PAPER

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The opercular syndrome, or, the Foix-Chavany-Marie syndrome

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ABSTRACT The opercular syndrome is due to bilateral, and sometimes unilateral, lesions of this area of the cerebral cortex, or its connections with the brain stem. The symptoms and signs of the syndrome are described, especially the type of facial palsy. They differ slightly in childhood, and may be accompanied by developmental delay. They can be reversible when linked to epileptic activity in the opercular areas. The symptoms can be suggestive of two well-established syndromes in childhood. Firstly, the Landau–Kleffner syndrome (and there may well be shared pathophysiological mechanisms) and secondly, those first described by Worster–Drought.

The investigations needed to establish the location and the possible causes of the opercular syndrome are discussed. The most common causes are vascular lesions, but other possibilities such as infections and trauma have to be considered. Also malformations of prenatal origin can result in the suprabulbar palsy so typical of the syndrome. If the cause can be established there is always the possibility of treatment.

KEYWORDS Causes, diagnosis, investigations, opercular syndrome, symptoms, treatment

LIST OF ABBREVIATIONS Computed tomography (CT), (EEG), (MRI)

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INTRODUCTION

The opercular syndrome is characterised by diminished voluntary control of facial, pharyngeal, lingual, and masticatory muscles. It usually follows bilateral lesions of the anterior opercular area surrounding the insula formed from gyri of the frontal, temporal and parietal lobes; and causes a cortical type of pseudobulbar palsy. It is most frequently the result of vascular lesions, infections of the nervous system or cortical dysplasias, although it can occur as the result of tumours, trauma,[1] and degenerative diseases.[2] In the case of a developmental lesion, the pathology has been described as polymicrogyria and perivascular grey-matter heterotropias or disorders of migration.[3] The syndrome can occur after unilateral lesions, but then the symptoms and signs are usually unilateral resembling multiple cranial nerve palsies.[4] This can lead to diagnostic difficulties, since in the opercular syndrome, the facial paralysis may not demonstrate 'forehead sparing' so that the paralysis is thought to be of peripheral origin.[5] A right-handed patient has been described who developed a unilateral opercular syndrome after a stroke, and on investigation with CT was found to have no evidence of a cortical lesion, but a subcortical area of decreased density involving the deep white matter of the left hemisphere reaching the head of the caudate nucleus.[6] A patient with a supranuclear bulbar palsy due to a subcortical degenerative lesion has been reported by Weller *et al.*[7] Special reference will be made to other syndromes in childhood: the Worster–Drought syndrome and the Landau–Kleffner syndrome.

SYMPTOMS AND SIGNS OF THE OPERCULAR SYNDROME

The affected patient may be unable to speak, and the mouth may be held half open with a tendency to drool and an inability to chew or to move the tongue. In spite of a lack of voluntary facial movements some involuntary movements are preserved. The typical suprabulbar palsy is characterised by this type of facial palsy, and by dysphagia, dysarthria, and often by hypophonia as well. Examination can confirm the poor movements of the facial muscles, and of the palate and tongue. Seizures are often an important part of the syndrome's presentation, especially when the symptoms fluctuate.

Nisipeanu *et al*[8] have reported an elderly man who had suffered all his life from worsening dysphasia, as well as from dysarthria and facial palsy. Computed tomography and MRI scans showed bilateral opercular lesions, and the patient was diagnosed as an example of the congenital form of the syndrome.

The clinical picture in children can differ slightly, as stressed by Gropman *et al.*[9] Developmental delay is a feature, and they also listed poor palatal function, hypotonia, arthrogryposis, hemiparesis, apnea, paraparesis, micrognathia, pectus excavatum, hearing loss, and epilepsy, but many of these findings will depend on the extent of the responsible lesion, but the symptoms of suprabulbar palsy are essential for the diagnosis at any age.

The opercular syndrome can be reversible as was the case in a patient reported by Fusco and Vigivano.[4] A girl with a left hemimegalencephaly suffered from right-sided seizures. The seizures had worsened, and the epileptic activity had spread from the left cerebral hemisphere to the contralateral central regions. She then lost voluntary control of facial, pharyngeal, lingual, and masticatory muscles, and her general condition deteriorated; so it was decided to perform a left hemispherectomy. Soon after surgery, the epileptic features and the symptoms of the opercular syndrome disappeared. A patient with a similar history has been reported by Shuper *et al.*[10] However, the syndrome can become permanent, as was the case in the patient recorded by Pascual-Castroviejo *et al.*[11] She developed severe epilepsy at the age of five-and-a-half, and when aged sixteen still showed evidence of the opercular syndrome, and still suffered from epilepsy. The epilepsy was considered to be the cause of her symptoms, although Grattan-Smith *et al*[12] thought that it might have been due to herpes simplex encephalitis.

A transient opercular syndrome has also been recorded after the surgical removal of a glioma from the right opercular area in a patient who had suffered a previous head injury which had caused cerebral damage on the left side. The patient fully recovered, but when there is the possibility of bilateral cortical damage in the opercular areas the risks of precipitating the syndrome should be considered.[13]

Very occasionally, after bilateral cortical lesions, a patient may show a selective loss of emotional facial movements with preservation of voluntary movements, and an otherwise typical opercular syndrome. As voluntary and automatic facial movements have distinct pathways this unusual finding must be due to the exact area of the cortex which is affected.[14, 15]

POSSIBLE RELATIONSHIPS BETWEEN SYNDROMES IN CHILDHOOD

There do seem to be recognisable overlaps between the opercular syndrome and other more easily recognised syndromes in childhood. Shafrir and Prensky[16] described a child who suffered from the opercular syndrome, the symptoms of which fluctuated in parallel with the severity of the epileptic seizures. This relationship particularly applied to language impairment, and during the exacerbations, the EEG showed electrographic status epilepticus during slow wave sleep. Some similarities to the Landau–Kleffner syndrome were noted, and it was suggested that there could well be a shared pathophysiological mechanism between the two syndromes. This might be due to disruption of normal connections or to an excessive inhibitory reaction to epileptiform discharges.

Christen *et al,*[17] when describing the clinical findings in a group of children with the opercular syndrome, identified the causes as a meningoencephalitis, a bilateral dysgenesis of the perisylvian region, and an intermittent form correlating with epileptic seizures, the last two having a common pathogenisis. The symptoms and signs were those of the suprabulbar paresis described by Worster–Drought,[18] and it was suggested that the opercular syndrome in childhood should in fact be known by his name.

Another study on eleven affected children by Rolland *et al*[19] found that six presented with suprabulbar palsy, seven with various motor disorders, six with severe learning disorders, and four with epilepsy. Malformations were demonstrated by MRI in all of them, and it was thought that these were caused by abnormalities of antenatal development between the twentieth and twenty-fourth weeks of pregnancy.

DIAGNOSIS OF THE OPERCULAR SYNDROME

The opercular syndrome differs from supranuclear palsy in the lack of sphincter disturbances, the rarity of pathological laughing, the decreased tone of the affected muscles, and the abolished gag reflex; and it differs from bulbar palsy in the lack of fasciculation, atrophy and denervation, and in the preservation of involuntary innervation and of reflexes except for the gag reflex.[20]

The opercular syndrome may have to be diagnosed from other syndromes in which the opercular cortex is involved but the cerebral damage is more widespread, such as the opercular cheiro-oral syndrome. In this disorder there is additional sensory impairment over the upper limb due to infarction affecting more of the territory of the middle cerebral artery.[21]

INVESTIGATIONS OF THE SYNDROME

The EEG can show focal abnormalities in the opercular areas, and continuous spike and wave activity during slow sleep may coincide with episodes of the syndrome, when these are transient as in the acquired epileptiform opercular syndrome.[22]

Computed tomography scans may be unhelpful, but MRI is likely to be more sensitive in showing opercular damage, depending on the aetiology.[23] Also the single photon emission computed tomoraphy may help to

differentiate infarction from primary cortical atrophy by showing if there is evidence of cerebral hypofunction.[22] This test can also show localised high perfusion in the acquired epileptiform opercular syndrome,[24] and in cases in which the MRI scan demonstrates only a unilateral lesion it can reveal a disturbance on the contralateral side due to diaschisis.[25]

CAUSES OF THE OPERCULAR SYNDROME

There are a variety of causes, such as vascular lesions, particularly in adults,[26] and including moyamoya disease,[27] infections of the nervous system, for example herpes simplex encephalitis,[28] and toxoplasmosis secondary to AIDS;[29] acute disseminated encephalomyelitis, trauma, perinatal difficulties; and perisylvian cortical dysplasias due to disorders of heuro migration which are under genetic control.[30] The operculum includes the area covering the island of Reil, and is made up of parts of the frontal, parietal, and temporal cortex, and any lesion which disrupted the connections between these areas and the brain-stem, containing the relevant cranial nerve nuclei, will cause the suprabulbar palsy of the opercular syndrome. This also explains the preservation of emotional and automatic bulbar reflex movements, which must use other neural pathways.[23]

Yamamoto *et al*[30] reported four patients with the opercular syndrome due to perinatal difficulties. They showed evidence of suprabulbar palsy, as well as severe learning disorders. Two of them had suffered from fetal bradycardia during labour, and had had neonatal convulsions and subarachnoid haemorrhages, one other had neonatal convulsion and a subarachnoid haemorrhage, and one of the babies with fetal bradycardia and one other had vacuum extractions. They all showed bilateral cortical atrophy in the opercular regions on MRI studies, and single photon emission CT confirmed the presence of hypoperfusion in these areas, which justified the diagnosis of the acquired type of the opercular syndrome. Koeda *et al*[31] reported a similar patient with the opercular syndrome due to perinatal difficulties.

Among the possible infections that can result in the opercular syndrome, especially in childhood, is herpes simplex encephalitis, as reported by Wolf *et al*,[32] the diagnosis being made by MRI and elevation of oligoclonal antibodies specific to herpes simplex virus in the cerebrospinal fluid.

If the cause is a degenerative disorder such as Pick's disease, the opercular syndrome can be progressive over a number of years.[33] Toriumi *et al*[34] have described a patient who suffered from both the opercular syndrome and distal myopathy with rimmed vacuoles, the latter condition being confirmed by genetic analysis. They speculate that the opercular syndrome may have been caused by destructive events *in utero* or in the perinatal period, but also that there is a possibility that the brain abnormalities were due to a gene mutation.

CONCLUSIONS

It seems that any combination of cortical or subcortical lesions of the operculum or its connections on both sides of the brain can present the symptoms and signs that merit their inclusion within the rubric of the opercular syndrome.[2] Establishing the exact site of the lesion is obviously important, and essential in identifying its nature, in the hope that this will lead to effective treatment.

The hallmarks of the syndrome are anarthria, and bilateralcentral facio-linguovelo-pharyngeo-masticatory paralysis with 'automatic voluntary dissociation'.[35]

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