Eur Arch Psychiatr Neurol Sci (1989) 238:196–198

European Archives of Psychiatry and Neurological Sciences

General Dysplasia of the Cerebral Arteries with Persistent Primitive Acoustic Artery and Giant Aneurysm

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Summary. A case of cerebrovascular systemic dysplasia is presented. The anomalies were: persistent primitive acoustic artery, giant aneurysm of the left internal carotid artery, aneurysm of the basilar artery, and an arteriovenous fistula. A review of the literature shows that the association of multiple malformations of cerebral vessels with a persistent primitive acoustic artery has not previously been described. Impairment of embryonic development is suggested as the origin of the malformations.

Key words: Persistent primitive acoustic artery – Giant aneurysm – Cerebrovascular dysplasia

Introduction

Carotid-basilar anastomoses are found during normal embryonic development. They disappear when the embryo reaches a length of 14 mm and are of functional significance for approximately 1 week. Normally there is involution of these vessels [4]. In rare cases these embryonic arteries persist, most frequently the primitive trigeminal artery, followed by the primitive hypoglossal artery [9, 20, 27]. Persistence of the proatlantal artery and the primitive acoustic artery has been described only rarely [11, 12, 14].

The association of a persistent embryonic carotid-basilar anastomosis with a cerebral aneurysm has been previously described [7, 10, 17], but not with persistence of the primitive acoustic artery. Few cases of arterial cerebral aneurysms with arteriovenous malformations have been observed [5, 22].

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In this paper we describe a case of cerebrovascular malformation with a persistent primitive acoustic artery, a giant aneurysm of the left internal carotid artery, an aneurysm of the basilar artery, and an arteriovenous fistula in the region of the left internal carotid artery, a combination apparently not previously reported.

Case Report

Over a period of 3 months, a Turkish male, age 30 years, developed weakness and a feeling of heaviness in the left arm and leg.

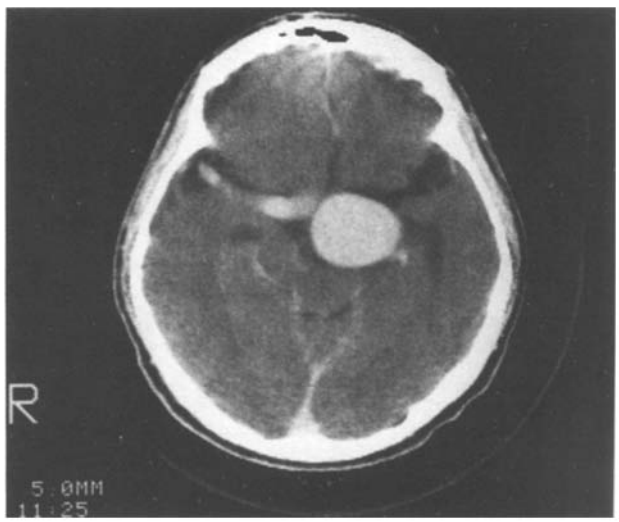

Fig. 1. Cranial CT scan with contrast material; giant aneurysm in the left temporo medial area; dilated right middle cerebral artery

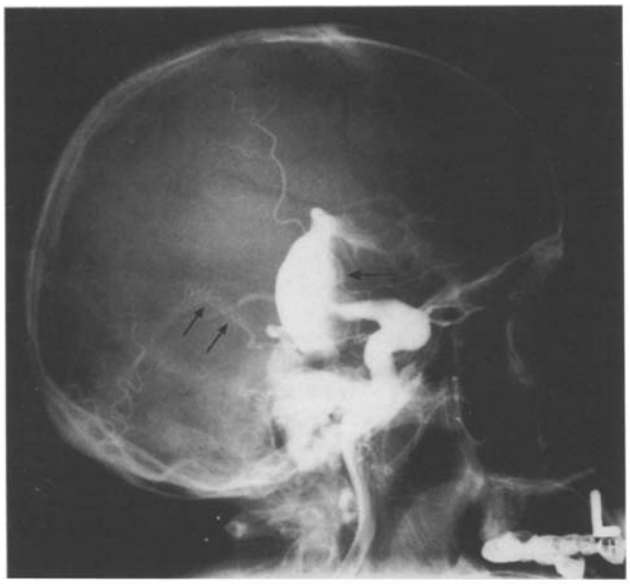

Fig.2. Left carotid arteriogram; giant aneurysm (*arrow*) and adjacent dilated internal carotid artery; distal to that an arteriovenous fistula between marginal tentorial artery left and sinus rectus (*double arrow*)

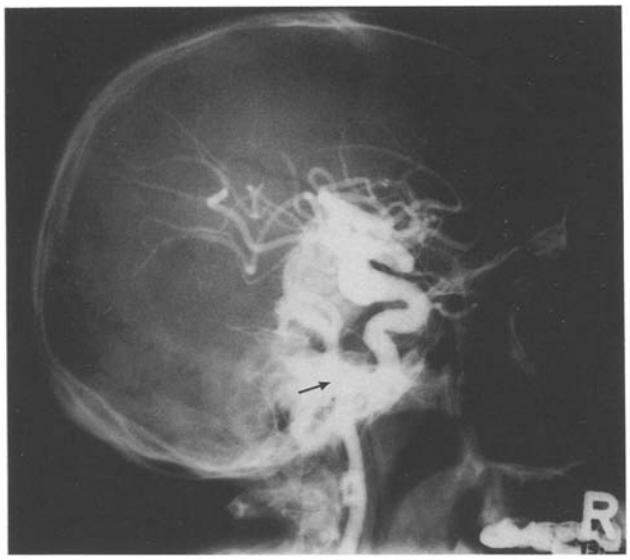

Fig.3. Right carotid arteriogram; basilar artery, which is fed by persistent primitive acoustic artery (*arrow*)

Neurological examination revealed hemiplegia, increased tendon reflexes with permanent clonus of the patella and the ankle, and Babinski's sign of the left side. All mental functions were impaired.

The *EEG* showed fluctuating alterations of moderate degree and a focus of paroxysmal theta acitivty in the left temporo parietal area.

CT with and without contrast material showed a vascular malformation located mainly in the left hemisphere (Fig. 1).

Bilateral carotid angiography demonstrated a giant aneurysm situated on the supraclinoid part of the left internal carotid artery with substantial ectasia of the adjacent vessels. The size of the aneurysm was $3 \times 5 \times 6$ cm (Fig. 2). In the final section of the right internal carotid artery and the right middle cerebral artery fusiform dilatations were seen (Fig. 3). In addition, a marginal tentorial artery on the left side with arteriovenous fistula to the sinus rectus (Fig. 2), as well as a persistent primitive acoustic artery on the right side supplying a dilated basilar artery, could be seen (Fig. 3).

Dramatic progression of the neurological deficits occurred soon after hospital admission. Because of the progressive symptomatology with a high risk of subarachnoid haemorrhage and poor long-term prognosis [26], surgical intervention was undertaken. Initially, clipping of the giant aneurysm was successful, but death occurred a few weeks later from subarachnoid haemorrhage from a small aneurysm of the basilar artery, which had been seen in postoperative angiography. This aneurysm was not found in the first arteriogram.

Discussion

Conventionally, a giant aneurysm is defined as having a diameter of more than 20 mm [16]. The aneurysm in our case was of extraordinary size, measuring $3 \times 5 \times 6$ cm. Typically for aneurysms, as also seen in this case, neurological deficits develop late in the course of the disease [18, 24]. Our patient showed a progressive hemiplegia and a psychosyndrome.

The combination of a giant aneurysm of the left internal carotid artery, an aneurysm of the basilar artery, an arteriovenous malformation (AVM) in the form of an anastomosis connecting the marginal tentorial artery and the left sinus rectus and a persistent primitive acoustic artery has, as far as we know, not previously been described in the literature. According to Anderson and Blackwood [1] and Arieti and Gray [2], embryonic maldevelopment can lead to vascular cerebral malformation. In our case the constellation of malformations suggests a systemic arterial dysplasia. The specific disturbance of embryonic development of the cerebral vascular system, which resulted in a persistent primitive acoustic artery might also be responsible for the additional vascular dysplasia [6, 25, 28].

The nature of this embryonic disorder is not known. It is possible that maldevelopment led to structural insufficiency of the arterial wall, especially of the media, inducing massive arterial ectasias and later the fusiform giant aneurysm. Trauma or perinatal infection, which has been regarded as a cause of congenital intracranial aneurysms [12], was not reported in this case. The right-sided embryonic arterioarterial anastomosis could have created a haemodynamic relief to the following fusiform dilated internal carotid and middle cerebral artery. Thus haemodynamic stress appears unlikely to be the only cause of the arterial giant aneurysm. As in our case, the occurrence of arterial aneurysms and AVM is usually

characterized by a more distal position of the AVM [5, 8, 15, 23]. This association is believed to be due to increased blood flow or haemodynamic stress in the supplying arteries of the AVM [19].

However, another arterial aneurysm was located on the basilar artery, which is not a supplying vessel of the AVM. This also implies a generalized affliction of the cerebral arterial vessel walls. Moreover, hypertension, which is frequently associated with multiple aneurysms in juveniles, was not found in our case [21]. Mere coincidence of the different malformations cannot be excluded with certainty [3]; however, the persistence of a primitive acoustic artery as a relict of embryonic development is extremely rare. In our case the persistent primitive acoustic artery could indicate a very early disorder of the cerebral arterial vascular system, which could account for the other malformations found.

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Received May 27, 1988