Sickle-cell Disease with "Leather Pouch" Uterus

Brit. med. J., 1964, 1, 1360

Eisenstein, Posner, and Friedman (1956) reviewed the literature on sickle-cell disease in pregnancy and failed to find any evidence of uterine abnormality in this condition. Anderson, Went, MacIver, and Dixon (1960) found that the mean birth-weight of infants born to mothers with sickle-cell anaemia was only 5 lb. 10 oz. (2.6 kg.), while the mean birth-weight of infants of mothers with the sickle-cell haemoglobinopathy was 6 lb. 9 oz. (3 kg.). These compared with a mean birth-weight of 7 lb. 4 oz. (3.3 kg.) in infants born to normal mothers. As this difference could not be accounted for on the grounds of prematurity, they postulated that erythrocytosis of sickled red blood-cells in the utero-decidual arterioles might be responsible by bringing about a reduced chorio-decidual blood-flow and placental insufficiency.

A case of sickle-cell disease in pregnancy is described which required the performance of a caesarean hysterectomy, and in which the uterus showed marked pathological changes.

CASE REPORT

A 23-year-old woman had a previous history complicated by several hospital admissions for sickle-cell crises, and in 1953 an arthrodesis of the right hip had been performed for avascular necrosis of the head of the femur. There was no abnormality in her gynaecological history; her menstrual cycle had always been a regular one of 28 to 30 days.

In 1959 she had been delivered by caesarean section of a baby who weighed 4 lb. 3 oz. (1.9 kg.) and survived. The indication for the operation had been a severely contracted and deformed pelvis.

The patient was seen on 31 December 1962 in her second pregnancy, when she was eight weeks pregnant. Her haemoglobin at that time was 8 g.%. Pregnancy was complicated by a sickle-cell crisis at 29 weeks, for which she was admitted to hospital. During this crisis, which was of the haemolytic variety, her haemoglobin fell to 5.8 g.% and she was given a blood transfusion of two pints. Further progress was uneventful until 34 weeks, when she went into labour. In view of the severe degree of contracted pelvis and the previous caesarean section, a further caesarean section was carried out after two pints of blood had been cross-matched. A lower-segment operation was performed and a living male infant weighing 3 lb. 4 oz. (1.5 kg.) delivered. The mother was given 0.5 mg. ergometrine intravenously, but the placenta failed to separate and had to be removed manually, though no difficulty was encountered in finding a plane of cleavage. The lower segment was closed in the usual manner.

It was now noted that the uterus was of a most unusual consistency, the upper segment being extraordinarily thin and feeling like a pouch of leather. In spite of repeating the intravenous ergometrine, and making several attempts at rubbing up a contraction, this thinned-out uterus failed to contract and repeatedly filled with blood. As the patient's haemoglobin had been 6.6 g.% at the beginning of the operation it was decided to prevent any further haemorrhage by performing a hysterectomy. A transfusion of two pints of blood was started and a subtotal hysterectomy was done.

The post-operative course was complicated by a mild paralytic ileus which cleared up with gastric aspiration and intravenous fluid therapy. The haemoglobin and reticulocyte count were checked each day, as it was anticipated that the patient might have a sickle-cell crisis. However, this did not occur, the wound healed well, and the patient was discharged from the hospital with her baby on 17 July 1963.

Pathological Findings.—The specimen received in formalin was a post-partum uterus measuring 13.5 by 9 by 5 cm. There was a sutured lower segment surgical incision. The cavity contained considerable clot but no placental tissue could be identified. The

wall of the uterus had a peculiar leathery feel suggesting an unusual degree of sclerosis.

Sagittal sections of the entire uterus were cut and stained with haematoxylin and eosin, Mallory's trichrome method, and an elastic van Gieson combination. In addition selected blocks were treated routinely. The larger vessels in the subserosa and external half of the myometrium were not remarkable. The smooth muscle in the outer half was also normal, though there was possibly some excess of interstitial connective tissue. The inner half to one-third of the myometrium showed blurring of detail due to a peculiar hyaline degeneration of the myometrium. This effect was seen throughout most of the uterus, and in places the demarcation between essentially normal and abnormal myometrium was quite abrupt (see Fig.). Connective-tissue stains showed that the necrotic tissue retained its staining properties for smooth muscle but in places there was a marked increase in collagen. The necrosis was not acute as there was remarkably little inflammatory reaction. A few arterioles showed fibrinoid necrosis with a mild inflammatory reaction. Everywhere the small vessels were stuffed with sickled red blood-cells. The general impression was of a low-grade, slowly progressive necrosis of the inner myometrium associated with sickle-cell disease.

Demarcation of necrotic and viable myometrium with small venous thrombosis. (H. and E. $\times 80$.)

COMMENT

The case presented is interesting in that it illustrates that the erythrocytosis of sickled red blood-cells in the smaller uterine arterioles postulated by Anderson *et al.* (1960) can occur. What is difficult to explain in the present case is an ischaemic process sufficiently severe to cause extensive myometrial necrosis, but not severe enough to result in placental insufficiency and intra-uterine death of the foetus.

I wish to thank Mr. Colin Benjamin, F.M.T. (W.I.), for the histological preparations, and Dr. W. B. Robertson for his pathology report and the photomicrographs.

PETER CURZEN, B.Sc., M.B., M.R.C.O.G.,
Lecturer, Department of Obstetrics and Gynaecology,
University of the West Indies, Kingston, Jamaica.

REFERENCES

- Anderson, M., Went, L. N., MacIver, J. E., and Dixon, H. G. (1960). *Lancet*, 2, 516-521.
Eisenstein, M. I., Posner, A. C., and Friedman, S. (1956). *Amer. J. Obstet. Gynec.*, 72, 622.