Hyperglycæmia is well recognized in many cerebral conditions such as trauma, subarachnoid hæmorrhage and tumours, but it is suggested here that the clinical picture of pallor, abdominal distension, tachycardia and sweating may be due to overactivity of the sympathetic autonomic nervous system. This view is to some extent sustained by the increased adrenaline metabolites found in the urine during the first twelve hours of a collapse period, and during the terminal phase, as compared with excretion in the resting state. The tachycardia and normotension would suggest an adrenaline rather than a noradrenaline effect.

Micropolygyria has been associated with fits and severe mental retardation, but, as far as we are aware, the picture of what appears to be episodic sympathetic autonomic overactivity has not been described.

It is of interest that adrenaline injected into the subarachnoid space in man 'produces no rise in blood pressure, but a great rise in blood sugar' (McCulloch 1964).

Acknowledgments: We are indebted to Dr Vincent Marks for the chemical catechol amine determinations.

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Dr Vincent Marks said it was impossible on the evidence to establish that adrenaline released during an 'autonomic fit' was causally related to the hyperglycæmia observed in these twins. However, it was made extremely likely by the constant association of hyperglycæmia and glycosuria with increased catechol amine excretion, both when compared with basal excretion rates in the infants themselves and with normal controls; by the known ability of minute amounts of adrenaline to produce comparable degrees of hyperglycæmia in infants (Kinsbourne & Woolf 1959, *Arch. Dis. Childh.* 34, 166) and its demonstration in one of these children; and, finally, by the absence of any other known physiological mechanism adequate to account for such rapid and profound changes in blood glucose concentration.

Dr J A Black asked whether an estimation of the level of blood pyruvic acid had been done, as this was raised in beri-beri; and whether the effect of giving thiamine had been tried. These cases were somewhat similar to two children, whom he had seen at another hospital, in whom the levels of pyruvic acid had been high but had not responded to thiamine.

Dr Wyatt replied that neither of these had been done.

Hamman-Rich Syndrome with Encephalitis, Hepatosplenomegaly, Anæmia and Thrombocytopenia

H J Liebeschuetz MRCP
(for N R Butler MD MRCP)

G P, girl, born 16.7.58

Healthy until aged 1 year 7 months. She then developed a chest infection associated with anæmia, purpura and hepatosplenomegaly.

Investigations: Hb 5 g/100 ml, reticulocytes 5%, WBC 8,000, platelets 92,000 per c.mm. ESR 10 mm in 1 hr (Wintrobe). Bone marrow hypercellular but otherwise normal. Percutaneous trans-splenic venograph normal.

As hypersplenism was suspected the spleen was removed (Mr D A Waterston). Histologically normal.

Following this, the platelet count became permanently elevated at 300,000–700,000 per c.mm; WBC 20,000–40,000 with 20% monocytes. Apart from developing transient arthritis on two occasions, she remained well until aged 3 years 6 months.

She then presented with measles, and having recovered from this was found to have signs of chronic lung disease.

Investigations: Chest X-ray showed segmental collapse and consolidation in both lower zones. Lung biopsy (Dr B Ockenden, Dr L Reid): alveolar walls thickened, mainly due to increase in reticulin and dilated capillaries. Small amount of collagen. Several collections of lymphoid tissue are also present. Appearances are those of interstitial pulmonary fibrosis, Hamman-Rich syndrome.

Her condition remained unchanged until March 1964 when she was aged 5 years 9 months. Two weeks after fracturing her right elbow she developed vomiting, severe headache and anorexia over the course of two days.

On examination: Drowsy, ill child. Bilateral papilloedema, with retinal hæmorrhages and exudates. No localizing neurological signs. Chest: rales at both bases.

She gradually recovered after ventricular drainage but was now found to be blind with bilateral optic disc atrophy, thought to be due to cerebro-macular degeneration.

Investigations: CSF protein 70 mg/100 ml; no cells. Chest X-ray (Fig 1): widespread patchy shadowing more marked on right; some pleural shadowing in right phrenic angle. Repeat lung biopsy was similar to the previous biopsy but more fibrosis and more marked infiltration with mononuclear cells. Renal, muscle and skin

biopsies all normal. Antinuclear factor negative. Thyroid cytoplasmic antibodies negative. No LE cells found in several preparations. Thyroglobulin antibody titre 1 : 5; sheep cell agglutination titre 1 : 16.

Treatment was started with prednisone. However, she developed hypertension and is now also receiving methyl dopa. Despite this her respiratory function is continuing to deteriorate.

Fig 1 Chest X-ray showing widespread patchy shadowing

Discussion

The condition of interstitial pulmonary fibrosis was originally described by Hamman & Rich (1944). Initially there is infiltration of the alveolar walls with mononuclear cells and reticulin followed later by fibrosis and scarring, leading to respiratory failure and pulmonary hypertension. The condition may occur at any age and the duration is very variable. In the patients described initially the disease was confined to the lungs. However, Rubin & Lubliner (1957), Ellman (1956) and others have shown that very similar lung lesions occur in such collagen diseases as lupus erythematosus and dermatomyositis. The fact that the lesions often respond to steroid therapy and that experimentally in rats similar lesions can be produced with anti-rat-lung serum (Read 1958) also adds support to the theory that autoimmune processes play an important part in the aetiology of this condition.

Our patient also had an attack of encephalitis causing permanent brain damage and it seems

most likely that this was an acute allergic encephalopathy. This may be one of the manifestations of lupus erythematosus (Vessasiva 1965).

The hepatosplenomegaly, anaemia and thrombocytopenia with which our patient presented originally would also favour this diagnosis (Larson 1961). However, biopsy of the spleen and kidney were normal and neither antinuclear factor nor LE cells were demonstrated. Nevertheless, she appears to have a closely related autoimmune disease.

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Congenital Hypoplastic Anæmia with Transfusion Siderosis

S M Tucker MRCPED DCH
 (for C P Pinckney MD FRCP)

K K, boy, aged 15 years

History: Seen initially at 9 weeks of age because of extreme pallor. The pregnancy and perinatal period were entirely normal, and the birth weight was 5 lb 12 oz. No neonatal bleeding; no icterus.

Investigations on admission revealed a severe anaemia (Hb 24%), with absence of reticulocytosis. Platelets were present in normal amounts and the white blood count fell within normal range. Bone marrow studies showed an erythroblastic hypoplasia with normal appearing lymphocytes, myeloid cells, and megakaryocytes.

Blood transfusions were subsequently administered at approximately six-week intervals. From 1949 until 1954, 71 transfusions were given. Examination of the marrow at frequent intervals showed no evidence of erythropoiesis, despite attempts to stimulate red cell activity by the use of various measures, which included folic acid, vitamin C, vitamin B₁₂, low oxygen tension, cobalt, ACTH, cortisone and testosterone.

In May 1953, when the boy was aged 4 years, splenectomy was performed in an effort to prolong the period between transfusions and because of evidence of hypersplenism. Haemosiderin was present in very large quantities in the spleen.

By July 1964, over 200 transfusions had been administered. This resulted in approximately