Matters arising

gomyelia may have been secondary to the abnormality at the craniocervical junction which may prevent the normal egress of CSF from the fourth ventricle. There was no history of any difficulty in pregnancy or birth, infection or trauma that may have resulted in adhesions in this area. As discussed by Busis and Hockberg, the possibility of this condition being inherited must be entertained.

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Isolated central facial palsy

Sir: With great interest we read Drs Huang and Broe's article Isolated facial palsy: a new lacunar syndrome. They reported three cases of isolated upper motor neuron facial palsy without significant limb weakness. Two of their cases showed some degree of dysarthria and dysphagia. Their CT scans were consistent with small infarcts in the region of the corona radiata bilaterally in one case, the right corona radiata in another case, and the anterior corona radiata bilaterally in the third case. Recently we encountered such a case of central facial palsy with mild dysarthria and clumsiness of the hand. The CT scan showed a small infarct in the region of the left corona radiata as in their cases. We believe the symptoms of our case and their cases are identical to those of Fisher's dysarthria-clumsy hand syndrome, as shown in our case reported below.

A 58-year-old right-handed man developed sudden dysarthria on 20 July 1984. A right central facial palsy with dysarthria was noted and treated as a cerebral infarction by his doctor. Five days later, he visited our hospital for CT scan. On examination he showed right central facial palsy and mild dysarthria and showed normal tongue and soft palate movement. Tendon reflexes and strength of extremities were normal. He complained of difficulty writing, although his writing appeared normal. CT scan was consistent with a small infarct in the region of the left corona radiata (fig). On 5 September, his symptoms improved. CT scan was unchanged.

In 1967, Fisher reported 20 cases of the dysarthria-clumsy hand syndrome. He stated, "The cardinal features of our cases have been: moderate to severe dysarthria; central facial weakness on one side of the face; deviation of the tongue on protrusion; impairment of la-la-la; a trace of dysphagia; clumsiness, awkwardness, and slowness of the manipulations of the affected hand; questionable weakness of the hand; difficulty in writing..." One pathologically studied case revealed a pontine laque, which was responsible for the symptoms, he said. He speculated that a similar syndrome might also result from a lesion in the internal capsule or corona radiata. Spertell and Ransom reported a case of the dysarthria-clumsy hand syndrome associated with acute infarction of the genu of the internal capsule. Although Huang and Broe did not mention Fisher's report, we consider their cases and our case to be examples of his speculation.

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Fig CT scan without contrast enhancement; 5 days after onset. Arrow indicates low density in the left corona radiata.

References


Huang and Broe reply:

Although Fisher was less than exact in his original definition of the clumsy hand later reports have interpreted the hand movement defect as being incoordination. Fisher has supported this interpretation subsequently by classifying the syndrome as a variant of the ataxic-hemiplegia syndrome. Supranuclear facial palsy is seen in both the dysarthria-clumsy hand syndrome and the faciobrachial variant of pure motor stroke. In view of the fact that the patient was seen at a stage when no weakness or incoordination was present and difficulty in writing could result from either syndrome in the resolving phase, it is difficult to be certain whether the case described by Dr Nishimoto et al should be classified as the former or the latter. Involvement of the hand would however exclude isolated supranuclear facial palsy which should only be diagnosed when the patient has deficits limited to the face and bulbar muscle and has no limb involvement. Whilst pure motor stroke is due to a lesion limited to the corticospinal tract, ataxia would imply involvement of the corticobulbar pathways. Although the lacunar syndromes share many common features, distinction between each is necessary if useful anatomico-clinical correlation is to be made.

References