OLGU SUNUMU CASE REPORT

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Coincidental Diagnosis of an Orbital Encephalocele-Beyond an Eyelid Myokymia

Bir Orbital Ensefaloselin Tesadüfi Tanısı-Kapak Miyokimisinin Ötesi

Pınar ALTIAYLIK ÖZER[a], Serdar ÖZER[b], Gökçe KAAN ATAÇ[c], Refah SAYIN[d], Ahmet ŞENGÜN[a]

[a]Ufuk University Faculty of Medicine, Department of Ophthalmology, Ankara, TURKEY
[b]Hacettepe University Faculty of Medicine, Department of Otorhinolaryngology-Head and Neck Surgery, Ankara, TURKEY
[c]Ufuk University Faculty of Medicine, Department of Radiology, Ankara, TURKEY
[d]Ufuk University Faculty of Medicine, Department of Neurology, Ankara, TURKEY

This case report was presented as a case presentation in scientific evening meeting of Turkish Ophthalmological Association Ankara Regional Branch, 28th November 2018, Ankara, Turkey.

ABSTRACT A 38-years-old woman presented to our hospital for routine eye examination. She had no complaints but it was learnt that she has been followed up with the diagnosis of a left upper eyelid myokymia for more than 10 years in various ophthalmological centers. Since her eyelid movement was atypical for a myokymia, but in form of an orbital pulsation instead she has undergone an orbital magnetic resonance imaging (MRI) scan. She was coincidentally diagnosed to have a left frontoorbital encephalocele detected by MRI. She had no trauma or chronic illness in history, so the condition was diagnosed as a 'congenital encephalocele' which is lately detected since due to the absence of obvious proptosis or an active clinical complaint.

Keywords: Eyelid myokymia; encephalocele; orbital pulsation; proptosis

ÖZET Otuz sekiz yaşında bir bayan hasta rutin göz muayenesi için polikliniğimize başvurdu. Herhangi bir şikayeti yoktu ancak sol üst göz kapağında uzun yıllardır atım hareketi olduğu, miyokimi tanısı ile çeşitli göz merkezlerinde 10 yıldır takip edilmekte olduğu öğrenildi. Ancak hastanın göz kapağındaki sürekli atım hareketinin miyokimi için atipik olması ve orbital pulsasyona benzer formda olması nedeni ile Orbital Manyetik Rezonans Görüntüleme (MRG) planlandı. Orbital MRG sonucunda hastada insidental olarak sol orbital ensefalosel tespit edildi. Hastada kafa travması ya da kronik hastalık öyküsü yoktu. Olguya, belirgin proptoza ve aktif şikayete neden olmadığından geç saptanan insidental 'konjenital ensefalosel'tanısı koyuldu.

Anahtar Kelimeler: Göz kapağı miyokimisi; ensefalosel; orbital pulsasyon; proptoz

A 38-years-old woman, who has been followed up with the diagnosis of a left upper eyelid myokymia for more than 10 years was admitted to our outpatient clinic, due to a routine check-up of her myopia. She was diagnosed to have a left frontoorbital encephalocele, determined by an orbital MR scan performed - because of atypical eyelid movement for a myokymia but in form of an orbital pulsation instead.

CASE REPORT

Informed concent form was obtained from the patient prior to her involvement in this case presentation. A 38-years-old woman was admitted to our outpatient clinic, for a yearly check-up of her myopia. It was learnt that she had been followed up with the diagnosis of a left upper eyelid myokymia for more than 10 years in other eye care centers. She did not report any complaints, including headache or visual disturbance. She had no history of trauma or chronic disease. Her best corrected visual acuities (BCVA) were 20/20 in both eyes with -2.00 spheric lenses OU. She had no diplopia or strabismus. Slit lamp examination and dilated fundus examination in both eyes were normal. Her

Correspondence: Pınar ALTIAYLIK ÖZER
Ufuk University Faculty of Medicine, Department of Ophthalmology, Ankara, TURKEY/TÜRKİYE
E-mail: drpinar@yahoo.com

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pupils were isocoric, with normal light reflexes (direct and indirect), without any relative afferent pupillary defects. She had no ptozis, proptosis or enophthalmus. But her upper eyelid on left side was observed to move up and down all through the examination involuntarily. She was requested to perform a valsalva and the movement was observed to increase in amplitude. This movement was thought to be atypical for a myokymia, but in form of an orbital pulsation instead. It was detected that no MRI or CT (computerised tomography) scan was applied to the patient previouly. She was ordered to undergo an orbital MR scan which revealed a 5x5 mm defect in the left superior orbital roof including dura, leptomeninges, and a small portion of frontal lobe parenchyma herniating through the bony defect (Figure 1). The orbitofrontal encephalocele was in close contact with the left globe but without and depression or deformation of the orbital structures. A CT scan was also performed to detect the bony structural defect demonstating the origin of the meningoencephalocele (Figure 2). She was consulted to otorhinolaryngology and neurology departments and it was reported that this bony defect in orbital floor was not associated to any other maxillofacial pathology or neurological sequelae. The patient was referred to department of neurosurgery for close follow up and surgical management.

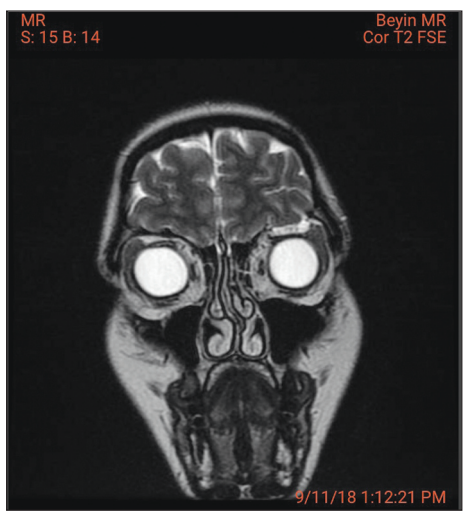

FIGURE 1: A coronal scan of orbital magnetic resonance image which revealed a defect in the left superior orbital roof and the meningoencephalocele.

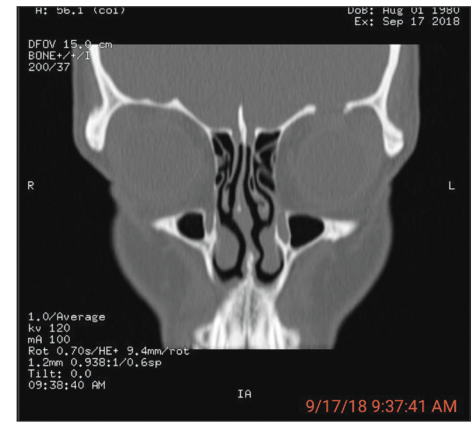

FIGURE 2: A coronal scan of computerised tomography image demonstrating bony structural defect of the left orbital roof.

DISCUSSION

Eyelid myokymia is a benign condition that is due to the contractions of the orbicularis oculi muscle and is mostly self-limiting.[1] It is generally observed unilaterally and intermittently takes not more than several hours. It is transient and disappears in a few days, although a case exists in the literature that lasts for several months.[2] It is known to rarely progress to facial myokymia, which might be the presenting sign of a neurological disorder like Multiple Sclerosis. Eyelid myokymia is composed of involuntary, fine, rhythmic contractions of the upper or lower eyelid. Unlike eyelid myokymia, orbital pulsation constitutes a continious movement and is a phenomenon due to pulsatile lesions of the orbita like orbital varices or encephaloceles.[3] Meningoencephaloceles are rare congenital malformations of the skull base, resulting in herniations of the meninges and cerebral tissue through the defects.[3] It is reported to occur in 10,000 to 15,000 live births, and can be observed associated to other central nervous system abnormalities like Dandy-Walker anomaly, holoproscencephaly, and neurofibromatosis.[4] A congenital defect in the orbital roof generally lead to pulsatile exophthalmus and are diagnosed at young ages.[5-7] Our patient was checked for any underlying systemic abnormalities but there were no disorders detected.

The preferred radiological imaging methods in orbital meningoencephaloceles are computed tomography to detect the defects in orbital bony structures and magnetic resonance imaging to diagnose the

composition and relation of herniated tissue to adjacent orbital structures. It's crucial to exclude pathologies like tumors and vascular abnormalities in these cases. In the presented case, MRI scans revealed clearly the herniation of cerebral tissue without any adjacent abnormalities of central nervous system.

Management of intraorbital meningoencephaloceles depends on the size and structure of the orbital bony defect. The surgical approach includes exposition of the bony defect, removal of the herniated tissues, followed by the reconstruction of the cranium. Either a bone graft from the exposed skull or a synthetic material (e.g titanium mesh) may be used for the reconstruction.[4,8,9]

The potential risk of optic nerve damage by compression of the orbital encephalocele can be minimized by early diagnosis and treatment of the condition. It is reported in the literature that the repair procedure is with minimal neurological complications, since the brain tissue within the encephalocele is nonfunctional but sometimes seizures may be observed if the adjacent brain tissue is scarred after the procedure.[10]

The presented case is unique, since it's a very late diagnosed case due to the mild pulsations that can easily be overlooked in an unattentive examination. Considering orbital pulsatile lesions in the differential diagnosis of involuntary eyelid movements is vital. A head trauma in history or an obvious proptosis in external examination are not obligations for arising a strong clinical suspicion of congenital orbital encephalocele in suspected cases.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Pınar Altıaylık Özer;* ***Design:*** *Gökçe Kaan Ataç;* ***Control/Supervision:*** *Ahmet Şengün, Refah Sayın;* ***Data Collection and/or Processing:*** *Serdar Özer, Pınar Altıaylık Özer;* ***Analysis and/or Interpretation:*** *Serdar Özer;* ***Literature Review:*** *Pınar Altıaylık Özer;* ***Writing the Article:*** *Pınar Altıaylık Özer;* ***Critical Review:*** *Ahmet Şengün, Refah Sayın;* ***References and Fundings:*** *Serdar Özer, Pınar Altıaylık Özer;* ***Materials:*** *Serdar Özer, Pınar Altıaylık Özer, Gökçe Kaan Ataç.*

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