Early Presentation of Right Adrenal Mass, Hepatoblastoma and Hepatic Cavernous Haemangioma in Beckwith-Wiedemann Syndrome

S H Teh, MRCPCH, G B Ong, MRCP, MMed

Department of Paediatrics, Hospital Umum Sarawak, Kuching, Sarawak

SUMMARY

Beckwith-Wiedemann Syndrome (BWS) is associated with early development of embryonal tumours usually in the first four years of life. We describe a patient who presented with a right adrenal cyst in the first month of life and hepatoblastoma in the third month of life. A cavernous haemangioma was subsequently found in the resected tumour.

KEY WORDS:
Beckwith-Wiedemann Syndrome, Hepatoblastoma, Adrenal cyst, Cavernous haemangioma

INTRODUCTION

Beckwith-Wiedemann Syndrome (BWS) is a disorder of growth associated with macrosomia, hemihypertrophy and development of embryonal tumours. A male infant with Beckwith-Wiedemann presented early with a right adrenal cyst followed by a solitary hepatoblastoma of segment 6 of the liver. He was successfully treated with chemotherapy and had the hepatoblastoma completely resected. Histopathological examination of the resected tumour found a previously undiagnosed cavernous hamagioma within the tumour mass.

CASE REPORT

GSL was delivered vaginally at term with good Apgar scores to a 41 year old Bidayuh lady with essential and pregnancy-induced hypertension and impaired glucose tolerance in pregnancy. His birth weight was 3.80kg, birth length was 51cm and head circumference was 34cm. Facial congestion was noted at birth. He was subsequently admitted for abdominal distension and vomiting and treated for presumed neonatal sepsis.

Newborn examination revealed macroglossia and right sided hemihypertrophy involving the right half of the face, right upper limb and right lower limb. He had a hepatomegaly of 6cm and splenomegaly of 3cm. There was no exomphalos nor transverse ear creases/pits. Bedside monitoring of random blood sugar was normal. A clinical diagnosis of Beckwith-Wiedemann syndrome was made. No other anomalies were found, and he was discharged well upon completion of antibiotics and resolution of neonatal jaundice.

There was no family history of BWS. He was the only child of the current union, with four other sibs from his mother's previous marriage. An abdominal ultrasound was done on Day 23 of life. A right adrenal nodule measuring 1.8 x 1.8 x1.9cm was seen. There was no evidence of Wilm's tumour nor hepatoblastoma.

A contrasted computerized tomogram (CT) of the abdomen was done at Day 40 of life. The right adrenal cystic mass was described measuring 1.5 x 1.7cm with no calcification. There was no lesion in the liver nor the kidneys. The radiological differentials for the adrenal mass were an adrenocortical cyst, cystic neuroblastoma or cortical adenoma. Magnetic Resonance Imaging (MRI) was performed five weeks later. The liver showed an ill-defined lesion in segment VI, enhancing following gadolinium measuring 4cm x 4cm, most likely to be a hepatoblastoma. The right adrenal cyst was 1.1cm x 1.1 cm x 1.0cm; the impression was an adrenal cortex cyst or an adrenal adenoma.

An ultrasound-guided liver biopsy was done with an 18G trucut biopsy needle under general anaesthesia six days later with two good samples obtained. The tissue was reported as hepatoblastoma based on blastoid-looking cells with rosette-like appearance with mitoses, staining positive for alpha-foetoprotein and weakly positive for human chorionic gonadotropin. They were negative for cytokeratin,epithelial membrane antigen, vimentin and carcinoembryonic antigen. The cell showed mainly embryonal features.

Serum alpha-foetoprotein was 21860 µg/L (dil 1:50) at diagnosis. His bone marrow aspirate and trephine were normal. His chest radiograph was clear. Urine cathecholamines were normal. Baseline liver and renal functions were normal. Pre treatment haemoglobin was 7.9 g/dl, total white cell count 8110 cell/mm^3 and platelets were 112 000/ mm^3.

The patient was started on chemotherapy at three months of age with the International Childhood Liver Tumour Strategy Group (SIOPEL) 3 Low Risk Protocol using Cisplatin as a single agent at two weekly intervals with ultrasound monitoring of the tumour response. He underwent four courses of chemotherapy before surgery. Serial alpha fetoprotein showed a decreasing trend with treatment down to 1745 µg/L after the 4th course of chemotherapy.

This article was accepted: 29 July 2007
Corresponding Author: Teh Siao Hean, Department of Paediatrics, Hospital Umum Sarawak, Kuching, Sarawak Email: tehshean@gmail.com

Table I: Serial imaging of the right adrenal mass and hepatoblastoma

Date	Age	Mode of Imaging	Size of right adrenal mass	Size of hepatoblastoma
20/2/06	Day 23	Ultrasound	1.8cm x1.8cm x1.9 cm	Absent
9/3/06	Day 40	Computerised Tomography (CT)	1.5cm x 1.7cm	Absent
12/4/06	11 weeks	Magnetic Resonance Imaging (MRI)	1.1cm x 1.1cm x 1.0cm	4cm x 4cm
18/4/06	3 months	Biopsy of liver mass		
26/4/06	3 months	Chemotherapy started		
10/5/06	4 months	Ultrasound	1.7cm x 1.4cm x 2.0 cm	1.8cm x 3.2cm x 3.2cm
22/5/06	4 months	Ultrasound	1cm	2.5cm x 2.2cm
14/6/06	5 months	64-slice High Resolution Computerized Tomography	0.9cm x 1.1cm x 1.4 cm	2.5cm x 1.7cm x 1.4cm
28/6/06	5 months	Surgical Resection of Hepatoblastoma		

Intraoperatively a well encapsulated liver tumour measuring 3cmx 3cm x4 cm was completely resected. The right adrenal gland was inspected on table and noted to be grossly normal and hence was left in situ. Histopathological examination of the resected tumour revealed no residual malignancy and had features suggestive of cavernous haemangioma. Following surgery, he had completed another two courses of chemotherapy and is currently under follow up. His latest alpha-foetoprotein at two months post treatment was 98.58 µg/L.

DISCUSSION

Beckwith-Wiedemann Syndrome (BWS) is a disorder of growth characterized by neonatal macrosomia, macroglossia, visceromegaly, hypoglycaemia, ear creases/pits and exomphalos. It is well known that children with BWS have a higher risk of Wilm's tumour, hepatoblastoma and neuroblastoma. Hemihypertrophy, partial or complete, was noted in 12.5% of cases but in more than 49% of the children with neoplasms[1]. Data reported from the National Cancer Institute's Beckwith-Wiedemann support group indicate a relative risk of hepatoblastoma as 2,280, higher than that for other embryonal tumors, including Wilms' tumor. The relative risk for all cancers in this group was 676[2].

Our patient had an early abdominal ultrasound which detected a right adrenal cyst and subsequently a hepatoblatoma in the 3rd month of life. The earliest reported hepatoblastoma in the BWS in the above-mentioned National Cancer Institute's BWS registry was at four months of age, while the youngest patient with Wilms' tumour was at two months of age. Early detection of hepatoblastoma would improve outcome as the prognosis depends on the resectability of the tumour. Stages I and II have a 91% five-year event-free survival as compared to Stages III and IV with 64% and 25% five-year event-free survival, respectively. Raised alpha foetoprotein in the presence of a liver mass in BWS strongly suggests hepatoblastoma[3]. Alpha foetoprotein levels by itself may not be useful for screening due to the high normal levels in infancy. In this patient it appears that the cavernous haemangioma had coexisted within the tumour mass of the hepatoblastoma. All the various forms of imaging in the prior workup did not reveal any features of haemangioma. We have not come across any instance of a hepatoblastoma with concomitant hepatic cavernous haemangioma in BWS in the literature.

Primary adrenocortical tumours are rare in children. A case of neonatal adrenal cysts in a BWS patient which subsequently developed into a virilizing adrenal tumour was reported by Sbragia-Neto *et al*[4]. In our patient, the right adrenal appeared normal intraoperatively. We intend to follow up on this radiologically and clinically for signs of virilization.

Infants with BWS should be screened for hepatoblastoma as early as the third month of life with abdominal ultrasounds at three monthly intervals. Alpha foetoprotein levels if elevated beyond the norm for age would warrant a biopsy for histological confirmation. Early treatment and total resection of hepatoblastoma has an excellent prognosis.

REFERENCES

1. Wiedemann HR. Tumours and hemihypertrophy associated with Wiedemann-Beckwith syndrome [Letter]. Eur J Pediatr 1983; 141: 129.
2. DeBaun MR, Tucker MA. Risk of cancer during the first four years of life in children from the Beckwith-Wiedemann Syndrome Registry. J Pediatr 1998; 132: 398-400.
3. Clericuzio, Carol L. MD, Chen, Emily MD, PhD; McNeil, Dawn Elizabeth *et al*: Serum [alpha]-fetoprotein screening for hepatoblastoma in children with Beckwith-Wiedemann syndrome or isolated hemihyperplasia
4. Sbragia-Neto L, Melo-Filho AA, Guerra-Junior G, Valente de Lemos Marini SH, Baptista MT, Sabino de Matos PS. Goncalves de Oliveira-Filho A. Bustorff-Silva JM. Beckwith-Wiedemann syndrome and virilizing cortical adrenal tumor in a child. Journal of Pediatric Surgery. 2000; 35(8): 1269-71.