

Case Report

Bilateral Adrenal Neuroblastoma; Stage IV - A Case Report

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Abstract: Bilateral adrenal neuroblastomas have always been considered rare with few cases presented in medical literature. Here we present a case of bilateral adrenal neuroblastoma with liver, skeletal, nodal and leptomeningeal metastasis presented at 3 years of age; which is a late presentation considering the fact that bilateral neuroblastomas present at a mean age of 9 months.

Keywords: Bilateral adrenal neuroblastomas, ultrasonography, computed tomography.

CASE REPORT

A 3 year girl child presented with 4 month history of on and off fever and abdominal distension with progressive proptosis over the last month. On examination a tender hepatomegaly with bilateral upper-quadrant/flank masses were noticed. Ultrasound of the abdomen demonstrated a bilateral, mixed echogenic mass, enveloping the upper poles of the kidneys. These masses flattened and displaced the kidneys inferiorly (Figure 1). A large echogenic lesion was also noted in the caudate lobe of liver (Figure 2).



Fig. 1: USG- Kidneys flattened and displaced by mass



Fig. 2: USG- Large echogenic lesion in the caudate lobe of liver

Computed tomography (CT) Abdomen showed bilateral large heterogeneously enhancing masses in the

paravertebral region showing multiple non enhancing areas of necrosis and subtle areas of amorphous calcification (Figure 3).

The mass was compressing the upper poles of the kidneys displacing the kidneys infero-laterally and crossing the midline, displacing vessels. This conglomerated necrotic retroperitoneal nodal mass was encasing the renal vessels, aorta and its major branches and IVC displacing them anteriorly. Hepatomegaly with a large heterogeneously enhancing metastatic lesion was seen in the caudate lobe of liver (Figure 4).

Lytic permeative destruction of bilateral iliac wings and vertebral bodies is seen with speculated periosteal reaction and adjacent enhancing soft tissue (Figure 6). Similar lesions were also seen in bilateral greater wings of sphenoid and frontal bones with large adjacent soft tissue mass which shows strong post contrast enhancement. The mass shows extension into extraconal space of bilateral orbits invading lateral rectus muscles causing proptosis. Multiple leptomeningeal metastasis were also seen (Figure 7, 8).

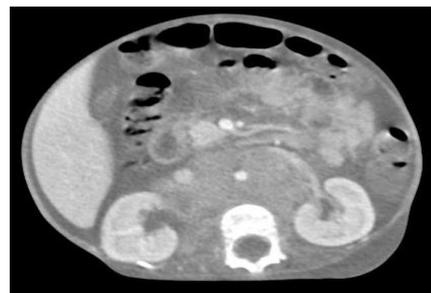


Fig. 3: CT- bilateral large heterogeneously enhancing masses in the paravertebral region with areas of necrosis and calcification.

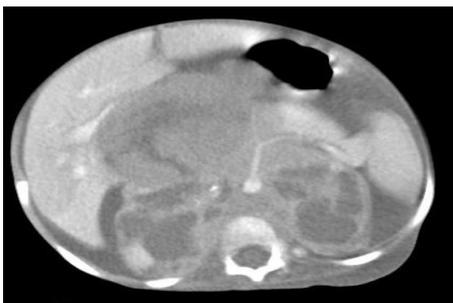


Fig. 4: CT- Hepatomegaly with a large heterogeneously enhancing metastatic lesion was seen in the caudate lobe of liver

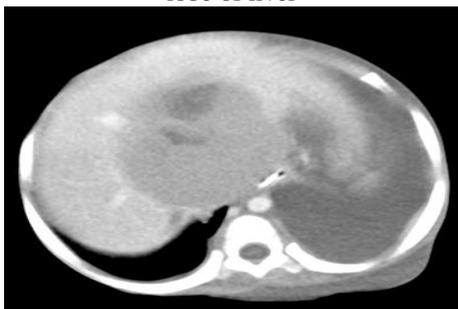


Fig. 5: CT- mass in caudate lobe

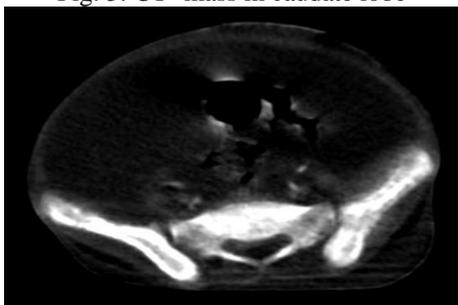


Fig. 6: CT- Lytic permeative destruction of bilateral iliac wings and vertebral bodies

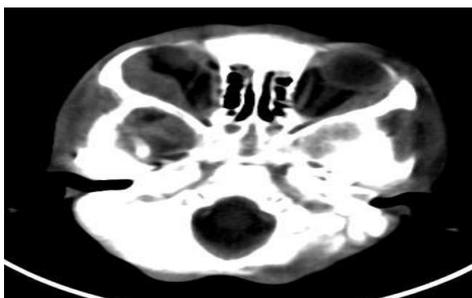


Figure 7: CT- Sphenoidal and frontal bone metastasis with post contrast enhancement

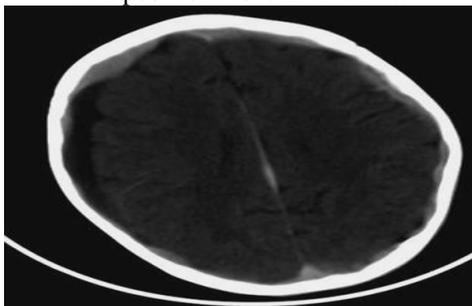


Figure 8: CT- Leptomeningeal metastasis

All the above mentioned findings suggested it as a case of bilateral adrenal Neuroblastomas (STAGE IV). Serum and urine catecholamine levels showed mild elevation of serum homovanillic acid (HVA) and urine vanillylmandelic acid (VMA). Histological study confirmed the diagnosis of neuroblastoma. Patient was referred to radiotherapy clinic for further management.

DISCUSSION

Neuroblastoma is a poorly differentiated neoplasm derived from neural crest ectoderm. More than one third of cases (36%) are diagnosed in children younger than 1 year [1]. It is one of the most common malignant tumors of infancy with most of cases arising in the adrenal glands [2]. Bilateral adrenal involvements from synchronous development or metastatic spread of the tumour are seen in less than 10% of children with neuroblastoma [3-4]. Male-to-female sex ratio is 1.2:1. Some patients exhibit a hereditary predisposition for neuroblastoma, such as in cases with apparent familial, bilateral or multifocal disease. In these subsets of patients, the median age at diagnosis is 9 months [4].

Neuroblastoma has been called "the great mimicker" because of its myriad clinical presentations related to the site of the primary tumour, metastatic disease and its metabolic tumor by-products. Most neuroblastomas produce catecholamines, which causes these varied clinical presentations [3]. The tumor cell can produce and excrete catecholamines substances such as dopamine, epinephrine and its metabolites - vanillylmandelic acid (VMA) and homovalnillic acid (HVA) [5, 6]. About 90-95% of neuroblastoma cases have high urine VMA and HVA at diagnosis. If the ratio between urine HAV and VMA is >1.5 , it is a good prognosis [5, 6]. Because the tumor cell can excrete epinephrine which can be detected in plasma, this can cause severe systemic hypertension at diagnosis [7]. The neuroblastoma cell can produce and excrete other substances such as neurone-specific enolase (NSE), vasoactive intestinal peptide (VIP), ferritin and lactic dehydrogenase (LDH) [5, 6]. As in our patient, serum NSE is very high. NSE is the tumor marker for neuroblastoma. It is useful for following the disease activity and the response to treatment in individual patients.

On ultrasonography, an adrenal neuroblastoma appears as a suprarenal heterogeneous echogenic mass. Ultrasonography can show urinary obstruction, vascular displacement and compression, nodal involvement, tumor extent, and liver involvement. However neuroblastomas are often large, and when they spread throughout the abdomen and/or into the chest, ultrasound may not be able to define their precise edges. It has limited ability to detect metastases in retroperitoneal and retrocrural lymph nodes and is usually unable to detect extradural extension of tumors [8, 9]. Color Doppler ultrasound has been reported to showed increase flow in neuroblastoma [10, 11].

On CT, neuroblastoma appears as a lobulated, soft-tissue mass, either homogeneous or heterogeneous, which is caused by hemorrhage, necrosis, and/or calcification. Calcification has been reported on CT in approximately 85% of patients. CT can show prevertebral extension of tumor across the midline as well as encasement of the celiac axis or superior mesenteric artery by the neuroblastoma. It can also show extension of tumor to retroperitoneal lymph nodes, to the liver, around central vessels, and into the vertebral canal. CT is excellent for demonstrating retrocaval and paravertebral tumor extension to the chest, common in abdominal neuroblastoma [12, 13]. The CT scan can demonstrate all accurate primary tumors and metastatic lesions. Abdominal neuroblastoma typically appears on CT as irregular suprarenal masses with a heterogeneous texture due to hemorrhage and necrosis. Calcification is detected in 85% of cases [14, 15].

Magnetic resonance imaging (MRI) is excellent for evaluating the location, extent and spread of neuroblastoma and the tumor shows equal or lower signal intensity on T1-W images and higher signal intensity on T2-W images compared with muscle. The center of the tumor is often heterogeneous, reflecting the presence of hemorrhage, necrosis or calcification. Advantages of MRI over CT and ultrasonography are: multiplanar imaging (useful for assessing invasion of adjacent organs); detection of extradural tumor extension; identification of bone marrow metastases (useful for staging); and delineation of intra-abdominal vascular displacement or encasement without using intravenous contrast media [12, 13].

The treatment of neuroblastoma depends on the age of the child and the stage of the tumor. Surgery is indicated for stage 1 and 2 tumors while chemotherapy and radiation are the primary treatment modalities for advanced tumors.

In conclusion the bilateral neuroblastoma is a rare entity. Medical imaging is very helpful in the diagnosis and the staging of this tumor.

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