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CASE REPORT

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Bilateral adrenal cystic neuroblastoma with superior vena cava syndrome and massive intracystic haemorrhage

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Abstract Bilateral cystic adrenal tumours are a rare presentation of neuroblastoma. Intratumoural haemorrhage is a frequent finding in neuroblastoma, but is rarely symptomatic. We present an 11-month-old girl with predominantly cystic bilateral neuroblastomas and distant lymph-node metastasis. Massive intracystic haemorrhage and superior vena cava (SVC) syndrome were ominous prognostic factors, leading to death. Large tumours with intracystic haemorrhage might require a conservative approach.

Keywords Adrenal neuroblastoma · Cyst · Haemorrhage · Superior vena cava syndrome · Ultrasound · CT · Child

Introduction

Neuroblastoma is the most common malignant tumour in infancy, with 25% of cases arising in the adrenal glands [1, 2]. Bilateral primary adrenal neuroblastoma is rare; most of the patients are less than 1 year of age and generally have a good prognosis [3-5]. Cystic neuroblastoma is an unusual variant [6-9], and only two cases of bilateral cystic neuroblastoma have been described [10, 11]. Massive haemorrhage into the adrenal tumour is uncommon beyond the neonatal period and is rarely symptomatic [12]. We report an 11-month-old girl with bilateral adrenal cystic neuroblastoma, distant lymph node metastasis and massive intratumoural haemorrhage.

Case report

An 11-month-old girl was admitted to hospital with marked abdominal distension. On physical examination, the patient was pale and a large left-sided abdominal mass was palpable. Ultrasonography of the abdomen demonstrated a 10×9×5 cm, oval cystic mass on the upper pole of the left kidney, and 2×2×3 cm, thick-walled cystic mass on the upper pole of the right kidney

(Fig. 1). CT showed displacement of the left kidney inferomedially and bowing of the splenic vein anteriorly because of a huge left adrenal mass. The mass had a large cystic component and showed no contrast enhancement. A further mass lesion was detected in the region of the right adrenal with similar CT findings. Multiple lymph nodes in the coeliac and para-aortic areas encircling the aorta, IVC and renal veins were also seen (Fig. 2). A chest radiograph revealed mediastinal enlargement and chest CT demonstrated multiple conglomerate mediastinal lymph nodes in the paratracheal, perihilar and subcarinal regions. The superior vena cava (SVC) was narrowed and displaced anteriorly. Some segmental atelectasis and pleural effusion were also detected in the right lower lobe as a result of bronchial compression (Fig. 3). ^{123}I-metaiodobenzylguanidine (mIBG) scintigraphy was not available.

Laboratory investigations showed haemoglobin 5 g/dl, urine vanillylmandelic acid (VMA) 0.45 mg/day (normal <9.8 mg/day), prothrombin time 14.6 s (NR 10–15), activated partial thromboplastin time 26 s (NR

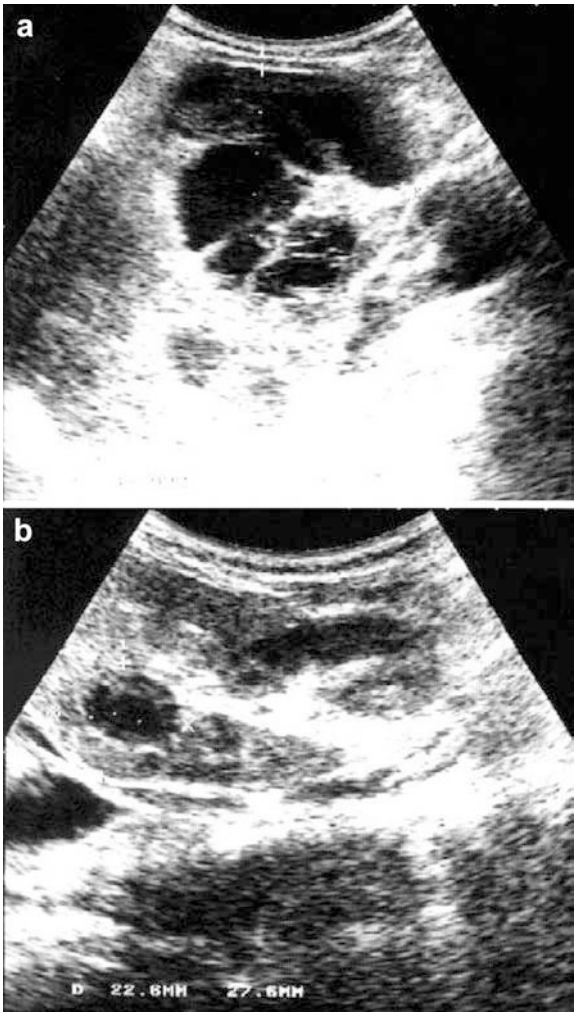

Fig. 1a, b Abdominal US. **a** A mass above the left kidney containing large cystic and solid components and septa. **b** A thick-walled cystic mass on the right side

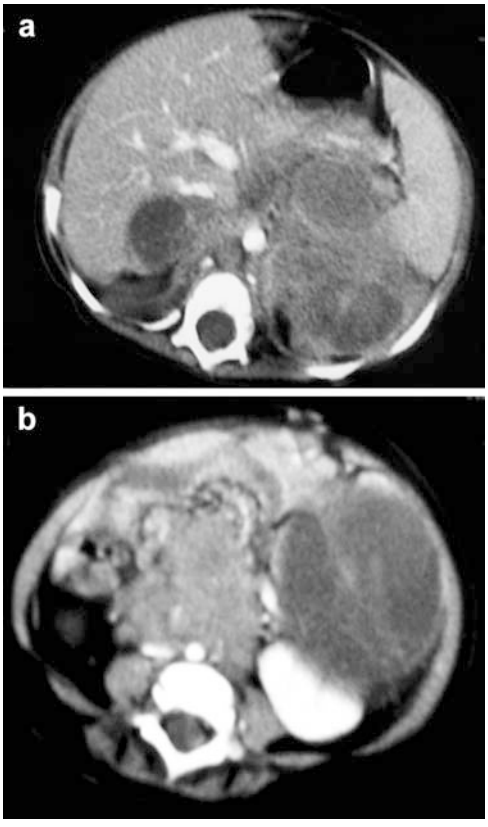

Fig. 2a, b Contrast-enhanced CT **a** at the level of the adrenals showing bilateral cystic adrenal masses and **b** more caudally, showing lymphadenopathy surrounding the aorta and a large cystic mass displacing the left kidney

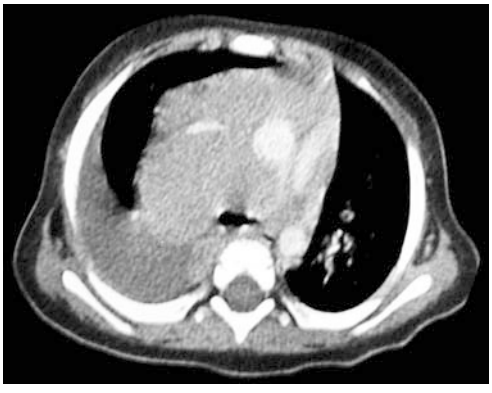

Fig. 3 CT demonstrating lymphadenopathy around the mediastinal vessels. The SVC is compressed. A pleural effusion is seen in the right thoracic cavity

26–36), serum LDH 2,285 U/l, serum ferritin 14.4 mg/dl (NR 13–150) and normal serum electrolytes, BUN, creatinine, SGOT and SGPT. Bone-marrow aspiration and biopsy showed no tumour infiltration.

Trucut biopsy was thought to be impossible because of a very thin capsule containing cystic material. We decided to do an incision biopsy of the cystic mass instead of Trucut biopsy because of the risk of rupture of the mass wall. At surgery, the wall of the tumour was very thin, and the tumour was full of haemorrhagic material. The para-aortic lymph nodes detected by abdominal CT were covered by the huge neuroblastoma mass and were not accessible and could not be biopsied. After harvesting an incision biopsy of the solid component of the wall, the extraordinarily vascularized surface of the tumour began to bleed. In spite of applying artificial bleed control material into the tumour and coagulating the bleeding surface, the bleeding was only partially controlled. The patient died of haemorrhagic shock despite massive blood transfusion. Histopathological examination revealed poorly differentiated neuroblastoma.

Discussion

Neuroblastoma is the most common malignant solid tumour in infancy [1, 2]. Bilateral adrenal neuroblastoma occurs in 10% of cases and may be due to metastasis or a second primary tumour [3, 11, 12]. Cystic neuroblastoma is located, almost exclusively, in the adrenal gland. It is diagnosed earlier than the solid form, rarely presents with metastases, and has a more benign course than solid neuroblastoma. VMA and homovanillic acid (HVA) levels are generally normal and gross surgical resection of tumour has been accomplished in the majority of cases [8]. In our case, there was distant lymph-node metastasis (mediastinum) despite the bilateral and cystic nature of the tumour and the age of the patient; stage IV neuroblastoma was considered. Metabolic activity such as increased secretion of catecholamines and/or mIBG uptake does not exclude the possibility of tumours such as ganglioneuroma or ganglioneuroblastoma [13], and the definitive diagnosis of neuroblastoma can be made if there is an unequivocal pathological diagnosis from tumour tissue or bone-marrow involvement and increased urinary catecholamine metabolites [14]. The findings were not sufficient for the diagnosis of neuroblastoma in our case and a tissue diagnosis was required. The low initial haemoglobin level and radiological findings suggested intracystic haemorrhage. Although spontaneous rupture and intratumoural haemorrhage is a recognized feature of neonatal neuroblastoma, massive life-threatening haemorrhage is known to be rare, especially beyond the neonatal period [11, 12, 15-17]. We thought that Trucut biopsy would not be appropriate because of the high risk for haemoperitoneum. Open biopsy from the solid component of the cystic tumour with direct control of haemorrhage was proposed, but unfortunately it was impossible to stop the bleeding.

In infancy, neuroblastoma is a rare cause of SVC syndrome [18, 19], which is known to cause cardiovascular and respiratory problems during general anaesthesia [20]. When our patient suffered from massive haemorrhage during surgery, marked hypotension developed, resulting in further cardiovascular distress and aggravating the SVC syndrome. The reduction of the lung volume and venous return of the heart associated with general anaesthesia may have contributed to the cardiovascular collapse of the patient.

In conclusion, the possibility of massive haemorrhage should always be considered in patients with a large cystic tumour and anaemia. Associated abnormalities, such as SVC syndrome in our case, further increase surgical mortality and morbidity in tumours with intracystic haemorrhage. In these cases, biopsy may be too hazardous and a more conservative approach might be required, such as preoperative chemotherapy or radiotherapy without an initial definitive diagnosis.

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