**CASE REPORT**

**Ganglioneuroblastoma presenting as dilated cardiomyopathy**

Y H Lee, H D Lee, Y A Lee, Y S Lee, J A Jung, G G Hwang, G W Jung, D W Kim, M S Roh

**We report an unusual presentation of ganglioneuroblastoma with features of dilated cardiomyopathy in a 22 month old girl. She was admitted with cardiomegaly; during echocardiography a suspicious abdominal mass was detected by chance. Further imaging studies, including abdominal ultrasonography and spiral computed tomography, revealed a solid mass originating in the right adrenal gland.**

**Abbreviations:** ALT, alanine aminotransferase; AST, aspartate aminotransferase; CK, creatinine kinase; CPK, creatine phosphokinase; CT, computed tomography; HVA, homovanillic acid; LDH, lactate dehydrogenase; NSE, neurone specific enolase; VMA, vanillylmandelic acid.

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**CASE REPORT**

A 22 month old girl with a negative past medical history was admitted to Dong-A University Hospital, Pusan, Korea for evaluation of cardiomegaly. For seven days prior to admission, she had been treated for fever and cough at a private clinic. The day before admission she developed dyspnoea. Chest radiography showed an enlarged heart and she was referred to our hospital.

On admission, she weighed 10.5 kg (10–25th centile) and was slightly dyspnoeic. Blood pressure was 140/90 mm Hg, pulse rate 138 beats/min, and respiratory rate 54/min. Physical examination revealed coarse breathing sounds and regular heart beat without murmur. The liver was palpated at 4 cm below the right costal margin and no mass was palpated on the abdomen. Chest x-ray examination showed moderate cardiomegaly (cardiothoracic ratio: 0.63) with increased pulmonary vascular marking. Electrocardiography was unremarkable except for sinus tachycardia; echocardiography revealed a dilated left ventricle (left ventricular end systolic/diastolic dimension: 32/36 mm) with poor contractility (ejection fraction 27%, fractional shortening 12%), suggesting dilated cardiomyopathy (fig 1).

**Figure 1** M-mode echocardiogram showing severely dilated (left ventricular end systolic/diastolic dimension: 32/36 mm) and poorly contracted left ventricle (fractional shortening 12%, ejection fraction 27%).

Results of laboratory blood studies were as follows: aspartate aminotransferase (AST) 536 IU, alanine aminotransferase (ALT) 390 IU, creatine phosphokinase (CPK) 675 U/l (normal 30–170), CK-MB 155 U/l (normal 0–16), lactate dehydrogenase (LDH) 1533 U/l (normal 120–520), troponin-I 4.49 ng/ml (normal <0.5), which are also compatible with cardiomyopathy.

During echocardiography, a suspicious abdominal mass was detected by chance. Further imaging studies, including abdominal ultrasonography and a spiral CT scan, were performed (fig 2). These showed a 5×5×6 cm solid mass originating in the right adrenal gland, which suggested neuroblastoma. Metabolic studies for catecholamine secreting tumours revealed increased serum neuron specific enolase (NSE 73.16 ng/ml; normal <12) and increased urinary vanillylmandelic acid (VMA 61.4 mg/day; normal 1.4–8.8) and homovanillic acid (HVA 30.3 mg/day; normal 0–9). Bone marrow examination was normal without tumour cells.

She was given dopamine, captopril, L-carnitine, diuretics, and amrinone (phosphodiesterase inhibitor) for cardiomyopathy, and her clinical condition and laboratory findings gradually improved over the next two weeks. The tumour originating in the right adrenal gland was then carefully exposed. The margin of the tumour was well demarcated and easily removed, sparing the right kidney. The mass was 8.5×8.0×6.0 cm in dimension and 206 g in weight. On section, the mass was a brownish gray solid with multifocal haemorrhagic foci and calcific spots (fig 3). Microscopic findings revealed focal aggregation of undifferentiated small round cells with hyperchromatic nuclei and differentiated small round cells with hyperchromatic nuclei and...
DISCUSSION

Ganglioneuroblastoma is a heterogeneous group of tumours with histopathological features spanning the extremes of maturation represented by neuroblastoma and ganglioneuroma. The signs and symptoms of neuroblastoma reflect the location of primary, regional, and metastatic disease. Since most primary tumours occur within the abdomen, sudden enlargement of the abdomen or a palpable mass with or without compressing signs are the usual presenting signs of neuroblastoma. Primary thoracic tumours are often diagnosed coincidentally when chest radiographs are performed to evaluate patients for trauma or infectious disease. Several classic signs and symptoms such as proptosis, bone pain, and bone marrow failure, have been associated with metastatic neuroblastoma. Para-neoplastic signs such as opsomyoclonus, hypertension, and intractable secretory diarrhoea have been associated with both localised and disseminated neuroblastoma; these are also manifestations of tumour secretion of peptide hormones.2,3 Most tumours secreting peptide hormones are mature histologically (ganglioneuroblastoma, ganglioneuroma), and these patients almost always achieve a favourable outcome.4

Dilated cardiomyopathy is a disease characterised by impairment of the systolic function, with progressive ventricular dilation. The causes of dilated cardiomyopathy are quite variable and acute infectious myocarditis is the most common. Among the rare well defined causes, catecholamine cardiomyopathy has been reported rarely.1,5 Although catecholamine secretion from a tumour could also cause hypertension, resulting in cardiomyopathy,5 our patient had hypertension only at presentation and this was easily controlled. Although we could not rule out a viral aetiology, something more than a simple temporal relation was suggested because complete normalisation of laboratory findings for dilated cardiomyopathy and neuroblastoma was associated with removal of the tumour mass.

Supportive treatment for cardiomyopathy with l-carnitine, diuretics, and amrinone (phosphodiesterase inhibitor) is necessary for gradual improvement before and after surgery, but complete normalisation could be achieved with surgery. The rapid reversal of cardiac function before surgery, as occurred in our patient, may be associated with an initial reversible stage and the use of captopril, which has a direct effect on the growth and activity of neuroblastomas.5 It is also important to control hypertension as soon as possible in patients with catecholamine cardiomyopathy in order to improve cardiac function and facilitate surgical intervention for diagnosis and treatment.

The prognosis of dilated cardiomyopathy depends on the underlying disease, if any, and idiopathic dilated cardiomyopathy has a poor prognosis. Thus, it may be necessary to search for underlying diseases and measure levels of catecholamines in dilated cardiomyopathy with unknown aetiology.

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