Short reports

Late functioning adrenocortical carcinoma in a 5-year-old girl

Adrenocortical carcinoma is a rare form of malignancy in childhood and its incidence has not been accurately recorded. The mode of presentation is usually the development of signs of virilisation or Cushing's syndrome. Very rare feminising tumours and occasional nonfunctioning ones have been reported (Lewinsky et al., 1974). The tumour may present in association with hemihypertrophy, urinary tract abnormalities, and other anomalies (Fraumeni and Miller, 1967). We present the case of a 5-year-old girl with a late functioning tumour, and an unusual presentation.

Case report

A 5-year-old girl presented in July 1976 with paraplegia of acute onset. There was associated retention of urine and a sensory level at D8. She had previously been a healthy child apart from having gluten-sensitive enteropathy which was diagnosed in 1972 and more recently she had developed recurrent abdominal pain and occasional nocturnal enuresis.

Plain x-rays of her dorsal spine showed a collapse of the vertebral body at D7 with a associated large paravertebral mass. At operation extradural tumour was found compressing the spinal cord and a laminectomy and biopsy were performed. This was initially reported as having the characteristics of a neuroblastoma. She was treated with radiotherapy and chemotherapy (vincristine, actinomycin D, and cyclophosphamide), after which she made a full recovery with return of normal motor function.

The child came under our care in December 1976. Chemotherapy was continued. The biopsy slides were reviewed and it was felt that they were more compatible with a primary tumour of carcinomatous histology, probably arising in the adrenal cortex. Blood levels of electrolytes, calcium, and glucose, and urinary catecholamine excretion were all normal. Intravenous pyelography and skeletal survey showed no abnormality apart from the vertebral lesion.

Over the following 4 months she twice developed an acute paraplegia. On each occasion cisternal myelography showed obstruction to the flow of Mydil and operative decompression was attempted. On the first occasion she made a complete recovery postoperatively, but after the second episode she unfortunately remained paraplegic. The major part of the tumour was at this time found to be anterior to the vertebral bodies. Further review of the histology confirmed the suspicion of an adrenocortical carcinoma as the primary tumour. There was still no clinical or radiological evidence of this and no evidence of increased production of oxo- or oxogenic steroids. The tumour was at this time enlarging radiologically.

In April 1977, 10 months after the onset of her illness, she rapidly developed plethora, coarsening of the skin with acne, trunkal obesity, and progressive virilisation with pubic and axillary hair. Urinary steroid levels were as shown in the Table, first estimation. The oxosteroid and oxogenic steroid levels rose further over the next week, second estimation. She then developed multiple lung metastases of cannonball type with an associated pleural effusion and died suddenly. Necropsy was not permitted.

Table Results of biochemical tests

<table>
<thead>
<tr>
<th></th>
<th>First estimation</th>
<th>Second estimation</th>
<th>Normal range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Urinary 17-oxosteroids (nmol/24 h)</td>
<td>73</td>
<td>118</td>
<td>0-5-6.9</td>
</tr>
<tr>
<td>Urinary 17-oxogenic steroids (nmol/24 h)</td>
<td>13</td>
<td>17</td>
<td>13-9-34.7</td>
</tr>
<tr>
<td>Plasma testosterone (nmol/l)</td>
<td>4.9</td>
<td>4.9</td>
<td>&lt;1-0</td>
</tr>
<tr>
<td>Midnight plasma cortisol (nmol/l)</td>
<td>37</td>
<td>133</td>
<td>50-120</td>
</tr>
<tr>
<td>9.00 a.m. plasma cortisol (nmol/l)</td>
<td>168</td>
<td>203</td>
<td>248-607</td>
</tr>
</tbody>
</table>

Urinary steroids and plasma testosterone estimated at onset of virilisation and 3 weeks later showing progressive rise in levels. Cortisol levels show reverse of diurnal variation.

Conversion: SI to traditional units—17-oxosteroids and 17-oxogenic steroids: 1 nmol/24 h = 0.288 mg/24 h. Testosterone: 1 μmol/l = 20.88 ng/ml. Cortisol: 1 nmol/l = 0.036 μg/100 ml.

Discussion

Adrenocortical carcinoma is a rare tumour in childhood. In one large review (Hutter and Kayhoe, 1966) 40 cases presenting under the age of 20 years were reported out of a total of 186. The mean age was 36 years with a range of 6 months to 72 years and the female to male ratio was 2:1. The majority of cases in children manifest with signs and symptoms of adrenal hypercorticism, and a neurological...
presentation is very rare. In Hutter and Kayhoe’s series there were only 9 nonfunctioning tumours.

Dumb-bell tumours due to neuroblastoma can cause an acute onset paraplegia, but they are not usually associated with any vertebral collapse. The bony lesions seen in disseminated neuroblastoma are rarely single and are more frequent in the limb bones and calvarium. Metastatic adrenocortical carcinoma in the spinal extradural space has rarely been described (Lipsett et al., 1963). Vertebral metastases are also very rare, occurring in only 7 of Hutter and Kayhoe’s 138 patients. Metastases usually manifest the same hormonal pattern as the primary tumour but a few cases have been reported in which functioning tumours recurred as non-functioning types (Rapaport et al., 1952).

The absence of any clinical or radiological evidence of primary adrenocortical tumour is very unusual, but in the absence of a necropsy we were unable to rule out a small tumour which was not causing renal displacement. Aortography, renography (Craig et al., 1975), and adrenal scintigraphy (Jorgensen et al., 1975) might have elucidated this problem but rapid deterioration in the patient’s clinical condition did not allow their performance.

The response of this tumour to radiation and chemotherapy is much better than in adult tumours of the same type and we have previously reported survival of metastatic cases (Stewart et al., 1974). Initial response to radiotherapy and chemotherapy was demonstrated here.

Summary

A 5-year-old with adrenocortical carcinoma presented with acute paraplegia. The tumour was initially nonfunctioning but finally showed rapid dissemination and the patient then developed Cushingoid features and virilisation.

References


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Unusual case of adrenal cortical carcinoma in a female infant

A 44-month-old girl remains well 39 months after removal of a left adrenal cortical carcinoma. The clinical presentation of this tumour differed from those previously reported.

Case report

A girl weighing 2480 g was born at term to a 27-year-old gravida 2 primiparous mother. Growth progressed at the 3rd centile. At 3 months transient bloody stools resolved after change of milk formula. At 4½ months a left abdominal mass was noted after a 2-week history of weight loss, irritability, and increasing abdominal girth. Positive physical findings included blood pressure 190/140 mmHg and a tender left abdominal mass filling the entire side of the abdomen. Congenital anomalies, hirsutism, Cushing’s syndrome, and inappropriate feminisation, the most commonly reported presentations of this tumour in this age group and during childhood, were absent (Reidel, 1952; Knight et al., 1960; Bacon and Lowrey, 1965; Cooper et al., 1967; Fraumeni and Miller, 1967; Kenny et al., 1968; Gilbert and Cleveland, 1970; Stewart et al., 1974). Initial clinical impression was either neuroblastoma or nephroblastoma.

Preoperative investigation could not be done due to the sudden onset of respiratory distress secondary to rapid, progressive expansion of the abdominal mass. At surgery, a 10 × 18 × 10 cm haemorrhagic mass occupied the entire left retroperitoneum, encasing the left kidney and invaded by direct extension the mesocolon at the splenic flexure. Microscopic