MEDICAL MEMORANDA

Addison's Disease Presenting as a Crisis in the Puerperium

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British Medical Journal, 1971, 2, 566

A case of Addison's disease presenting in the puerperium is described, and it is suggested that where maternal adrenal insufficiency is present or develops during pregnancy cortisol may be transferred from the fetus to the mother.

Case Report

A white European woman aged 31 was admitted to the Horton General Hospital, Banbury, on 2 April 1969 in the first stage of labour after a normal 39-week pregnancy. A healthy infant was delivered vaginally on 3 April and she was discharged home on the 5th. Her blood pressure at that time was 140/90. On 8 April she began to feel weak, tired, breathless on exertion, and anorectic. These symptoms persisted and increased in severity until 17 April, when she developed vomiting and recurrent syncpne. She was found to have peripheral circulatory failure, with a blood pressure of 75/50, and she was readmitted to hospital.

She had been previously healthy, and in 1967, after a straightforward pregnancy, delivered a normal infant. The puerperium was uneventful.

On examination she was stuporous, and peripheral cyanosis was noted. No pulses other than the carotids were palpable and her blood pressure was unrecordable. She was pigmented to an abnormal degree, particularly around the eyes and in the skin creases of the hands, and buccal pigmentation was pronounced. Her heart rate was 120/min. The heart sounds were normal, the lung fields were clear, and examination of other systems showed nothing contributory. Her respiration was Kussmaul in type. A diagnosis of acute adrenal insufficiency, probably due to Addison's disease, was made.

Investigations on Admission.—Haemoglobin 17·2 g/100 ml; packed cell volume 53%; W.B.C. 15,000 (neutrophils 52°, lymphocytes 44°, monocytes 2°, eosinophils 2°). Plasma electrolytes: sodium 122 mEq/l., potassium 6·4 mEq/l., chloride 86 mEq/l., bicarbonate 17 mEq/l. Blood urea 102 mg/100 ml; blood sugar 80 mg/100 ml; plasma cortisol 2 µg/100 ml (using the method described by Mattingley (1962)).


Plasma P.B.I. 10·1 µg/100 ml. T-3 resin uptake 32·7°. (normal range 25-35°). Serum antibodies by immunofluorescent technique: thyroid microsomal positive, adrenal positive, stomach parietal cell negative, smooth muscle negative, kidney negative, antinuclear factor negative. A tetracosactrin stimulation test showed basal plasma cortisol to be 1·5 µg/100 ml, with no evidence of a response to the tetracosactrin after 60 minutes. Adrenal tomography showed no evidence of adrenal calcification, and Mantoux 1:1,000 was negative.

She was treated with intravenous hydrocortisone and fluids and made an uneventful recovery. She was discharged two weeks after admission taking cortisone acetate 37·5 mg daily and 9α-fluorohydrocortisone 0·1 mg daily. Her blood pressure was 120/80. Subsequently she had another uneventful pregnancy, but she suddenly died at home two months after this pregnancy soon after developing a respiratory infection. Necropsy was not performed and I have been unable to acquire further information about events at that time.

Comment

There seems little doubt that this patient had Addison's disease of "autoallergic" type. Previous case reports indicate that the patient with untreated Addison's disease feels much improved in the latter half of pregnancy but is particularly at risk in the puerperium (Fitzpatrick, 1922; Hunt and McConahey, 1953; Plotz, 1953; Simcock, 1966). This suggests a contribution of cortisone-like substances to the mother from the fetal-placental unit. The fetal adrenal cortex can produce cortisol, probably using placental progesterone as a precursor (Villee, 1969). Though the level of free blood corticoids in the newborn is low (Klein et al., 1954) cortisol binding in the fetus is mainly to albumin (Mills et al., 1959), which has a much lower affinity for cortisol than maternal transcortin, which is an a-globulin (Daughaday, 1958). It is therefore likely that free cortisol is transferred from the fetal to the maternal circulation during the last trimester of pregnancy if maternal transcortin is not saturated. If this is so fetal cortisol production should protect a mother with adrenal insufficiency, but the withdrawal of this supply at delivery would result in a critical situation in the first week of the puerperium. The course followed by the above case lends support to this view.

References