Case 11108

Congenital cystic neuroblastoma of the adrenal gland
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Section: Paediatric radiology
Area of Interest: Adrenals
Procedure: Diagnostic procedure
Imaging Technique: MR
Imaging Technique: CT
Imaging Technique: Ultrasound
Special Focus: Neoplasia Case Type: Clinical Cases
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Patient: 14 days, male

Clinical History:

A 14-day-old boy, delivered by an uncomplicated Caesarian section at 38 weeks gestation of a twin pregnancy, was transferred from another institution due to a left suprarenal mass detected in the 20-week pre-natal ultrasound scan. Laboratory tests, including bone marrow and urine catecholamine level measurements were unremarkable.

Imaging Findings:

Ultrasound (US) revealed a circumscribed, round, complex predominantly cystic mass, with thin septations and some calcifications, measuring 4.4 x 4.1 x 4.0 cm in the area of the adrenal gland, displacing the left kidney but showing a cleavage plane with the upper pole of the kidney. Doppler-US imaging revealed internal vascularisation. Computed Tomography (CT) demonstrated a hypodense mass with thin wall, but post-contrast images did not show unequivocal enhancement. Magnetic resonance imaging (MRI) showed a septated cystic mass with high signal intensity content on T1WI and T2WI, due to haemorrhage or proteinaceous fluid. Subtraction images showed enhancement in the cyst wall. There was retroperitoneal adenopathy encasing the coeliac trunk and the superior mesenteric artery. No abnormality of the liver was seen. 

[123I]meta-iodobenzyl guanidine (123I-MIBG) scan did not show MIBG uptake in the mass.

Follow-up, two weeks later, demonstrated that the cyst was unchanged.

Discussion:

Neuroblastoma is the most common perinatal malignancy, and the adrenal gland is the most common site of origin [1, 2]. Haemorrhage and necrosis are often seen in neuroblastomas, but cystic neuroblastoma (CN) is a very rare form [1-4]. The content of cysts is usually haemorrhagic, although it may be serous or gelatinous [2, 5]. The usual appearance of an adrenal CN is that of a complex cystic mass compressing or displacing the kidney inferiorly and laterally [3]. The main differential diagnosis is adrenal haemorrhage (AH) which is the most common cause of adrenal mass during the perinatal period presenting a similar appearance [6]. The spectrum of appearances of AH depends on the age of the haemorrhage. Sonographic follow-up shows cystic transformation within several days, evidence of gradual decrease in size, and ultimately disappearing or being replaced by small calcifications in a median follow-up period of 90 days [1, 7].

Prenatal detection in fetal US was reported in AH, but is more common and usually much earlier in CN [1]. AH is
frequently associated with renal vein thrombosis [8].
Elevation of serum or urine catecholamines is uncommon in cases of CN, thus, a negative test cannot exclude these possibility [1, 4, 5].
There are some findings, which may suggest CN:
- failure of a mass to resolve on follow-up examination [1, 7];
- initially detected calcifications are unusual during the neonatal period, because they appear as a late finding in the haemorrhage [1];
- irregular, thickened walls or of prominent soft tissue component [5];
- vessels within the mass on Colour Doppler US (seen only in neuroblastoma) [1, 7];
- enhancement of the mass in MRI or CT [1];
- hepatic or nodal metastasis (highly suggestive of neuroblastoma) [1].
All the imaging techniques can potentially show the vascular nature of the lesion, but Doppler US and subtraction MRI have more sensitive. US and TC are useful in the detection of tiny calcifications. MRI is especially useful to determine the stage of the haemorrhage. US is the modality of choice in the follow-up.
The sensitivity and specificity 123I-MIBG-scintigraphy in newborns is unknown [7].
An excellent prognosis was reported for cystic neuroblastomas in infancy [6].
In our case, the persistence of the mass, the presence of internal flow blood and a concurrent retroperitoneal adenopathy were the crucial features for the diagnosis.
US-guided biopsy was performed. Haemorrhagic fluid was aspirated but the material was insufficient for cytological diagnosis. Surgery was performed and the pathologic diagnosis was CN. Due to irresectable disease the patient underwent chemotherapy.

Differential Diagnosis List: Cystic neuroblastoma, Adrenal haemorrhage, Cystic teratoma, Extralobar sequestration, Dilation of upper-pole renal calyces of a duplex kidney, Congenital adrenal cyst, Adrenal abscess, Enteric cyst, Cystic Wilms’ tumour, Mesoblastic nephroma, Multilocular cystic nephroma, Choristoma

Final Diagnosis: Cystic neuroblastoma

References:

Description: On Doppler-US blood flow was seen in the cyst. Origin: Department of Radiology, Centro Hospitalar de São João, Porto, Portugal
Description: Axial unenhanced CT of the abdomen shows thin wall of the left cystic mass and calcification. Origin: Department of Radiology, Centro Hospitalar de São João, Porto, Portugal
Description: Axial contrast enhanced CT did not demonstrate clear enhancement of wall and internal septa (arrow). Origin: Department of Radiology, Centro Hospitalar de São João, Porto, Portugal
Description: Coronal fast spin-echo T2-weighted MRI of the cystic tumour shows high signal intensity. Note the median retroperitoneal adenopathy encasing the coeliac trunk and the superior mesenteric artery (arrow). Origin: Department of Radiology, Centro Hospitaarl de São João, Porto, Portugal
Description: Axial spin-echo fat-saturated T1-weighted MRI of the cystic tumour shows high signal intensity. Origin: Department of Radiology, Centro Hospitalar de São João, Porto, Portugal
Figure 6  

**Description:** Subtraction images show enhancement in the cyst wall. **Origin:** Department of Radiology, Centro Hospitalar de São João, Porto, Portugal
Description: 123I-MIBG scan did not show MIBG uptake in the mass. Origin: Department of Radiology, Centro Hospitalar de São João, Porto, Portugal
Description: US of the left upper quadrant shows a complex multiloculated mass (arrow) with thin septations, internal echoes and some calcifications in superior and medial location to the left kidney (asterix). Origin: Department of Radiology, Centro Hospitalar de São João, Porto, Portugal