Considerations Concerning a Case of Fibromuscular Hyperplasia of the Carotid Arteries

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Summary. The authors describe a case of F.M.H. of the carotid arteries. The typical angiographic appearance is diagnostic. The relationship between this disease and the neurological signs is discussed.

A proposito d’un cas d’hyperplasie fibromusculaire des artères carotides

Résumé. L’auteur rapporte un cas d’hyperplasie fibromusculaire carotidienne chez une jeune femme.

Fibromuscular hyperplasia of the renal arteries, first described by Leadbetter and Burkland [6], is at the present time a well-known radioanatomical and clinical syndrome. On the other hand, the recognition of this lesion on the carotid arteries is relatively recent. In fact, the first description is that Connett and Lansche [2]. The disease is characterized by areas of hypertrophy of the media alternating with zones of fragmentation and reduction. Its angiographic appearance is very typical: it is characterized by narrowing and dilatation of the arterial lumen.

The purpose of this article is to call attention to this particular angiographic picture once again.

Case history

A 41-year-old woman was admitted to the neurology department of the university hospital at Geneva in December 1969, her chief complaint being attacks of loss of consciousness, a grand mal seizure having been observed on one occasion. A trauma with a brief period of unconsciousness was reported in 1962. In the last few months the patient had experienced a change of character with a depressive state. The results of the physical examination were unremarkable. No carotid bruits were noted. An E.E.G. showed discrete signs of disturbance on the left hemisphere, but no epileptic activity. The pneumencephalography showed a discrete cortical atrophy on the left hemisphere. The percutaneous bilateral carotid angiography of the common carotids showed the “string of beads” appearance on the two internal carotids, which is typical of fibromuscular hyperplasia (Figs. 1 and 2). Nothing pathological was noticed intracranially. In order to exclude all artefacts which might have been produced as a result of the direct puncture of the arteries, a right retrograde brachial angiography was performed 12 days later (Fig. 3) and showed the same angiographic picture. The right vertebral artery was normal. No renal angiogram was performed. During the patient’s stay in hospital the psychiatric symptoms became worse and she was transferred to the psychiatric hospital with a diagnosis of endogenous depression in a schizoid personality.

Discussion

Since the first description of Connett and Lansche [2], observations of cases of fibromuscular hyperplasia of the carotid arteries have become quite frequent in recent years [1, 3, 4, 5, 7, 8, 9, 10, 11, 12]. The condition has been seen in children and in the elderly of both sexes. Nevertheless, young adult females seem more often affected.

The angiographic picture with narrowing and dilatation of the arterial lumen is pathognomonic. The differential diagnosis with artifactual radiologic images is generally easy. However, in case of doubt, a repeat angiographic examination, avoiding the direct puncture of the artery, is desirable.

The internal carotid artery is most often involved, while the common and external carotids are generally spared. The F.M.H. may involve the whole length of the internal carotid artery, but it does not usually extend intracranially. Only in the two cases reported by Huber and Fuchs [5], and in the case of Bergan and MacDonald [1], were the intracranial portion of the carotid and its branches affected by the lesion. In our case an extension of the F.M.H. into the petrous canal was visible on the left side.

The lesion is often bilateral, in 85% of cases according to Houser and Baker [4], and 75% according
to Morris, Lechter and De Bakey [8], who are the authors with the longest series. Of the other vessels of the neck, the vertebral arteries can also be involved [8, 1].

First authors [12] always carry out a cerebral angiogram when F.M.H. is discovered on the renal arteries. We think that in these cases a carotid angiogram might be valuable for identifying F.M.H. of the carotids.

Fig. 1. Percutaneous bilateral carotid angiography. Lateral views (Subtraction). Right (A). Left (B). Typical narrowing and dilatation of the internal carotid arteries (→). Note the extension of the F.M.H. to the petrous canal on the left.

Fig. 2. Same. A.P. views. (Subtraction)

Some authors (Wylie et al. [12], Palubinskas et al. [10]) have noted the high frequency of intracranial aneurysms in the case of F.M.H. For that reason the lesion can be asymptomatic or there may be variable neurologic symptoms. In the literature there are frequent references to a bruit over the carotid.