

CASE REPORT

Adrenal cytomegaly associated with diaphragmatic hernia: Report of a case

BB ONG, MBBS, MPATH and KT WONG, MBBS, MRCPATH

Department of Pathology, Faculty of Medicine, University of Malaya, Kuala Lumpur, Malaysia.

Abstract

A rare case of a 22-week-old foetus with unilateral adrenal cytomegaly and left diaphragmatic hernia is reported. Typical cytomegalic cells were found focally in the left adrenal but the right adrenal was normal. There was no stigmata of the Beckwith-Wiedemann syndrome. The association of adrenal cytomegaly with various congenital malformations, the significance and possible pathogenesis of this condition is discussed.

Key words: Adrenal cytomegaly, diaphragmatic hernia.

INTRODUCTION

Adrenal cytomegaly is a rare but well-known entity first described in 1927.¹ It may occur in either one or both adrenals, and is known to be associated with the Beckwith-Wiedemann syndrome, pancreatic islet hyperplasia and cytomegaly, erythroblastosis foetalis, exposure to exogenous steroids prenatally and various congenital malformations.² We report a case of a foetus with unilateral adrenal cytomegaly associated with diaphragmatic hernia.

CASE REPORT

A 28-year-old women, gravida 4, para 3, was admitted with the complaint of per vaginal bleeding associated with abdominal pain and fever. She has no previous antenatal follow-up. Her previous 3 pregnancies were uneventful. She had no history of diabetic mellitus or hypertension. On admission, her uterus was about 22 weeks in size. An ultrasound examination showed a singleton pregnancy with femur length and biparietal diameter corresponding to about 22 weeks of gestation. After admission, the baby died in-utero and was delivered spontaneously.

Autopsy findings

The 500 g foetus had a crown-rump length of 18 cm and rump-heel length of 10.5 cm. No dysmorphic features such as macroglossia or omphalocele were noted. The umbilical cord was normal and was attached to the edge of an unremarkable placenta. There was a left diaphragmatic herniation of the intestines and

left lobe of the liver into the left thoracic cavity, displacing the left lung and heart to the right (Fig. 1). The left hemidiaphragm was absent, and the left lung was atrophic. Both the left kidney and adrenal were displaced superiorly. The combined weight of the adrenals was within normal limits. There was no evidence of visceral organomegaly.

Histological examination of the left adrenal gland showed the foetal cortex to be replaced by cells with giant nuclei and abundant eosinophilic cytoplasm (Fig. 2). Some nuclei were hyperchromatic with chromatin clumping, while others were vesicular with prominent nucleoli. Nuclear inclusions were observed in some cells. Mitoses, cellular necrosis and lymphocytic infiltration were absent. The right adrenal was normal. There was no evidence of pancreatic hyperplasia or cytomegaly, or any abnormality in the other organs.

DISCUSSION

Adrenal cytomegaly associated with diaphragmatic hernia is very rare. In a reported series of 9 cases of adrenal cytomegaly with congenital malformations, there was only one case with diaphragmatic hernia.³

The Beckwith-Wiedemann syndrome (comprising classically of umbilical hernia, macroglossia, visceral hypertrophy and adrenal cytomegaly)⁴ and other congenital malformations such as cardiovascular and gastrointestinal abnormalities, have been associated with adrenal cytomegaly.³ In addition, it may be associated with congenital intraviral infections e.g.



FIG. 1: Herniation of the liver (thin arrow) into the left hemithorax. Note the absence of macroglossia, the atrophic left lung (arrowhead), and the normal umbilical cord (broad arrow).

congenital rubella syndrome and extensive haemolytic states e.g. Rhesus isoimmunization.⁵

Adrenal cytomegaly may be unilateral or bilateral, diffuse or focal. In the present case, there was only focal involvement of the left adrenal cortex. As in previous reports, the cytomegalic cells which were typically hyperchromatic, may or may not contain nucleoli or inclusions.^{2, 3, 6} Furthermore, mitoses were generally not observed. Occasionally, surrounding necrosis and lymphocytic infiltration have been reported.³

Ultrastructurally, nuclear inclusions have been shown to be cytoplasmic intrusions sun-ounded

by a double membrane.² Others have reported that the inclusions may contain numerous organelles.⁶ Flow cytometry and image analysis have revealed increased amounts of DNA in the nuclei of cytomegalic cells.^{5, 7, 8}

Many causes of adrenal cytomegaly have been postulated. Initial hypotheses that implicated viral infections have fallen out of favour as electron microscopic studies have so far failed to demonstrate any virus.⁹ Prenatal exogenous steroid therapy as a cause appears to be unsubstantiated.¹⁰ There is no proof that the cytomegalic cells are precursors of neoplasia either.⁷

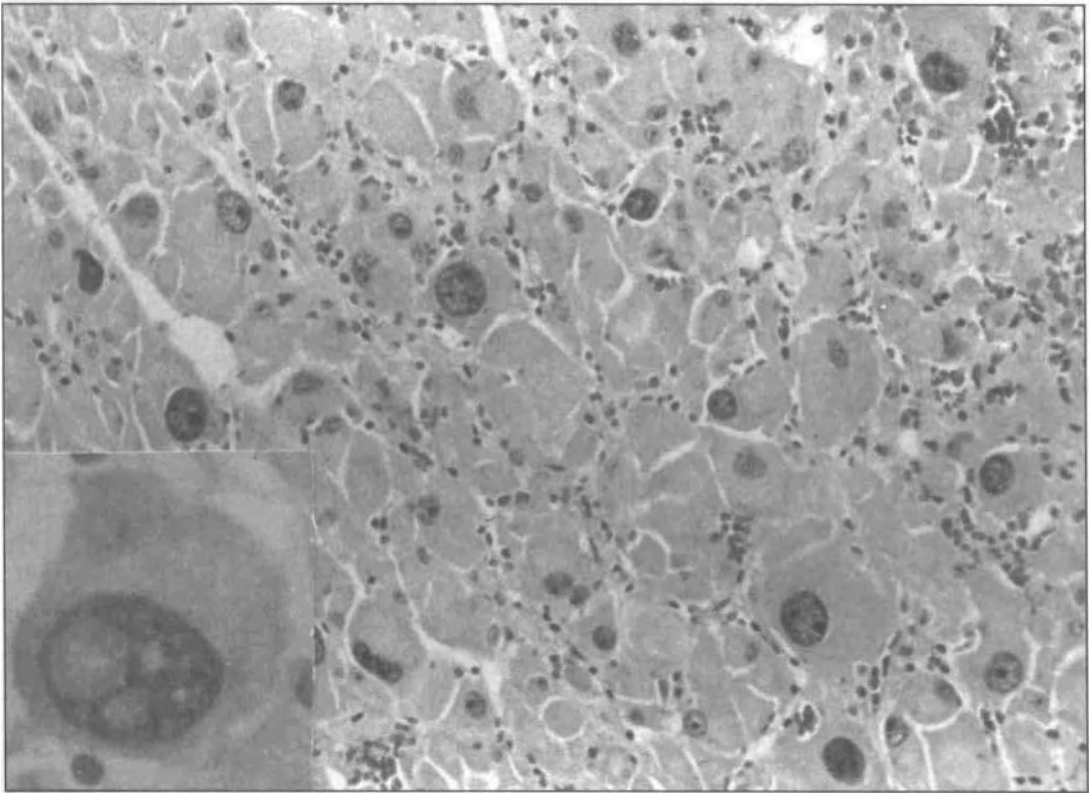


FIG. 2: Typical cytomegalic cells with giant nuclei, prominent nucleoli and abundant cytoplasm. Inset shows intranuclear inclusions. (H & E).

Based on ultrastructural and imaging studies, it is postulated that cytomegaly and polyploidy occurs when adrenocytes are provoked by some unknown stimulus to undergo incomplete mitosis. The hyperstimulated adrenocytes are unable to produce proteins such as cyclin (which controls mitosis) despite being able to produce other proteins at an accelerated rate. Consequently, there is increased nuclear content of DNA but no cell division.⁵

ACKNOWLEDGEMENT

We are grateful to the Medical Illustration Unit of Faculty of Medicine, University of Malaya for their photographic assistance.

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