Adrenal masses associated with Beckwith Wiedemann syndrome in the newborn

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ABSTRACT

Adrenal cystic lesions are rare and may be associated with both complete and incomplete Beckwith syndrome (BWS). Because the adrenal gland often houses malignant lesions, differentiation between benign and malignant lesions of the gland, although usually difficult, is very necessary from the point of view of management. Here we present our experience in a case of incomplete BWS with adrenal cystic lesion and review of the literature.

Key words: Adrenal gland, Beckwith Wiedemann syndrome, haemorrhage

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INTRODUCTION

Adrenal cystic lesions are rare and may be associated with incomplete Beckwith syndrome. Beckwith Wiedemann syndrome (BWS) consists of macroglossia, abdominal wall defects, visceromegaly, gigantism, hemihypertrophy, hypoglycaemia, ear crease, nevus flavus, adrenal cystomegaly, midface hypoplasia and associated various tumours.[1-3] Incomplete BWS refers to a condition where two or three of these features are present.[4]

The prevalence of malignancy in this syndrome is around 7.5% to 11%.[3,5] Here we present a case report of a 2-month-old baby boy with incomplete BWS and an antenatal diagnosed bilateral adrenal cystic lesion and also a review of the similar association in the literature.

CASE REPORT

A 2-month-old baby boy was referred with antenatal diagnosed of bilateral adrenal cysts at 32nd week of gestation. A postnatal left-sided lesion reduced significantly with persistence of cystic right suprarenal mass on ultrasound. The patient had right-sided hemihypertrophy. CT scan of the abdomen showed a right adrenal thick walled cystic lesion (2.3 × 2.4 cm), no calcification. There was a small left adrenal nodular lesion with unsure calcification. [Figure 1] His blood pressure and blood parameters were normal except for the urinary vanillylmandelic acid (VMA) raised (17 mg/dl). Repeat CT scan after 1 month did not show any significant reduction in size of the right-sided suprarenal cystic lesion. Because the patient was residing very far (more than 600 km) from the hospital and was would have been difficult for regular follow up, surgical removal of the lesion was done at 2 month of age. It was a well-defined cystic lesion which was removed intact [Figure 2]. Histopathology report showed a benign cyst with adrenal rests suggestive of hemorrhagic cyst.

DISCUSSION

The incidence of somatic hemihypertrophy is reported as 1 in 86,000 neonates.[4] Hemihypertrophy is considered as an incomplete form of BWS.[2,5] BWS, whether complete or partial, is associated with development of various tumours such as Wilms' tumour, neuroblastoma, hepatoblastoma and rhabdomyosarcoma. Various adrenal lesions are associated with both complete and partial BWS which included cortical hyperplasia, foetal cortical cytomegaly, pheochromocytoma, adrenal calcification, cystic adrenal haemorrhage, and neuroblastoma.

Congenital adrenal haemorrhage and cystic neuroblastoma are difficult to differentiate by ultrasonography.[6] Haemorrhage appears solid and echogenic in the initial phase and then becomes hypoechoic and cyst like and later may develop marginal calcification. Differentiation of congenital adrenal haemorrhage from cystic neuroblastoma is difficult even with MRI.[2] Colour-coded Doppler ultrasonography may be useful.[7]

According to Sauvat *et al*. in a series of 53 patients on

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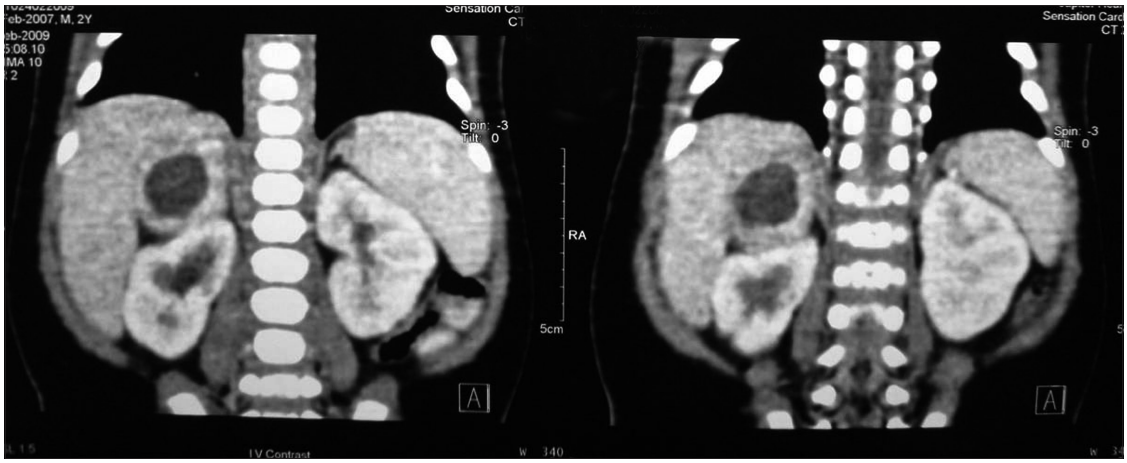


Figure 1: CT scan of the abdomen showing a cystic adrenal lesion on the right

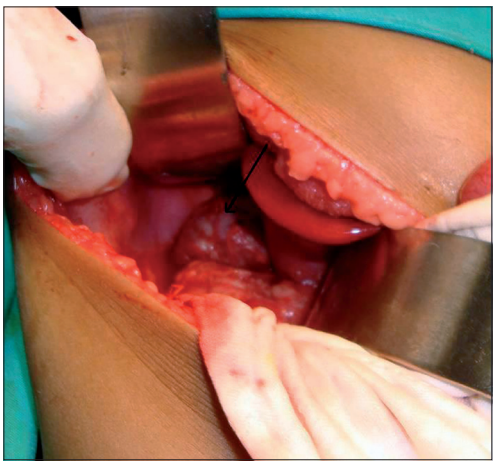


Figure 2: Operative photograph, arrow showing the right adrenal lesion

outcome of suprarenal localized masses when diagnosed during the perinatal period, localized neuroblastoma was found to be the most common suprarenal lesion in the perinatal period. Most of the lesions were located on the right side.[8]

Our review of the literature revealed 10 cases of incomplete BWS associated with either unilateral or bilateral adrenal cysts.[1,2,4,9] Most of the cystic adrenal lesions were on the right side (five), followed by left side (three) and bilateral lesions (two). Most of these masses showed adrenal cystomegaly with multiloculated hemorrhagic cysts in the adrenal gland without any evidence of tumour. Sauvat *et al*. reported that the most common lesion was localized neuroblastoma. In the patients with incomplete BWS all the surgically removed glands showed hemorrhagic macrocysts initially, but later developed secondary malignancies.[1]

In patients with BWS either complete or incomplete there is increase of occurrence of malignant lesions, but in cases of cystic adrenal lesions with BWS most of them are hemorrhagic cysts. These can be regularly followed up conservatively. But close monitoring of size of the lesion is required along with search for other malignancies. Surgical excision can be considered if the size does not decrease.

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