Fetal alcohol syndrome with hydrocephalus

A case report

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Summary

An unusual case of the fetal alcohol syndrome with hydrocephalus caused by a Dandy-Walker cyst is reported. It is postulated that aberrant neuroglial tissue due to prenatal maternal alcohol ingestion may have caused the Dandy-Walker deformity.

The fetal alcohol syndrome (FAS) is a common cause of central nervous system dysfunction. It manifests as microcephaly in over 80% of affected infants. Hydrocephalus in FAS is rare and follows obstruction of the ventricular cerebrospinal fluid (CSF) outflow due to structural disorganization of the brain. This report describes a case of FAS associated with hydrocephalus caused by a Dandy-Walker cyst of the fourth ventricle.

Case report

A female infant was delivered prematurely by caesarean section because of fetal distress. The mother admitted having smoked heavily and drunk at least one bottle of wine daily throughout pregnancy. The birth weight was 1 560 g and the infant was clinically assessed as being at 29 1/2 weeks' gestation. The initial medical records make no mention of the infant's head circumference. Because of the antenatal history and certain dysmorphic features, the diagnosis of FAS was entertained. The postnatal course was uneventful, and the baby was discharged after 7 weeks weighing 2 050 g.

The patient first presented at the Red Cross War Memorial Children's Hospital at the age of 3 months with diarhoea. She was a marasmic, dehydrated infant with obvious dysmorphic features which included short palpebral fissures, prominent epicanthic folds, maxillary hypoplasia, micrognathia, aplasia of the middle phalanges and pseudo syndactyly, and a cardiac murmur heard. Chest radiography showed a bulky heart with plerhoric lung fields and an ECG showed left axis deviation suggestive of an atrioventricular defect. The infant required prolonged hospitalization for persistent diarhoea. During this period her head enlarged progressively and 7 weeks after admission the occipitofrontal circumference measured 44 cm. Ultrasound examination demonstrated marked hydrocephalus involving the third ventricle, a large posterior cyst with delineation of the cerebellar hemispheres and aqueduct, and an anterior bulging of the tentorium. The findings suggested the presence of a Dandy-Walker cyst but did not exclude an arachnoid cyst. A percutaneous ventriculogram confirmed the presence of a cyst of the fourth ventricle typical of the Dandy-Walker syndrome.

Initially a ventriculoperitoneal shunt was unsuccessful because the high CSF outflow caused significant abdominal distension. A ventricular drain was substituted and later replaced by a ventriculoperitoneal shunt after the head had decreased in size. The infant's further course was uneventful. At the time of discharge she was still generally hypotonic.

Discussion

The fetal alcohol syndrome is well described and has many characteristic clinical features. Our patient had the typical facial features of FAS, hypoplastic nails and a cardiac murmur probably due to an atrioventricular defect. These clinical features, together with the mother's history of considerable ingestion of alcohol during pregnancy, confirm the diagnosis of FAS.

Microcephaly and mental retardation occur frequently in FAS. Clarren and Smith reported these features in over 80% of affected infants. Clarren et al. have also suggested that in a small number of cases gross structural derangement of the central nervous system (CNS) may be the only manifestation of excessive antenatal alcohol ingestion. The exact mechanism of CNS damage in the fetus is not known, but it has been postulated to result from release of acetaldehyde, a disturbed amino acid balance or possibly alcohol-induced hypoglycaemia.

The unusual feature in our patient was the associated hydrocephalus. This appears to be an uncommon finding in infants with FAS since very few such cases have been reported in the literature. Autopsy findings are limited, but they have reported structural brain disorganization with aberrant neuroglial tissue covering the brain and causing distortion and obstruction to normal CSF outflow.

The hydrocephalus in the present case was associated with a cyst of the fourth ventricle typical of the Dandy-Walker syndrome. The pathogenesis of the Dandy-Walker cyst is uncertain but it appears to result from failure of the foramina of Magendie and Luschka to open. This leads to enlargement of the fourth ventricle and failure of the vermis to develop. Whether the cyst and subsequent malformation in this infant were related to the maternal ingestion of alcohol antenatally is unknown. It is

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conceivable that aberrant neuroglial tissue was responsible for the ventricular CSF outflow obstruction and formation of a Dandy-Walker cyst.

REFERENCES

2. Constrictive pericarditis following myocardial revascularization

A case report

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Summary

The occurrence of constrictive pericarditis after coronary bypass surgery is rare and clinical manifestations may appear at variable intervals after surgery. Three possible causes have been postulated, all of which were probably involved in the case which we describe. The clinical diagnosis of postoperative constriction is difficult and not often considered. It is best confirmed by means of cardiac catheterization, which shows typical haemodynamic features. Surgical treatment is both difficult and a threat to the coronary bypass grafts, when present. Conservative management with diuretics is preferred unless constriction is severe.

Constrictive pericarditis is a rare complication of cardiac surgery. The aetiology is unknown and the interval between surgery and evidence of constriction is variable. The diagnosis should be considered if unexplained right heart failure develops after cardiac surgery.

Case report

A 46-year-old housewife with a history of hypertension was admitted to hospital in December 1978 with unstable angina. Her condition was initially controlled by means of medical treatment, but again became unstable in August 1979, at which stage the patient was readmitted for coronary angiography. This revealed double-vessel coronary artery disease with severe stenosis of the proximal left anterior descending (LAD) artery, a large diagonal branch with a significant lesion proximally and a significant lesion in the middle third of the right coronary artery. Left ventricular angiography demonstrated normal function with an end-diastolic pressure (EDP) of 16 mmHg. Four weeks later saphenous vein bypass grafts to the LAD artery, its diagonal branch and the posterior descending artery were performed. After surgery the jugular venous pressure (JVP) was elevated, but this responded to treatment with diuretics. Four months after surgery the patient’s angina recurred, and in March 1980 she was re-investigated by means of cardiac catheterization; this showed the grafts to the LAD and posterior descending artery to be patent, but the graft to the diagonal branch could not be demonstrated. A left ventricular angiogram was normal and the EDP was 16 mmHg.

In January 1982 she was readmitted with a 2-week history of rapidly progressive swelling of both legs, right upper quadrant abdominal discomfort and tiredness. On examination she was afebrile and short of breath, and oedema of the legs and sacrum was present. The pulse rate was 80/min and regular, while the blood pressure was 140/95 mmHg with 15 mm of paradoxus. The JVP was elevated 20 cm above the sternal angle with equal 'a' and 'v' waves and rapid 'x' and 'y' descents. Examination of the precordium revealed a diastolic lift at the left sternal edge. A third heart sound was audible. The patient’s breath sounds were normal and a 4 cm non-pulsatile hepatomegaly with ascites was present. Chest radiography showed a normal heart with upper lobe venous distension. The ECG was of normal voltage with nonspecific ST-T wave changes in leads II, III and aVF.

An echocardiogram showed no pericardial effusion; the right ventricle was dilated and there was paradoxical septal motion (Fig. 1A). A simultaneously performed apex cardiogram confirmed the diastolic lift (Fig. 1B). Cardiac catheterization...