

Research Article

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Reversible Foix-Chavany-Marie syndrome after right operculo-insular contusion from a spear gun trauma through the cranial base

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Abstract: *Background and object:* Foix-Chavany-Marie syndrome (FCMS) is a rare type of pseudobulbar palsy, which is characterized by anarthria or severe dysarthria and bilateral central facio-linguo-velo-pharyngo-mastigatory paralysis with “automatic voluntary dissociation”. We report on a patient who suffered a reversible FCMS following a spear gun trauma through the cranial base leading to right operculo-insular contusion. *Case Report:* This 28-year-old lady attempted suicide by shooting a spear gun into the head through her right submandibular region. Major vessel injury was ruled out and the patient was taken to the operating room for shaft removal. Postoperatively, we observed the mouth half open, drooling saliva, inability to move her tongue, anarthria, bilateral facial weakness, and loss of the gag reflex. Yawning was otherwise preserved resulting in a clinical diagnosis of FCMS. Postoperative imaging demonstrated a right operculoinsular contusion. Symptoms were fully recovered after two years of follow-up. *Conclusion:* FCMS is a rare and severe form of pseudobulbar palsy. Unilateral lesions are exceptional but should be recognized, as we presented. Generally, the outcome is moderate to poor but the occurrence in brain trauma can be associated with complete functional recovery.

Keywords: Foix-Chavany-Marie syndrome, head trauma, spear gun

1 Introduction

Foix-Chavany-Marie syndrome (FCMS) is a rare type of pseudobulbar palsy, which is characterized by anarthria or severe dysarthria and bilateral central facio-linguo-velo-pharyngo-mastigatory paralysis with “automatic voluntary dissociation” [1] The term was initially coined by Weller¹ in 1993 who recognized five clinical types, namely cerebrovascular disease, infection, neurodevelopmental, children with rolandic epilepsy, and neurodegenerative. We report on a patient who suffered a reversible FCMS following a spear gun trauma through the cranial base leading to right operculo-insular contusion.

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2 Case report

This 28-year-old left-handed lady attempted suicide by shooting a spear gun into the head through her right submandibular region. On admission, the patient was awake and cooperative, but verbalization was not possible. Left hemiplegia was also noted. Computed tomography (CT) and CT-angiography ruled out intracranial hemorrhage and major vessel injury and confirmed the shaft entry point adjacent to the carotid canal (Fig.1). The patient was taken to the operating room and submitted to tracheostomy, neck dissection, and decompressive craniectomy. After careful dissection of C1 segment and microsurgical temporary clipping of the intracranial segment of the carotid artery, the shaft was removed in a caudal direction. Intraoperative brain angiography revealed no abnormalities after shaft removal. Postoperatively, we observed the mouth half open, drooling saliva, inability to move her tongue and anarthria. Further examination identified bilateral central facial weakness and loss of the gag reflex. Yawning was otherwise preserved resulting in a clinical diagnosis of FCMS. Postoperative imaging demonstrated a right operculo-insular contusion, which was treated conservatively (Fig.1).

The patient improved gradually to dysarthria with the assistance of speech therapist and was discharged on postoperative day 25 with preserved tongue movements and efficient swallowing for soft foods, which were fully recovered after two years. The patient gave informed consent for the publication of this study.

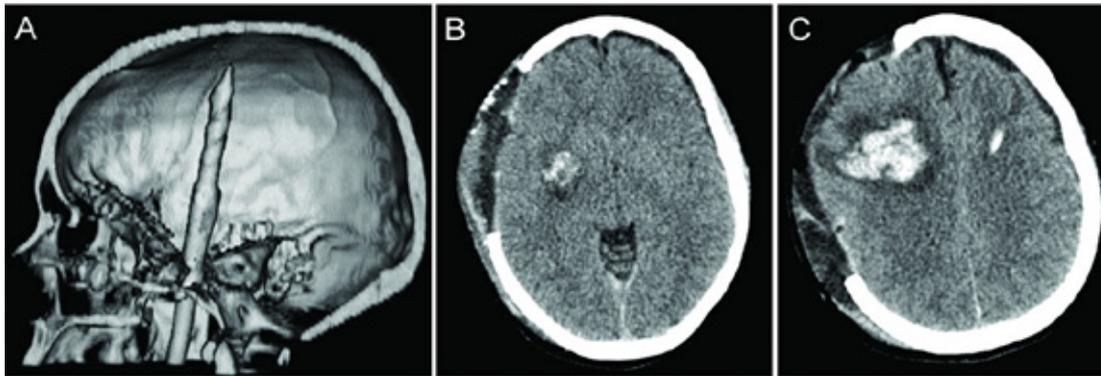


Fig. 1. Preoperative 3D reconstruction (A) showing craniocerebral penetrating injury caused by the shaft of a spear gun. Note the entry point through the cranial base towards coronal suture. Postoperative sequential (B, C) CT scans demonstrating acute traumatic insular (B) and opercular contusions (C) on the right side throughout missile trajectory.

3 Discussion

Of the five FCMS distinct clinical types, cerebrovascular disease is by far the most common cause [1]. FCMS in the scope of brain trauma is represented by scarce reports in the literature. To the best of our knowledge, there are only three detailed descriptions of FCMS in pure brain trauma (Table) [2-4]. According to Laurent-Vannier et al. [2], one additional case was described by Richard-Cremieux in her doctoral thesis in 1990. This patient was a 35 year-old female who also attempted suicide by a gunshot injury through her left temporal region. Brain imaging revealed bilateral frontotemporal lesions. Unfortunately, further information was not available.

Interestingly, two patients were affected by bilateral fronto-opercular Lesions [2,3], which is the topographical hallmark of the syndrome [1]. Of note, however, is that bilateral lesions occurred as a result of sequential brain trauma since left frontal injuries were diagnosed with an interim of 1 day [2] and 22 months [3] after right frontal contusions. Then, FCMS developed as a potential effect of the cumulative result of different lesions [3]. Nitta et al. [4] provided the first report of a unilateral right frontal contusion associated to FCMS. Their study was also remarkable because of low uptake area demonstration in the right frontal lobe on single photon emission computed tomography (SPECT) [4].

Table 1. Summary of published cases of Foix-Chavany-Marie Syndrome of traumatic origin.*

Author, Year	Age (yr), Gender	Hand dominance	Brain Trauma Mechanism	Imaging Findings	Symptoms	Treatment	Outcome
Laurent-Vannier et al. ² , 1999	10, M	Left	Bilateral Closed Car accident	Right frontopolar, right anterior temporal cortex contusions, brain edema and SAH (day 1) Left subdural collection and extradural hematoma (day 3)	Automatic-voluntary dissociation Loss of voluntary control of muscles innervated by cranial nerves V, VI, VII, IX, X, and XI	Surgical evacuation of the left extra-axial hematoma	Poor recovery limited orofacial abilities (18mo)
Campbell et al. ³ , 2009	24, M	NA	Sequential bilateral Closed Assault	Right frontal and temporal contusions, midline shift (day 1) Left frontal contusion (22mo later)	Automatic voluntary dissociation Bilateral central facial palsy, loss of gag reflex, loss of voluntary movements of the tongue Mouth half open, drooling saliva	Surgical evacuation of the right contusion Conservative (SLT)	Good recovery
Nitta et al. ⁴ , 2013	24, F	Right	Unilateral Closed Car accident	Right subdural hematoma, midline shift, opercular Premotor cortex contusion on intraoperative inspection	Automatic-voluntary dissociation Unable to swallow and speak Mouth half open, mastigatory diplegia	Surgical evacuation of the right contusion	Partial recovery (18mo)
Acioly et al., 2017	28, F	Left	Unilateral Penetrating Spear gun injury	Right operculo-insular hematoma (day 1)	Automatic-voluntary dissociation Loss of voluntary control of muscles innervated by cranial nerves V, VI, VII, IX, X, and XII	Decompressive craniectomy SLT	Full recovery (24mo)

* F – female, M – male, mo – months, NA – not available, SAH – subarachnoid haemorrhage, SLT – speech and language therapy, yr – year

FCMS is assumed to be caused by bilateral injury of the corticonuclear tracts in the cortical or subcortical region [2]. Our patient is unique in the way that FCMS-associated unilateral lesions are believed to be exceptional [1]. Weller identified just 2 out of the 62 reviewed patients, in whom FCMS could be attributed to unilateral lesions [1]. Despite the controversy behind the existence of a unilateral opercular syndrome, its apparent rarity is ascribed to a bilateral corticobulbar innervation of the lower cranial nerves [1]. Thus, the pathogenesis of unilateral opercular syndrome remains largely unknown.

One potential explanation for unilateral lesions causing bilateral anterior opercular syndrome is diaschisis [5]. Von Monakow introduced this theory in 1914 in order to describe the phenomenon of neurophysiological changes that occurs in remote areas from a focal brain lesion [5-7]. It was suggested that the loss of excitatory inputs from the original lesion contribute to functional “shock” or deactivation of such remote brain areas, which are connected to the primary injury [5,7]. This assumption was confirmed by SPECT studies in stroke patients who developed FCMS after unilateral opercular injury [5].

As previously reported, FCMS outcome is poor and depends directly on the clinical natural history of the underlying disease, which is unlikely to be Reversible [1,2]. Thus, the role of speech and language therapist (SLT) for anarthria remains to be determined for most of the patients [1]. Brain trauma, on the other hand, was associated to a temporary FCMS, from which a moderate to full recovery is possible. This was observed in 3 of the 4 patients reported, including our case. Such recognition is of utmost importance given that the assistance of SLT should be initiated as soon as possible in order to accelerate functional recovery. Moreover, this observation gives further support for diaschisis as the potential pathogenesis since, by definition, it means a reversible process in which functional improvement with time is a necessary requirement [6,7].

4 Conclusion

FCMS is a rare and severe form of pseudobulbar palsy, which is mostly caused by bilateral fronto-opercular lesions. Unilateral lesions are exceptional but should be recognized, as we presented. Generally, the outcome is moderate to poor but the occurrence in brain trauma can be associated with complete functional recovery.

Authors contribution: Study concept and design: Acioly, Barbosa, Pontes, Müller, Aguiar; Acquisition of data: Acioly, Barbosa, Pontes; Analysis and Interpretation of the data: Acioly, Barbosa, Pontes, Aguiar; Drafting of the manuscript: Acioly; Critical Revision of the manuscript: Acioly, Barbosa, Pontes, Müller, Aguiar; Study supervision: Acioly.

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