OLGU SUNUMU CASE REPORT

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Isolated Unilateral Oculomotor Nerve Palsy Due to Hematoma in Mesencephalone

Mezensefalondaki Hematomdan Kaynaklanan İzole Tek Taraflı Okülomotor Sinir Felci

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ABSTRACT Oculomotor nerve palsy causes reduced adduction, supraduction, infraduction, and ptosis with or without mydriasis. We report a case of isolated unilateral oculomotor nerve palsy due to hematoma in mesencephalone. A 30-years- old male patient presented with sudden onset of left ptosis, vertical diplopia, and difficulty in reading with his left eye that had persisted for one week. Direct and indirect light reflexes were negative on his left eye. There was incomplete ptosis on his left eyelid. In primary position, the left eye was deviated outward and downward. Supraduction of the left eye was absent, infraduction and adduction were reduced. MRI demonstrated hematoma in mesencephalone. Three months later, patient's symptoms decreased, MRI demonstrated resolution of the hematoma and cavernoma in pons and gamma knife therapy was recommended. Isolated unilateral oculomotor nerve palsy due to hematoma related to cavernoma in mesencephalone is seen rarely and complete or incomplete recovery may be observed.

Keywords: Hematoma; mesencephalon; oculomotor nerve diseases

ÖZET Okülomotor sinir felci azalmış addüksiyon, supradüksiyon, infradüksiyon ve kapakta düşüklüğe neden olur, midriasis de olaya eşlik edebilir. Biz, mezensefalondaki hematomdan kaynaklanan izole tek taraflı okülomotor sinir felci olan bir vakayı takdim ediyoruz. Bir hafta devam eden, ani başlangıçlı sol gözde kapakta düşüklük, dikey çift görme ve sol gözle yakını görememe şikayetiyle polikliniğimize başvuran 30 yaşındaki erkek hastada, yapılan muayenede sol gözde direkt ve indirekt ışık reflekslerinin negatif olduğu gözlendi. Sol göz üst kapakta kısmi düşüklük mevcuttu. Primer pozisyonda sol göz dışa ve aşağı doğru kaymış durumdaydı. Sol gözün supradüksiyonu yoktu, addüksiyon ve infradüksiyonu ise azalmıştı. MR görüntülerinde mezensefalonda hematom izlenmekteydi. Üç ay sonra hastanın belirti ve bulguları azaldı, MR görüntülerinde hematomda çözülme ve ponsta kavernom izlendi ve hastaya gamma knife terapisi önerildi. Mezensefalondaki hematomdan kaynaklanan kavernom ile ilişkili izole tek taraflı okülomotor sinir felci nadiren görülür ve tam veya kısmi iyileşme gerçekleşir.

Anahtar Kelimeler: Hematom; mezensefalon; okülomotor sinir hastalıkları

The oculomotor nerve (the third cranial nerve) innervates the medial, inferior, superior recti muscles, the inferior oblique muscle, the levator palpebrae superioris muscle, the iris sphincter muscle and the ciliary body.[1]

The third nerve nucleus is located in the midbrain near the cerebral aqueduct at the level of the superior colliculus. Each extraocular muscle

that receives innervation from the third nerve has a corresponding subnucleus. A single central nucleus (central caudal nucleus) innervates both levator palpebrae muscles. Distinct, bilateral subnuclei exist for the extraocular muscles. An additional bilateral subnucleus, the Edinger-Westphal nucleus, provides parasympathetic input to the ciliary body and pupillary sphincter. Each subnucleus innervates the ipsilateral corresponding extraocular muscle, with two exceptions, the single central caudal nucleus sends projections to both levator muscles and the superior rectus subnucleus innervates contralateral superior rectus muscle.[2]

Oculomotor fascicles travel ventrally through the midbrain, each passing through the red nucleus, substantia nigra and crus cerebri before they exit into subarachnoid space of the interpeduncular fossa. Then the third nerve enters the cavernous sinus, wherein it is separated into superior and inferior divisions. The superior division innervates the superior rectus and levator muscles, while the inferior division supplies the inferior rectus, inferior oblique, medial rectus, ciliary body and pupillary sphincter muscles. After entering the superior orbital fissure, the oculomotor nerve lies within the annulus of Zinn and muscle cone and reaches the respective muscles.[3]

Damage to the third cranial nerve causes reduced adduction, supraduction, infraduction, and ptosis with or without mydriasis.[4] The aim of this case report is to represent the course of this rare case.

CASE REPORT

A 30-years-old male patient presented with sudden onset of left ptosis, vertical diplopia and difficulty in reading with his left eye that had persisted for one week. He had no systemic diseases. He never used tobacco or alcohol. On examination, he was conscious and well oriented, his vital signs were normal.

His uncorrected visual acuities were 20/20 on both eyes, his intraocular pressures were 16 mmHg in right eye and 15 mmHg in left eye respectively, and fundus examination was normal bilaterally. Direct and indirect light reflexes were positive on his right eye, and negative on his left eye. Left pupil was mydriatic and unresponsive to light. There was incomplete ptosis on his left eyelid. In primary position, the left eye was deviated outward and downward. Supraduction of the left eye was absent, infraduction and adduction were reduced. All movements of the right eye were normal, as shown in Figure 1. Forced duction test was negative. Patient's diplopia complaint was getting worse, when he was forced to look upwards. His visual field analysis revealed no abnormalities. He had no other neurological signs or symptoms.

Complete blood count and biochemical analysis of the blood were normal. MRI demonstrated hematoma in mesencephalone on T1 and T2 weighted images, as shown in Figure 2. Patient also has arachnoid cyst seen in anterior part of the left temporal lobe on the MRI images, but that was not related to the patient's clinical findings. Patient was consulted to Neurology and Neurosurgery Clinics and he was followed-up by neurologists and neurosurgeons, too. Since central caudal nucleus (the levator palpebralis nucleus) projects bilaterally and the superior rectus subnucleus projects contralaterally, in our patient the pupillary involvement, unilateral ptosis and unilateral reduction in the movements of inferior, superior,medial recti and inferior oblique muscles, indicate that the fascicles within the midbrain were affected.

A gradual improvement of oculomotor nerve functions was observed during the subsequent follow-ups. Meanwhile he did not take any treatments. Three months later, patient's symptoms decreased but didn't resolve completely. Left pupil was responsive to light but lazy, ptosis was recovered, eye movements were better, but still they were reduced, as shown in Figure 3. MRI at that time demonstrated resolution of the hematoma and cavernoma or cavernous angioma (arterio-venous malformation) in pons on T1 and T2 weighted images, shown in Figure 4. Neurosurgeons recommended gamma knife therapy (radiation therapy) for the patient to eradicate or minimize the lesion.

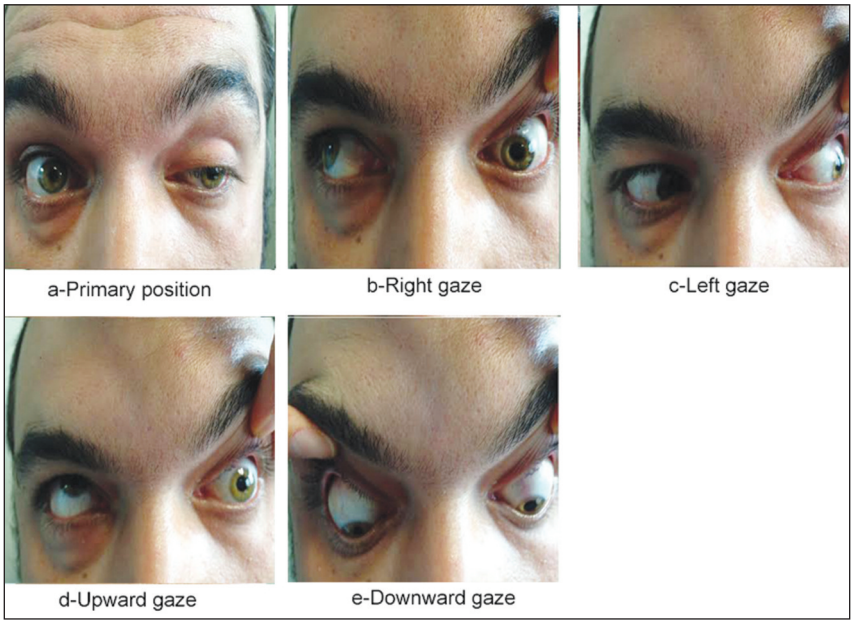

FIGURE 1: The primary position and the movements of right and left eyes of the patient.

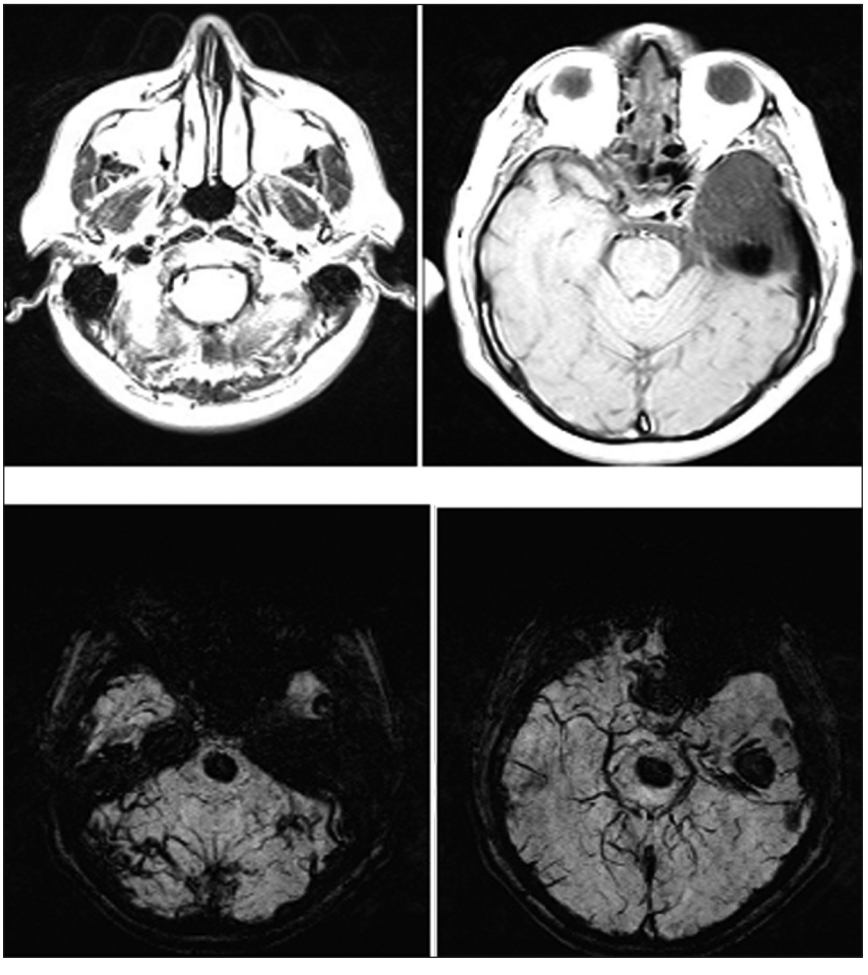

FIGURE 2: T1 and T2 weighted MRI images.

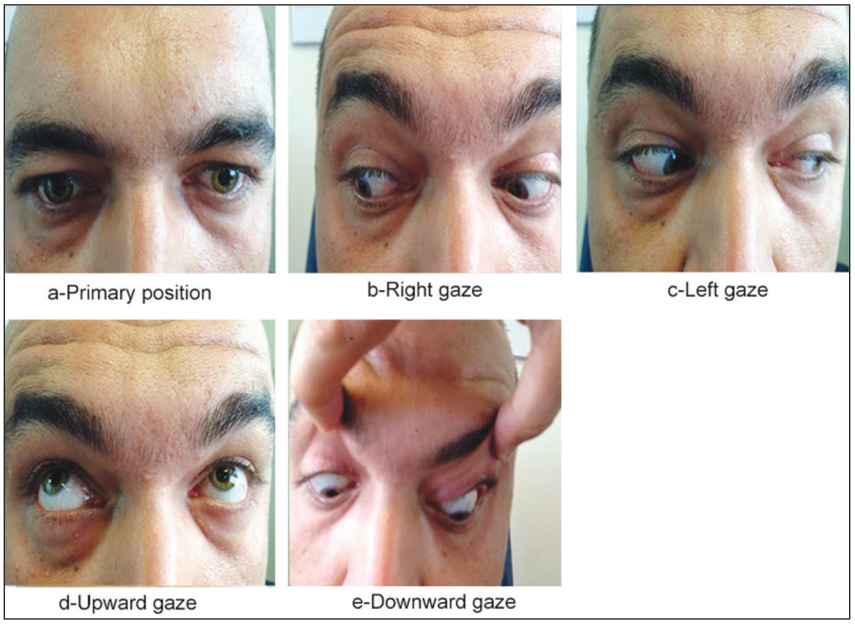

FIGURE 3: The primary position and the movements of right and left eyes of the patient after three months.

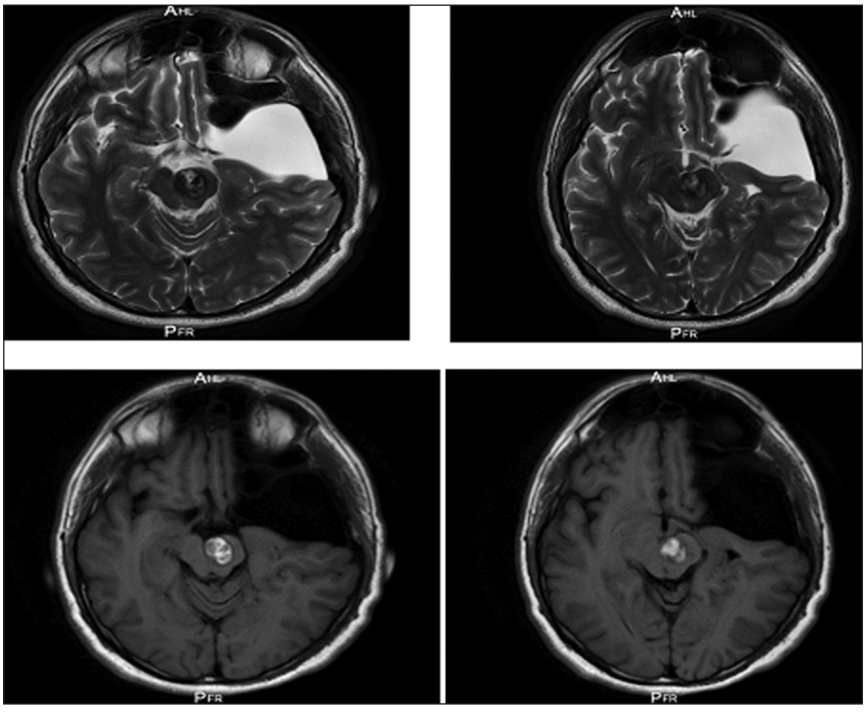

FIGURE 4: T1 and T2 weighted MRI images after three months.

DISCUSSION

Oculomotor nerve palsy may be congenital or acquired. Most acquired causes in adults are vascular diseases such as ischemia or infarction due to diabetes, hypertension and atherosclerosis, and hemorrhage from arterio-venous malformation and hypertension, demyelinization and compression by intracranial aneurysms or other mass lesions like primary or metastatic tumors. Infectious, inflammatory

and traumatic causes are less likely.[5-14] In our patient the cause was hematoma due to hemorrhage from cavernoma (arterio-venous malformation).

Identification of a total third nerve palsy is typically straightforward on examination, when there is ptosis, an eye that is deviated 'down and out' and a dilated pupil.[1,5] Our patient had total third nerve palsy, since he had ptosis, 'down and out' deviation and a dilated unreactive pupil. But it was incomplete third nerve palsy, because infraduction and adduction of the left eye were not absent but reduced and the ptosis was not complete.

Systemic hypertension and arterio-venous malformations represent the most common etiologies of primary hematomas of the mesencephalone.[15] De Mendonca et al.[16] reported a 70-years-old hypertensive woman suffering from sudden onset of bilateral blepharoptosis, within few hours she developed focal signs attributable to a lesion of the mesencephalone and a stuporous state, from which she did not recover, neuropathological examination showed a hematoma in mesencephalone. Ramirez-Moreno et al. presented a case of hypertensive woman with an oculomotor nerve syndrome associated with limitation of horizantal gaze of the contralateral eye and reactive head inclination due to mesencephalic hematoma.[17] Durward et al. reported two patients presenting with a mesencephalic hematoma, the etiology was a proven arteriovenous malformation in one case and the other one was suspected to be due to hypertensive arteriolar rupture.[18] In our patient, the etiology was also arteriovenous malformation (cavernoma in pons).

The crossed projection of the superior rectus subnucleus indicates that unilateral third nerve lesions with contralateral superior rectus involvement obligate a nuclear lesion, whereas a third nerve palsy with no contralateral superior rectus abnormality can not be caused by a nuclear lesion, furthermore, unilateral ptosis also shows that the lesion is not nuclear, because a single central nucleus (central caudal nucleus), innervates both levator palpebrae muscles. Nuclear and fascicular nerve palsies often are associated with other neurological signs such as pyramidal tract dysfunction, abnormalities of coordination and impaired consciousness, because of the large number of structures located nearby, isolated palsies are very rare.[2,15] Due to unilateral ptosis and unilateral extraocular muscular involvement in our patient, the lesion was thought to be fascicular in origin. And other cranial nerves, motor, sensory, coordination and reflex systems were spared in our patient.

Edinger-Westphal nucleus provides parasympathetic input to the pupillary sphincter and ciliary body. This nucleus and its fascicles are located at the top of the nuclear complex. Bilateral pupillary involvement with nuclear oculomotor palsy, therefore, is only expected in lesions with extension to the rostral midbrain.[3,15] Pupil-sparing oculomotor palsy is most likely caused by microvascular ischemia resulting from diabetes, hypertension and atherosclerosis.[5] Unilateral difficulty in reading and unreactive pupil in our patient were due to the destruction of the pupillomotor fibers located medially among the oculomotor fascicles.

Neurosurgical intervention (surgical evacuation) may be required in severe cases of mesencephalic hematoma.[18] Careful observation for at least 6 months is essential before planning a strabismus and/or ptosis surgery, as some patients will show a partial or full spontaneous recovery. Most oculomotor nerve palsy due to microangiopathy resolves completely in the first few months. When strabismus surgery is needed, the extent and severity of extraocular muscle involvement and the presence of aberrant regeneration play a crucial role in surgical planning.[5] For our patient, neurosurgeons recommended gamma knife therapy, which is a radiation therapy.

CONCLUSION

In conclusion, isolated unilateral oculomotor nerve palsy due to hematoma related to cavernoma in mesencephalone is seen very rarely and complete or incomplete recovery may be observed in such 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: *Servet Çetinkaya;* ***Design:*** *Servet Çetinkaya;* ***Control/Supervision:*** *Emine Mestan;* ***Data Collection and/or Processing:*** *Servet Çetinkaya;* ***Analysis and/or Interpretation:*** *Servet Çetinkaya;* ***Literature Review:*** *Servet Çetinkaya;* ***Writing the Article:*** *Servet Çetinkaya;* ***Critical Review:*** *Emine Mestan.*

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