Cerebral arteriovenous aneurysm presenting with heart failure

Report of three cases

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Three infants with cerebral arteriovenous aneurysms developed severe heart failure. The diagnosis was suggested by hearing a murmur when listening over the skull. The vascular abnormality was successfully shown by cerebral arteriography in two of the cases. The heart failure proved difficult to treat and two of the infants died. Some of the problems of diagnosis and management are discussed.

Silverman *et al.* (1955) described two examples of cerebral arteriovenous aneurysm presenting with cardiac failure in the neonatal period.

The Children's Hospital, Birmingham, provides a cardiological service for a population of 5½ million, and until 1971 only 3 cases had been recognized – all at necropsy: 2 of these have been reported (Corrin, 1959).

This paper describes 3 more cases all seen in the last 2 months of 1971: 2 were diagnosed during life; in the third the condition was suspected clinically but was not proved until after death.

Case reports

Case 1

A female Indian baby born at full term (birthweight 2.6 kg) presented with severe vomiting at 9 days of age and was found to be in heart failure. She was slightly centrally cyanosed and had a large heart with an easily felt apical impulse, systolic ejection murmur heard best at the left sternal edge, and some accentuation of the pulmonary component of the second heart sound. The most striking sign was the pronounced arterial pulsation, especially dramatic in the carotid arteries. A loud continuous murmur could be heard over the cranium suggesting an arteriovenous shunt.

A large heart and plethoric lung fields were seen on radiography. The electrocardiogram showed right ventricular and right atrial hypertrophy (Fig. 1).

Cerebral arteriography confirmed the clinical impression and demonstrated the anatomy of the vascular malformation (Fig. 2). The condition was considered to be unsuitable for operation and the child died 5 weeks later in heart failure at another hospital. At necropsy the heart

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was reported to be normal apart from hypertrophy of the right ventricle and the right atrium. The brain was examined at this hospital. There was a cerebral arteriovenous aneurysm involving the left anterior and posterior cerebral arteries and the vein of Galen, with secondary atrophy of the left occipital lobe (Fig. 3).

Case 2

A male infant (weight 3.5 kg) was delivered at term by caesarian section because of placenta praevia. When 2 days old he was found to be in heart failure. Unlike Case 1 he was not cyanosed and the arterial pulses were of normal volume. There was a loud continuous murmur over the cranium.

Cardiac catheterization showed a pulmonary artery pressure of 48/25 mmHg. The arterial systolic pressure was 60 mmHg and the oxygen saturation was 91 per cent. Cineangiogram with injection into the right atrium showed a normal circulatory route. Cerebral arteriography showed a lesion very similar to that of the previous case. The situation and nature of the lesion were considered to be contraindications to operation. The congestive heart failure was treated with digoxin and diuretics, and the child improved. He has since developed satisfactorily and at the age of 3 years has shown no neurological complications, and cardiac failure is controlled by digitalis and diuretics.

Case 3

A full-term female infant weighing 4.0 kg showed evidence of heart failure and peripheral cyanosis within a few hours of birth. On the second day the congestive failure was severe, the heart noted to be large, and the apex beat diffuse and dynamic. Two distinct murmurs were heard – pansystolic below the left clavicle and a short systolic at the left sternal edge. Radiography

FIG. 1 Electrocardiogram (Case 1) showing right ventricular and right atrial hypertrophy (patient on digitalis).

showed a large heart with engorged pulmonary vessels, and the electrocardiogram showed right axis deviation with right ventricular hypertrophy.

A very faint systolic murmur was audible over the left temporal region and the possibility of cerebral arteriovenous aneurysm was discussed though the diagnosis was not seriously entertained. However, when a venous angiogram revealed a normal circulatory route the skull was screened immediately, but no arteriovenous malformation was seen. Despite vigorous medical treatment the child died before further investigations could be performed.

At necropsy the heart was found to be normal but there was a large cirroid aneurysm overlying the right cerebral hemisphere. This was fed by the middle cerebral artery and drained into the right transverse and straight sinuses (Fig. 4).

Discussion

Since Silverman's original description a further 32 cases of cerebral arteriovenous aneurysm presenting with congestive heart failure in the neonatal period have been reported (Brown and Alexander, 1967; Claireaux and Newman, 1960; Corrin, 1959; Deverall *et al.*, 1969; Falcone, Friedman, and Peker, 1965; Glatt and Rowe, 1960; Gold, Ransohoff, and Carter, 1964; Gomez *et al.*, 1963; Hirano and Solomon, 1960; Holden *et al.*, 1972; Lehman *et al.*, 1966; Levine *et al.*, 1962; Fyler, 1968; Pollock and Laslett, 1958; Stern, Ramos, and Wiglesworth, 1968). Of these, 17 were diagnosed in life (see Table 1). From this Table it will be seen that the aneurysm usually involves the great vein of Galen, that the infant almost always presents for

(a)

(b)

FIG. 2 Cerebral angiogram (Case 1) (PA and lateral) showing dilatation of the vein of Galen.

FIG. 3 Inferior surface of the brain (Case 1) showing a cut cross-section of the temporo-occipital region with shrinkage due to atrophy on the left. The dilated carotid arteries are arrowed in white and the aneurysm of the vein of Galen arrowed in black. (P.M. number 72-48.)

treatment in the first week of life, and that cyanosis is common.

It is noteworthy that, of the 14 case reports, no cranial bruit was heard in 2. Our 2 cases with aneurysmal dilatation of the great vein of Galen demonstrated easily audible continuous bruits. In the third, however, where there was a cirroid aneurysm overlying the area supplied by the right middle cerebral artery the bruit was faint and confined to systole.

Various authors (Moore and Baumann, 1969; Gellis, 1971) have cast doubts on the diagnostic value of hearing a bruit over the skull because it is said that a murmur can be heard in 14 to 15 per cent of normal infants (Still, 1921; Hughes and Todd, 1953). Our experience has been different, and we feel that in an infant with unexplained cardiac failure the presence of continuous or systolic murmurs over the cranium should suggest the possibility of intracranial aneurysm. There are, however, a number of other conditions reported in association with murmurs over the skull. These are listed in Table 2. We have heard a loud continuous murmur over the skull conducted from a persistent ductus arteriosus in an infant with congestive heart failure. However, in this situation the murmur is best heard over the praecordium and there may be additional peripheral radiological and electrocardiographic signs of the lesion. The diagnosis of arterio-

FIG. 4 Lateral view of the right cerebral hemisphere (Case 2) with frontal lobe to the right showing the cirroid aneurysm overlying the parietal lobe. (P.M. number 71-199.)

TABLE 1 Previous cases diagnosed in life

Author	Sex	Birthweight (kg)	Onset symptoms (dy)	Cyanosis	Cerebral bruit	Anatomy	Surgery	Survival
Brown <i>et al.</i> (1967)	F	NK	5	o	o	Vein of Galen	o	o
	M	NK	7	o	+		+	o
Falcone <i>et al.</i> (1965)	F	2.7	1	o	+		o	o
Gomez <i>et al.</i> (1963)	M	1.8	105	+	+	Vein of Galen	o	o
	M	1.9	1 hr	+	NK		o	o
	M	3.4	1	+	NK		o	o
	M	3.0	2	+	NK		o	o
	F	NK	3	o	+		+	+
Holden <i>et al.</i> (1972)	M	4.9	6	7 out of 8	7 out of 8	Vein of Galen	+	+
	F	2.5	60			Cirroid	+	+
	F	2.3	1			Vein of Galen	+	o
	F	3.1	7				+	o
	M	NK	3			Vein of Galen	+	+
	M	3.9	35			Cirroid	+	+
	M	2.2	1			? Cirroid	o	o
	M	3.4	1			Vein of Galen	o	o
Lehman <i>et al.</i> (1966)	M	NK	90	+	+	Vein of Galen	+	o
Our series	F	2.6	9	+	+	Vein of Galen	o	o
	M	3.5	2	o	+	Vein of Galen	o	+
	F	4.0	6 hr	+	+	Cirroid	o	o

NK = Not known

TABLE 2 Occurrence of cerebral bruit

- (a) In normal child
- (b) Underlying pathology;
 - angioma;
 - AV malformation;
 - meningioma;
 - meningitis
- (c) Systemic causes;
 - high output states
 - e.g. anaemia
- (d) Conducted;
 - persistent ductus arteriosus

venous aneurysm is confirmed by cerebral angiography. In infants where there is doubt about a congenital heart lesion full cardiac catheterization is necessary, followed by cerebral angiography if the former does not confirm the diagnosis.

Surgery was attempted in 9 of the 17 children previously reported and 5 children survived. The quality of survival, however, is not well documented though Holden *et al.* reported that 2 of their 6 patients were progressing normally after one year. In one of these, operation had been deferred until one year after medical treatment of congestive heart failure, a good response being obtained to digitalis and diuretics. We considered operation in Cases 1 and 2. From previous reports however (Gomez *et al.*, 1963) and from our own experience (Corrin,

1959), we know that most cases have pronounced cerebral damage caused primarily by reduced local tissue perfusion secondary to shunting of blood through the aneurysm. Indeed Case 1 at necropsy had a wide area of cerebral atrophy. This unknown degree of cerebral damage plus the damage likely to be incurred in the necessary corrective operation on a small neonatal brain would, in our opinion, have made an acceptable quality of survival unlikely at the time of diagnosis. We adopted a policy of medical management hoping for survival with the possibility of operation at a later date. Case 2 is well at 3 years, with no apparent neurological complications. It is felt that further postponement of surgery will confer more benefit than potential dangers.

Intracranial arteriovenous aneurysm is a rare cause of heart failure in infancy. The occurrence of cyanosis and right ventricular hypertrophy are unexpected features in this condition. Cyanosis may be central, as in one of our three cases, or peripheral. Central cyanosis theoretically could result from a shunt from right to left through a patent foramen ovale if the normal pressure difference between the two atria is reversed. This would be supported by the finding of right ventricular hypertrophy which suggests pulmonary hypertension, and information from the published material (Rudolph, 1970; Holden *et al.*, 1972) suggests that this is often present. Pulmonary hypertension may be due to the persistence of a high pulmonary vascular resistance after birth, in the face of an increased pulmonary

blood flow caused by an obligatory shunt (Rudolph, 1970).

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