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Case Report

Giant Retroperitoneal Neuroblastoma in a Teenager Demonstrated by Split-Bolus MDCT

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Abstract

Neuroblastoma represents the most common extra cranial solid tumor among the childhood malignancies. It is rare in adolescents and very rare in adults. Neuroblastoma occurs most commonly in the abdomen, rather than in thorax. For the first time in literature, we have demonstrated the occurrence of a giant retroperitoneal Neuroblastoma in a 14-year-old young teen, by split-bolus Multidetector-Row Computer Tomography (MDCT). This innovative technique, based on the splitting in two boluses of the intravenous contrast medium, combines two phases in a single scan, with significant reduction of the radiation dose.

Keywords: Neuroblastoma; Retroperitoneal tumor; Multidetector computer tomography (MDCT); Split-bolus technique

Introduction

Neuroblastoma represents the most common extra cranial solid tumor among the childhood malignancies. It is the third most common tumor after leukemia and brain malignancies, accounting for about 15% of childhood cancer-related deaths and in the 50% of cases is already metastatic at time of diagnosis [1]. Neuroblastoma is rare in adolescents and very rare in adults and this explains the small series and limited number of reports on the topic in literature. The great majority (88.5%) of cases is seen in infancy, while the age at diagnosis exceeds the 5 years in the 10% of patients and the 14 years in only the 1.5% [2]. The tumor has a neuroectodermic origin and it occurs most commonly in the abdomen, rather than in thorax. The intra-abdominal specific sites of its origin are: adrenal glands (35%), retro peritoneum (30-35%), Zuckerkandl's organ, coeliac axes, paravertebral sympathetic chains [3]. The clinical presentation of abdominal neuroblastoma is usually characterized by discomfort and distension, due to local mass effect. It can be the cause of syndromic conditions, such as Hutchinson syndrome (pain and limping due to skeletal metastases), Pepper syndrome (hepatomegaly due to liver metastases), blueberry muffin syndrome (purpura due to extramedullary hematopoiesis in course of metastatic disease) and *dancing eyes - dancing feet* syndrome (opsomyoclonus) [4]. Abdominal Computed Tomography (CT) typically shows a heterogeneous mass, with calcifications and necrotic areas of low attenuation, and it allows an accurate TNM staging [1]. We have implemented an innovative split-bolus protocol for the TNM staging by 64-detector row CT, in order to ensure diagnostic accuracy and to obtain reduction of the radiation dose in oncologic patients [5,6]. Here, we report the value of this novel split-bolus MDCT technique in the assessment of retroperitoneal neuroblastoma in a 14-year-old young teen.

Case Presentation

A 14-year-old young teen was admitted to the hospital for an abdominal palpable mass associated with discomfort, worsened in the last two weeks. The laboratory analyses revealed high serum levels of Neuron Specific Enolase (NSE, 312 µg/L), Lactate Dehydrogenase (LDH, 930 U/L) and ferritin (754 µg/L), together with a high urinary concentration of Vanillylmandelic Acid (VMA, 214 mg/24h) and Homovanillic Acid (HVA, 415 mg/24h). After a preliminary Ultrasound (US), which showed a voluminous heterogeneous mass containing calcifications at the left upper abdomen, a split-bolus MDCT of the chest and abdomen by 64-detector row scanner was performed.

Method

Split-bolus MDCT is an innovative method that in a single pass combines arterial and venous phases, allowing the detection of hypo-

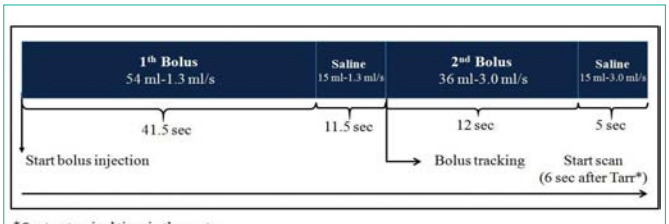

Figure 1: Procedural scheme of the split-bolus whole body MDCT in our 45-Kg patient, resulting in a simultaneous contrast enhancement of the arterial and venous system. The volume of the contrast material is calculated as 2 mL/Kg, with a maximum of 90 mL. First bolus, at the start of bolus injection (or time zero): 54 mL of contrast material at 1.3 mL/sec, followed by 15 ml of saline solution at same flow rate, is injected to obtain an adequate parenchymal and venous enhancement. Second bolus: 36 mL of contrast material at 1.3 mL/sec followed by 15 ml of saline solution at the same flow rate is injected to obtain the late arterial phase.

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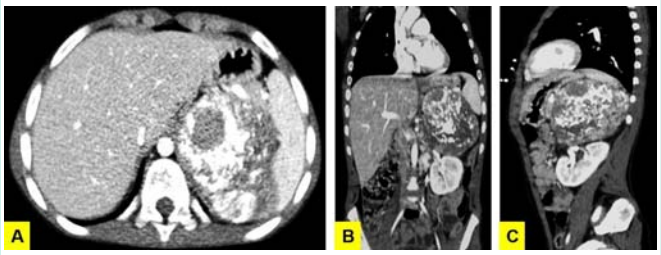

Figure 2: Split-bolus whole body MDCT: the axial (A), coronal (B) and sagittal (C) multiplanar reconstructions show a retroperitoneal mass in the left upper side. The voluminous neoplasia is heterogeneous with diffuse calcifications and defined contours. Note the compression and displacement of the neighboring structures and vessels by the mass.

or hyper-vascular and mixed lesions, together with related lymph node involvement and distant metastases. For a 45-Kg patient, the split-bolus MDCT protocol is based on a single acquisition of the chest-abdomen-pelvis after intravenous injection of 90 mL of contrast medium (Iopamiro 350 mg/mL; Bracco, Milano, Italy), splitted by an automatic power injector (Medrad Stellant, Indianola, PA, USA) into two boluses, as reported schematically in Figure 1. The first bolus consisted in the injection of 54 mL of contrast material at 1.3 mL/sec, followed by 15 ml of saline solution at same flow rate, in order to obtain an adequate parenchymal and venous system enhancement. The second bolus, which consisted of 36 mL of contrast material at 1.3 mL/sec followed by 15 ml of saline solution at the same flow rate, was injected to obtain late arterial phase. A manual bolus tracking was set up, raising the threshold value at 500 HU, by placing a circular Region of Interest (ROI) in the descending aorta. The scan was cranio-caudally performed, starting from the pulmonary apex toward the pubic symphysis, after a 6-second delay from the arrival of the contrast material in aorta. The inherent delay in the bolus tracking was necessary to move the scan table, give breath-hold instructions to the patient, and tune the gantry parameters. For the split-bolus MDCT protocol, the following acquisition parameters were used: gantry rotation speed 0.75 seconds; slice thickness 2.5 mm; reconstruction index 1.25; pitch 0,935:1; tube voltage 120 kVp with automatic tube current (mA) using z-axis modulation. The examination was completed with axial, coronal and sagittal Multiplanar Reconstructions (MPR).

The split-bolus CT showed a left-sided retroperitoneal heterogeneous mass (11 cm in maximum size) containing calcifications, which compressed and displaced the adjacent organs. No infiltration of the nearby structures or lymph nodes enlargement was revealed (Figure 2A, 2B & 2C). The dose length product (DLP) was 98.1 mGy.cm. The final diagnosis of neuroblastoma was achieved by the histological examination of the tumor resection specimens (Figure 3).

Discussion

In the clinical radiologic practice, in order to detect and to define the extent of an abdominal neuroblastoma, US, CT and Magnetic Resonance Imaging (MRI) are used. US usually show a heterogeneous mass with internal vascularity, but also areas of low echogenicity, due to necrosis or calcifications, can be revealed [7]. MRI commonly demonstrates a heterogeneous iso-hypointense mass at T1-weighted images, variable and heterogeneous enhancement,

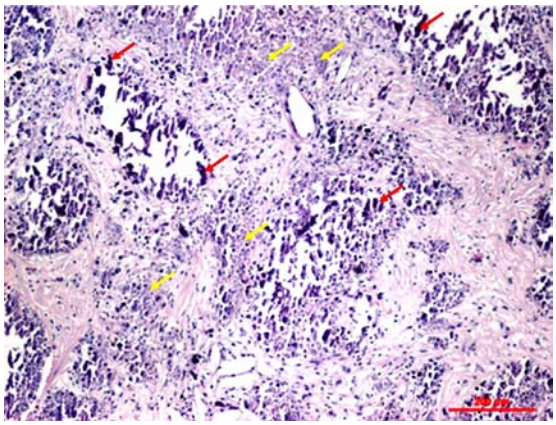

Figure 3: Histologic capture of the giant retroperitoneal neuroblastoma: the atypical neoplastic nests, at the center of the mass, have undergone a necrotic process, with consequent deposit of calcium salts. The red arrows point out the atypical neoplastic cells, while the yellow ones the necrotic areas (hematoxylin/eosin stain).

after intravenous administration of Gadolinium (Gd) with Diethylenetriaminepentaacetic Acid (DTPA), and hyperintense cystic/necrotic areas at T2-weighted images [8]. However, the staging of the abdominal neuroblastoma, as reported by International Society of Pediatric Oncology Europe Neuroblastoma Group (SIOPEN), differentiates the locoregional tumors in resectable or unresectable on the basis of a series of 'Imagine-Defined Risk Factors' (IDRF), detected by CT [9]. In fact, the CT is the only high-definition technique able to investigate the bone structures, and their relationship with the tumor, and to highlight the intra-tumoral calcifications. On CT the 80-90% of cases of abdominal neuroblastoma present some hypodense areas of necrosis into a mass, which insinuates itself beneath the aorta and lifts it off the vertebral column. Moreover, the mass tends to displace adjacent organs, encasing or compressing the nearby vessels. In the most aggressive forms, the lymph node enlargement and the infiltration of psoas muscle or kidney can be revealed [10]. In this last case, the differential diagnosis with Wilms tumor can be difficult [7]. Split-bolus intravenous contrast material administration is an innovative multidetector-row CT technique that in a single pass combines arterial and venous phases, allowing the detection of hypo- or hyper-vascular and mixed lesions, together with lymph node involvement and related distant metastases. If compared with the standard bi- or multiphasic CT, the split-bolus MDCT ensures diagnostic accuracy and efficacy, reducing significantly the radiation dose at the same time. Our double split-bolus MDCT protocol, by means of an optimal enhancement of the arterial, venous system and abdominal parenchymas, has allowed the detection of the mass and has clearly depicted its extension and relationship with the neighboring structures for an adequate staging. The split-bolus single pass whole body MDCT resulted in a DLP of 98.1 mGy.cm and in a reduction of the number of data sets, when compared to multiphasic CT.

Conclusion

The split bolus technique is currently used in patients with hematuria, for its specificity and accuracy in the detection of upper urinary tract tumors [11], and in the study of focal liver lesions [5]. It is intended to increase its fields of application, because it aims to

reduce the radiation dose with health benefits for pediatric and adult patients in the ALARA (as low as reasonably achievable radiation) era. The presented case supports the rationale of the split bolus MDCT technique, related to its high accuracy in the detection and staging of retroperitoneal neuroblastoma and to its radiation dose reduction for the young patients.

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