ADRENAL HEMORRHAGE VERSUS CONGENITAL NEUROBLASTOMA IN NEWBORN WITH RIGHT SUPRAARENAL MASS AND HYPERTENSION: A CASE REPORT

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ABSTRACT Adrenal haemorrhage is a potentially life-threatening condition rarely present in the neonatal period. The large size and increased vascularity of the average infant adrenal gland increase the bleeding tendency. Predisposing factors may include macrosomia, birth asphyxia, perinatal injuries, septicemia, and coagulopathy. Nevertheless, it can also occur without apparent risk factors. The most common findings are poor feeding, vomiting, persistent jaundice, anaemia, and abdominal mass. However, the event may be asymptomatic and be detected incidentally. Ultrasound imaging is the cornerstone of diagnosis and follow-up monitoring. Herein, we report an unusual case of a male newborn with an adrenal haemorrhage associated with hypertension. The lesion was detected by ultrasound as an echogenic cystic mass at the right suprarenal area. The mass was initially mimicking neuroblastoma. Nevertheless, the normal levels of urine catecholamine metabolites and the regression of the mass size verified the diagnosis. The infant was managed conservatively, and the mass entirely resolved without calcification or adrenal insufficiency.

KEYWORDS adrenal haemorrhage, neonatal hypertension, neuroblastoma, suprarenal mass, adrenal calcification

Introduction

Adrenal haemorrhage (AH) is a relatively uncommon clinical problem in the neonatal period. The large size and increased vascularity of the neonatal adrenal glands may increase the bleeding vulnerability. Males and term infants are affected more frequently than females or preterm infants [1]. Minor haemorrhage may remain asymptomatic with the diagnosis made incidentally. Whereas, an extensive haemorrhage may become symptomatic shortly after birth. The typical clinical findings are a palpable abdominal mass, unexplained anaemia, and prolonged jaundice. AH is frequently associated with large fetal size, birth trauma, perinatal asphyxia, septicemia, or coagulation defects. However, in many cases, the aetiology of bleeding cannot be established [2]. Abdominal ultrasound facilitates the diagnosis of adrenal haemorrhage, particularly when the clinical presentation is subtle. AH commonly appears as a round cystic lesion with no blood flow. Hypertension is an uncommon problem in neonates, and AH is an exceedingly rare acquired cause of neonatal hypertension [3].

Case report

A full-term baby boy was a product of difficult vaginal delivery associated with shoulder dystocia at a peripheral hospital. The mother was a 30-year-old primigravida with an uneventful pregnancy apart from amniotic fluid leaking for the last 2 days. The parents were nonconsanguineous with unremarkable family history. The birth weight was 3.4 kg, and the Apgar score was 8, 9, and 9 at one, five, and ten minutes, respectively. The baby was observed to have severe right Erb’s palsy and hypoaevity; therefore, he was shifted to the NICU for admission. Basic laboratory investigations, chest and abdomen x-ray, and blood
cultures were taken then intravenous antibiotics and fluids were commenced. After stabilization, oral feeding was started and initially tolerated. However, at 20 hours, the baby developed poor feeding, vomiting, and more lethargy. Urgent brain and abdominal ultrasound revealed cerebral oedema with multi-foci of petechial haemorrhages. The abdominal ultrasound study was normal.

On the 2nd day of life, the baby was transferred to our hospital for further management. The baby was stable on room air on arrival with normal vital signs. The physical examination was unremarkable except for the severe right Erb’s palsy. Laboratory data were normal, including complete blood counts, blood chemistry, coagulation profile, and C-reactive protein. Repeated blood culture was negative. He developed unexplained indirect hyperbilirubinemia and received phototherapy. On the 4th day of life, follow-up brain US was normal. However, the abdominal US (figure 1) showed a heterogeneous cystic mass (2.6x2.3 cm) in the right suprarenal region.

The mass was not connected to the renal collecting system, and both kidneys were normal. The colour flow Doppler study (figure 2) showed no blood flow inside the mass, and the renal arteries were normal. On the 6th day of life, the baby started to have sustained hypertension, and IV hydralazine was commenced. An echocardiogram revealed a normal heart and aorta. An adrenal mass was reported, and a CT scan abdomen was advised for proper identification. CT scan abdomen with an adrenal protocol (figure 3) revealed a well-defined 2.8 x 1.6 x 2 cm right adrenal mass that showed slight heterogeneous pre-contrast hypodensity and heterogeneous enhancement in post-contrast images. There were no foci of calcifications within the mass, and the left adrenal gland was normal. The history of high blood pressure, persistent mass size with abnormal echotexture, and the enhancement in post-contrast CT images necessitated the exclusion of adrenal tumours like neuroblastoma and ganglioneuroma. Therefore, the levels of catecholamine metabolites (vanillylmandelic acid and homovanillic acid) in 24-hour urine collection were tested, and the results were normal. Urine microscopy and chemistry were also normal. The subsequent abdominal US showed the resolution of the cystic area, but the mass size was the same. Brain MRI was done, and it was essentially normal.

Based on the clinical findings, imaging studies, and laboratory results, the diagnosis of the right AH was made, and conservative management was considered.

After 14 days of hydralazine treatment, the blood pressure normalized, and the dose was gradually reduced and discontinued. The infant was on regular physiotherapy with progressive improvement in his right Erb’s palsy. On the 38th day of life, the abdominal US showed complete resolution of the adrenal mass. The patient was discharged home in good condition and given regular follow-up. During the subsequent visits, there was neither recurrence of hypertension nor the adrenal mass. When writing this article, the child was 15 months old, healthy with normal growth and development.

Discussion

AH is the most common adrenal mass in neonates and is more frequent than in older children. The large size of the neonatal adrenal gland, and its excessive vascularity, may contribute to the bleeding vulnerability. The incidence of AH varies from 1.7 per 1000 autopsied neonates to approximately 3% of the infants undergoing the abdominal US. The right side is involved in 70% of the cases due to compression between the liver and the kidney, and it is bilateral in 10% of the cases [4].

The pathophysiology of adrenal haemorrhage is not yet fully known, and it is deemed to be multifactorial. Acute fluctuation of blood pressure and abnormalities in autoregulation occur-
AH can be challenging and inconclusive. The adrenal gland is usually excellent. Laparotomy is indicated when haemorrhage in doubt [14].

Clinical presentation can vary from asymptomatic minimal bleeding to fulminating bleeding resulting in severe hypovolemic shock, adrenal failure, or death. Physical findings include palpable abdominal mass, unexplained anemia, prolonged jaundice, hematuria, and bluish scrotal discoloration. The adrenal gland has a considerable regenerative capacity, and adrenal insufficiency rarely occurs [6]. AH is a rare cause of neonatal hypertension. The possible mechanisms may include direct stimulation of the adrenal medulla to release catecholamines by humeral factors derived from the hematoma, transient compression of the renal artery, and obstruction of the collective renal system secondary to the displacement of the kidney by the mass [7].

Abdominal ultrasound remains the modality of choice for both the initial screening and the follow-up. Computed tomography (CT) and magnetic resonance imaging (MRI) may be used to confirm the presence of the haemorrhage or to exclude malignancies [8]. The ultrasound appearance depends on the stage of the haemorrhage ranging from an echogenic lesion in the acute stage to a cystic or multicystic appearance in the subacute and chronic stages. Ultimately, the adrenal mass becomes anechoic, shrinks then resolves completely or is left with a rim of calcification. AH shows consistently no vascularization in the colour flow Doppler study [9]. The main differential diagnoses of neonatal AH are congenital neuroblastoma, adrenal abscess, congenital adrenal hyperplasia, lung sequestration, mesoblastic nephroma, and urinary or intestinal tract duplications. However, neuroblastoma is the most important entity requiring immediate attention due to its notorious extension and the need for urgent interventions [10].

Neuroblastoma is the most common neonatal tumour with an incidence of 0.61 per 100,000 live births. It accounts for 28–39% of all malignancy in the first month of life. The sonographic appearance of neuroblastoma is commonly a homogeneous echogenic solid mass, which will continue to increase in size. Moreover, blood flow signals can be detected by the colour flow Doppler study [11]. Neuroblastoma can also have the sonographic appearance of a hemorrhagic adrenal cyst. The distinction from AH can be challenging and inconclusive. The adrenal gland is the most common site of primary localization. Adrenal tumours should be suspected if the mass is not resolved within 90 days [12]. The level of urinary catecholamine metabolites does not fulfill the role of a screening test for neuroblastoma. However, increased levels of these metabolites may suggest a diagnosis other than AH [13].

A noninvasive approach is strongly recommended in the management of neonatal AH. Infants with acute haemorrhages need urgent blood replacement and administration of vitamin K. Hydrocortisone should be given in all bilateral haemorrhages and cases not responding adequately to blood or intravenous fluid administration. The outcome of the adrenal haemorrhage is usually excellent. Laparotomy is indicated when haemorrhage seems to be continuing, especially when the exact site of haemorrhage is in doubt [14].

Conclusion

Neonatal AH imposes diagnostic and management difficulties. It should be considered and looked for in at-risk newborns with macromasia, perinatal asphyxia, traumatic deliveries, overwhelming sepsis, and bleeding diathesis. Still, abdominal sonography is the best modality for diagnosing and monitoring AH. Adrenal hematomas should be scrutinized in hypertensive newborns with a history of delivery complications. Serial ultrasound studies along with specific laboratory tests can establish the diagnosis in many cases. We present this case owing to the rare presentation of neonatal AH with hypertension. We aim to emphasize the importance of careful blood pressure monitoring and the effectiveness of conservative therapy in AH.

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Conflict of Interest

There are no conflicts of interest to declare by any of the authors of this article.

References


