Paediatric Pathology Society

Fragments of the anterior pituitary of young rats were cultured in contact with fragments of the adrenal cortex of newborn and fetal rats, and the corticosteroids present in the medium were estimated. The results of these experiments showed that the 17-day-old fetal rat adrenal can react to corticotrophic hormones.

Effects of Chromosomal Anomalies on Fetal Development. D. I. Rushton (Department of Pathology, University of Birmingham).


Solitary Rhabdomyoma of the Heart. J. Hakosalo, O. Rasanen, and F. Stenback (Oulu, Finland). The patient, a boy, had slight postnatal asphyxia, but made normal progress until at 7 months of age there was a 7-day episode of convulsions. At that time the first chest x-rays and ECGs were taken. Left ventricular enlargement with an abnormal, egg-shaped, cardiac contour was seen. The ECG showed left bundle-branch block, which was a constant finding for the rest of the patient’s life. Physical examination of the heart, and physical and mental development were normal. At the age of 4-7 years the boy contracted pneumonia, followed by convulsions and respiratory arrest. Some days later he died, after a cardiac arrest. At necropsy no tuberous sclerosis or other malformations were found. In the lateral wall of the left ventricle, at the apex of the heart, a spherical, pale, spongy, and sharply-bordered tumour was found, measuring 3 × 3 × 4 cm. The histological sections presented a typical picture of a rhabdomyoma of the heart. In this case three unusual features of the disease were evident: the tumour was single, the disease was not associated with tuberous sclerosis, and the life span of the patient was quite long. A triad of an abnormal ECG, an abnormal cardiac contour, and convulsions should always arouse suspicions of a heart tumour.

Three Cases of Testicular Adenocarcinoma of Infancy. T. E. Parry (Cardiff). The following cases were reported. Case 1. A soft grey translucent tumour was removed from the scrotum of a 14-month-old boy. It had been present for 11 months. A recurrence was excised 3 months later. The child died at the age of 19 months; at necropsy the pelvis was filled with greyish translucent brain-like tissue which extended into the abdomen. There were extensive peritoneal metastases, and small secondary nodules were present in the left lung. Case 2. A 5-year-old boy was admitted in April 1960 with enlargement of the left testis which had been noticed for 6 weeks. He was otherwise well. A testicular tumour measuring 5 × 4 × 3.5 cm. and consisting of soft friable greyish material was removed. Recovery was uneventful. In June 1960 he was readmitted with acute appendicitis and an acutely inflamed appendix was removed. No evidence of metastasis was seen. He was alive and well 6 years later. Case 3. A boy aged 2-5 years presented with a swelling of the left testis in August 1967. Apart from some anaemia (Hb 9.6 g./100 ml.), nothing abnormal was found on clinical examination and the chest x-ray was normal. The tumour was excised. Examination under anaesthesia 2 months later was normal. He remained well until Christmas 1967, when he complained of pain in the abdomen and legs, and began to lose weight. By January 1968 a large mass was present in the left hypochondrium. Treatment with ‘provera’—a progesterone compound—was given, but the patient died one month later. At necropsy a large soft retroperitoneal mass was found overlying the left kidney. On section the tumour presented a greyish translucent appearance resembling neonatal brain. Secondary deposits were present in the para-aortic glands and in both lungs.

All three tumours presented similar microscopical appearances. Each showed an adenocarcinomatous pattern, with well-marked tubular and glandular areas as well as solid sheets of undifferentiated cells, many of which were vacuolated. The cells were rich in glycogen, mucin was present in many of the acinar spaces, and fat stains were weakly positive. The macroscopical features of soft homogeneous greyish translucent brain-like tissue appear to be sufficiently characteristic to suggest the correct diagnosis.

Case 1 was included—as their Case 4—in the series reported by Teoh, Steward, and Willis (1960).

REFERENCES

Lymphangioma of Bone. H. B. Marsden (Royal Manchester Children’s Hospital, Manchester).

Blast Cell Proliferation in Children with Untreated Acute Lymphoid Leukaemia. H. P. Wagner (Berne).

A Case of Dys-γ-globulinaemia. D. I. K. Evans (Booth Hall Children’s Hospital, Manchester). A girl now aged 10 years first developed atopic eczema at 3 months of age. She had had recurrent infections including three attacks of generalized herpes simplex, four attacks of pneumonia, and five attacks of pneumococcal meningitis. γ-globulin treatment was ineffectual.

The tonsils and lymph nodes were small. Circulating lymphocytes averaged 1000/cu. mm. Platelets were normal. Bone-marrow lymphocytes and plasma cells were normal. Immunoglobulins showed high IgG, high IgA, and low IgM values (e.g. IgG 3900, IgA 400 and IgM 27 mg./100 ml. serum). Anti-A titre 1/16, anti-B titre 1/8. There was no response to 0·1 mg. DNCB after a 1 mg. sensitizing dose, and no delayed hypersensitivity to candida antigen.

Two injections of TAB produced antibody titres to S. typhi H of 1/250 and S. paratyphi B H of 1/125,
but no O antibody response. Three doses of PTAP produced a normal Schick conversion and an antibody rise to between 0·01 and 0·1 A.U. per ml. Salk vaccine produced poliovirus neutralizing antibody titres to 1/16 or more, but with evidence of an impaired response.

In vitro lymphocyte transformation with PHA showed 20–25% blast cells at 3 days. No blast cell transformation occurred when the patient's lymphocytes were cultured in the presence of heat-killed pneumococci isolated from the CSF during an attack of meningitis two weeks previously: a control showed 74%. Chromosome analysis showed a normal female karyotype.

The tendency to recurrent infections associated with eczema, low serum IgM, low circulating lymphocytes, impaired delayed hypersensitivity reactions, and impaired response to polysaccharide antigens recalls the Wiskott-Aldrich syndrome (Cooper et al., 1968; Blaese et al., 1968). The immunological defect is identical, but the patient is a girl with low isohaemagglutinins and normal platelets.

REFERENCES

A Possible Case of Niemann-Pick Disease. H. J. Degenhart, H. E. Zoethout, W. C. de Bruijn and G. J. M. Hooghwinkel (Rotterdam). A 5-month-old boy was admitted to hospital with mild cyanosis, early finger clubbing, dyspnoea, and tachypnoea. A chest x-ray showed increased hilar shadows and reticulation extending into the periphery of the lung fields. There was no calcification of the adrenals. During hospitalization liver and spleen gradually increased in size. Cherry-red spots were not present in the fundi. He showed moderate general mental retardation, with poor motor development. He died at the age of 18 months. Bone-marrow biopsies showed numerous large, foamy cells with a vacuolated cytoplasm. These vacuoles by electron microscopy appeared to consist of concentric lamellae, around an electron-lucent centre. In the lamellae the unit-membrane structure could be observed. Storage of sphingomyelins at these places is likely. Liver sphingomyelin was only twice normal and amounted to 19% of the phospholipids. Liver cholesterol was increased 2–3 times. Plasma and erythrocyte lipids were normal. In the bones a highly abnormal amount of liquid fat was found: 96% triglyceride, principally triolein, 3% cholesterol.

The clinical and electron microscope features were similar to those in Niemann-Pick disease. The chemical data suggest a new syndrome.


The electron microscopic study of a biopsy of the frontal lobe of a patient suffering from this disease shows characteristic lesions in the different cell types: cyto-membranous bodies in the nerve cells, membrano-vesicular bodies in the oligodendroglia and astrocytes and pericytes, empty large vacuoles in the endothelium of the capillaries.

The intracellular localization of the ganglioside and the lysosomal origin of the different inclusions are discussed.

Neonatal Hepatitis. J. F. Saltet (Rotterdam). 5 cases of neonatal giant cell transformation are described. The first 4 are 2 pairs of sibs, one of the cases being anencephalic. The last case is the sixth child in a family, the other children being normal.

There is a morbid entity in the newborn with the following characteristic features: (1) familial appearance in sibs; (2) survival for a few days only; (3) prominent dyspnoea and cyanosis, with low blood sugar levels; (4) haemolysis may be present; (5) jaundice is absent; (6) the placenta is enlarged, probably as a result of interference with blood flow in the cirrhotic liver.

At necropsy the following features are found. The liver is smaller than normal and has a rough surface. There are large cells in the liver (containing 20 to 40 nuclei) derived from the normal liver cells. There is striking iron storage in these giant cells and in other organs such as the pancreas, thyroid, renal tubule cells, and heart muscle cells, but not in the spleen. In most cases there is an increase of islet cell tissue in the pancreas.

Only in the proved cases of virus infection is it justified to speak of giant cell hepatitis. In other cases it is better to use the term giant cell transformation.

These changes have been referred to as neonatal haemochromatosis, but in view of recent work indicating a specific defect of gastroferrin in this condition, the term is better avoided when referring to giant cell transformation in the newborn, where the lesion may well be an enzyme defect in the liver.

Recent Progress in the Knowledge of Histocytosis X. E. Baskett and C. Nezeloff (Paris). Tissues from 39 cases of histiocytosis X have been studied since 1965. They include 6 specimens of lung tissue, 4 skin biopsies, and 20 fragments of bone.

In each case, on electron microscopic study, characteristic granules were observed within the cytoplasm of the pathological cells. The granules appeared as rod-shaped profiles, 420 Å thick, composed of a central osmiophilic core and a double outer sheath. A transverse striation of 100 Å periodicity was seen on the central core.

After 6 days, tissue cultures of cosinophilic granuloma material showed the presence of large polykaryocytes. Rod-like profiles were observed as long as 3 months after the explantation.
A case of dys-gamma-globulinemia.

D. I. Evans

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