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Diffusion Weighted MRI on Diagnosing Abdominal Ganglioneuroma Surrounding the Mesenteric Artery

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Abstract

Ganglioneuroma arise from sympathetic ganglions and belongs to the group of neurogenic tumors including ganglioneuroblastoma and neuroblastoma. Ganglioneuroma can be considered a benign form of neuroblastic tumors, and is thought to be either a primary ganglioneuroma or follows a spontaneous or treatment-induced differentiation of neuroblastoma or ganglioneuroblastoma. Histopathology is the gold standard for diagnosis. Complete excision cannot always be done because of surgical complications. Therefore, imaging methods are required for distinguishing malignity and benignity. Benign and malign lesions such as ganglioneuroblastoma, neuroblastoma and ganglioneuroma have different ADC on diffusion weighted MRI. We report a 10-year-old-male with abdominal ganglioneuroma surrounding the superior mesenteric artery in which diffusion weighted MRI was used for diagnosis.

Introduction

Neuroblastoma, ganglioneuroblastoma and ganglioneuroma are a heterogeneous group of tumors derived from the primordial neural crest cells that form the sympathetic nervous system. Neuroblastomas are mainly composed of immature neuroblast and neuropil, whereas ganglioneuroblastoma contain both mature gangliocytes and immature neuroblasts. Ganglioneuroma is a benign, consisting of gangliocytes and mature stroma. Under therapy neuroblastoma and ganglioneuroblastoma can mature into a ganglioneuroma [1,2].

Abdominal regions other than adrenal gland or paravertebral areas are rare locations for a GN and retroperitoneal GNs are often lately diagnosed. In addition, a small number of GNs may undergo malignant transformation [3]. Thus, early detection leading to a complete resection of the tumor is clinically important. Computed tomography (CT) and magnetic resonance imaging (MRI) have been found to be useful in detection and diagnosis of GN [4-8]. Surgical treatment can sometimes be risky, in particular when the great vessels are involved [9]. This study describes a case with abdominal GN surrounding the mesenteric artery by using MRI and diffusion weighted MRI.

Case Report

A 10 year-old-male was admitted with abdominal pain. He has not any complaint prior to admission. The pain was sudden onset, severe and lasting up to 1 hour. He had a history of trauma to the lower region of the chest on the previous day. On physical examination, there was a palpable 7x8 cm mass with smooth surface on the right upper quadrant of abdomen which starting from the bottom of rib, and extending to the umbilical level. Complete blood count, blood smear, erythrocyte sedimentation rate, serum electrolytes and coagulation profile was normal. Lactate dehydrogenase (LDH) level was 422 U/L (N:220-450), urea: 21 mg/dL, creatine: 0.67 mg/dL, uric acid: 2.19 mg/dL (N:3.6-8.2), aspartate aminotransferase (AST) : 19 U/L, alanine aminotransferase (ALT) : 10 U/L, alkaline phosphatase (ALP) 301 U/L, α-fetoprotein (αFP) : 1.72 ng/mL (N:0-9), β-human chorionic gonadotrophin (β-hCG) : 0.3 mIU/mL (N:0-2.67), 24-hour-urinary vanilmandelic acid (VMA) : 6.1 mg/g creatinine (N:<8.2), and homovanillic acid (HVA) : 5.5 mg/g creatinine (N:<11.3). Abdominal ultrasonography revealed a 9x8 cm solid lesion inferomedial to the liver. Abdominal CT and MRI confirmed the sonographic findings with encasement of superior mesenteric artery by the mass. Mean apparent diffusion coefficient (ADC) value of the tumor was 2.34×10^{-3} mm^2/s (Figure 1a, 1b, 1c). The lesion has high signal intensity both on diffusion weighted image and ADC map consistent with a benign pathology. Adrenal glands were normal. Tru-cut biopsy of the lesion resulted as GN after pathologic examination of the specimen. There was an increased risk of complications including vascular complications, short bowel, liver atrophy, and spleen injury. Thus, the mass was planned to close follow up of the patient. The patient was good in condition and no increase on the mass size for 9 months.

Discussion

Ganglioneuroma is a benign form of peripheral neuroblastic tumor. Either spontaneous or chemotherapy-induced differentiation from malignant to benign forms can be seen in neuroblastic tumors. Ganglioneuroma can be considered a benign form of neuroblastic tumors, and is thought to be either a primary ganglioneuroma or follows a spontaneous or treatment-induced differentiation of neuroblastoma or ganglioneuroblastoma. These tumors occur in older children rather than in the first 5 years of life that more aggressive neuroblastomas diagnosed [1,9]. Our case was ten-years-old.

Ganglioneuroma is commonly localized in thoracic region (41.5%) especially in posterior mediastinum. Abdominal, nonadrenal tumors (37.5%) and adrenal GN (21%) were slightly rare. Ganglioneuromas represent 0.7-1.6% of all primary retroperitoneal tumors and are often asymptomatic even they got in large dimensions [10,11]. Occasionally, abdominal pain or a palpable abdominal mass is the only clinical finding. Cai et al. [12] reported 13 patients with retroperitoneal GN had no obvious clinical symptom or sign, while the other 4 presented diarrhea,

Figure 1: T2 (a), diffusion weighted image (b) and ADC map (c) MRI of upper abdomen shows a well defined mass encasing superior mesenteric artery (arrow in a). It is hiperintense both on the diffusion image (b) and ADC map, which suggests benignity.

hypertension or palpable masses. The secretion of catecholamines or vasoactive intestinal polypeptides could be responsible for such symptoms as hypertension or diarrhea [13]. Our patient had not got any symptom prior to admission and his catecholamine levels were normal.

Detection of the retroperitoneal GNs usually depends on imaging modalities. Ultrasound is regarded as the preferred initial screening and diagnostic modality for children because it is noninvasive, safe, and readily available. Sonographic appearance is not specific, the mass showing a heterogeneous solid structure for ganglioneuroblastoma and neuroblastoma, homogenous solid structure for GN. US can be helpful in localizing the origin of the mass and in visualizing the relationship to the vessels. Doppler US may be used to evaluate flow in encased vessels.

Surrounding of major vessels is also observed in both GNs and more undifferentiated neurogenic tumor, but occlusion it usually related to malignant forms [12]. Otal et al. [4] reported in three of seven cases with retroperitoneal GN consisting of a tendency of the tumor to surround major blood vessels, resulting in absent or mild compromise of the lumen. Our patient has a GN surrounding superior mesenteric artery without any occlusion.

Additional diagnostic tools include CT and MRI, which can provide more excellent visualization of tumors, organ of origin, regional invasion, vascular encasement, adenopathy, calcification and reveal helpful information for surgical approach. On CT and MRI, GNs usually present an oval, well-defined mass. CT is the most sensitive method in detecting calcifications which are present in 20-60% of cases [12]. The calcification is a commonly observed sign in neurogenic tumors, other than a feature special for GN. One of the main disadvantages of CT is ionizing radiation. GNs are low attenuation and homogeneous on unenhanced CT scans and demonstrate slight to moderate enhancement, which may be heterogeneous or homogeneous [8,14].

Ganglioneuromas were previously reported to have homogeneous low signal intensity on T1-weighted images and inhomogeneous high signal intensity on T2-weighted images [4-6]. Several reports indicate that relatively high, heterogeneous signal intensity on T2-weighted images correlates with GN; the appearance is presumed to be caused by a combination of myxoid material and relatively low amounts of ganglion cells. MRI enhancement varies from mild to marked; early enhancement at dynamic MRI is not typically seen in GN. GN does appear to accumulate contrast material over time, however, so delayed images may show increasing enhancement [8,14,15].

Under chemotherapy or spontaneously neuroblastoma can mature into a more benign form such as ganglioneuroblastoma or GN. So far, only histology can differentiate these tumors but not CT or MRI

studies. Diffusion describes a passive process of random thermal motion of millions of individual water molecules. Subtle changes in the degree of restriction to diffusion, for example due to an alteration in cell membrane integrity or permeability to water are reflected in changes in the diffusion-weighted signal. A low ADC in vivo represents restriction of the free diffusibility of water molecules, which supports a model in which increasing cellularity and more densely packed tissues have a relatively low ADC. Neuroblastomas consist of densely packed, small, immature cells that contain little cytoplasm, whereas in GN mature ganglion cells with abundant cytoplasm surrounded by more stroma are seen [16].

Gahr et al. [16] reported when performing additional diffusion weighted imaging they found a significantly different ADC of ganglioneuroblastoma and GN compared to the ADC of neuroblastoma. In their study, all but one neuroblastoma had an ADC lower than 1.13×10^{-3} mm^2/s, whereas the ADC of ganglioneuroblastoma and GN were all higher than 1.13 up to 1.99×10^{-3} mm^2/s. Kocaoglu et al. [17] used 1.11×10^{-3} mm^2/s as cut-off value and found that mean ADC values of benign and malignant pediatric abdominal lesions were $2.28 \pm 1.00 \times 10^{-3}$ mm^2/s and $0.84 \pm 1.7 \times 10^{-3}$ mm^2/s, respectively. They also assessed signal intensity changes and found that all hyper intense mass lesions on ADC map is benign as in our case.

Biopsy is gold standard for the diagnosis of this group of neuroblastic tumors. However, in inoperable cases biopsy can be performed only from a specific location of the tumor, other parts of the tumor can be in a malignant biology. Although USG, CT or MRI have been mostly practiced, preoperative diagnosis of GN is difficult from other tumors because of the lack of specific imaging [8,9].

Prognosis after surgical resection without further therapy seems to be excellent. However, surgery has increased complication rate in some conditions like great vessel invasion, massive hemorrhage, aortic artery rupture, renal insufficiency, paraparesis, intestinal infarction and death. In these limited group of patients follow-up till arterial occlusion symptoms develop come to an option. In such inoperable cases, the exact diagnosis cannot be possible. Thus, diffusion weighted MRI may be useful in malignity and benignity.

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