Nishimoto-Takeuchi-Kudo disease: case report

K. URBÁNEK, H. FÁRKOVÁ, AND E. KLAUS

From the Clinic of Neurology and Clinic of Paediatrics, University Hospital, Olomouc, Czechoslovakia

SUMMARY The case is reported of a 7-year-old Czech boy with an obliterating disorder of the large intracranial arteries with formation of an extensive collateral network of blood vessels of the 'rete mirabile' type. It is analogous to cases described under the names 'abnormal cerebrovascular network', 'occlusion of the circle of Willis', or the 'moyamoya disease' by Japanese authors.

The Japanese authors Kudo (1968), Nishimoto and Takeuchi (1968), and Suzuki and Takaku (1969) have recently drawn attention to a particular vascular disorder characterized by narrowing or occlusion of both internal carotid arteries at the level of the siphon, combined with a highly specific abnormal cerebrovascular network at the base of the brain which evidently plays the role of a collateral circulation. The disease has been described under different names such as 'telangiectasia', 'vascular malformation', 'occlusion of the circle of Willis', 'abnormal cerebrovascular network', or the 'moyamoya disease'. It is most frequently manifested in juveniles, as a rule by a transitory hemispherical paretic symptomatology, attacks of unconsciousness, or by convulsive seizures.

While more than 100 cases of this vascular affection have been reported in Japan, analogous findings are almost unknown in other countries. Weidner, Hanafée, and Markham (1965) and Leeds and Abbott (1965) presented similar case reports in their studies on the intracranial collateral circulation. Both communications were written without familiarity with the Japanese literature and it is of still greater interest that all three cases referred to American-born Japanese. On the basis of these data, Kudo (1968) considers the disease to be a disorder confined to the Japanese race.

Recently, similar angiographic findings have been reported by Taveras (1969) in non-Japanese cases. This vascular disorder has been reported without stating the race and clinical picture under the name 'kapilläre Teleangiektasie', by Krayenbühl and Yasargil (1965), in their textbook. Simon, Sabouraud, Guy, and Turpin (1968) demonstrated at the Société Française de Neurologie a case of 'Nishimoto

disease' in a 10-year-old French boy with typical angiographic findings in both carotid systems.

We had the opportunity of examining a child showing all the typical signs of this vascular disorder. We would like to present our findings in this communication, as this is one of the few non-Japanese cases.

CASE REPORT

A 7-year-old boy was admitted to the Clinic of Paediatrics on 29 September 1969. He comes of a Czech family, none of the members of which had visited Asia. His mother is of below-average intelligence as is his 12-year-old sister. His 3-year-old sister shows retarded psychosomatic development; she has not yet begun to speak. The patient's birth and further post-natal development was uneventful according to his mother, but he had to repeat the first class of elementary school because of a bad school record. In March and July 1969 he experienced paroxysms of unconsciousness without seizures. On 25 July 1969, he awoke in the night and complained of pains in the left extremities. In the morning his parents found that the boy was hobbling on the left leg. His condition improved during the week but on walking and running the child fell frequently towards the left side. He was examined in the Department of Paediatrics of one regional hospital and because of a pathological EEG finding he was referred to this clinic. Examination revealed a slight psychological and somatic retardation and a mild left hemiparesis with increased tendon reflexes. Slight muscular hypotonia and spastic pyramidal signs were present in the left extremities. A complete blood count, sedimentation rate, urinalysis, serum antistreptolysin-O titre, and serum lipids were within normal limits. Serum protein electrophoresis showed total protein 7.2 g/100 ml., albumin 56%, α_1 globulin 7.3%, α_2 globulin 15%, β globulin 8.7%, γ globulin 13%, and A/G ratio 1:27. The serum protein immunoelectrophoresis showed in-

creased β -lipoprotein and absent IgM fraction. Routine examination of the cerebrospinal fluid (CSF) was within normal limits. The Wassermann reaction in the blood and CSF, the latex-RA test, and the LE-cell test were negative.

Radiographs of the skull were normal. Pneumography revealed internal hydrocephalus of the lateral ventricles with the left larger than the right and a dilated third ventricle, the transverse projection of which was 13 mm. The electroencephalogram showed generalized slowing with dominant diffuse theta and delta activity maximal in the right temporal area. The result of ^{99m}Tc -pertechnetate brain scanning was negative. Bilateral carotid angiography disclosed almost identical appearances on each side (Fig. 1). There was considerable slowing of the circulation, an undulated extracranial portion of the internal carotid artery, and tapered stenosis of its supraclinoid segment. The proximal portion of the middle cerebral artery was thin and its distal course was not visualized. A very thin anterior cerebral artery showed poor branching. A hypertrophic ophthalmic artery had a tortuous middle portion and exhibited no visible collaterals with the external carotid artery. The greatest amount of contrast medium was localized in a non-homogeneous hazy formation between the anterior and middle cerebral arteries, consisting of a collection of tortuous and disorganized small blood vessels, forming a picture like a bush or a bouquet. Vertebral angiography (Fig. 2) demonstrated slowed circulation, a weakly outlined trunk of the basilar artery with irregularities. In the area of the cortical portions of the posterior cerebral artery there were similar fused vascular clusters resembling clouds of smoke. This was most evident in the region of the posterior choroidal arteries. The cerebellar areas were considerably avascular, only the posterior inferior cerebellar artery was strongly outlined. Some contrast

FIG. 1 Left carotid angiogram.

FIG. 2 Vertebral angiogram.

medium was localized in irregular foci in the nutritive region of the anterior and middle cerebral arteries.

DISCUSSION

There is no doubt that our case is identical with the vascular disorder described by Japanese authors. Taking into account the papers of Taveras (1969) and Simon *et al.* (1968), the disease can then be considered as not being exclusive to the Japanese race. As for the aetiology, previous authors have discussed the possibility of congenital vascular disorder or of a certain form of arteritis. The assumption of congenital dysplasia is particularly disproved by those cases which could be followed up for several years. Undoubted progressive changes were found in all of them (Suzuki and Takaku, 1969). It is thus in all probability a chronic progressive disease with a tendency to advance in attacks which can, however, cease at any stage. The findings of normal values of body temperature, pulse rate, erythrocyte sedimentation, blood picture, and of the CSF examination argue against an arteritic process. In our opinion it is possible that the clinical manifestation of the illness is a sign of a functional decompensation of a not quite adequate collateral circulation which need not coincide directly with the active phase of an arteritic process. Even secondary changes can participate in its origin, such as mural thromboses and sclerotic foci described in a few necropsy cases as well as intimal thickening of the arterial wall (cited by Nishimoto and Takeuchi, 1968) which may be considered a result of endarteritis. We think that the cause and initial stages of the disease must thus be looked for

in the infant and, perhaps, even in the prenatal period.

The angiographic picture of this disease is characteristic to such a degree that it can hardly be confounded with another disorder. The rich 'moyamoya' vessels might be considered to be pathological vasculature in a neoplasm. Stenosing or even obliterating changes in large intracranial arteries are, however, a marked manifestation pointing to vascular origin of the disease. In an angiomatous malformation, hypertrophy of afferent blood vessels, acceleration of the circulation, and a rapid filling of the venous system would be expected. The pneumographic picture is non-specific but by demonstrating a possible dilatation of the ventricles it can provide information about the extent and localization of changes in the cerebral parenchyma. Such a pneumographic finding together with normal values of the cerebrospinal fluid associated with a certain focal hemispherical symptomatology may lead to the assumption of perinatal cerebral palsy. We believe that a number of cases of the 'moyamoya disease' have been diagnosed as cerebral palsy if angiography was not performed.

REFERENCES

- Krayenbühl, H., and Yasargil, M. G. (1965). *Die zerebrale Angiographie*. Thieme: Stuttgart.
- Kudo, T. (1968). Spontaneous occlusion of the circle of Willis. A disease apparently confined to Japanese. *Neurology (Minneapolis)*, **18**, 485-496.
- Leeds, N. E., and Abbott, K. H. (1965). Collateral circulation in cerebrovascular disease in childhood via rete mirabile and perforating branches of anterior chorooidal and posterior cerebral arteries. *Radiology*, **85**, 628-634.
- Mount, L. A., and Taveras, J. M. (1957). Arteriographic demonstration of the collateral circulation of the cerebral hemispheres. *Arch. Neurol. Psychiat. (Chic.)*, **78**, 235-253.
- Nishimoto, A., and Takeuchi, S. (1968). Abnormal cerebrovascular network related to the internal carotid arteries. *J. Neurosurg.*, **29**, 255-260.
- Simon, J., Sabouraud, O., Guy, G., and Turpin, J. (1968). Un cas de maladie de Nishimoto. A propos d'une maladie rare et bilatérale de la carotide interne. *Rev. neurol.*, **119**, 376-383.
- Suzuki, J., and Takaku, A. (1969). Cerebrovascular 'moyamoya' disease. Disease showing abnormal net-like vessels in base of brain. *Arch. Neurol. (Chic.)*, **20**, 288-299.
- Taveras, J. M. (1969). Multiple progressive intracranial arterial occlusions: a syndrome of children and young adults. *Amer. J. Roentgenol.*, **106**, 235-268.
- Weidner, W., Hanafey, W., and Markham, C. H. (1965). Intracranial collateral circulation via leptomeningeal and rete mirabile anastomoses. *Neurology (Minneapolis)*, **15**, 39-48.