Clinical Neurology and Neurosurgery 155 (2017) 12–13

Contents lists available at ScienceDirect

Clinical Neurology and Neurosurgery

journal homepage: www.elsevier.com/locate/clineuro

Case Report

Reversible opercular syndrome secondary to osmotic demyelination

Shuchit Pandey, Vikram V. Holla, Imran Rizvi, Rakesh Shukla*

Dept. of Neurology, King George's Medical University, Lucknow, Uttarpradesh, India

ARTICLE INFO

Article history:
Received 27 June 2015
Received in revised form 2 February 2017
Accepted 5 February 2017
Available online 11 February 2017

Keywords:
Opercular syndrome
Osmotic demyelination
Hyperosmolar hyperglycaemic state

ABSTRACT

Opercular syndrome (OPS) is characterized by weakness of facial, masticatory, pharyngeal, laryngeal, tongue and brachial muscles on voluntary command with preservation of emotional and reflexive movements. We report a case of 45 year old female who developed the features of OPS due to lesions of bilateral frontal opercular region induced by osmotic demyelination secondary to hyperosmolar hyperglycaemia. On follow up at 6 months, she had complete recovery.

1. Introduction

Opercular syndrome (OPS) also known as Foix-Chavany-Marie syndrome is facio-labio-glosso-pharyngo-laryngo-brachial paralysis with intact autonomic, involuntary and reflexive movements. It is considered as a cortical equivalent of pseudobulbar palsy. The most common cause of OPS is ischemia although it can be due to various other causes [1].

Herein we are describing a 45 year old female who presented with acute onset altered sensorium due to hyperosmolar state, later on after regaining consciousness she developed inability to execute movements of face, masticatory, laryngeal, pharyngeal and tongue muscles on command with preservation of emotional and autonomic movements due to extra pontine myelinolysis in bilateral frontal opercular regions. She had complete recovery and had no features of OPS at 6 months follow-up.

2. Case history

A 45 year old right handed female, not a known case of diabetes mellitus or hypertension, presented in emergency department with 5 days history of mental confusion progressing to stuporous state. On examination, her heart rate was 120/min, respiratory rate 18/min, blood pressure 120/70 mmHg, with profound dehydration. Patient's Glasgow coma scale was E2M5V1, with normal pupillary reaction and doll's eye response was intact. Plantars were bilateral extensor. She was immediately intubated and given respiratory support. Her routine investigations showed random blood sugar −625 mg/dl with normal serum sodium, potassium, chloride, bicarbonate, pH, renal and liver function tests. Serum acetone was absent, serum osmolality was 340 mOsm/ml and glycosylated haemoglobin was 9%. She was treated as hyperosmolar hyperglycaemic state due to type 2 diabetes mellitus (T2DM) with intravenous normal saline and insulin. Patient gradually improved and was extubated within 5 days. Her serum sodium was normal during the entire course of treatment. After regaining consciousness patient developed difficulty in swallowing, chewing food and speaking. She also had weakness in right half of the body in form of difficulty in gripping the objects tightly, raising hands above shoulder and difficulty in wearing footwear with right foot. On neurological examination she had anarthria, but could understand spoken and written commands and responded by writing, signs and gestures. Pupillary and corneal reflexes were normal with intact extraocular movements. She had inability to raise eyebrows, close the eyes, show the teeth, whistle, and move tongue inside as well as outside the mouth. She was able to follow commands directed to limbs but not to face. Tongue was immobile and without any atrophy or fasciculation or deviation. Taste sensation, gag reflex and blinking were normal with intact spontaneous smiling and yawning movements. Power was normal on left side but was medical research council grade 4-/5 in right upper limb and lower limb. Deep tendon reflexes were brisk in right upper and lower limb with absent jaw jerk and bilateral extensor plantar response. The sensations were normal and there were no exrapyramidal or cerebellar signs. 2D echocardiography of heart and bilateral carotid artery Doppler study were normal. Cerebrospinal fluid analysis was non-contributory. Magnetic resonance (MR) imaging of brain revealed hyperintensity on T2 and FLAIR sequences (Fig. 1) and hypointensity on T1-weighted images in bilateral frontal opercular

* Corresponding author.
E-mail addresses: drshuchit@gmail.com (S. Pandey), vikramvholla@gmail.com (V.V. Holla), imranrizvi09@gmail.com (I. Rizvi), rakeshshukla.rakesh@rediffmail.com (R. Shukla).

http://dx.doi.org/10.1016/j.clineuro.2017.02.001
0303-8467/

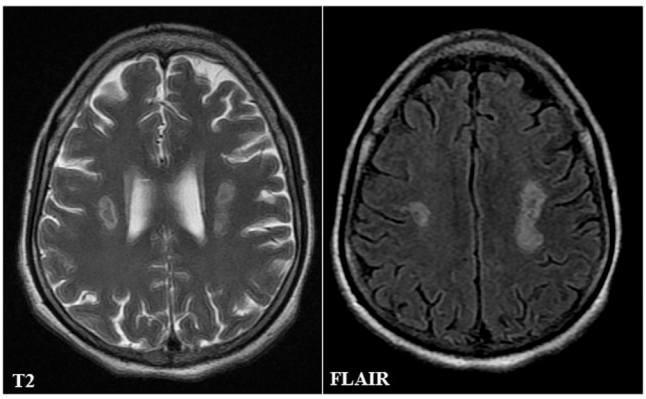

Fig. 1. Axial T2 weighted and fluid-attenuated inversion recovery images of the patient showing hyperintensity in bilateral frontal opercular subcortices.

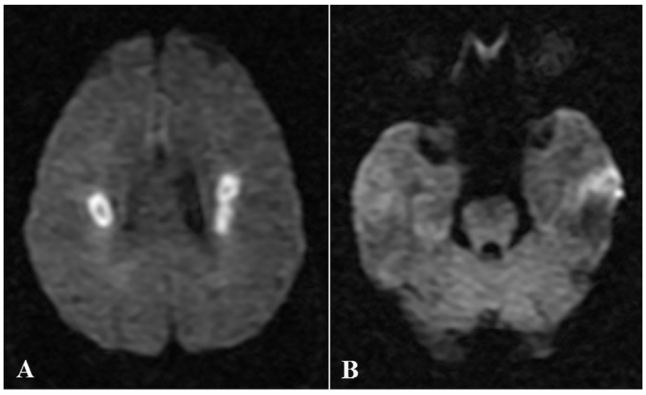

Fig. 2. Axial diffusion weighted images of the patient showing restricted diffusion in bilateral frontal opercular subcortices (A) and normal diffusion in pons (B).

subcortices including corona radiata with restricted diffusion on diffusion weighted images (DWI) (Fig. 2A) which was maximal in peripheral zone than in central region indicating osmotic demyelination (OD). There was no evidence of demyelination in pons or other areas of brain on MRI (Fig. 2B). MR angiography of brain was normal. She was subjected to speech and swallowing therapy. At the time of discharge three weeks later she was still on nasogastric tube and showed no improvement in swallowing, speech and chewing functions.

On follow up at tenth week, patient had marked improvement and was able to clench the teeth, raise the eyebrow and protrude the tongue on command. However, dysarthria and dysphagia was still present and she required nasogastric tube feeding. At the last follow up, 6 months after the illness patient was able to respond to commands by moving facial, bulbar and lingual muscles and there was no dysarthria or dysphagia, thus completely recovering from OPS. The blood sugar was controlled on oral hypoglycaemic agents.

3. Discussion

Our patient was a case of newly diagnosed T2DM who developed weakness of facial, masticatory, tongue and laryngeal muscles. This was associated with right sided pyramidal weakness; jaw jerk was absent but gag reflex as well as reflexive movements were intact. She was moving limbs on commands but was not able to move facial musculatures on command. Automatic movements like smiling, yawning and coughing were normal. This clinical picture can be explained by OPS due to lesions in bilateral frontal opercular regions caused by OD due to hyperglycaemic hyperosmolar state. Osmotic demyelination was first described in chronic alcoholic or malnourished patients. Now it is most commonly associated with rapid correction of hyponatremia. Various other uncommon conditions ascociated with OD include but not limited to patients who underwent liver transplant, hypernatremia, hypoglycaemia, uncontrolled diabetes mellitus, hyperosmolar hypoglycemic state, diabetic ketoacidosis, Wilson disease, Celiac disease and primary adrenal insufficiency [2,3]. In our patient the cause was correction of hyperglycaemic hyperosmolar state. Serum electrolytes were in normal range throughout the course of treatment. OD commonly involve pontine region hence earlier called central pontine myelinolysis. Involvement of areas other than pontine can occur when it is called extrapontine myelinolysis. Other sites to be affected are basal ganglia, thalamus, extreme and external capsule and other cortical areas. Osmotic demyelination is the preferred term which encompasses both pontine and extrapontine myelinolysis [2]. In a necropsy series of 58 cases by Gocht et al. isolated central pontine myelinolysis was present in about half, pontine with extrapontine myelinolysis in about one third, and isolated extrapontine myelinolysis in about one fifths of cases. In our case, the patient had isolated extrapontine OD changes in bilateral frontal opercular subcortical regions and corona radiata which on DWI sequences showed restriction with sparing of pons. OD can also cause restriction in DWI sequences but differentiating point from ischemic infarct is that diffusion restriction is maximal in peripheral zone than in central region of the lesion. MRA of brain, bilateral carotid Doppler imaging and echocardiography was normal in our patient.

Operculum is part of the brain which is made up of insular cortex, inferior frontal gyrus, pre and post central gyrus, supramargainal gyrus and superior temporal gyrus. OPS was originally described due to lesions of posterior part of inferior frontal gyrus and inferior part of precentral gyrus bilaterally. Voluntary activities are controlled by primary motor cortex and pyramidal system while reflexive and expressive movements are executed by pathways originating outside the primary motor cortex, which is the reason for automatic-voluntary dissociation in OPS [4]. OPS can occur with involvement of bilateral corticobulbar tract regions (subopercular region) without damage to opercular cortex bilaterally, also known as subopercular syndrome due to involvement of white matter tract (corticobulbar tracts) carrying fibers from opercular cortices. OPS is also recently reported in a patient with bilateral corona radiata infarcts similar to seen in our case [5].

Most common cause of OPS is infarct in bilateral opercular region. Other less common causes include encephalitis, trauma, tumors, developmental perisylvian dysplasia, vasculitis, and degenerative [1]. Most of the aetiologies causing OPS are non reversible. Till date there are no reports of reversible OPS due to OD of bilateral frontal opercular regions.

Our case emphasize that hyperglycaemic hyperosmolar state can be associated with isolated extrapontine osmotic demyelination and osmotic demyelination of bilateral frontal opercular regions can be a cause of reversible OPS.

References

[1] M. Bakar, H.S. Kirshner, F. Niaz, The opercular-subopercular syndrome: four cases with review of the literature, Behav. Neurol. 11 (1998) 97–103.
[2] J.D. King, M.H. Rosner, Osmotic demyelination syndrome, Am. J. Med. Sci. 339 (June (6)) (2010) 561–567.
[3] S.S. Kote, A. Khandelwal, D.G. Pathak, et al., An unusual case of osmotic demyelination syndrome without electrolyte changes in a patient with diabetes, J. Neuroanaesthesiol. Crit. Care 3 (2016) 145–148.
[4] H.C. Hopf, W. Müller-Forell, N.J. Hopf, Localization of emotional and volitional facial paresis, Neurology 42 (1992) 1918–1923.
[5] N. Bradley, N. Hannon, C. Lebus, et al., Bilateral corona radiata infarcts: a new topographic location of Foix-Chavany-Marie syndrome, Int. J. Stroke 9 (2014) E39.