

Intracranial Fibromuscular Dysplasia and Stroke in Children

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Summary. Fibromuscular dysplasia (FMD) is an arterial lesion of unknown origin which may sometimes affect intracranial arteries. In recent years a few rare cases, mostly involving younger people, have been described. The present case concerns a child of 8 years in whom the finding of hemiplegia led to a diagnosis of intracranial fibrodysplasia which was proven by arteriography and biopsy¹. This arterial lesion is described within the context of stroke in children.

Key words: Fibromuscular dysplasia – Intracranial fibrodysplasia – Pediatric stroke

Fibromuscular dysplasia (FMD) was first described in 1938 by Leadbetter and Buckland [15] in a hypertensive boy with a pelvic kidney. Since then, fibroplastic dysplasia of the renal arteries has been recorded fairly frequently, as has the association with renal hypertension. From 1965 onwards, involvement of arteries other than renal was described by Palubinskas [17] and Connett [3]. In addition to the involvement of the various abdominal arteries, more and more cases were recorded in which arteries in the extracranial carotidovertebral region were affected [2–4, 10, 12, 16]. This carotidovertebral location was the subject of important studies by Houser and Baker [12] which was based on 52 cases, and by Manelfe [16] which involved 70 cases. Typical intracranial locations were not established in either study but the latter did mention two which were demonstrated only

on autopsy. In 1967, Huber and Fuchs [13] described the radiological demonstration of intracranial involvement in two patients and in the last 10 years an intracranial location has been described only sporadically [1, 5, 8, 11, 13, 14, 16, 18, 19, 21, 23, 24]. If from these, one disregards the radiologically atypical cases, especially those in which the lesion was not histologically confirmed, the number dwindles rapidly.

Case Report

A girl aged 8, complained suddenly of severe headache and subsequently developed right hemiparesis. There was no previous illness and, in particular, no throat infection. The girl was one of twins born on 4 September 1967; the delivery had been normal. Strabismus was noted and psychomotor development was somewhat retarded; the child was being educated at a special school.

On admission the child was stuporous and right hemiparetic with right hyperreflexia and a pathological extensor plantar reflex on the right. The girl was right-handed and aphasic but there was no sensory disturbance. Further investigations were unremarkable except for the EEG which showed diffuse, delayed delta dysrhythmia, mainly on the left side. There had been no epileptiform incidents in the past history. Lumbar puncture revealed no unusual abnormalities and tests for syphilis were negative. Vision tests carried out later were normal.

Left carotid angiography was carried out on 6 February 1976 and on 1 March 1976. On both occasions a slight tubular narrowing of the cervical internal carotid artery was noted, and obvious strictures of a generally dilated supraclinoid carotid. In particular, there were striking deviations of the mid-

¹ Of course, there is still a little doubt about the definitive diagnosis, as long as the histology is made by biopsy of the a. temporalis superficialis and not of the affected intracranial vessels.

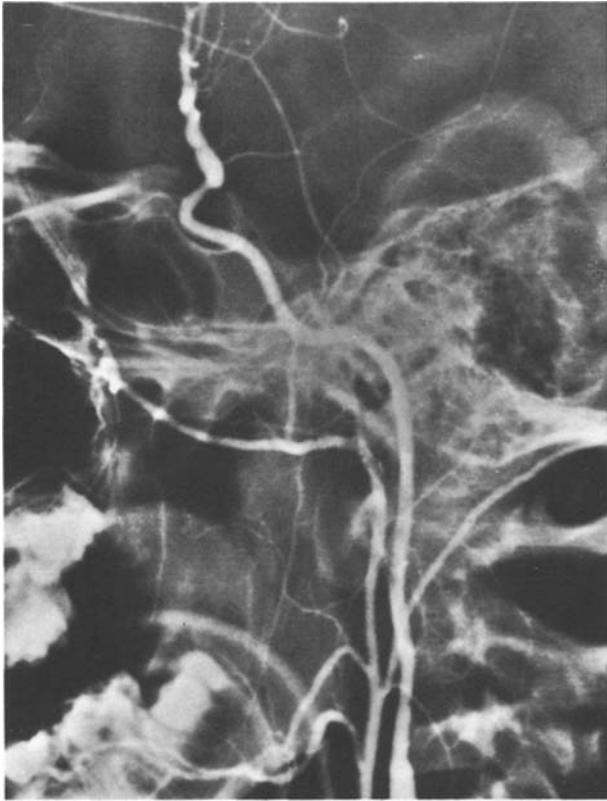


Fig. 1. Lateral cervicocranial view of left carotid artery. Slight tubular narrowing of cervical internal carotid artery, and strictures on dilated supraclinoid carotid artery

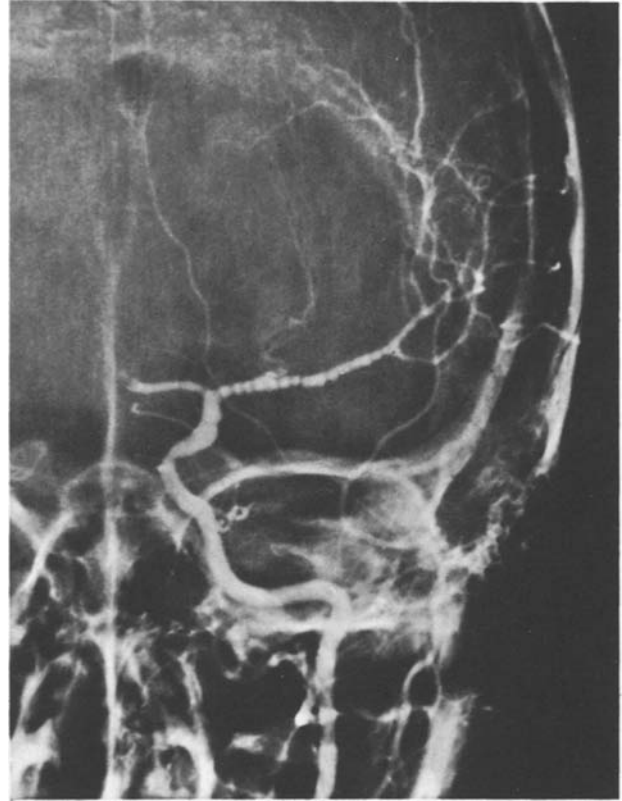


Fig. 2. Magnification AP view of left intracranial carotid artery. Typical 'string of beads' pattern of entire proximal segment of middle cerebral artery, and occlusion of anterior cerebral artery

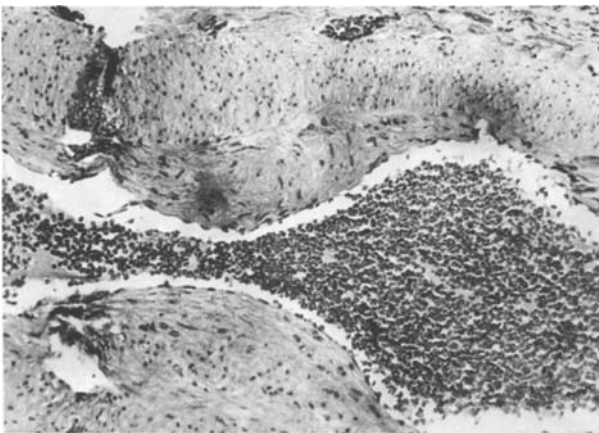


Fig. 3. Biopsy specimen of superficial temporal artery. Arterial dysplasia of primary intimal dysplasia type; elastica is fragmented

dle cerebral artery: the first segment M_1 showed about twenty microdilations separated from one another by fine strictures. The distal branches of the middle cerebral artery were poorly filled, and the A_1 - A_2 transition of the anterior cerebral artery was completely occluded.

Arteriography of the right carotid and vertebral arteries was completely normal. Selective arteriography of the two renal arteries and of the celiac artery was also normal.

This is a typical case of plurifocal FMD of the left internal cervical carotid artery and particularly of intracranial involvement of the supraclinoid carotid and sphenoid segment of the middle cerebral artery coupled with signs of occlusion of several distal branches.

The diagnosis of FMD was histologically confirmed by biopsy of the superficial temporal artery (as has been recommended by several authors). The further course of the patient was relatively favorable; she recovered quickly from the hemiplegia and examination 6 months and 1 year later revealed moderate right spastic hemiparesis as residual damage.

Radiological Comment

Intracranial FMD raises few specific problems from the radiological point of view. Intracranially – as elsewhere in the body – one finds the well-known

'string of beads' or 'bead roll' feature. This radiological appearance is due to the formation of a chain of arterial dilatations (microaneurysms or microsacculations) which are separated from one another by localized strictures resulting in narrowing of the lumen by approximately 40%. The saccular dilatations are mostly broader than the normal lumen.

The tubular form is seen less frequently; it more or less takes the form of a long, concentric stenosis and is usually seen on the cervical internal carotid.

Finally, there are also signs of occlusion – which as such are not specific for FMD – but when they occur in association with typical FMD must logically be ascribed to the same etiology. Generally speaking, FMD is multifocal and therefore extensive arteriographic studies should always be performed. On the other hand, it seems from the literature [16] that intracranial FMD cannot always be detected by arteriography even when its presence is definitely demonstrable histologically. This raises the question as to whether or not 'infraradiological' forms of FMD exist and if so, how frequent they are.

The radiological differential diagnosis depends upon the arteriographic appearance. The classical 'bead-roll' image must be distinguished mainly from the so-called 'stationary arterial wave' image (also called the 'pearl necklace' image) which is an anomaly characterized by an exceedingly regular rippling of the intra-arterial contrast column. The anomaly is due to a kind of resonance phenomenon, the exact origin of which is uncertain since it may also be seen on arteriography in cadavers [22].

It is more difficult to establish a differential diagnosis for the rarer stenosing form of FMD, since it must be distinguished from atheromatous stenosis, tubular occlusion due to all kinds of arteritic conditions [7, 20], congenital hypoplasia and arterial spasm.

Associations

FMD, both renal and extrarenal, is frequently associated with intracranial aneurysms. Aneurysms may also occur in the dysplastic segment of the artery.

It seems from important reviews that intracranial aneurysms are recorded in approximately 30% of patients with FMD. For this reason, Frens [8] recommends that systematic intracranial arteriography should be carried out.

Rare associations include the existence of a carotid cavernous fistula as described by Zimmerman [24] in a woman with diffuse extra- and intracranial FMD. The association of intracranial FMD with mul-

tipple enchondromata has also been described [19] as has convoluted 'looping' of the cervical internal carotid with FMD [4].

Discussion

From the pathological point of view, FMD has always been classified according to which arterial lamina is mainly involved [9]. Fibroplasia of the intima occurs only rarely. The overwhelming majority of cases involve fibroplasia of the media. Fibroplasia of the adventitia is exceedingly rare.

On the basis of 196 cases, Stanley et al. [20] have tried to obtain a better understanding of the histopathology. According to them, fibroplasia of the intima is seen in about 5% of cases and this form appears chiefly in children and younger persons and is distributed equally between the sexes. Fragmentation of the internal elastic lamina is often noted histologically. This characteristic must be emphasized because classical FMD is practically exclusively seen in women of about 50 and older, including the extracranial lesions in the carotidovertebral region [12, 16].

On the other hand, it is statistically evident that the rare cases of intracranial FMD are detected mainly in children and young adults. There is still insufficient histopathological evidence to establish a connection with intimal fibroplasia, even though in the case described here, the correlation does exist.

A third very important aspect of intracranial FMD is its role in pediatric stroke. Slagsvold [19] quite rightly notes "... extracranial carotid fibromuscular dysplasia may be an incidental finding; intracranial involvement is more serious ...". Intracranial FMD causes serious morbidity in children and young persons.

Clearly the question of cerebrovascular accident in children is involved here. It is postulated that in this much discussed problem, about half of the cases can be attributed to neither cardiac nor vascular cause [6]. FMD is seldom named as a possible cause of stroke in children. In the vast majority of these cases, common arteritic conditions are noted, often in association with pharyngitis or tonsillitis and seldom in connection with systemic arteritis [11, 20]. Harwood-Hash [11] with reference to 40 such cases wrote: "... fibromuscular dysplasia has yet to be reported in the intracranial arteries ...", and demonstrated a very typical case.

We would like to draw attention to the fact that the existence of intracranial FMD is a reality and that this location within the broad context of FMD has its own specific characteristics, particularly in connec-

tion with stroke in children, as is clearly demonstrated in the case described and in recent literature.

Treatment

The precise etiology of FMD is as yet unknown. The interesting hypothesis put forward by Stanley et al. [20] cannot really apply to intracranial involvement in children. For this location, treatment is conservative and purely symptomatic.

According to Stanley, extracranial FMD in adults might be due to the following: female hormonal influences, mechanical stretch-traction stresses and arterial mural ischemia caused by insufficiency of the vasa vasorum.

Therapeutic surgical intervention may be considered in cases of extracranial FMD. The paper by Collins [2] is noteworthy: he suggests that intracranially affected arteries could be treated by use of a dilating probe.

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