The operculum syndrome

A case report

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Summary

A 48-year-old hypertensive man with signs and symptoms of pseudobulbar paralysis was ultimately found to be suffering from the operculum syndrome. Recognition of this syndrome is important in that: (*a*) it may be caused by a localized cortical lesion amenable to treatment; (*b*) some of the symptoms carry a good prognosis as opposed to those in pseudobulbar paralysis; and (*c*) the operculum syndrome usually has a more favourable prognosis since there is no associated mental impairment or sphincter disturbances.

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The operculum insulae is a small region of the telencephalic mantle which covers the insula (Reili). Its connections and functions have been only partially clarified, but the clinical symptomatology of opercular lesions has been well documented; however, studies of these lesions since 1837 have been almost exclusively carried out by the French; the syndrome appears to have escaped Anglo-Saxon attention.

Case report

A 48-year-old hypertensive man presented with an 18-month history of dysphagia, dysarthria and dysphonia. His hypertension was longstanding and he had been on various medications in the past. He had been well until 2 years previously when he had had an acute cerebrovascular accident which left him with mild right-sided hemiparesis and mild expressive dysphasia.

On examination there were no abnormal cardiovascular, respiratory or abdominal signs. The peripheral pulses were present and there were no carotid bruits. His blood pressure was 175/105 mmHg, and the pulse rate 72/min and regular. He had no neck stiffness and was well orientated in time and space.

Funduscopy revealed stage II hypertensive retinopathy but the optic disc appeared normal. Frontal lobe reflexes were exaggerated and the glabellar tap was positive. Pupillary responses were present and his eye movements were full, with some limitation of upward gaze. The corneal reflex was present bilaterally. There was mild bilateral ptosis. Voluntary movements of the facial muscles were frozen and he was unable to show his teeth on command but was able to smile and laugh at a joke. He was unable to wrinkle his forehead on upward gaze or to close his eyelids tightly on command. However, the eyelids closed easily when tested for the presence of the corneal reflex or during sleep. Hearing and understanding were intact. His jaw muscles were weak bilaterally and he was drooling from one side of his mouth. There was no masseter atrophy and his jaw jerk was brisk. His tongue was small and immobile and there was no atrophy or fasciculations. Chewing and swallowing were weak and the gag reflex was absent. His speech was dysarthric and dysphonic and he had mild expressive dysphasia. The sternomastoid muscles were weak bilaterally. Muscle tone was increased bilaterally, perhaps slightly more on the right and a positive Babinski sign was present on the right. Hoffmann's reflex was negative bilaterally. His gait was slow with no ataxia. There was no mental impairment and no sphincter disturbance. Pathological crying and laughing were not observed. There was no sensory loss and his cerebellar functions were intact.

The cerebrospinal fluid was normal. Computed tomography (CT) revealed prominent basal cisterns, marked tissue loss in the region of the sylvian fissure and insulae bilaterally, several low-density areas in the left frontal and right occipital regions (multiple infarcts) and moderately dilated ventricles. The tissue loss involved predominantly the operculum insulae (Fig. 1).

In summary, the patient had: (*i*) signs of pseudobulbar palsy involving the 5th, 7th and 9th through 12th cranial nerves, associated with marked automatovoluntary dissociation, absence of mental impairment and sphincter disturbances and absence of pathological crying and laughing; (*ii*) marked dysarthria, dysphonia and expressive dysphasia; (*iii*) facial weakness including the upper portion of the face and forehead muscles; (*iv*) no signs of muscle denervation and no palatal and pharyngeal reflexes; and (*v*) had previous cerebrovascular accidents and marked tissue loss of both operculum regions as evidenced on CT.

Discussion

The opercular syndrome, being the cortical type of pseudobulbar palsy, includes the clinical manifestations of pseudobulbar palsy of the 5th, 7th and 9th through 12th cranial nerves. There are, however, specific features that distinguish it from pseudobulbar palsy. The operculum syndrome may follow unilateral lesions, while a transient and predominantly distal pyramidal-type paresis of the arm on the same side as the cranial nerve involvement invariably accompanies it. Lesions of the parietal operculum may also cause hemi-anaesthesia. However, the dissociation between the discrete sensory symptoms and the marked motor deficit of the 5th cranial nerve suggests a supranuclear location of the lesion. A further remarkable feature is that the central-type facial palsy includes the upper portion of the face. Mental symptoms or sphincter impairment never occur, in contrast with pseudobulbar palsy, and pathological crying and laughing are rare. The patient's history often reveals a cerebrovascular accident (in the great majority of cases either embolic or thrombotic) leading to well-defined encephalomalacia. The patients are usually hypertensive and over 50 years of age. The opercular syndrome ultimately results from an acute attack (although it may appear gradually) and the signs and symptoms characteristically regress within a few days.

It has been suggested that ischaemia in the area of the smaller sylvian branches of the middle cerebral artery is the immediate cause[1] but other causes have been reported, e.g. astrocytoma and meningo-encephalitis.[1]

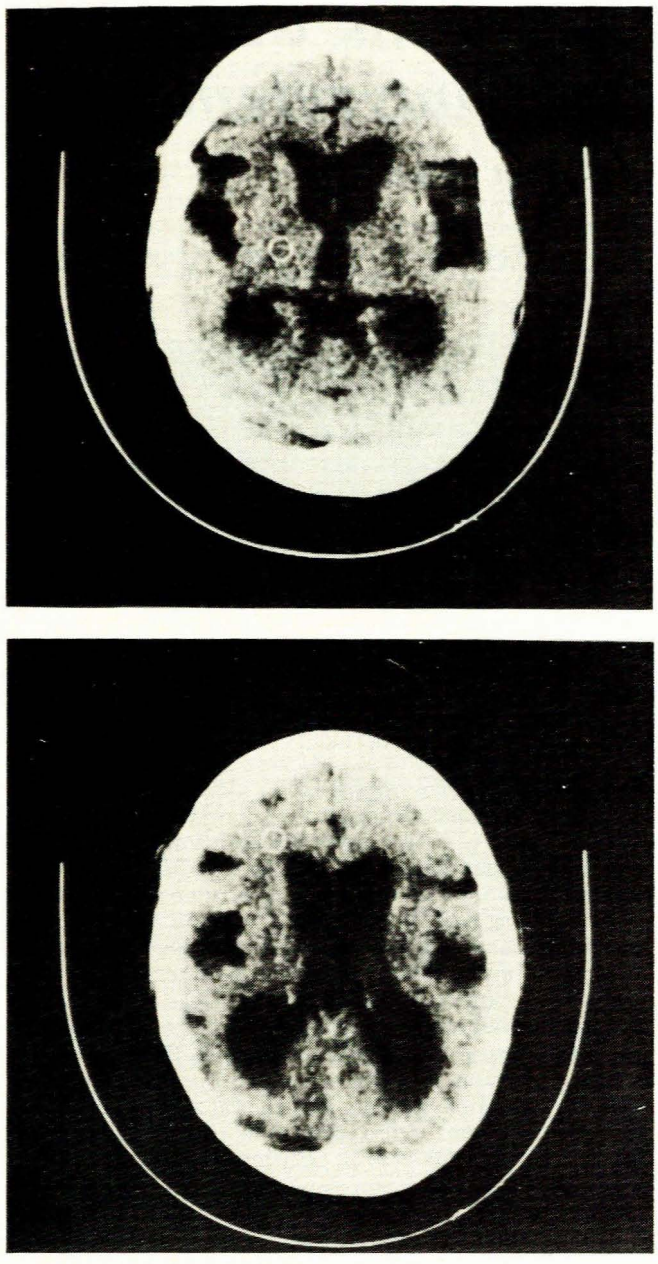

Fig. 1. CT reveals prominent basal cisterns, marked tissue loss in the region of the sylvian fissure and insulae bilaterally and moderately dilated ventricles. The tissue loss involves predominantly the opercular regions bilaterally.

Recognition of the operculum syndrome not only offers an opportunity for the study of cortical functions but is also of practical importance since it may point to a localized cortical lesion (such as a tumour) in the operculum area as opposed to the bilateral lesion of pseudobulbar palsy, and since some of the symptoms, such as impaired deglutition (salivation, difficulty with eating), tend to regress rapidly in contrast to their prolonged presence in patients with pseudobulbar palsy. Finally, the operculum syndrome usually has a better prognosis than pseudobulbar palsy since there is no associated mental impairment or sphincter disturbance. The operculum syndrome, mainly analysed by the French school of neurology, still requires detailed investigations which may well be correlated with the extent of the lesion visualized on CT.

REFERENCE

1. Bruyn GW, Gathier JC. The operculum syndrome. In: Vinken PJ, Bruyn GW, eds. *Handbook of Clinical Neurology*, vol. 2. Amsterdam: North-Holland, 1969: 776-783.