PREGNANCY IN LEON ISRAEL SYNDROME

(A Case Report)

by

SHASHANK V. PARULEKAR

and

MINA BHATTACHARYYA

Introduction

Leon Israel syndrome is rare. There are only 6 cases reported in world literature Israel (1967). This is the 7th case, and the first with an unfavourable outcome.

Case Report

Mrs. S.D., a 30 years old woman, married for 3 years presented with primary amenorrhoea and primary sterility a year ago. Her secondary sex characteristics had developed completely by the age of 14 years. Her general examination revealed no abnormality. Her breasts were well developed (Tanner's stage 5) with no secretions; the axillary and pubic hair were normal. The external genitalia were normal, the uterus anteverted, normal sized, smooth, firm and mobile, the fornices were clear. There was no hirsutism. There was no genital outflow tract abnormality. Chest radiograph-AP view and skull radiograph lateral view were within normal limits. Serial vaginal smears showed cyclical changes, with a progesterone dominance, but no parabasal cells. Progesterone challenge test and estrogen challenge tests were both negative. Laparoscopy with dilatation and curettage was performed, which showed normal ovaries, normal uterus and normal and patent tubes. The endometrial histopathological examination showed secretory endometrium. A diagnosis of Leon Israel syndrome was made.

The patient was advised planned relations depending on the results of serial cervical mucus and serial ultrasonic scans of ovaries for follicular size. She was subsequently lost to follow up and presented with 30 weeks sized pregnancy. The fetus was in vertex presentation, FHS regular. An ultrasonic scan using multiple parameters of fetal maturity showed the fetal age to be 29-30 weeks. One week later, patient presented with the fetus in frank breech presentation. Fetal heart rate was 148/minute, regular. Tocolytic therapy failed to control preterm labour and the cervical dilation and she delivered a fresh stillborn male child weighing 1.8 kilogram vaginally at 5.30 a.m. The fetal heart sounds were regular and in the range of 140 to 150 per minute till the breech of the fetus had delivered. The assisted breech delivery had been uneventful. A loop of the umbilical cord was found around the baby's right thigh, but was loose enough and was unlikely to have caused intrapartum fetal death. The fetus showed a maturity of 35 weeks and features of intrauterine growth retardation. A necropsy of the baby was refused by the relatives. The patient had an uneventful recovery and was sent home on 5th postpartum day.

Discussion

With availability of ultrasonic scanning facilities and serum progesterone assays, ovulation can be easily diagnosed and monitored, so that the management of Leon Isreal syndrome is not difficult. However the endometrial defect which prevents normal cyclical bleeding of menstruation may also cause a defect in placentation and placental insufficiency, as was observed in the present case, though not reported so

From: Department of Obstetrics and Gynaecology, K.E.M. Hospital and Seth G.S. Medical College, Parel, Bombay 400 012.

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far in world literature. It would have been better diagnosed and managed, had the patient come for early and frequent antenatal visits as advised. The last nonstress test was performed 1 week before fetal death. Lack of continuous intrapartum electronic fetal heart rate monitoring and fetal scalp blood pH monitoring facilities were also partly contributory to the fetal death during labour.

Acknowledgements

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Reference

1. Israel, S. L.: Menstrual disorders and sterility. Fifth edition. Now York, Harper and Row, 1967, p. 269.