

Occlusion of the Cerebral Arteries in Recklinghausen's Disease

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Summary. Two children, aged 18 months and 6 years, who had Recklinghausen's disease, had occlusion of cerebral arteries. One child had no motor deficit but the other had right hemiparesis and partial occlusion of the left posterior cerebral artery, a fact not found in the literature.

Key words: Arterial occlusive disease – Neurofibromatosis – Recklinghausen's disease

Introduction

Occlusion of the cerebral arteries in Recklinghausen's disease was first described by Hilal et al. in 3 patients in 1971 [4]. Subsequently, Tomsick et al. [8] compiled all the published cases and found a total of 14 cases, including two patients of their own. From 1972 to 1978 among our patients with Recklinghausen's disease we found two children who had occlusion of the cerebral arteries and basal telangiectasias. In one of the cases the posterior cerebral artery was affected, a fact which has not been reported in any of the published cases.

Case Reports

Case 1

Female, admitted to hospital at the age of 18 months (21. 4. 72) because of gastroenterocolitis.

Personal history: Normal pregnancy and delivery. Weight 3.7 kg. Numerous cafe-au-lait spots from birth. No convulsions. No motor deficit.

Family history: Mother of 35 years affected by Recklinghausen's disease. This was the fourth pregnancy; had two miscarriages. A younger brother with Recklinghausen's disease was studied angiographically and will be referred to in the discussion. No other record of neurofibromatosis in the family.

Examination: Macrocephaly p.98. Size p.3. Weight p.3. Normal arterial pressure. Strabismus in the left eye. Ophthalmoscopy normal. Slight psychomotor retardation. Did not begin to walk until the age of 21 months. This retardation continues at present, although not very marked. Multiple cafe-au-lait spots distributed mainly on the trunk and abdomen, some of them large, about 14 in number. At present the patient continues without convulsions or motor deficit.

Brain scan normal. Echoencephalography (A-scan) normal. During these years there has been a slowing tendency in the EEG.

Radiological study. Plain skull film: macrocephaly with Cronqvist index of 62; enlarged chiasmatic groove. These findings have not been modified over the years.

Pneumoencephalography. (25. 4. 72) Moderate dilatation of the lateral ventricles, particularly the left, with an Evans index of 0.30. Dilatation of interpenduncular and optochiasmatic cisterns, assuming the form of a cyst. Large cisterna magna.

Left carotid angiography (25. 4. 72). Partial stenosis of the internal carotid artery 5 cm above the posterior communicating artery. Complete occlusion of the middle cerebral artery with marked hypertrophy of all basal branches which have a telangiectatic, pseudoangiomatous formation (Fig. 1).

Hypertrophy of all branches of the posterior cerebral artery, mainly the temporal branches. Occlusion



Fig. 1. Case 1. Left carotid angiography. Complete occlusion of middle cerebral artery with marked hypertrophy of all basal branches which have a telangiectatic, pseudoangiomatous appearance



Fig. 2. Case 2. Left carotid angiography. Occlusion of internal carotid artery with multiple telangiectatic, pseudoangiomatous branches

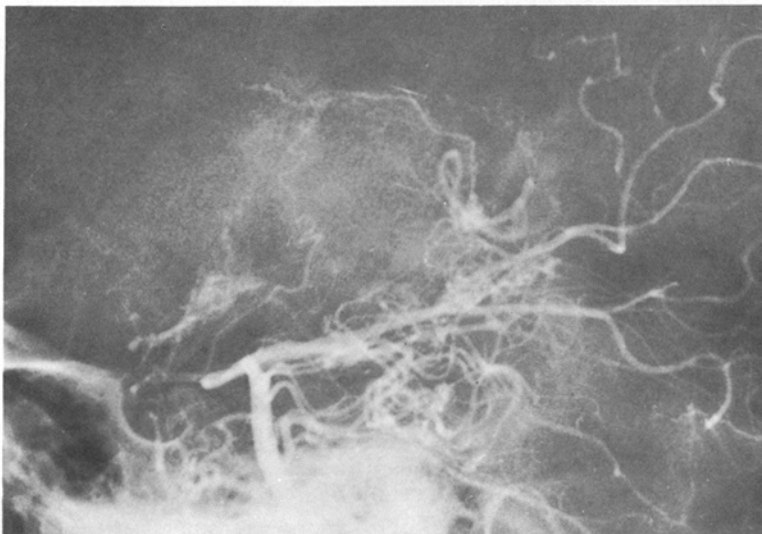


Fig. 3. Case 2. Vertebral angiography. Telangiectatic branches coming from left posterior cerebral artery. Complete occlusion of left posterior cerebral artery

of the pericallosal artery, which is replaced by the posterior pericallosal. Numerous collateral connections at all levels with retrograde filling of the branches of the middle cerebral artery, principally from the territory of the posterior cerebral.

Right carotid: Narrowing of primitive carotid artery at cervical level, measuring 2 cm in length. Spasm cannot be ruled out.

Left carotid. (June 1974) The same findings persist.

Vertebral angiography. Fine telangiectasic branches coming from the first portions of the posterior cerebral artery, with hypertrophy of the posterior pericallosal, posteromedial and lateral choroid and the thalamoperforating arteries.

Urography. Normal. Skeleton normal (6. 6. 78).

Computed tomography. No zones of cerebral infarct. The slight increase in size of the ventricular system persists, particularly the left and of the interpenduncular and optochiasmatic cisterns.

Case 2

Male, 6 years old, admitted to hospital because of acute right hemiplegia 1 week old.

..... Normal pregnancy and delivery. Weight 3.3 kg. Multiple cafe-au-lait spots from birth.

Family history. Father aged 31 years. Mother aged 30 years has Recklinghausen's disease, also a brother aged 2. No other family record.

Examination. Head circumference normal (P50). Normal weight and size. Normal psychomotor development except for slight speech retardation. Numerous cafe-au-lait spots all over the body, particularly on the trunk, about 25 in number. Right hemiparesis. Ophthalmoscopy and blood pressure normal. EEG: Spikes in the right parietal region. Echoencephalography, normal.

Radiological study. Plain skull film normal, with index of Cronqvist of 55. Normal sella and optic foramens.

Urography. Normal. Skeleton normal.

Computed tomography normal. No infarct.

Left carotid angiography. Complete occlusion of the internal carotid 4 mm from the posterior communicating artery (Fig. 2). Multiple telangiectasic, pseudoangiomatous branches of fine caliber in entire basal area.

Right carotid angiography. Stenosis of the anterior cerebral artery at its very beginning with small telangiectasic branches filling the left anterior cerebral.

Vertebral angiography. Almost complete occlusion of the left posterior cerebral artery with multiple telangiectasic branches coming out at the level of the stenosis (Fig. 3).

Discussion

The involvement of the posterior cerebral artery has not been described in any of the 14 cases found in the literature [1–6, 8–12]. Case 2 had incomplete occlusion of the left posterior cerebral artery with projection of multiple telangiectasic branches.

The occlusion of the cerebral arteries in Recklinghausen's disease does not appear to be progressive. We checked our second case at the age of 2 years and did not find any angiographic changes. Neither does Recklinghausen's disease appear to affect all members of one family since we studied the brother of our first case at the age of 1 year and at the age of 6 years and did not find occlusion of any cerebral artery. The first case did not have any motor deficit. This fact has been found in three other cases published [4, 8]. Our two patients did not have any accompanying intracranial tumor, nor had they received radiation therapy. Thus, our two patients, together with the other cases in literature, support the theory that the occlusion of the cerebral arteries in Recklinghausen's disease is primary.

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