Opercular syndrome: Case reports and review of literature

Thapa Lekhjung, Paudel Raju, Rana PVS

Department of Neurology, College of Medical sciences-Teaching Hospital, Bharatpur, Chitwan, Nepal

Abstract

Five cases of “bilateral opercular syndrome” of vascular etiology are reported. Cortical pseudobulbar palsy (i.e. spastic anarthria and inability to swallow) with dissociation of automatic voluntary movements in the affected muscles are the essential features of this syndrome. Additional motor and sensory symptoms differentiate its subtypes. All 5 patients had bilateral opercular syndrome. The unusual features was its occurrence as the presenting feature of preeclampsia in a young lady, and the development of the transient syndrome following a right focal seizure with generalization at high altitude in a young female trekker who had an old unilateral infarct in left opercular region. Whereas the limb motor weakness recovered well, the recovery was unsatisfactory for speech and for swallowing.

INTRODUCTION

The opercular syndrome is a rare disorder due to bilateral lesions of opercular cortex surrounding the insula, which is separated by the ascending and the posterior rami of the lateral sulcus into (a) Frontal operculum formed by posterior part of the inferior frontal gyrus (i.e. pars-triangularis, pars-opercularis and even by the caudal portion of the pars-orbitalis); (b) Fronto-parietal opercula formed by the lowermost part of the precentral and postcentral gyrus and the anterior and lowermost part the inferior parietal lobule; and (c) Temporal opercula formed by the superior temporal gyrus. The syndrome was first described by Magnus in 1837 and is also known as Foix-Chavany-Marie syndrome (FCMS), facio-labio-glosso-pharyngeal-branchial paralysis or cortical type of pseudobulbar paralysis. Further, it is classified based on the site of lesion as bilateral anterior opercular syndrome (lesion in both anterior or frontal operculum),1,5 opercular-subopercular syndrome (lesion in opercular cortex one side and the subopercular lesion on the other side),6 subopercular syndrome (lesions in subcortical corticobulbar projections only)4,7, unilateral anterior syndrome involving frontal operculum1,4, and posterior syndrome involving frontoparietal operculum.1,8 Its manifestations includes central type of facial paresis, difficulty in opening of mouth, chewing the food, protrusion of tongue and inability to speak and swallow. Additional features include trismus9, ageusia10 and pseudo-opthalmoplegia (i.e. ptosis with weakness of conjugate gaze to opposite side and deviation of head and eyes to the side of lesion). In addition unilateral anterior syndrome is reported involving frontal operculum and presenting with contralateral and upper limb paresis and inability to speak (Piere Marie’s anarthria).1 It differs from bulbar palsy by preservation of jaw jerk, pharyngeal reflex and by the absence of fasciculation, atrophy and phenomenon of denervation and unlike pseudobulbar palsy the pathological laughter and emotional disturbances are lacking.

The etiology in most of the reported cases is vascular (thrombosis or embolism) involving branches of middle cerebral artery supplying the opercular area.1,5 Other lesions producing the syndrome include astrocytoma11, developmental bilateral perisylvian cortical dysplasia12,13, herpes simplex encephalitis14-16, progressive supranuclear motor system degeneration17, bilateral toxoplasmosis in AIDS18, and in multiple sclerosis.19 Weller et al20 reviewed 62 cases and classified opercular syndrome based on etiology; i.e. (a) Classical form most often related to vascular etiology; (b) Subacute form due to central nervous system infections; (c) Developmental form most often related to neuronal migration disorders; (d) Reversible form in children with epilepsy; and (e) Rare type related with neurodegenerative disorders. Some authors consider Foix-Chavany-Marie syndrome in children a different entity.21 We report this rare syndrome with some previously unreported observations with review of the literature.
CASE REPORTS

Patient 1
A 35 years old lady, a resident of U.K, while trekking in Nepal at height of 3,000-4,000 meters, had a right focal seizure with secondary generalization lasting for 10-15 minutes. Following this episode she developed difficulty in swallowing and inability to speak and mild weakness on the right half of the body predominantly affecting the upper limb. She had an ischemic stroke 2 years back, manifesting with right hemiplegia and non-fluent aphasia. She was diagnosed as left middle cerebral artery infarction with no evident risk factors for stroke. She was treated conservatively with aspirin and made full neurological recovery. At admission she was afebrile, with BP 140/90 mms Hg, pulse 64/minute. Neurological examination revealed normal higher mental functions. She could not vocalize but, she could follow spoken and written command and could copy normally. She could recognize and name the objects. She could not open the mouth or protrude the tongue or swallow. However, opening of mouth was noted during yawning and laughing. She also had facial paresis of central type, sluggish palatal and pharyngeal reflexes, pronator drift in right arm, brisk deep tendon reflexes and extensor plantar response on the right side. Fundus and other examinations were normal. Hematological and biochemical parameters, ECG and echocardiography were normal. EEG revealed focal slowing and epileptic discharges from left frontotemporal region. CT scan (Figure 1) revealed old opercular infarcts in left opercular and sub opercular areas with ipsilateral dilatation of lateral ventricles. Carbamazepine was added to aspirin. When transferred back to U.K. after 7 days, she had marked improvement in her speech but only partial improvement of swallowing.

Patient 2
A 24 years old lady, a primigravida with 6 months history of amenorrhea, was admitted with acute neurological illness manifesting with moderately severe global headache and inability to speak.
There was no history of loss of consciousness, vomiting, diplopia, seizures, transient ischaemic attack or of a similar episode in the past. She denied history of trauma, fever or vaccination, diabetes mellitus and hypertension. She was not a booked case and had not attended antenatal clinic services. At admission she was afebrile with pulse of 110/min, BP 180/124 mm Hg, respiration 24/min and had mild bilateral pitting edema. No icterus, cyanosis or lymphadenopathy were detected. She was conscious and responding to commands. However, she could not vocalize. She could not open her mouth, protrude her tongue or swallow. Involuntary movements were preserved. She had central facial paresis, mild hemiparesis (upper limb affected more than lower limb) and brisk reflexes on right side and extensor plantar response on both sides. All other systems including the cardiorespiratory system were normal. The fundal height was commensurate with the period of amenorrhea. Evaluation for risk factors for ischemic stroke was negative. Except for slight albuminuria and mild rise in hepatic enzyme, other liver functions, hemogram, metabolic parameters, coagulation profile, plasma fibrinogen level, ECG and ultrasonography of abdomen were normal. Antinuclear factor and tests for antiphospholipid antibodies were negative. CT scan (Figure 2) revealed large bilateral cortical infarct in the posterior frontal region involving both opercular areas. She was treated as a case of severe preeclampsia with magnesium sulphate, antihypertensive drugs (calcium channel blockers), aspirin, anticerebral edema measures and termination of pregnancy. Anticonvulsant was added later for seizure prophylaxis. She improved gradually and when discharged after three weeks, she could speak a few syllables but the swallowing did not improve.

Patient 3
A 75 years old lady, a known case of atrial fibrillation, presented with one day history of weakness right half of the body (upper limb more than lower limb), inability to speak and swallow. There was no history of loss of consciousness, headache, vomiting or seizures. She had left sided weakness one year back from which she recovered and was ambulatory without support and independent for acts of daily living. At admission she was conscious, afebrile, with pulse 92/min.
irregularly irregular (pulse deficit of 15/minute) and BP 180/120 mm Hg. She could utter a few syllables and had preserved comprehension of spoken commands and could recognize the objects. Other neurological deficit included central facial palsy, inability to open the mouth and swallow, weakness of right grip, and deep tendon reflexes were brisk more on left side with extensor plantar response both sides. The fundus showed grade II hypertensive changes. No primitive reflexes could be elicited. The jaw jerk was normal. The involuntary facial movements were preserved. She had cortical sensory deficit on the right side. The hematological and metabolic parameters were normal. ECG revealed atrial fibrillation with rapid ventricular rate. Echocardiography revealed dilated left atrium with concentric left ventricular hypertrophy. No thrombus was detected. MRI revealed multiple infarcts in the left opercular, posterior parietal, occipital and right parietal region. Marked atrophy of sylvian cortex (arrow) was noted on right side. (Figure 3) She showed no improvement in speech and in swallowing over 4 weeks follow-up.

**Patient 4**

A 50 year old female, a known case of mitral stenosis, was admitted with history of acute onset instability of gait, truncal ataxia and left hemiparesis of two days duration. There was no history of headache, vomiting, seizure, diplopia or loss of consciousness. She was a thinly built lady with BP 110/70 mm Hg, pulse 88/min, and respiration 20/min. She had gaze evoked non-fatigable nystagmus with fast component to left side, left hemiparesis (upper limb more
than lower limb), truncal ataxia with left sided cerebellar signs. She had marked difficulty in speaking. She could utter only a few syllables but could understand simple commands and could identify the objects. She could not protrude the tongue, open the mouth or swallow. Involuntary facial movements were present. Cardiac examination revealed mitral stenosis. Lung fields were clear. All other systems were normal. Hematological and metabolic parameters were normal. Echocardiography confirmed the valvular heart lesion with dilated left atrium. There were no vegetations or left atrial thrombus. CT brain scan revealed a left cerebellar infarct and a left perisylvian infarct and perifocal edema. MRI done two weeks later (Figure 4) showed additional right insular infarct. Brain stem and ventricular systems were normal. She was treated conservatively with aspirin and anti-cerebral edema measures. She improved gradually with marked regression of cerebellar signs. She could stand and walk with support but there was no improvement in speech and swallowing.

**Patient 5**

A 60 year old lady was admitted with acute onset weakness of the right half of the body. There was no history of loss of consciousness, seizure, headache or vomiting. One year back she underwent mitral valve annuloplasty. On admission her BP was 110/70 mm Hg. She had mitral regurgitation with atrial fibrillation. She could not speak and neurological examination showed right central facial paresis with right sided grade 4/5 weakness (right upper limb more than lower limb), anarthria, inability to open the mouth and protrude the tongue. She could not swallow. Her palatal and pharyngeal reflexes were normal. Echocardiography showed moderate mitral regurgitation. There was no evidence of clot, vegetations or pericardial effusion. CT scan (Figure 5) showed a small infarct in the right sylvian region and a large infarct in the left opercular region. At discharge two weeks later, the disabilities were persisting.

![Figure 4: MRI Flair image of Patient 4 showing bilateral lesion in opercular region](image-url)
DISCUSSION

All patients presented here had classical features of bilateral anterior opercular syndrome characterized by cortical pseudo-bulbar paralysis (facio-labio-pharyngo-glosso-masticatory paralysis) with automatic-voluntary dissociation as evident by preserved movements of facial muscles and opening of mouth during yawning and laughing. While the lesion was unilateral in one patient (Patient 1, Figure 1), the other 4 patients had typical bilateral opercular lesion (Figures 2 - 5) developing following an acute vascular episode. Additional subcortical infarcts were noted in 2 patients (Patients 1, 3).

An interesting feature was the development of transient bilateral opercular syndrome in a young female patient (Patient 1) following right focal seizure with generalization while trekking at 3,000-4,000 meters. CT brain scan (Figure 1) showed only a solitary old infarct in left opercular area. Many neurological syndromes develop at high altitude. This patient had no features of acute mountain sickness, cerebral or pulmonary edema. The onset and the transient character of the syndrome suggested that it was the result of seizure itself. It is likely that hypoxia at high altitude activated the latent epileptic focus from old left opercular infarct.

Seizures associated with bilateral anterior opercular syndrome are reported mostly in children. Pascal-Castroveijo et al. reported a child who developed bilateral anterior opercular syndrome following prolonged status epilepticus. A CT scan done before onset of status epilepticus was normal. A follow up MRI done afterwards revealed lesions in both parietal cortex and in corpus callosum. Features of opercular syndrome persisted even after control of seizures, suggesting epilepsy induced cerebral damage as the cause of bilateral anterior opercular syndrome. Tachikawa et al. reported a child who showed a prolonged episode characterized by almost continuous diffuse sharp and slow wave complexes during sleep and development of bilateral anterior opercular syndrome. The inter-ictal single photon emission tomography (SPECT) revealed a localized high resolution perfusion area in the left posterior frontal area. The computer assisted EEG analysis suggested that epileptic focus was on the left side which produced secondary bilateral synchronous sharp wave slow wave complexes. Clonazepam

Figure 5: CT brain scan of Patient 5 showing large left opercular infarct and a small infarct in right opercular region
therapy controlled both the seizure discharges and neurological deficits. Fusco and Vigevano\(^{25}\) reported a child with hemimegalencephaly who initially had right focal epilepsy. Later the epileptic discharges spread to left side producing bilateral opercular syndrome. Reversal of neurological deficits and control of seizure occurred after hemispherectomy. Transient bilateral opercular syndrome was also reported following surgical removal of glioma from the right opercular area in a patient who already had head injury induced lesion on left side. This patient recovered fully.\(^{26}\)

Our patient was similar to that reported by Tachikawa et al\(^{24}\) who also had a unilateral epileptogenic focus in left opercular area demonstrated by SPECT, while our patient had only an unilateral lesion (i.e. old infarct) in the left opercular area. Steiner-Birmanns et al\(^{27}\) reported a 55 years old patient with chronic renal failure and sub-opercular lacunar infarcts who developed bilateral opercular syndrome while on dialysis and maintaining normal hemodynamic status. His EEG showed a continuous seizure activity consistent with nonconvulsive status. No fresh lesion was seen in CT scan. Treatment with antiepileptic drugs led to reversal of EEG changes and complete neurological recovery. However, unlike our patient, lesions were bilateral and in the subcortical regions. A better prognosis can be predicted in cases where seizure is the cause of the syndrome. Patients with seizures and stroke developing at high altitude usually have a good prognosis on evacuation to lower altitude.\(^{28}\)

Bilateral anterior opercular syndrome with unilateral lesions have been reported.\(^{5,6,7}\) However, a report of the cases by Tothghi et al\(^{29}\) where a second lesion was missed on CT scan but detected later by MRI and SPECT cast doubt on strict unilaterality of the underlying lesion. This is further supported by Patient 4, where CT showed lesion on one side but follow-up MRI revealed lesions on both sides. While MRI can visualize lesions missed on CT scan, SPECT has the additional advantage that it can differentiate the infarction from cortical atrophy by showing a localized area of hyperperfusion during the ictal phase\(^{25}\), and reveals an area of diachisis in ischemic stroke.\(^{30}\)

Preeclampsia is usually apparent in the later part of the third trimester, but may develop earlier in the later half of the second trimester as in our Patient 2.\(^{31}\) It complicates approximately 3 to 14 percent of all pregnancies worldwide and about 3-8 percent in the United States.\(^{32}\) Common central nervous system manifestations of preeclampsia include headache, blurred vision, scotomata, and rarely cortical blindness. Cerebrovascular manifestations of severe preeclampsia are poorly understood, but may represent a form of reversible posterior leukoencephalopathy syndrome (RPLS). Brain imaging is normal in approximately 70% of patients with the reversible cerebral vasoconstriction syndrome, and the rest have border-zone ischemic strokes, parenchymal hemorrhage, vasogenic edema, and nonaneurysmal subarachnoid hemorrhage overlying the cortical surface\(^{33}\) with loss of cerebrovascular autoregulation and cerebral edema. The neuroimaging findings are similar to hypertensive encephalopathy where MRI usually shows T2 hyperintense signal intensity in white matter and a corresponding reduced density in CT scan (representing edema) usually concentrated in the posterior part of the cerebral hemisphere. Their presence upstages the diagnosis from mild to severe preeclampsia. The angiographic abnormalities in reversible cerebral vasoconstriction syndrome are dynamic, often subtle and typically resolve within three months.\(^{33,34}\) Most patients recover completely, although neurologic impairment (and even death) from progressive vasoconstriction, stroke, and brain edema has been reported.\(^{35}\) Pregnancy related stroke has an incidence of 34.2 cases per 100,000 deliveries.\(^{36}\) Incidence of stroke increases further with development of preeclampsia. Our Patient 2 had severe preeclampsia presenting as stroke where CT scan showed symmetrical large infarcts in both opercular regions. This was atypical as no lesions were noted in the parieto-occipital region and add another condition presenting as bilateral anterior opercular syndrome.

REFERENCES

6. Bakar M, Kirshner HS, Niaz F: Opercular-