BRIEF REPORTS

Hypothyroidism with Multiple Congenital Anomalies

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The features of hypothyroidism during neonatal life are usually never striking or conclusive and its diagnosis is often delayed for six to twelve weeks after birth(1). We here report the chance finding of hypothyroidism in an eighteen day old with an unusual presentation-that of very prolonged gastric atony following laparotomy. This child also had multiple congenital anomalies which have not been reported together with hypothyroidism, previously.

Case Report

A new born female child, with a birth weight of 2400 g was born by cesarean section, to a mother with a bad obstetrical history. The mother had conceived twice previously, the first was a vesicular mole which had to be evacuated and the second time she had an abortion. During the delivery of this child it was noted that her placenta was literally in contact with the peritoneum through a thinned out part of the uterus.

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The child was born with a lumbosacral kyphosis and a rough systolic murmur in the second and third left parasternal spaces. She also had a small umbilical hernia. Jaundice was noticed fortyeight hours after birth. Serum bilirubin rose upto a maximum of 25 mg/dl. on the fourth day and then began to fall by itself. When the child was transferred here, on the fifth day, the bilirubin had come down to 20 mg/dl. Her blood group was O-Rh positive, as was that of her mother. The direct Coombs test was negative.

Child was referred here on account of a suspected intestinal obstruction. She was bringing up small quantities of bilious vomitus from the second day, and the skiagram of the abdomen showed a double bubble air shadow with the rest of the abdomen empty even five days after birth. There was no gross distension of the abdomen.

After the jaundice had abated and her general condition had been built up with the help of intravenous alimentation, she was taken up for surgery on the ninth postnatal day. At laparotomy she was found to have a malrotated bowel with bands causing duodenal obstruction. The stomach and duodenum were hugely distended. The duodenojejunal junction was to the right of the midline; cecum and appendix were in the left hypochondrium; and the duodenojejunal junction was tethered to the posterior abdominal wall by a fibrous band. These were surgically corrected.

Four days after surgery the child's gastric aspirate was still copious and bilious-raising the suspicion of an associated intraluminal obstruction, missed at operation. This prompted a dye study. 5 ml. of Gastrografin was given by the nasogastric tube. A skiagram, after four hours showed all the dye in the stomach.

However, another taken twenty hours later showed the dye in the small intestine. The child was thus presumed to have persistent gastric atony on account of excessive pre-operative dilatation.

This situation persisted till the child was eighteen days old. It was then that a skiagram of the spine was taken for the associated spinal deformity. This picture showed lumbosacral hemivertebrae. In addition, the centres of ossification of the lower end of the right femur and the upper end of both tibiae had not appeared. A clinical diagnosis of hypothyroidism was thus made and L-thyroxine was started. The ileus disappeared within forty eight hours of initiating thyroxine therapy. The serum sample taken prior to starting thyroxine showed total thyroxine (T4) 9.3 mg/dl., (Normal 11 to 21 mg/dl.) Free Thyroxine Index (FTI) 9.5 mg/dl. and TSH 100 μu/ml. (abnormal >25 μu/ml.)

Discussion

This child with hypothyroidism had associated, a malrotated gut with Ladd's bands, hemivertebrae with kyphosis in the lumbosacral region and a cardiac murmur, most probably a patent ductus arteriosus. This combination has not previously been recorded together.

In a review of the subject(2) it was noted that most authors reporting cases of athyrotic cretins do not mention any associated malformation though only a few have specified that the child was normal except for the thyroid deficiency. Recording of associated malformations in all cases of hypothyroidism may aid in evaluation of the developmental disturbance leading to the thyroid anomalies.

Neel *et al.*(3) have observed one case of absence of cervical vertebra and a case of a large hemangioma of the right face combined with a large pilose nerves of the left scalp region and another case of cavernous hemangioma of the back associated with hypothyroidism.

With reference to the-spinal deformity, developmentally, the notocord, which forms the vertebral column, and the alimentary canal are separated by the mesodermal cell mass. If a connection persists between the notocord and the alimentary canal, formation of the vertebrae and discs may be disturbed and remanants of the endoderm may develop into cysts. Fallon *et al.*(4) reports such an association between mediastinal cysts of the foregut and malformation of the spine and note that although the connection between the notocord and the mediastinal cyst may have disappeared at the time of birth, an anterior cleft in the vertebral body can remain as testimony to a previous connection. The bands seen in this child with hemivertebra may also be of similar etiology. Ingalls *et al.*(5) subjected pregnant female mice to low pressure chambers from the second to the eighteenth day of gestation and found asignificant number of newborn mice had hemivertebre, fused ribs, cleft palate and cranioschisis. It is subject to speculation if the hemivertebra and other congenital anomalies seen in this child have to do with a hypoxic insult consequent to placental dysfunction as a direct result of the faulty placental attachment noted during the cesarean operation.

In the absence of conclusive evidence it would not be possible to comment categorically on the exact cardiac anomaly producing the heart murmur in the child reported here. Thyroid therapy did not seem to reduce the intensity of the murmur. Thibeault *et al.*(6) have recorded 3 cases of hypothyroidism with patent ductus arteriosus, one of whom had a surgical repair of PDA before hypothyrodism was diagnosed.

To conclude it may be worthwhile remembering hypothyroidism in the differential diagnosis in a child with prolonged ileus following neonatal intra-abdominal manipulations.

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Plasmapheresis in Acute Guillian Barre Syndrome

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Guillian Barre Syndrome is a monophasic illness from which complete or partial recovery usually occurs. It has a multifactorial etiology. Irrespective of cause it culminates in neuronal injury, via immunological mechanisms(1). Pathological changes indicate a cell mediated selective attack on nerve myelin. In vitro, the sensitized circulating lymphocytes of patients result in demyelination of peripheral nerves in tissue cultures(2). The humoral limb is also implicated and complement fixing antibodies against nerve tissue are demonstrable in almost half the patients(3). Cell free serum of patients can produce demyelination in vitro. Both myelinotoxic and precipitating antibodies have been demonstrated(4,5). Circulating immune complexes have been detected in patients. The immune complexes demonstrated contain IgG and IgM antibodies(6). Reduction of IgM serum antibodies from 50 to 30% resulted in significantly decreased myelinotoxic activity in vitro.(4) An increased binding of IgG of patients to peripheral nerve tissue was demonstrated in almost half the patients investigated. There is, therefore, evidence to implicate both humoral and cellular mechanisms in the pathogenesis of the disease.

This raises interesting therapeutic implications. Corticosteroids have proved ineffective in a recently carried out controlled trial(7). In fact, the outcome was worse. Immunosuppression with azathioprine(8) and cyclophosphamide(9) have also proved ineffective. Based on the immunological mediation in the disease, Plasmapheresis was tried in this disease. The outcome was good(3,10). In this communication we report the improvement following Plasmapheresis in the first pediatric case.

Case Report

D.K. a four years old girl was admitted to the Pediatric Service of our Hospital

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