CASE REPORTS

BILATERAL ADRENAL HYPERTROPHY IN INFANCY

BY

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In 1940 Dijkhuizen and Behr drew attention to a condition they called 'adrenal hypertrophy in infants; a new clinical entity of the neonatal period'. They described three boys and a girl, all under three weeks of age, who fell ill with severe vomiting, suggestive of pyloric stenosis or other intestinal obstruction, all of whom died. Autopsies were performed on three, and in each case there was a great bilateral enlargement of the adrenal glands, but no sign of obstruction to the intestinal tract. It was claimed that this represented a new clinical entity or syndrome: severe vomiting in early infancy, associated with bilateral adrenal hypertrophy, and progressing to a fatal issue.

Their first case was a male pseudohermaphrodite (with perineal hypospadias), aged two weeks, admitted to hospital with intractable vomiting for twenty-four hours. Diagnosed as pyloric stenosis, the child was operated upon, but no stenosis was found. The child died 'after a few weeks' during which vomiting occurred frequently. Permission for autopsy was not given. Fourteen months later this child's brother, a normal male, was brought to hospital at the age of two weeks with an exactly similar history of vomiting. This child died in twenty-four hours, no operation having been done. An autopsy was performed and revealed enormously enlarged adrenals weighing together 22 gm. (normal average = 5.25 gm.). The large adrenals had a convoluted surface 'like the gyri and sulci of the brain'. The third case was also a two-weeks-old boy with intractable vomiting, the child died and at autopsy the only abnormality was, again, enormous enlargement of the adrenals (weight 24 gm.) with an appearance exactly like that of the second case. The fourth child was a girl five weeks old, with a history of vomiting beginning at three weeks; after two months in hospital this child also died. Post mortem showed adrenals weighing 34 gm., and of a similar appearance to those of the other two children.

Dijkhuizen's and Behr's cases have been quoted at some length that they may be compared with the following:—

Clinical history

A normal male child, aged six weeks, was brought to the casualty department of

the Bristol Children's Hospital with the history that he had not taken feeds well recently and had lost weight. The child's condition had not been alarming until the previous evening when the child suddenly became extremely pale, but recovered a few minutes later. Following this transient pallor he began to vomit violently, and had continued to do so intermittently throughout the night. By the following morning his condition had become so alarming that he was brought to hospital. On examination the child was cyanosed and collapsed, the pulse imperceptible, the heart beat was slow but regular. The infant was evidently moribund; oxygen was administered, but he died a few minutes after admission.

Autopsy was performed three-and-a-half hours after death. The body was that of a fairly well nourished male infant. There was no abnormality of the external genitalia. There was no jaundice, ascites, or abdominal distension, the lips and skin were cyanosed. The abdominal, pleural and pericardial cavities were normal. The heart was of normal size, the myocardium and valves showed no

FIG. 1.

FIG. 2.

lesions. The mucous membrane of the larynx and trachea was slightly injected and both lungs showed small areas of collapse and a few petechial haemorrhages beneath the pleural surfaces. The liver was normal, the gall bladder and bile ducts appeared healthy; there was no stenosis in stomach or intestines. The appearance of the adrenals, however, was striking, and is shown in the accompanying photographs (figs. 1 and 2). They were both enormously enlarged, the left adrenal weighed 11.0 gm., the right 12.1 gm.—a combined weight of 23.1 gm. as compared with the average combined weight at this age of 5.25 gm. In the gross they showed an exaggerated lobulated structure, the surface being thrown up into folds with deep crevices between them, exactly as described by Dijkhuizen and Behr as resembling 'gyri and sulci of the brain'. The kidneys, pancreas, spleen, pituitary and testes were all examined in the gross and appeared normal. The thymus, however, was smaller than usual, weighing 8.1 gm. as against the average normal weight for this age of 13.5 gm. The thyroid weighed 5.1 gm.

Histology. **ADRENALS.** The whole cortex was wider than normal due to an increase in number, rather than of size, of the cortical cells. The zona glomerulosa was for the most part poorly defined, but the architecture of the zona fasciculata was normal, except that the number of cells was greater than usual. The zona reticularis was of usual width but hyperaemic. The cytoplasm of the cortical cells stained darkly and the nuclei also were

small and dark. No mitoses were seen. The medulla was broader than normal, with many small pale cells with round vesicular nuclei arranged in irregular groups.

THYMUS. The appearance of the gland was normal for this age.

THYROID. The acini are small and only a few contained a little poorly staining colloid.

PARATHYROID appeared normal.

SPLEEN showed no change.

PANCREAS. Both islets and acini were normal.

LIVER. There was considerable cloudy swelling of the hepatic parenchyma particularly in the central cell of the lobules. The sinusoids were congested. Some of the bile canaliculi contained small masses of bile.

KIDNEY. The glomeruli were normal. Many of the proximal convoluted tubules show marked hyaline droplet degeneration of the tubule cells.

TESTIS AND EPIDIDYMIS. Both appeared normal for this age. The interstitial cells of the testis appeared normal in size and number.

LUNGS. There were large areas of recent collapse but no evidence of infection.

Discussion

This is the case of a six-weeks-old, apparently normal male child, who died of a rapidly fatal disease, characterized by severe vomiting, and also presenting signs suggestive of cardiovascular disturbance (the attack of extreme pallor which ushered

in the disease) and of respiratory involvement (the collapse of lungs and the severe cyanosis).

At autopsy there were two abnormalities, the huge adrenals and the relatively small thymus.

The nature of this condition is obscure. Dijkhuizen and Behr in their original paper collected references to seven other cases of adrenal hypertrophy in infancy which seemed to resemble their own. In a search of the literature two other cases of adrenal hypertrophy in infancy have been discovered to add to these, Fraser's and Dickson's case described by Berry Hart (1914), and Evans's and Sheldon's case in 1937.

There appear to be two classes of children with bilateral adrenal hypertrophy; in the first class we may put children who exhibit features of pseudohermaphroditism; they may be male or female pseudohermaphrodites, and often die with severe, even projectile, vomiting.

The second class of children have a similar history to that of the child described in detail above. They are normal children, more commonly male than female, who in early infancy, usually between three

and six weeks of age, are taken ill with severe vomiting and ultimately die. The vomiting in this disease is thus common to both groups. What the underlying nature of the disease may be is quite unknown.

Summary

A case of bilateral adrenal hypertrophy in an infant is reported.

The condition compared with previous reports in the literature and two other references to the condition are added.

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