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


H. B. Marsden, P. Morris Jones, P. D. Lees, and I. M. Hann

Royal Manchester Children's Hospital, Pendlebury, Manchester M27 1HA.

Correspondence to Dr P. Morris Jones

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
sections showed massive tumour necrosis with marked vascularity and extensive haemorrhage. The viable portions of the tumour were arranged in sheets and columns of cells around dilated and ectatic veins. Cytoplasm of many cells contained many droplets of neutral lipid. Marked nuclear and cytoplasmic gigantism were present, suggesting hypoxic degenerative changes. Mitotic figures were numerous and abnormal. A small portion of normal adrenal cortical tissue lacked evidence of the cytomegaly associated with Beckwith's syndrome. Microscopic findings were compatible with adrenal cortical carcinoma.

Transient postoperative elevations of 24-hour 17-hydroxy- and 21-oxosteroids normalised during chemotherapy. Ortho, para' DDD (1, 1-dichloro-2-(o-chlorophenyl)-2-(p-chlorophenyl)-ethane; mitotane, an isomer of DDD which causes haemorrhagic necrosis in the dog adrenal cortex and which has been used as chemotherapy for this tumour) was not used initially because of the patient's age, anticipated difficulties from side effects of medication, and uncertain effectiveness in prolonging life (Hoffman and Mattox, 1972; Schein, 1972). Instead, a solid tumour protocol consisting of actinomycin D (9 mg/kg per dose), cyclophosphamide (7 mg/kg per dose), and 5 fluorouracil (8 mg/kg per dose) was given on 5 consecutive days every 3 weeks for 13½ months (the length of pregnancy plus time at diagnosis). She remains well 21 months after completing chemotherapy with no evidence of primary or metastatic tumour.

Discussion

Adrenal cortical carcinoma is a rare neoplasm in infancy (Cooper et al., 1967). Schein (1972) reports that most cases have presented with Cushing's syndrome, hirsutism, precocious puberty or rarely inappropriate feminisation since these tumours may be endocrinologically active. Associated congenital anomalies and concurrent central nervous system and hepatic tumours have also been reported (Fraumeni and Miller, 1967). Any of the above may favour early diagnosis, before metastatic disease has occurred. Nonfunctioning tumours have also been described, and these cases may present with signs of metastatic disease, thus indicating a poorer outlook (Knight et al., 1960).

While the curative value of surgery in the absence of metastatic disease is proven (Cooper et al., 1967), the role of effective chemotherapy after surgery remains controversial. Ortho, para' DDD has been advocated, but its absolute value is unproven (Hoffman and Mattox, 1972). In addition, resulting adrenal insufficiency may be difficult to manage (Hoffman and Mattox, 1972; Schein, 1972). We therefore elected to use actinomycin D, cyclophosphamide, and 5-fluorouracil, a combination which has proved successful in selected epithelial tumours at the M.D. Anderson Hospital and Tumor Institute (unpublished data). Side effects from this combination were managed on an outpatient basis. While ultimate prognosis is still doubtful, the fact that transient rises of 17-hydroxy- and 21-oxosteroids disappeared during treatment suggests that this combination was successful in causing the active tumour to regress.

Our case differs from those previously reported because of the absence of clinical findings commonly associated with adrenal cortical carcinoma in this age group. Apart from hypertension, evidence of tumour hormone secretion was absent, and widespread metastatic disease was not present. The presentation thus mimicked neuroblastoma or nephroblastoma.

Summary

Adrenal cortical carcinoma in a 4½-month-old girl was treated by surgery in combination with actinomycin D, cyclophosphamide, and 5-fluorouracil given daily for 5 days every third week for 13½ months. Postoperative hypertension and raised 24-hour 17-hydroxy- and 21-oxosteroids suggested residual microscopical tumour activity. These findings resolved during chemotherapy. The patient is alive and well 22 months after completing chemotherapy. Adrenal cortical carcinoma may rarely mimic neuroblastoma or nephroblastoma when the tumour is not clinically secretory.

This case report is the combined work of Sheppard AFB Hospital and the M.D. Anderson Hospital and Tumor Institute. The views expressed herein are not necessarily those of the US Air Force.

References


Kenny, F. M.; Hashida, Y.; Askari, H. A.; Seiber, W. H.,
Loss of energy during continuous infusions of breast milk

Continuous feeding by intragastric or transpyloric milk infusion is currently popular in neonatal nurseries. We have noticed that human breast milk leaves a much greater residue in the burette and tubing at the end of an infusion than cows' milk formula. Since the residue looks fatty, we thought it worth investigating the change in total milk energy during infusions, in case appreciable amounts were being lost in discarded infusion sets.

Methods

Experiments were carried out with expressed human breast milk. This was delivered from a Metrisset burette to a series of beakers via a standard 2 m length of polyvinyl tubing, at flow rates ranging from 10–25 ml/h. During infusion the milk was kept at a room temperature representative of that found in special care baby units (30–31°C). Samples were collected for analysis at timed intervals (1–2 h) for 8–12 h. We included a well mixed sample of each milk as a base line, and also the residue in the burette and tubing at the end of the infusions, which was washed out with distilled water. The samples were of equal volume (15–25 ml), and the residue in the burette and tubing was approximately the same volume before dilution with the washings. All the samples were freeze dried. Total energy content of the dried milk was determined by bomb calorimetry (Miller and Payne, 1959). Each sample was analysed in triplicate.

Results

The results were essentially similar in each of 9 separate experiments: a decline in the energy content of the samples collected in the first 4–5 h of the infusion, followed by a rise which culminated in a peak energy value in the residual sample about 16% above that of the well mixed milk. This trend is shown in Fig. 1. The difference in energy content between the 2-hour sample and the residual sample was significant at the 2% level, using the t-test applied to paired comparison (t=2.9, P<0.02). The rate of decline in energy content of the early samples was not correlated with the rate of infusion. Since the timing of the sample with the lowest energy content ranged from 1 to 5 hours after the start of the infusion, we have compared initial energy and residual energy with the mean of the lowest energy values (Fig. 2). Mean minimum energy was 11% below that of the mixed milk, and 24% below the residual energy. These differences were highly significant using the paired t-test.

Fig. 1 Mean change (± SEM) in energy content of breast milk during infusion over 8–10 hours (9 experiments).
Unusual case of adrenal cortical carcinoma in a female infant.

E B Visconti, R W Peters, A Cangir, G L Zorn, Jr and S Fisher

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