ORIGINAL RESEARCH

PATHOLOGICAL LAUGHING FOLLOWING PONTINE INFARCTION DUE TO BASILAR ARTERY STENOSIS

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ABSTRACT

Pathological laughing-(PL) is a situation defined by uncontrollable episodes of laughter without motivating stimulus or appropriate emotional expression. Pathological laughing has been described with cerebrovascular disorders.

A 60-year-old man was admitted to the hospital with a right conjugated gaze paresis, left facial hemiparesis, nineth and tenth cranial nerve paresis, absent gag reflexes mild right sided hemiparesis.-Cranial MR angiography revealed giant dolichoectatic basilar artery and with severe stenosis at the mid portion of the basilar artery. Six months later he presented with pathological laughing. However, the exact mechanism of pathological laughing is unclear and disturbances in the coordination and modulatory functions of cerebellum and cerebro-pontocerebellar pathways might strongly explain the nature of this phenomenon.

Keywords: Pathological laughing, Basilar artery stenosis, Cerebro-ponto cerebellar pathways

BAZİLER ARTER TROMBOZUNA BAĞLI PONS İNFARKTI SONRASI GELİŞEN PATOLOJİK GÜLME OLGUSU

ÖZET

Patolojik gülme, uygun duygu durumu veya uyaran olmaksızın ortaya çıkan ve kontrol edilemeyen gülme ataklarıdır. Serebrovasküler hastalıklarda ortaya çıkabilen bir durumdur.
60 yaşında erkek hasta sağa konjuge bakış parezisi, sol fasyal parezi, 9 ve 10. cranial sinir parezisi ve sağ hemiparezi bulguları ile başvurdu. Kranial MR anjiografide dolikoektatik baziler arter ve midbaziler stenoz saptandı. Hastanın izleminin altıncı ayında patolojik gülme tablosu gelişti. Patolojik gülmenin mekanizmaları tam olarak bilinmese de serebellum ve serebropontoserebellar yolakların düzenleyici fonksiyonları bu fenomeni açıklamada yeni bir alternatif görüş oluşturmaktadır.

Anahtar Kelimeler: Patolojik gülme, Baziler arter trombozu, Serebropontoserebellar yollar

INTRODUCTION

Pathological laughing (PL) is a situation defined by uncontrollable episodes of laughter without motivating stimulus or appropriate emotional expression. Pathological laughing is distinguishable from mood disorders because laughter is not associated with feelings of happiness. The PL is a symptom of gelastic epilepsy, pseudobulbar palsy and the tumors of cerebellopontine angle[1-6]. Pathological laughing has also been described with cerebrovascular disorders which affect internal capsule, thalamus, cerebral peduncles and basis pontis[7-9].

We report of a case presenting with pathological laughing following pontine infarction due to basilar artery thrombosis.

CASE PRESENTATION

A 60-year-old, right handed man was admitted to the emergency room complaining of vertigo, recurrent nausea and vomiting, slurred speech and disphagia. His symptoms first began with slurred speech. Ten hours after the onset of slurred speech, his symptoms progressed and the patient developed dysphagia, dysphonia and right sided motor weakness.

Past medical history was significant for hypertension and uncontrolled diabetes mellitus type II and hyperlidemia. He had no personal history of neurological or psychiatric disease.

On admission he was conscious but drowsy and presented with a right conjugated gaze paresis, left facial hemiparesis, nineth and tenth cranial nerve

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paresis, absent gag reflexes mild right sided hemiparesis involving the arm and the leg with a bilateral brisk tendon reflexes and extensor plantar responses. Moreover, his examination revealed limb ataxia of both the upper and lower extremities, with trunckal and gait ataxia toward the right side.

A non-enhanced CT scan, performed in the first hour of admission was within normal limits whereas cranial MR angiography revealed giant dolichoectatic basilar artery and with severe stenosis at the mid portion of the basilar artery (Figs 1,2).

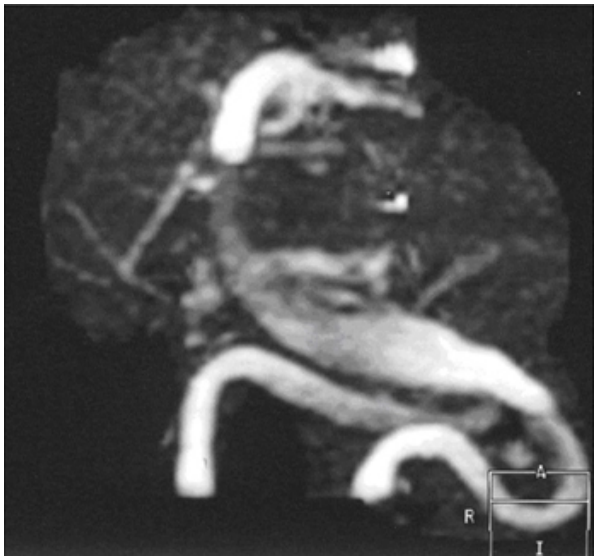

Fig 1: Cranial MRA showed a giant dolichoectatic basilar artery with severe stenosis at the mid portion.

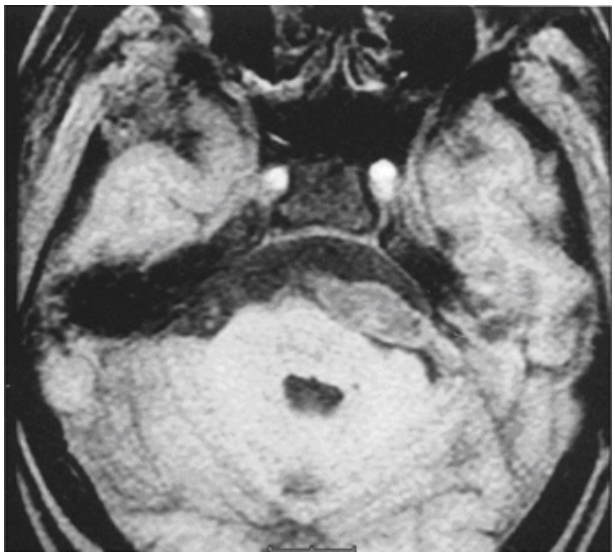

Fig 2: T1-weighted axial cranial MRI demonstrated a large, fusiform dilated basilar artery compressing the brainstem.

The patient was diagnosed to have progressive basilar artery thrombosis and he was treated with intravenous 0.9mg/kg tissue plasminogen activator(t-PA) followed by aspirin (300mg/day). His hemiparesis deteriorated progressively and intravenous heparin (1000IU/h) was started the following day. A cranial MRI repeated 2 days after the first symptoms revealed a left midpontine large infarct (Fig 3). His dysphagia, right sided hemiparesis, and ataxia deteriorated for 48 hours and then gradually improved. Three month later he was able to walk alone and returned to work.

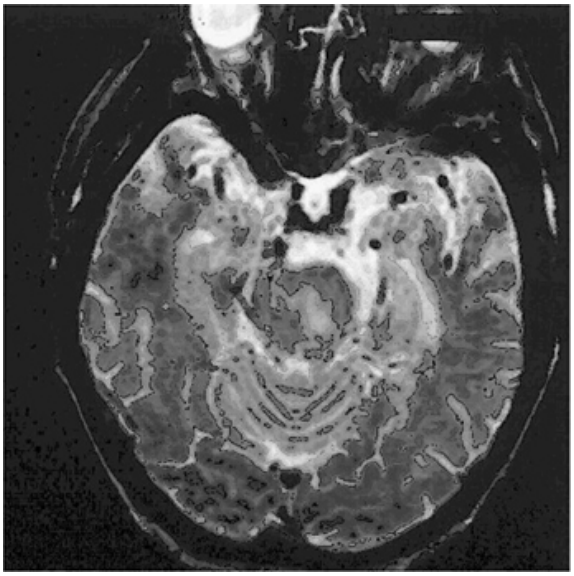

Fig 3: T2-weighted axial cranial MRI demonstrated a left mid-pontin large infarct

Six months later he presented to the neurology out-patient clinic complaining of unwilling laughter. He started to experience frequent episodes of inappropriate and involuntary laughing, triggered by talking -,-unaccompanied by mood elevation. He did not understand what was happening and was puzzled by it. He did not feel happiness during laughing attacks and could not control the paroxysms. The episodes did not have an apparent motivating stimulus for that emotional outburst. The patient was director in a large company and felt deeply embarrassed by the attacks particularly when occuring during professional activities. These attacks usually persisted for 30-seconds to 1 minute. His personal and social behaviour was appropriate except for the attacks of laughter and he had normal cognitive functions. The Beck Depression Inventory revealed mild depressive symptoms. Episodes of laughter, during which he was unable to control the tone of his voice and his breathing were observed during examination. Fluoxetine hydrochloride 10mg/day was started but the attacks were not controlled and the therapy was switched to paroxetine (20mg/day). Although the attacks did not resolve completely, severity of the symptom improved significantly.

DISCUSSION

Normal laughing is a complex motor action in which the facial and respiratory muscles are involved, and accompanied by mood elevation. Pathological laughing (PL) may be defined as uncontrollable, inappropriate, exaggerated laughing which is not related with a real emotional condition. Although the pathogenesis of pathological laughing is not clear yet, there are some speculative explanations. Wilson suggested that PL is the result of damaged voluntary inhibition of laughing and crying center (medullary effector center) located in the upper brainstem and defined this situation as a 'release phenomenon'10. This center is also controlled involuntarily by higher centers of the cerebral cortex and the limbic system. The medullary effector center can also control faciorespiratory functions during laughing and crying. According to Wilson's theory the location of this subcortical center must be above the faciorespiratory nuclei in upper pons or midbrain. On the other hand, Davison et al., reported that the hypothalamus is the primary center for laughing and crying, which is itself under the control of the inhibitory influences of the cortex 11. More recently, Arroya et al., reported that the main regulatory center for emotional responses may be located in the cingulate gyrus and the basal temporal cortex 2. However, these tempting speculations have been questioned by Parvizi et al., who presented an alternative hypothesis depending on Damasio's functional and neuroanotomical researches 12,13. They suggested that after the triggering stimuli is perceived, the ventromedial prefrontal cortex, anterior cingulate and amygdala are activated, in turn stimulating the effector sites as hypothalamus, periaqueductal grey mater, the cranial nerve nuclei and premotor regions. They postulated that the cerebellum plays an important role for adjusting laughing and crying behaviours12. The reason for this hypothesis is that the projections received by the cerebellum from telencephalic structures can convey social and cognitive stimuli, thus allowing the performed task to take into account such contexts. The cerebellum is capable of coordinating the complex facial, laryngopharyngeal and diaphragmatic rhythmic movements via the projections from the cerebellum to the brainstem and telencephalic structures. As a result Parvazi et al., believe that the occurence of pathological laughing and crying should be the consequence of distrupted cerebro-ponto cerebellar pathways.

Pathological laughing generally occurs following cerebrovascular lesions involving the descending corticobulbar pathways, especially with bilateral lesions at the level of the internal capsule, basis pontis and cerebral peduncles 9,14,15. Kim describes 13 patients with pathological laughing following cerebrovascular accident having mostly striatocapsular (- basal ganglia and subcortical white matter) infarction9. Pontine lesions are the second most common ones and such patients have been previously reported16. Our patient is the second one in the literature presenting with pontine infarction due to basilar artery thrombosis. Tei et al., have reported a patient who presented with hemiparesis accompanied by pathological laughing due to pontine infarction with basilar artery thrombosis 17. Like the latter, pathological laughing can herald a brainstem stroke, called 'fou rire prodromique' by Fere and can be the transient18. However, the immediate occurrence of pathological laughing after ischemic vascular lesions is rare and as it generally occurs after a period of time up to 1 year following stroke. Our patient started to experience episodes of inappropriate laughing six months ago following a cerebrovascular accident. Although the reason for the latent period between the onset of stroke and the occurrence of pathological laughing is still unclear, Kim hypothesized that recovery of the cortical dysfunction takes time and hyperexcitability can be seen in this period 9.

However, the exact mechanism of pathological laughing is unknown and the role of coordination and modulatory functions of cerebellum and cerebro-pontocerebellar pathways, as hypothesized by Parvizi et al., might be a strong alternative mechanism to explain the nature of this phenomenon. The other question is why this phenomenon is not encountered in all patients with the same lesion localization and if there are some individual differences between patients. Further neuroanatomical and neurophysiological studies are necessary to investigate pathogenetic mechanisms of post-stroke pathological laughing.

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