Curr Radiol Rep (2015) 3:28
DOI 10.1007/s40134-015-0111-2

MRI Evaluation of Pediatric Malignant Abdominal Tumors

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Published online: 10 June 2015

Abstract Mostly, pediatric abdominal tumors are benign. Eventually, malignant cancer may occur as abdominal tumor being the Wilms tumor, neuroblastoma, hepatoblastoma, and Burkitt's lymphoma found to be common on this age group. Abdominal tumors can have a wide etiology range that can be classified according to its location, age group, and associated signs and symptoms. Mostly, benign findings are hydronephrosis, infectious hepatosplenomegaly, and ascariasis, while neuroblastoma, Wilms tumor, and germ cell tumors can exhibit specific MRI findings helping on its determination. This article discuss some clinical and MRI presentation of main abdominal pediatric malignances, as well as imaging findings that can correlate to clinical aspects to help during staging and patients' treatment follow up.

Keywords Pediatric · Oncology · Abdominal tumors · Magnetic resonance imaging

Introduction

Mostly, pediatric abdominal tumors are benign. Eventually, malignant cancer may occur as abdominal tumor, being the Wilms tumor, neuroblastoma, hepatoblastoma, and Burkitt's lymphoma, found to be common on this age group. Abdominal tumors can have a wide etiology range that can be classified according to its location, age group, and associated signs and symptoms. Mostly, benign findings are hydronephrosis, infectious hepatosplenomegaly, and ascariasis, while neuroblastoma, Wilms tumor, and germ cell tumors (GCTs) can exhibit specific MRI findings helping on its determination [1••].

Objectives

The objective of this study is to summary MRI characteristics of the main pediatric abdominal malignancies, its differentials, and correlate with clinical aspects, helping with the imaging workflow for these patients.

Neuroblastoma

Neuroblastic tumors include all those that have been originated from the sympathetic nervous system from the primordial neural crest cells. They are the ganglioneuroma, ganglioneuroblastoma, and the neuroblastoma. Neuroblastoma is the more undifferentiated and aggressive containing neuroblasts, while the ganglioneuromas account for the more differentiated and less aggressive tumor containing

This article is part of the Topical Collection on *MRI for Pediatric Oncology*.

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ganglion cells [2]. They can occur at any region containing sympathetic nervous system, from the neck until the pelvis, at the midline, including the posterior mediastinum (20 %), retro peritoneum (30–35 %), and more frequently the adrenal (35 %). Its clinical presentation varies from the complete spontaneous regression until the disseminated and aggressive disease, in children, especially from 1 to 2 years old. In more than 90 % of cases, these tumors are found during pregnancy ultrasounds, typically at the right adrenal, usually at 32 pregnancy weeks [3•]. It accounts for the most common malignant congenital neoplasia, the second more common congenital tumor (30 %, behind of extracranial teratomas), the third more common malignancy during childhood (8–10 %, behind leukemia and central nervous system tumors), and accounting for 15 % of cancer deaths in pediatrics [4, 5••].

Neuroblastoma frequently presents at MRI as a variable sized lobulated mass of heterogeneous signal, due to necrosis and bleeding, low signal focal areas, representing calcifications are found on up to 90 % of cases, and a variable paramagnetic contrast enhancement are usually observed in every MRI sequence (Fig. 1).

Vascular encasement, focal psoas, paraspinal, and neural foramina invasion also can occur (Figs. 2, 3). Metastases are present in up to 70 % of cases at diagnosis. Hepatic metastases are common and characterized by diffuse parenchymal infiltration, or multiple focal nodules that could also be found on the lungs, and less frequently, in the meninges or cranial base. During staging, whole-body STIR MRI studies can be performed to help determine whether there is any other suspect area to be further evaluated for the presence of metastases (Fig. 4).

The international neuroblastoma risk group designated image defined risk factors (IDRFs) to suggest pre-operatory staging, as neuroblastoma conventional staging is based on surgical findings [4, 5••, 6, 7•, 8].

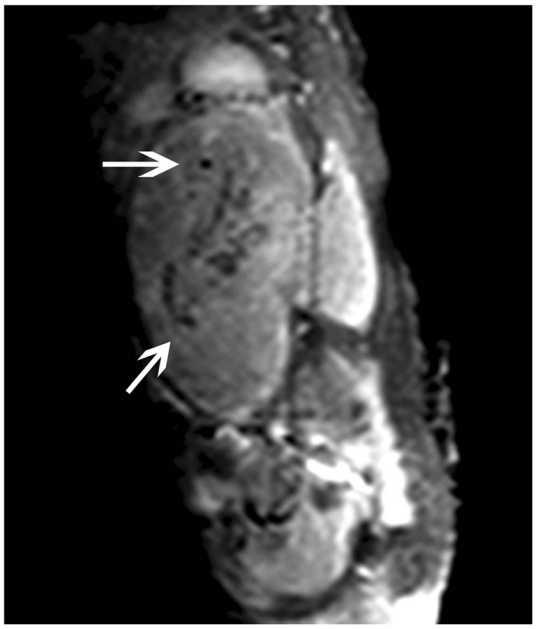

Fig. 1 Sagittal MRI demonstrating heterogeneous contrast media enhancement neuroblastoma mass with calcifications (*arrows*)

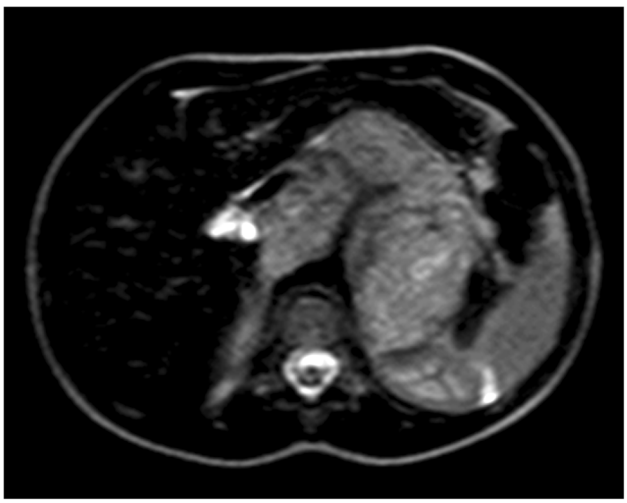

Fig. 2 Axial MRI demonstrating vascular encasement by the neuroblastoma

Wilms Tumor (Nephroblastoma)

The Wilms tumor is the most common childhood renal neoplasia (87 %); its incidence is frequently under 15 years of age, with 80 % of cases occurring under the fifth birth anniversary [9]. Various genetic events contribute to Wilms tumor, since 1990′s with the WT1 gene locus at the 11p15 chromosome, the WT2 locus description and the two family loci FWT1 (17q) and FWT2 (19q), and other loci have been investigated for the tumorigenesis. Syndromes as the WAGR syndrome (Wilms tumor, aniridia, genitourinary anomalies, and retardation) or the Denys–Drash syndrome (Wilms tumor, congenital nephropathy, and intersex disorders) are both related to the 11p chromosome disorder. The Beckwith–Wiedemann syndrome and the hemihyperplasia (formerly hemihypertrophy) are also associated with the Wilms tumor. It presents multiple microscopic characteristics composed by a variable proportion of the three normal renal development cells [10–16].

Its common clinical presentation is a non-movable unpainful palpable abdominal tumor, frequently found during bath or dressing by parents. Abdominal pain, macroscopic hematuria, or fever is also frequently found; hypertension is also present at 25 % of cases, and a small part of them can course with Intra-tumoral hemorrhage

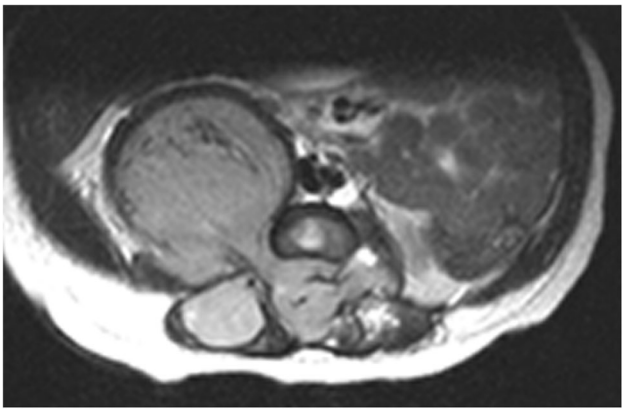

Fig. 3 Axial MRI demonstrating neural foramina invasion by the neuroblastoma

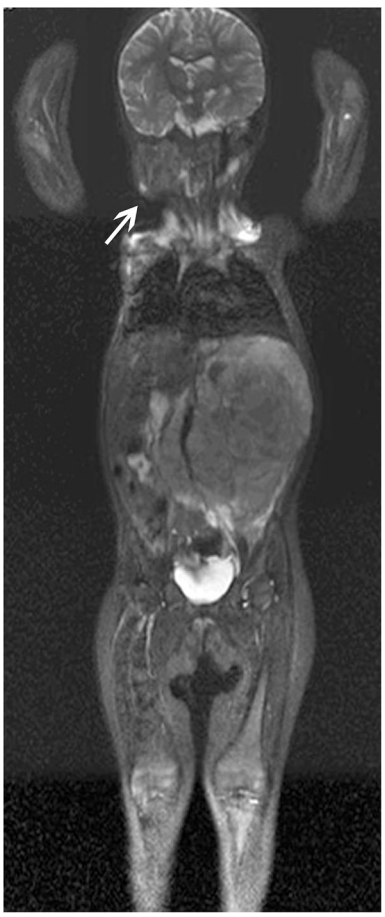

Fig. 4 Coronal STIR MRI of a neuroblastoma patient determining metastasis suspect area on right neck (*arrow*) to be further evaluated

followed by anemia. Some benign conditions can also have similar clinical presentation as the hydronephrosis, polycystic kidney disease, multilocular cystic nephroma, or renal abscess. The neuroblastoma is always the malignancy that should be included in their differentials [11].

MRI exams of the Willms' tumor present heterogeneous low signal T1 mass with high T2, which might contain hemoglobin degradation products' signs. It has slow enhancement due tortuous vessels within it, which might be helpful to differentiate from nodular nephroblastomatosis. Cystic areas of necrosis can also be found. The intravascular tumoral extension to the renal vein and inferior vena cava has to be scanned. MRI and CT scans have similar accuracy for the perirenal and linfonodal extension evaluation. The capsular rupture is considered as tumor extension to the perirenal fat. The most common metastasis sites are the lungs (present as isolated site in up to 80 % of cases), regional lymph nodes, and the liver. The NWTSG—National Wilm Tumor Study Group—has developed staging criteria according to tumor extension before chemotherapy, and these criteria have been later modified by the children's oncology group (COG) [17–21].

Germ Cell Tumors

Germ cell tumors are benign or malignant tumors derived from the primordial germ cells that might occur within or outside gonadal sites, these may result from embryo development errors. The GCTs have two different incidence peaks, one representing outside gonadal peak of tumors before 2-year old and another representing gonadal tumors peak between 8 and 12 years of age on girls and 11–14-year old on boys. They represent 3.3 % of malignancy on children under 15-year old. Due to its extremely variable clinical, histological, and radiological aspects, it is hard to generalize its behavior that should be individualized considering age group, primary site, and histology findings. The most used nomenclature classification includes the Precursor lesion, tumors of one histologic type (seminoma, spermatocytic seminoma, embryonal carcinoma, yolk sac tumor, choriocarcinoma, placental site trophoblastic tumor, GCTs, trophoblastic tumor, teratoma), tumors of more than one histologic type (mixed GCT), and testicular scar, consistent with regressed tumor [22, 23].

The teratoma has, as the main frequent site, the sacrococcygeal region, and other median or para median regions as the retro peritoneum, mediastinum, neck, and pineal gland. The teratoma is the most common histological GCT type under 5-year-old children, more frequent in girls, and the age of diagnosis represents its malignization risk that is between 7 and 10 % under 2 months of age and up to 47–67 % after this period. The teratoma presents on MRI as a solid or solid-cystic mass that can occasionally be completely cystic. They are usually well vascularized enhancing after contrast media injection. The presence of fat, which is characterized by

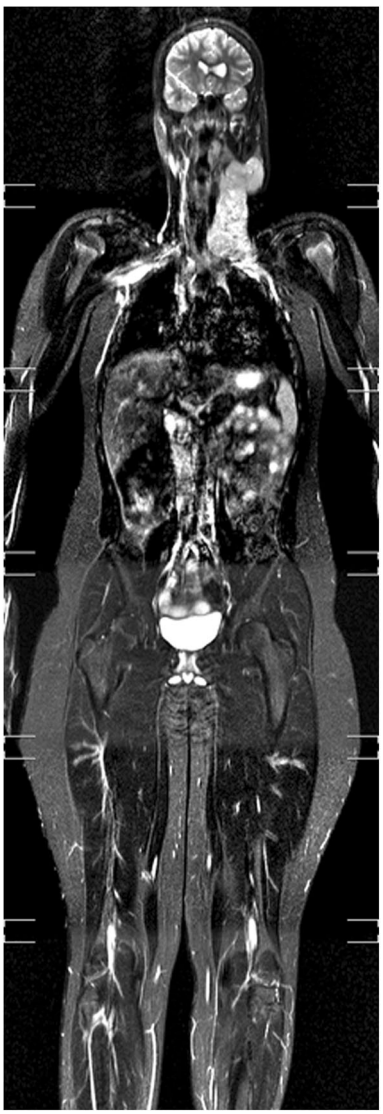

Fig. 5 Coronal STIR MRI of a lymphoma patient demonstration para aortic high signal due to lymphoma, primary from the left cervical node

the high T1 signal which is low on fat sat sequences, and calcification signal, low on all sequences, within the tumor on MRI, is pathognomonic of teratoma.

The dysgerminoma corresponds histologically and morphologically to testicular seminoma. Other than the seminomas, which always occur post-pubertal, the dysgerminomas occur before puberty at the ovaries. It is bilateral on 10–15 % of cases and the retro peritoneum is the most frequent site of dissemination. They have different sizes from little centimeters to large masses, capsulated; can have cystic areas or calcification, and present fibrovascular septation that enhances alter contrast.

Endodermal sinus tumor, also known as yolk sac tumor, was first described in 1939. It is the most common

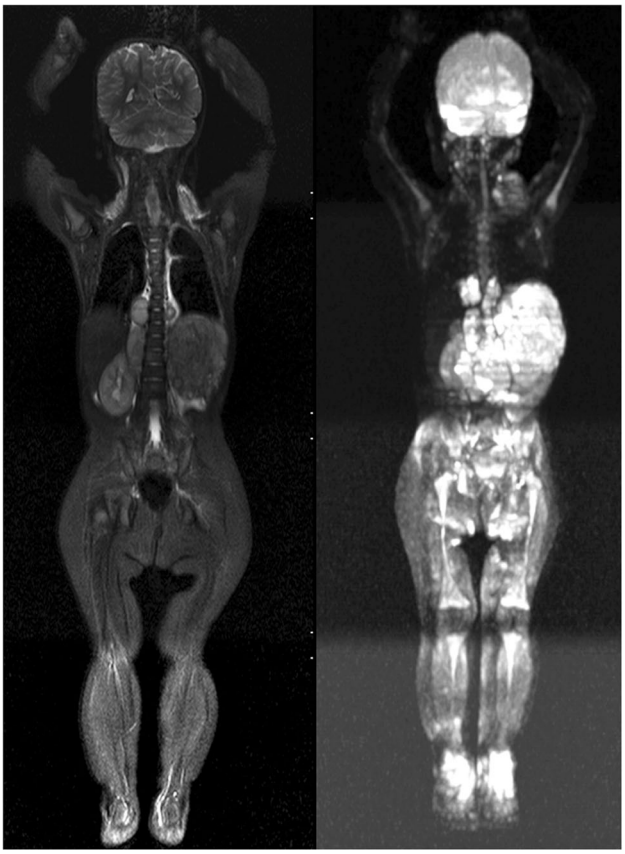

Fig. 6 Side by side coronal STIR and DWIBS images of a cervical lymphoma with abdominal mass, note the visible diffuse bone marrow infiltration on DWIBS (*right*)

testicular tumor in children under 3-year old and have similar to placental or yolk sac histology. The choriocarcinoma is another of the rare GCT that also can be found [24, 25].

Lymphoma

Lymphoma is the third most common pediatric cancer after leukemia and central nervous system tumors. Two-thirds of children's lymphomas are non-Hodgkin (NHL). Lymphomas are originated from either T or B lymphocytes with variable biological behavior that is composed by 70 different types.

Imaging studies come to determine the disease extension and tumoral size at diagnosis and its regression or not during treatment. The use of whole-body MRI (WB-MRI) has demonstrated good sensibility to lymphoma detection in both nodal and extra nodal sites, and also determine bone marrow infiltration, with good availability when compared to the gold standard FDG-PET/CT. Various MRI protocols have been used on WB-MRI, which usually

include a coronal STIR that shows high abnormal signal on lymphoma lesions (Fig. 5), and DWIBS that demonstrates water restriction on tumoral sites due to its high cellularity (Fig. 6) [26, 27].

Conclusion

This article summary some clinical and MRI presentation of main abdominal pediatric malignances, as well as imaging findings that can correlate to clinical aspects to help during patients' staging and treatment follow up.

Compliance with Ethics Guidelines

Conflict of Interest Jose Luiz de Oliveira Schiavon, Vivian Siqueira Tostes, and Rodrigo Regacini each declare no potential conflicts of interest. Henrique Manoel Lederman is a Section Editor for Current Radiology Reports.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

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