft-tissue mass in a child, site plays an important role in narrowing the differential diagnosis. Rhabdomyosarcomas can arise from any tissue, even those devoid of skeletal muscle. Knowledge of the common sites involved allows inclusion in the differential diagnosis where relevant and also has a limited potential to prognosticate. With this article we hope to give some clarity as to when this diagnosis is likely. Rhabdomyosarcomas are relatively rare, but radiologists should have a high index of suspicion at particular sites where this may be the most common childhood malignancy. Common sites are the head and neck, genito-urinary system and extremities.

## Introduction

Radiologists are often unsure whether a rhabdomyosarcoma should be included in a differential diagnosis of a solid mass found on imaging.

With this article we hope to give some clarity as to when this diagnosis is likely. Rhabdomyosarcomas are relatively rare, but radiologists should have a high index of suspicion at particular sites where this may be the most common childhood malignancy. Rhabdomyosarcoma is the most common soft-tissue sarcoma (50% of soft-tissue sarcomas)<sup>1</sup> of childhood, representing 5% of all childhood cancers.<sup>2</sup>

Rhabdomyosarcoma is the third most common extracranial solid tumour in children, after neuroblastoma and Wilm's tumour.<sup>3</sup> These tumours are thought to arise from primitive mesenchymal cells committed to skeletal muscle differentiation and can occur in a variety of organs and tissues, including those that lack striated muscle,<sup>2</sup> as well as from smooth-muscle cells, i.e. urinary bladder.<sup>4</sup> Tables I and II summarise the incidence of these tumours at specific sites and the prognosis related to the site involved.

**Table I. Sites of primary tumours**

Site	% of patients
Head and neck	35
• Orbit	10
• Parameningeal	10
• Other	15
Genito-urinary	26
Extremities	19
Other	20

**Table II. Favourable and unfavourable sites**

Favourable	Unfavourable
Non-parameningeal head and neck sites	Parameningeal head and neck sites
• Oropharynx	• Middle ear
• Scalp	• Nasal cavity
• Parotid	• Paranasal sinuses
• Neck	• Nasopharynx
• Larynx	• Infratemporal fossa
• Cheek	• Pterygo-palatine fossa
• Hypopharynx	• Parapharyngeal region
Genito-urinary system (other than bladder and prostate)	Orbits involving base of skull
Orbits and eyelids	Trunk
	Bladder and prostate
	'Other' sites

## Head and neck

Primary head and neck tumours are most commonly diagnosed in children younger than 8 years of age.<sup>4</sup>

### Non-parameningeal

**Orbital.** Overall orbital tumours have the best prognosis.<sup>1</sup> Orbital tumours are generally non-invasive and confined to the bony orbit.<sup>5</sup> Orbital tumours with intracranial invasion or bone destruction are for practical purposes treated as parameningeal disease<sup>5</sup> (Figs 1a and b).

### Parameningeal regions and paranasal sinuses

Rhabdomyosarcoma is the most common malignancy to arise within the nasal cavity or paranasal sinuses in children.<sup>3</sup> Tumours tend to be large and invasive at these sites.<sup>5</sup> Sites include the nasopharynx, paranasal sinuses, middle ear, mastoid and pterygoid and infratemporal sites. Tumours in parameningeal sites usually behave more aggressively than do tumours in other parts of the head and neck, primarily because of the proximity to the meninges and brain.<sup>6</sup> Rhabdomyosarcomas of the head and neck grow insidiously and often invade intracranial space through the numerous foramina leading to the brain (Figs 2a-d).

**Temporal bones** (Fig. 3). Paediatric rhabdomyosarcomas that arise in the temporal bone (middle ear or mastoid), although rare, are generally considered to be aggressive neoplasms by virtue of their proximity to vital structures, their tendency to spread intracranially, and their potential for meningeal involvement.<sup>6</sup> Patients with rhabdomyosarcomas of the middle ear and mastoid have a particularly poor prognosis.<sup>6</sup>

## Genito-urinary

Bladder and prostate are the most common genito-urinary sites.<sup>4</sup> Other common sites involved are the vagina, uterus and paratesticular region.<sup>7</sup>

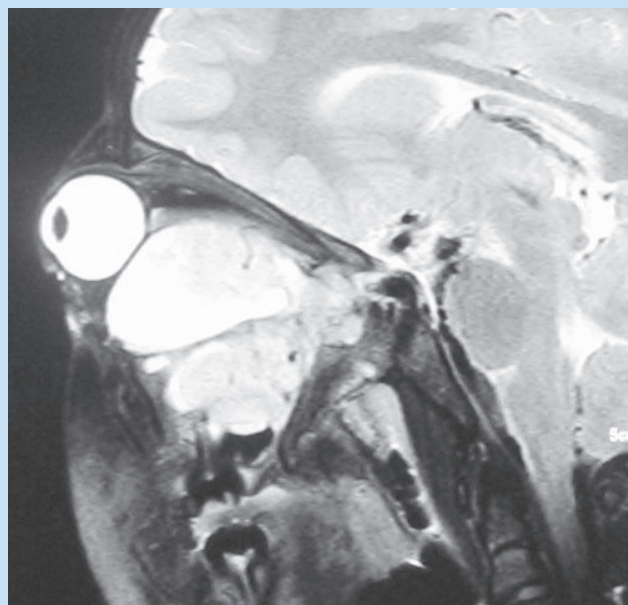


Fig. 1a. Sagittal oblique STIR MRI demonstrates a mass which has displaced globe and optic nerve superiorly. It appears to have expanded the orbit (indicating slow growth) and has also destroyed the orbital floor, extending into the maxillary sinus and unerupted teeth in the alveolar bone.

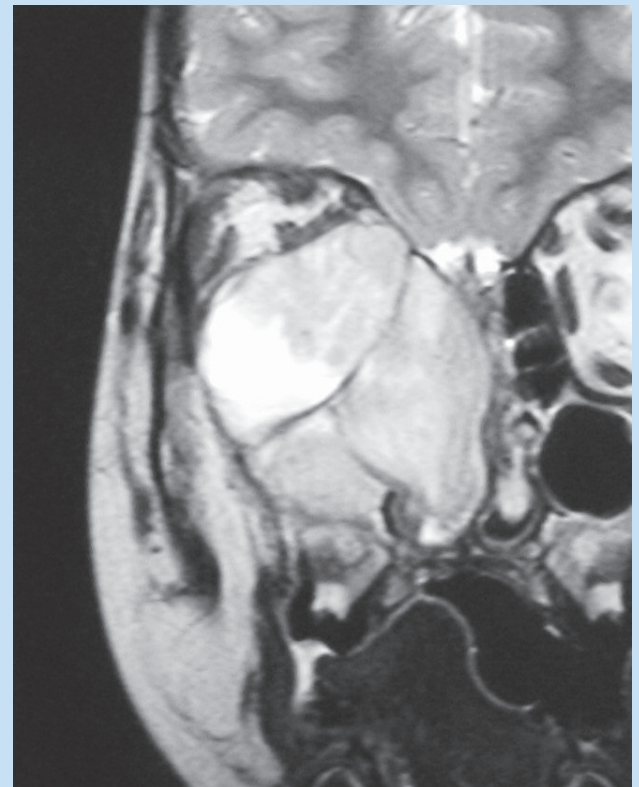


Fig. 1b. Coronal T2-weighted MRI demonstrating a mass near the orbital apex, extending through the lamina papyracea and orbital floor into the nasal cavity, ethmoid and maxillary sinuses.

Large pelvic masses are common at time of diagnosis.<sup>4</sup>

Most rhabdomyosarcomas of the bladder arise near the trigone or urethral orifice, are of embryonal histology and carry a good prognosis.<sup>8</sup> Tumours of the bladder and prostate generally have a worse prognosis than other sites such as the vagina.<sup>5</sup> Tumours originating in the prostate carry a significantly worse prognosis than tumours that involve the bladder only. When both are involved, the site of origin is often difficult to determine<sup>2</sup> (Fig. 4a and b).

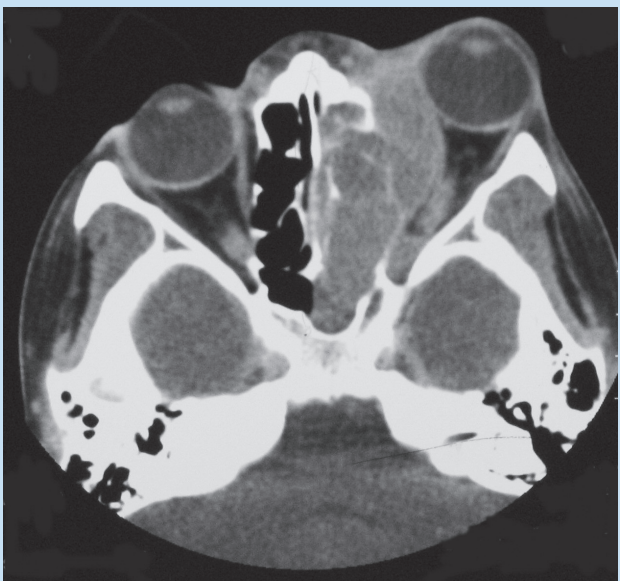
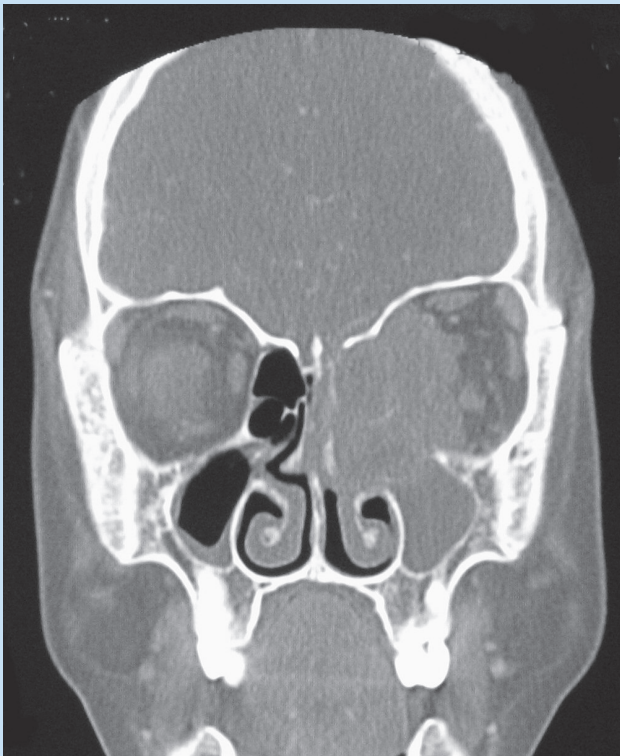
The term paratesticular rhabdomyosarcoma has been applied to primary tumours arising in the spermatic cord, testis, epididymis and penis.<sup>5</sup> Rhabdomyosarcoma is the most common paratesticular malignancy in children.<sup>2</sup> Most paratesticular rhabdomyosarcomas are composed of the spindle cell variant of embryonal type and this carries an especially good prognosis.<sup>2</sup> Prognosis of paratesticular rhabdomyosarcoma is generally good, but worse in children older than 10 years of age or if there is retroperitoneal lymph node involvement.<sup>5</sup> Paratesticular rhabdomyosarcomas usually occur in adolescent males.<sup>1</sup>

Rhabdomyosarcoma is the most common malignancy of the lower genital tract in young women but has an excellent prognosis. Cervical rhabdomyosarcoma is usually seen in the second decade of life, unlike vaginal lesions which usually present before the age of 4 years. Uterine rhabdomyosarcoma carries a worse prognosis than vaginal lesions.<sup>8</sup>

Prognosis is unfavourable for patients who have locally advanced pelvic tumours for which the exact site of origin cannot be determined.<sup>2</sup>



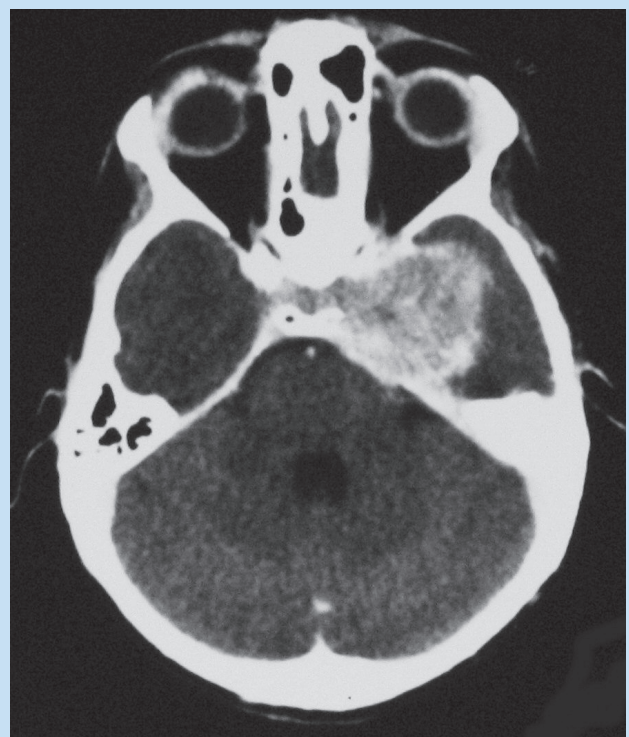
# PICTORIAL REVIEW



*Figs 2a and b. Coronal and axial CT scans of paranasal sinuses demonstrating a soft-tissue mass which has destroyed the wall of the ethmoid sinuses, the middle and superior turbinates, and the medial wall and inferior aspect of the orbit. The mass is seen to fill the superior portion of the nasal cavity, the left maxillary sinus and part of the left orbit. The inferior and medial recti are displaced laterally.*



*Fig. 2c. CT scan demonstrating a soft-tissue mass involving the anterior aspect of the nose and maxillary bones bilaterally with some extension into the right anterior choana.*



*Fig. 2d. An 8-year-old boy with rhabdomyosarcoma of the maxillary sinus, middle cranial fossa and base of the skull. Contrast-enhanced CT of the brain demonstrates a non-homogeneous enhancing mass in the left middle cranial fossa, involving the cavernous sinus and pituitary fossa.*

## Extremities

Extremity tumours are seen most commonly in adolescents.<sup>4</sup> Approximately half of extremity tumours are of alveolar subtype, accounting in part for the poor outcome in these patients. Lymph node involvement is common and imaging should include the entire nodal basin<sup>2</sup> (Figs 5a-c).

## PICTORIAL REVIEW

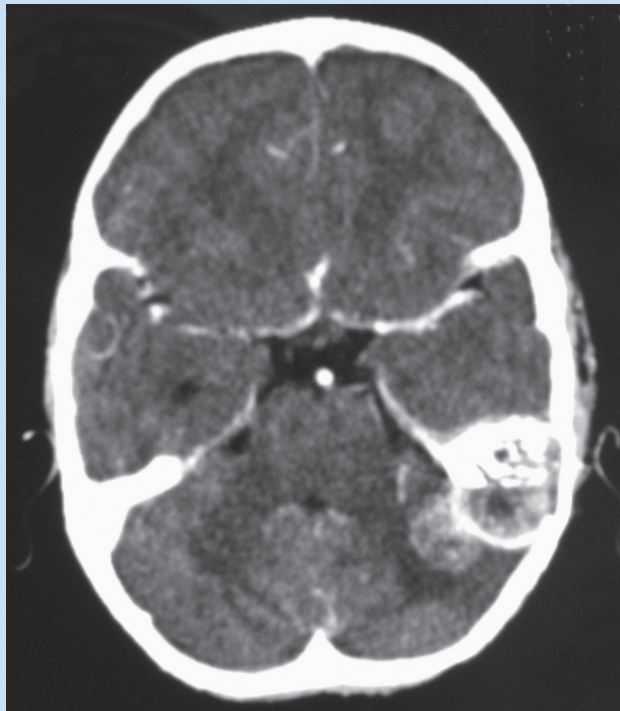


Fig. 3. Axial contrast-enhanced CT scan demonstrating a mass with destruction of the left petrous bone, extending to involve posterior and middle cranial fossa in the form of a soft-tissue mass. The mass also extends into the external ear canal (not shown here).

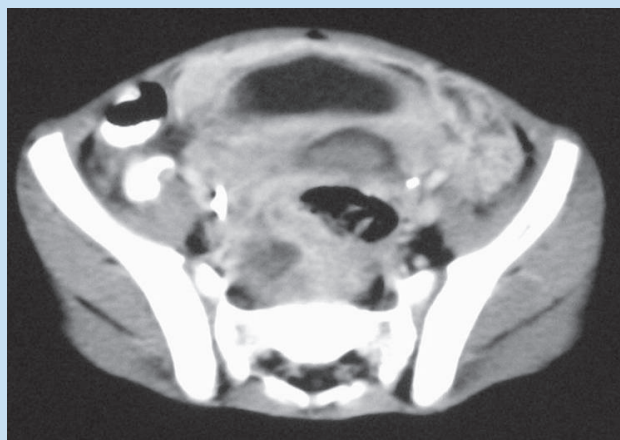


Fig. 4a. A 2-year-old boy with rhabdomyosarcoma in the bladder. T1-weighted axial MRI shows the mass to be iso-intense to muscle on T1 (mixed signal on T2). The rectum is seen separately from the mass.

### Other sites

Intrathoracic, retroperitoneal, perineal and peri-anal regions can harbour a primary rhabdomyosarcoma that can remain undiagnosed for a long period of time. These tumours can become very large and metastasise before they are diagnosed. Uncommonly rhabdomyosarcomas arise from the liver, brain, heart, breast or ovary<sup>4</sup> (Figs 6a and b).

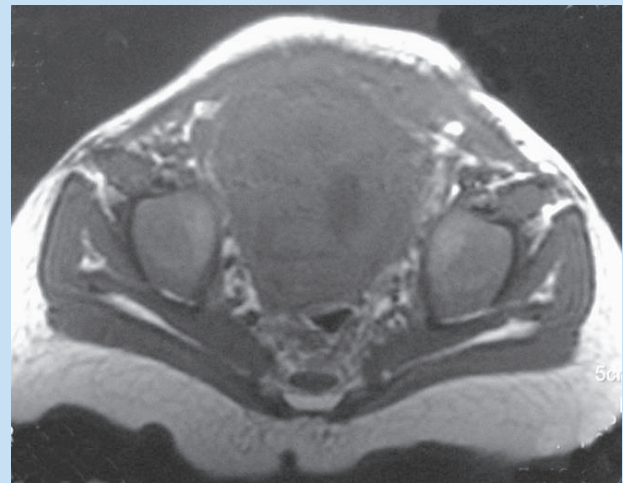


Fig. 4b. A 5-year-old girl with rhabdomyosarcoma in the pelvis. Contrast-enhanced CT scan of pelvis shows a non-homogeneous enhancing mass presacraly. The mass both encases and displaces the rectum. The tissue planes are also obscured between bladder and rectum.

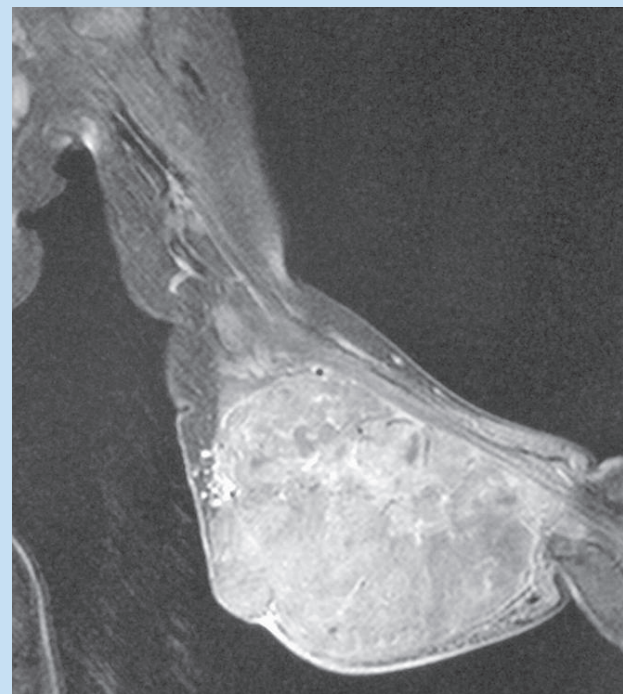


Fig. 5a. A 4-month-old boy with a rhabdomyosarcoma of the right forearm. Short Tau Inversion Recovery MRI of upper limb demonstrates an inhomogeneous, fairly well encapsulated mass of relatively high signal intensity in the forearm. The mass extends to the level of the olecranon but does not cross the joint space. The flexor tendons and vessels are seen separately.

### Site-related histology

**Embryonal.** This type often arises in mucosal-lined structures of the nasopharynx, auditory canal and genito-urinary and gastro-intestinal tracts.<sup>2</sup> Embryonal tumours usually occur before age 8.<sup>1</sup>





Fig. 5b. T2-weighted coronal MRI shows well-defined inhomogeneous mass in vastus lateralis.

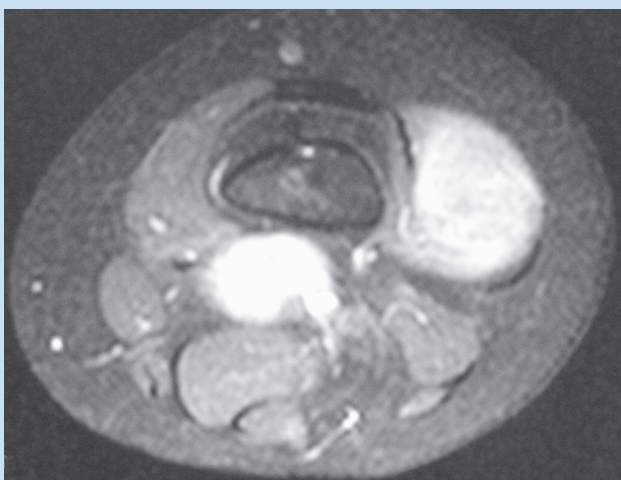


Fig. 5c. T1-weighted fat-saturated gadolinium-enhanced axial MRI shows two masses in distal thigh. Both demonstrate strong enhancement with one mass in the distal vastus lateralis and the second in the popliteal fossa, just anterior to the popliteal artery.

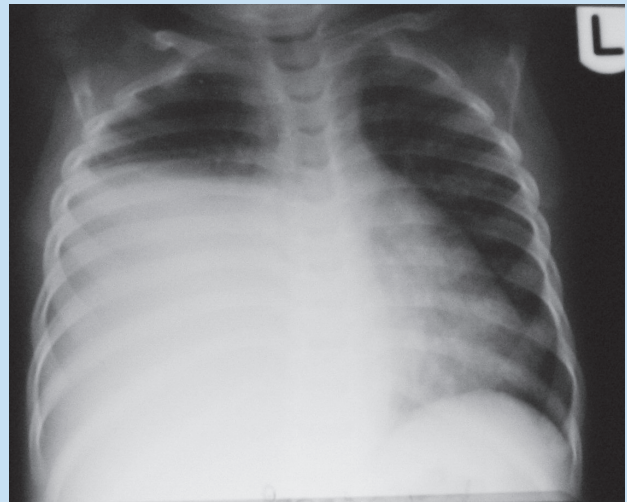


Fig. 6a. A 2-year-old boy with rhabdomyosarcoma of the right chest/abdomen (possibly arising from right diaphragm). Chest X-ray shows right-sided chest mass resulting in 'apparent' elevation of right hemidiaphragm, with displacement of heart to left.

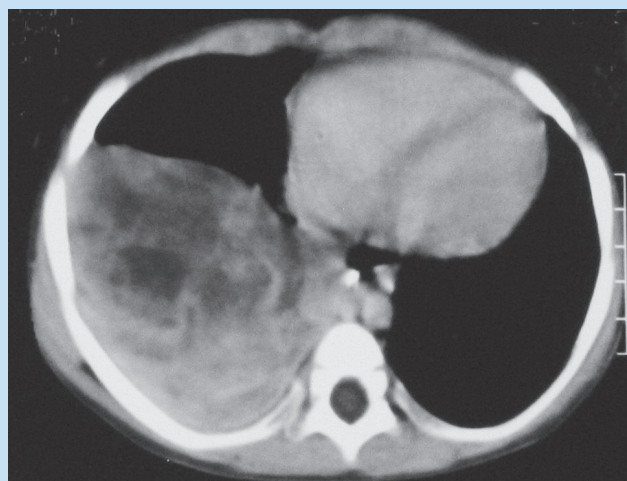


Fig. 6b. Contrast-enhanced CT of chest/abdomen shows large heterogeneous mass in right hemithorax. There are low density areas in mass compatible with tumor breakdown, and pleural thickening is also visible.

**Botryoid.** A variant of embryonal type occurs in hollow cavities such as the vagina, biliary tract and nasopharynx.<sup>1</sup>

**Alveolar.** Most commonly these occur on the trunk and extremities (and have a worse prognosis).<sup>1</sup>

## Metastases

Rhabdomyosarcoma commonly metastasises to lungs (36%), bone marrow (22%) or cortical bone (7%)<sup>2</sup> (Fig. 7).

## Conclusion

For the clinical radiologist imaging of a soft-tissue mass in a child plays an important role in narrowing the differential diagnosis. Rhabdomyosarcomas can arise from any tissue, even those devoid of skeletal muscle. Knowledge of the common sites involved allows inclu-

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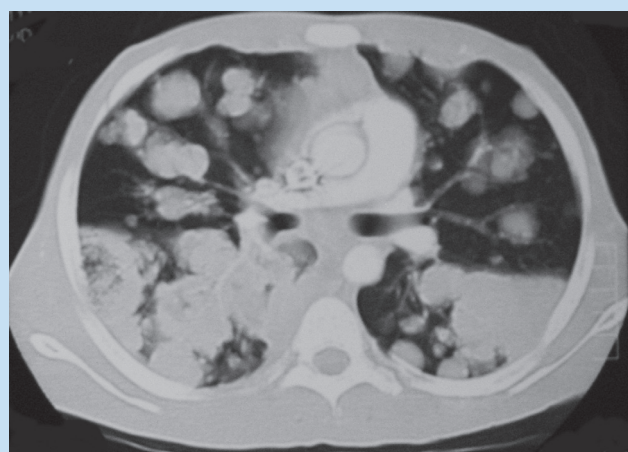
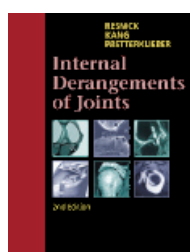


Fig. 7. Lung metastases on CT due to a pelvic rhabdomyosarcoma.

sion in the differential diagnosis when relevant, and also has a limited potential in determining prognosis.

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