Clinical Research

Cranial MRI Findings in Patients with Hemifacial Spasm

Hemifasiyal Spazm Olgularında Kranial MRI Bulguları

Eren GÖZKE¹, Zehra AKTAN¹, Sıdıka Sinem TAŞDEMİR¹, Hilal Taştekin TOZ¹ Pelin Doğan AK¹, Burcu Seher ANIL²

1. University of Health Sciences, Fatih Sultan Mehmet Training and Research Hospital, Neurology Clinic 2. University of Health Sciences, Fatih Sultan Mehmet Training and Research Hospital, Radiology Clinic

ABSTRACT

Background and Purpose: Clonic hemifacial spasm (HFS) is a disorder characterized by involuntary contractions of muscles innervated by facial nerve on one side of face and it negatively affects daily life. The aim of this study was to investigate cranial MRI findings in patients with HFS.

Methods: Cranial MRI findings of 92 patients (53 female, 39 male) who have HFS were evaluated radiologically.

Results: Age range and mean ages of cases were 23-81 and 55,5 \pm 13,1, respectively. Cranial MRI was normal in 40 (43,4%) patients. We detected small vessel disease in 46 (50%), dolicoectasic basilar artery in 7 (7,6%), cerebral atrophy in 6 (6,5%), benign tonsillar ectopi in 2 (2,1%), arachnoid cyst in pontocerebellar angle in 1 (1,08%), pontocerebellar angle tumor in 1 (1,08%), and periventricular demyelinating plaque in 1 (1,08%) of the patients.

Conclusions: Clonic HFS can occur due to any pathology in the course of facial nerve. In our patients cranial MRI showed direct irritation of facial nerve in only 9 (9,7%) cases (dolicoectasic basilar artery, arachnoid cyst and tumor in pontocerebellar angle). The presence of small vessel disease in half of the patients suggests relationship between atherosclerotic process and occurrence of clonic HFS.

Keywords: clonic hemifacial spasm; facial nerve; cranial MRI

ÖZET

Amaç: Klonik hemifasiyal spazm (HFS) yüzün bir yarısında fasiyal sinir inervasyonlu kaslarda istemsiz çekilmelerin olduğu ve hastanın günlük yaşamının olumsuz olarak etkilendiği bir durumdur. Bu çalışmada klonik HFS tanısı konan olgularda kranial MR bulgularının değerlendirilmesi amaçlandı.

Yöntem: Klinik olarak klonik HFS tanısı konulan 53'ü kadın (%57,6), 39'u erkek (%42,3) olmak üzere toplam 92 hastanın kraniyal MR görüntülemeleri incelendi.

Bulgular: Olguların yaş dağılımı 23-81 ve yaş ortalaması 55,5 \pm 13,1 idi. Kırk olguda (%43.4) kraniyal MR normal bulundu. Kırkaltı olguda (%50) küçük damar hastalığı ile uyumlu bulgular, 7 olguda (%7,6) dolikoektazik baziler arter, 6 olguda (%6,5) serebral atrofi, 2 olguda (%2,1) benign tonsiller ektopi, 1 olguda (%1,08) pontoserebellar köşede araknoid kist, 1 olguda (%1,08) pontoserebellar köşe tümörü, 1 olguda (%1,08) periventriküler demiyelinizan plaklar görüldü.

Sonuç: Klonik HFS fasiyal sinirin seyri boyunca oluşan herhangi bir lezyona bağlı olarak ortaya çıkabilir. Bizim olgularımızın sadece 9'unda(%9,7) kranial MR ile direkt olarak fasiyal siniri irrite eden patolojik bulgu (dolikoektazik baziler arter, pontoserebellar köşede araknoid kist, pontoserebellar köşe tümörü) gösterilebilmiştir. Olguların yarısında küçük damar hastalığının varlığı aterosklerotik sürecin klonik hemifasiyal spazmın oluşumuna zemin hazırladığını göstermektedir.

Anahtar Kelimeler: klonik hemifasial spazm; fasial sinir; kranial MRG

Contact

Corresponding Author: Eren GOZKE Address: Acibadem Mh. Cecen Sokak, Almond-hill Site, B-18, D:24, Uskudar, Istanbul, Turkiye Phone Number: +90 (532) 616 54 52 E-mail: egozke@hotmail.com Submitted: 23.12.2016 Accepted: 04.01.2017

INTRODUCTION

Clonic hemifacial spasm (HFS) is a condition characterized by involuntary twitchings involving hemifacial muscles innervated by facial nerve which also effects daily life of the patient adversely. Generally, it starts from periorbital region, and gradually spreads to the lower part of the face. Occasionally, tonic muscle contractions can emerge. Nuclear part of the facial nerve which is also termed as seventh cranial nerve is situated in the pons. It arises from the junction between pons, and medulla oblongata, and exits intracranial space through meatus acousticus internus. Motor fibers innervating mimic muscles exit through stylomastoid foramen, and spread all over the face. Clonic HFS can manifest itself with nuclear, and infranuclear lesion of the facial nerve (1, 2).

In this study we aimed to evaluate cranial MRI findings in cases diagnosed as clonic HFS.

METHOD

Cranial MR images of a total of 92 [53 (57.6%) female, and 39 (42.3%) male patients] cases with clinical diagnosis of hemifacial spasm who were under botulinum toxin therapy were evaluated. T1, and T2-weighted axial FLAIR, and T2 weighted coronal, and sagittal sections were examined.

RESULTS

Mean age of the patients was 55.5 ± 13.1 years (range, 23-82 yrs). In 40 cases (43.4%) cranial MRI findings were within normal limits. On MRI findings consistent with microvascular disease (n=46; 50%), dolicoectasic basilar artery (n=7; 7.6%), cerebral atrophy (n=6; 6.5%), mastoiditis (n=5; 5.4%), benign tonsillar ectopy (n=2; 2.1%), arachnoid cyst of the pontocerebelar corner (n=1; 1.08%), and periventricular demyelinating plaques (n=1; 1.08%) were seen. Cranial MRI findings of two cases with clonic hemifacial spasms are seen in Figure 1.

DISCUSSION

The first case of clonic HFS was reported by Schultze in the year 1875, and it was described by Gowers in detail in the year 1884. It generally becomes manifest during the fifth and sixth decades of life. It frequently starts by affecting m. orbicularis oculi, and gradually spreads to other muscles innervated by facial nerve. It is not a painful clinical picture. Its prevalence is 14.5, and 7.4 per 100.000 female and male populations, respectively (1, 2).

Needle electromyograms demonstrate onset of irregular motor unit potential discharges with higher frequency during clinically observed clonic contractions in affected muscles. In its etiology, vascular compression (dolicoectasic basilar artery, ectasic anterior, and posterior cerebellar artery, venous angioma, aneurysms, fistulas), demyelinating diseases as multiple sclerosis, previously experienced Bell's paralysis, structural anomalies of the posterior fossa (Chiari malformation), infections (otitis, meningitis), tumors of the cerebellopontine corner (acoustic neurinoma, meningioma), parotid tumors, peripheral inflammation, and stroke (brainstem) can be enumerated (3-10).

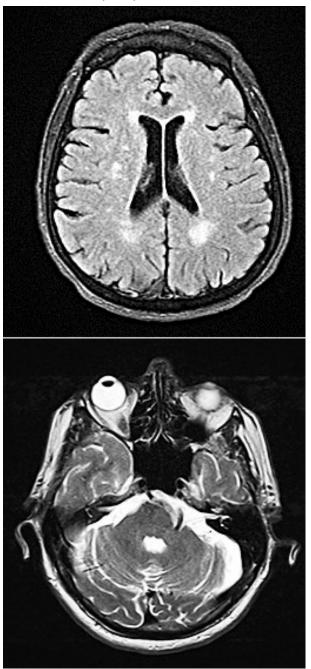


Figure 1: The cranial MR samples of two cases with clonic HFS. Left: Small vessel disease (T2 FLAIR, axial); Right: Dolichoectasia of basilary arter (T2, axial).

In many cases an apparent etiology cannot be revealed. In 40 % of the cases, hypertension is observed. Its pathogenetic mechanism has been explained by ephaptic transmission, reverberant activity at a nuclear level and kindling. Root entry/ exit zone is the most vulnerable region to myelin damage because of communication between oligodendrocytes and Schwann cells (11, 12).

Because of its typical characteristics and appearance, its diagnosis can be easily established, while in its differential diagnosis facial myokymia, unilateral blepharospasm, hemimasticatory spasm, facial chorea, facial tics, craniofacial tremor, oromandibular dystonia, and psychogenic contractions may be considered. In its treatment antiepileptic drugs; mainly valproic acid and clonazepam, levetiracetam, and baclofen may be mildly beneficial. However the most effective alternative is botulinum toxin injections despite recurrent treatment sessions at an average of 3 monthly intervals (1, 2). Botulinum toxin inhibits acetylcholine release at motor end plate to prevent convulsions. In appropriate cases, surgical decompression on facial nerve may be planned. Clonic HFS can manifest itself secondary to any lesion along the course of the facial nerve. On cranial MR, in only our 9 (9.7%) cases a pathology (dolicoectasic basilar artery, arachnoid cyst, and tumor of the pontocerebellar corner) which directly irritated facial nerve could be demonstrated. Since MR angiography cannot demonstrate its vicinity with surrounding tissues, it cannot provide additional information. Thin sections obtained during cranial MR from the exit site of the facial nerve may provide additional information.

More frequent detection of clonic HFS in hypertensive patients supports its association with atherosclerosis. Presence of microvascular disease in half of our patients indicates that atherosclerotic process predisposes to the development of clonic hemifacial spasms.

REFERENCES

1. Chaudhry N, Srivastava A, Joshi L. Hemifacial spasm: The past, present and future. J Neurol Sci 2015; 15:27-31.

2. Yaltho TC, Jankovic J. The many faces of hemifacial spasm: differential diagnosis of unilateral facial spasms. Mov Disord 2011;1:1582-92.

3. Jespersen JH, Dupont E, Gelineck J, Lundorf E. Hemifacial spasm: magnetic resonance angiography. Acta Neurol Scand 1996;93:35-8.

4. Jäger L, Reiser M. CT and MR imaging of the normal and pathologic conditions of the facial nerve. Eur J Radiol 2001;40:133-46.

5. Du C, Korogi Y, Nagahiro S, Sakamoto Y, Takada A, Ushio Y, Hirai T, Higashida Y, Takahashi M. Hemifacial spasm: three-dimensional MR images in the evaluation of neurovascular compression. Radiology 1995;197:227-31.

6. Chan LL, Lee E, Fook-Chong S, Tan EK. Case control MR-CISS and 3D TOF MRA imaging study of medullary compression and hypertension in hemifacial spasm. Mov Disord 2008;15:1820-4. doi: 10.1002/mds.22034.

7. Uluduz D, Karaali-Savrun F, Gunduz A, Kiziltan ME. An unusual case of vascular loop syndrome. J Headache Pain 2007;8:242-4. Epub 2007 Sep 24.

8. Deleu D, Mesraoua B. Hemifacial spasm resulting from vertebral artery dolichoectasia. Neurosciences (Riyadh) 2005;10:304-6.

9. Tan EK, Chan LL. Clinico-radiologic correlation in unilateral and bilateral hemifacial spasm. J Neurol Sci 2004;15:59-64.

10. Garibaldi DC, Miller NR. Tortuous basilar artery as cause of hemifacial spasm. Arch Neurol 2003;60:626-7.

11. Defazio G, Berardelli A, Abbruzzese G, et al: Primary hemifacial spasm and arterial hypertension: a multicenter case–control study. Neurology 2000; 54:1198-1200.

12. Tan EK, Chan LL, Lum SY, Koh P, Han SY, Fook-Chong SM, Lo YL, Pavanni R, Wong MC, Lim SH. Is hypertension associated with hemifacial spasm? Neurology 2003; 28:343-4.