ADDISONS'S DISEASE IN PREGNANCY

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SUMMARY

The Authors show a rare case of Addison's disease in pregnancy, in a 27 year old patient, under cortisonic therapy. They propose the current diagnostic therapeutic orientations relating to maternal foetal risks.

INTRODUCTION

Addison's disease is a rare endocrinopathy wich strikes 0.05-0.1‰ of the entire population ([1]) and is quite exceptional in pregnancy: one case into 12,000 pregnancies, according to Mason ([2]).

In most of the cases, it is a syndrome acquired even if some congenital familiar type-forms are pointed out.

Forms of obstetric interest are essentially the idiopathic ones, almost certainly of auto-immune, origin, while tuberculous forms relating to the improved preventive and therapeutic possibilities are always rarer ([3]).

Cortisonic therapy has completely changed both maternal survival (once mortality reached 35%) and gestational ability today, once deeply compromised by the high incidence in the very rare pregnancy-forms in the course of Addison's disease ([4]). Now pregnancy is not only possible but is actually without risks if suitably followed.

A case recently observed is described both for its rarity and for the evaluation of current maternal fetal therapeutic orientations.

CLINICAL CASE

Mrs. A. G., para 2113, has been admitted to the Obstetrics and Gynecology Department on 11/18/1983, for labour, in the 38th week of pregnancy, suffering from Addison's disease.

Familial anamnesis was negative and menarche appearance happened at 13 from physiologic anamnesis, with subsequent menstruation, regular for quantity, flow and duration.

Common childish esanthems were presented remote pathological anamnesis, a tonsillectomy in 1969, a viral pneumonia in 1982, an admission to hospital in 1977 during the 1st pregnancy in the 25th amenorrheal week for Addisonial crisis. The pregnancy was resolving into spontaneous abortion and checks done at that moment, to the discovery of a cronic cortico-surrenaled insufficiency.

After a brief hospitalization period, the patient was released in healthy condition, with a consistent cortinosic therapy (Cortone 75 mg/die).

After the admission, the patient was never subjected to clinical or lab checks in spite of

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doctors' repeated solicitations and did not change the cortisonic therapy prescribed in 1977, which can be explained by her low social-cultural condition.

After that, she had two full-term pregnancies with live and viable fetuses, 2,800 and of 2,700 g weight respectively; and one terminated in a premature birth at the 30th week with a live and viable fetus of 1,240 g; the newborns are all in good healt now. The patient had physiological confinements and gave bottle feedings.

The next pathologic anamnesis (at this current admission) the patient showed advanced labour at the 38th week in apparently good general condition. At a general objective examination skin showed up normotrophic, normoelastic, dark-coloured, the mucosae were normally-supplied with blood, muscles were normotrophic-normotonic, non-palpable superficial lymphnodes, standard pulmonary and cardiocirculatory apparatus, arterial tension at 120/80, no oedema-signs on lower limbs.

Gynecological examinations showed multipara external genitals, normal vagina; the neck showed itself to be levelled with 4 cm of dilation, with clear amnyotic fluid discharge. The part presented was cephalic, presenting the vertex, at the higher strait in O.I.S.A.

After almost two hours of labour there was the birth of a female foetus, alive and viable, 2,800 g of weight, with Apgar 9 at the 1st minute. The after-birth was spontaneous with the expulsion of a placenta of 750 g, the confinement was fully normal, without fever and hypocortico adrenalism with standard genital puerperal involution. Normal dosage (cortisolemya, ACTH, 17 KS, T3, T4) and routine-examinations confirmed normal values with a good metabolic balance. The patient was discharged in good general conditions on the 8th day. The newborn was admitted, soon after the birth, to the Paeditric Department of the University for the necessary checks. It showed standard haematoclinical values and was discharged on the 8th day, after having been submitted to phototherapy for a light neonatal icterus.

DISCUSSION

In this case the AA. could only be surprised at the happy evolution both of the pregnancy and the birth, given the poor follow-up and for checking. The patient brought her pregnancy to an happy conclusion, without any complication, getting over the birth stress successfully without any adjustment of the cortisol dose.

This completely exceptional fact, which can mostly be ascribed to an eventful therapeutic balance, in an Addisonian patient's therapeutic problems and maternal and fetal risks must absolutely not be forgotten.

Maternal risks, on the one hand of a corticosteroid insufficiency, are bound to the loss of electrolytes, on the other hand to hypoglicemia for the non-increase of cortisol which prevents the hepatic dismission of glucose ([5]); a shortage due to the non accumulation of fatty acids from the organism is combined in this condition ([4, 6]). These complications are greater during the 1st trimester and decrease during the 2nd and 3rd trimesters, probably due to the production or steroidal placental hormones with light glyco-mineral corticol action ([7, 8]).

Fetal risks are strictly connected to maternal metabolic balance and are difficult to evaluate by hormonal dosages ([9]). In fact E3 dosage is not very important, because of the hesogenic inflow of corticosteroids which lead to a reduced synthesis of androgens and, particularly, of DHA into the fetus, with subsequent reduction of placental biosynthesis ([8, 9]). On the contrary, HPL values are more reliable: they tend to be lower than the normal ones, permitting a forecast of delay of the fetal growth ([10]) and must be ascribed to the reduced placental metabolic function which often shows infarcts.

Furthermore in an Addisonian patient pregnancy lasts 14 days more than normally on average ([10]). The event did not occur in this case. The fetus weight 500 g less, on average. The reason for the low weight at birth and for the protracted pregnancy is caused, according to Osler ([11]), by the maternal low glycemic rate.

For the effect of the steroid therapy during pregnancy, the possibility, although rare (1%), of fetal deformities is shown; the most frequent of all is the labiopalatoschisis ([9]). Furthermore the newborn

experiences marked hygoglycemia soon after the birth for the sudden lack of cortisol.

The birth is, of course, a moment of risk for an Addisonian patient, being a stress which may lead to an obstetric shock. Anaesthesia, analgesia or even the light emathic losses during labour can, in fact, show the reduced resistance of these patients to any kind of stress, in the case of a imperfect pharmacologic compensation.

The use of oxytocin during labour is discussed by some, asserting the possibility of intoxication by water, seeing the molecular affinity between oxytocin and adiuretin ([12]).

The birth must be completed through the vaginal duct if obstetric conditions imposing a C.G. do not exist. In this case all anaesthetic care must be put into operation to reduce surgical stress.

Breast feeding must be avoided for fear of corticosteroid passing into maternal milk ([10]) and for the occurring electrolytes loss ([12]).

Finally the exceptionality of the reported case must be once again underlined. The 4 living children of an Addisonian patient with no medical checking during her pregnancy are its evidence but the delicacy of the obstetric problems of this endocrinopathy must not be forgotten. Once again the total prognostic change due to the introduction of the cortisonic therapy must be remembered, because pregnancy was made safe and possible for these patients. But it must be always considered as a risk and calls for all checks and cares which can possibly be used today.

Particularly, therapy must be put into practice by these modalities: the substitutive therapy with glyco and mineral corticoids (acetate cortisone and hydrocortisone are mostly used) must be generally continued unchanged during pregnancy; in case of stress, infection or vomiting, the dose must be increased; in case of Addisonian crisis electrolytic balance and blood-volume must be restored by opportune administration of glucosal tinctures with the addition of KCL and hemisuccinate cortisone must be supplied i.v. for the hormonal deficit; cortisone dosage must be increased before during and after the birth.

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