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Olgu Sunumu

Schizencephaly Associated with Chronic Subdural Fluid Collection

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Schizencephaly is an uncommon congenital structural disorder of cerebral cortex caused by abnormal neuronal migration. Patients usually present with seizure disorders, hemiparesis, quadriparesis, cognitive deficits and mental troubles. Very rarely, schizencephaly is also associated with subdural fluid collection, and may present with mass effect. In this study we present a schizencephaly case with chronic subdural fluid collection. In our view, clefts may promote subdural fluid collection in schizencephaly patients and careful follow up of these patients is needed.

Key words: Schizencephaly, subdural fluid, cleft, mass effect

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Kronik Subdural Sıvı Koleksiyonu ile İlişkili Şizensefali

Şizensefali anormal nöral migrasyon sonucu gelişen serebral korteksin konjenital yapısal bozukluğu olup, oldukça ender görülmektedir. Hastalar klinik olarak genellikle epileptik nöbetler, hemiparezi, kuadriparezi, kognitif ve mental bozukluklar ile karşımıza çıkmaktadır. Çok ender olarak şizensefali, subdural sıvı koleksiyonu ile ilişkili olup, kitle etkisi ile klinik oluşturabilir. Biz bu olgu sunumunda şizensefali ile ilişkili kronik subdural sıvı koleksiyonu olan hastamızı sunmaktayız. Görüşümüze göre şizensefali hastalarında yarıklar subdural sıvı koleksiyonuna zemin oluşturmaktadır. Bu nedenle şizensefali hastalarının klinik olarak dikkatli takip edilmeleri gerekmektedir.

Anahtar kelimeler: Şizensefali, subdural sıvı, yarık, kitle etkisi

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Schizencephaly was originally described by Yakovlev and Wadsworth [9,10]. It's an uncommon congenital structural disorder of cerebral cortex caused by abnormal neuronal migration [1-3,5,6]. The disease is characterized by pia-covered, grey matter-lined clefts spanning cerebral hemispheres from the pial surface to the lateral ventricles [2,9,10]. These patients usually present with seizure disorders, hemiparesis, quadriparesis, cognitive deficits and mental troubles [2]. Very rarely, schizencephaly is also associated with subdural fluid connections, and may present with mass effect [6]. In this study, we report a patient with subdural collection overlying cleft of schizencephaly, and discuss effects of schizencephaly clefts on occurrence of subdural fluid collection.

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CASE REPORT

A 33 year-old- woman was admitted to Dokuz

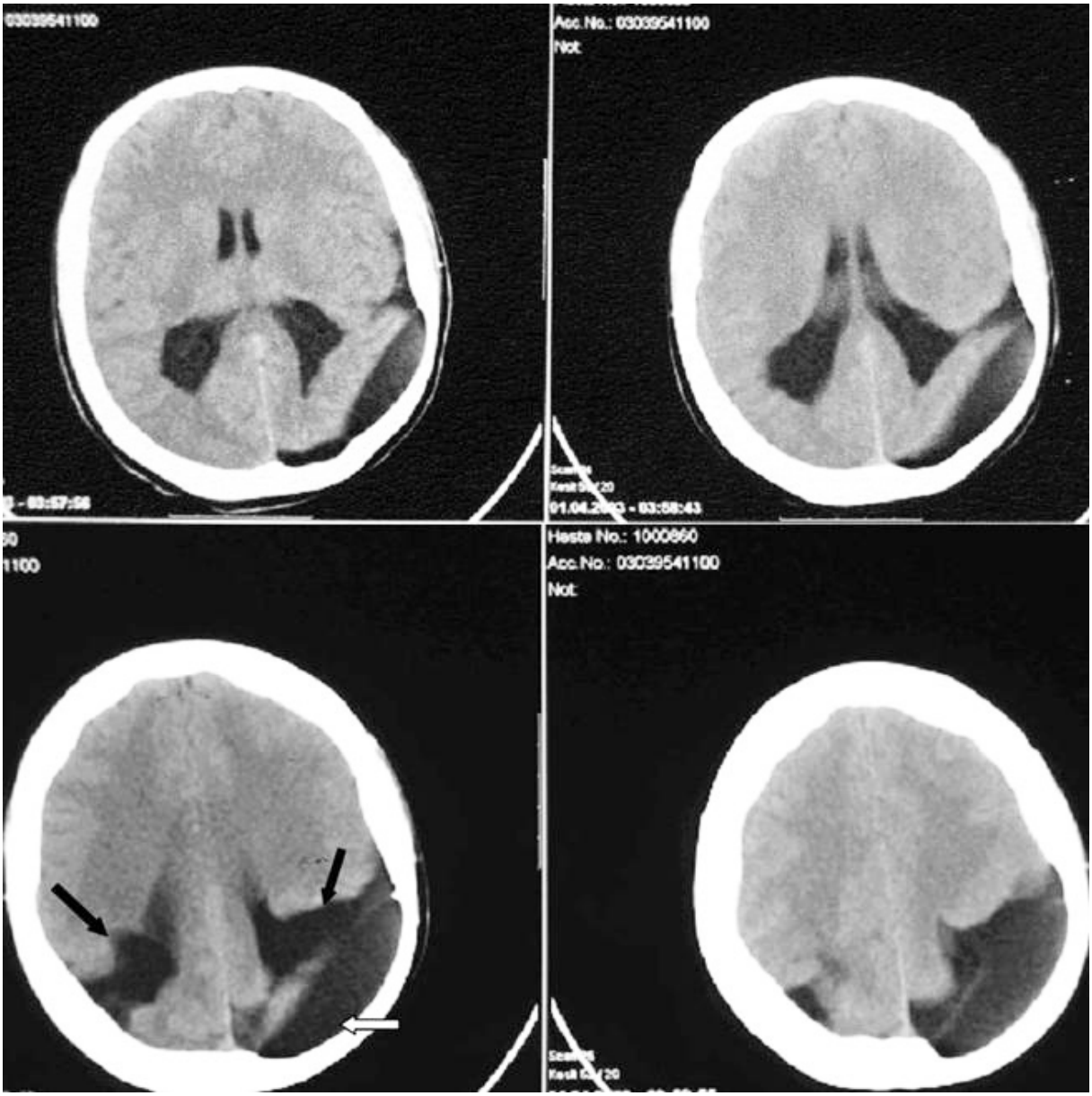

Figure 1. Serial axial CT images of the patient show bilateral separated lip schizencephaly and chronic subdural fluid collection overlying left cleft of schizencephaly at the parieto-occipital region. Black arrows point clefts and white arrow points subdural collection.

Eylul University Hospital Emergency Service because of loss of consciousness. She was schizophrenic and lived with her family. She was found on the floor in the house after she had lost her consciousness. When patient was admitted to the emergency service, she was cyanotic and hypotensive. Neurological examination at the admission revealed that she was in coma. She opened her eyes and localized painful stimuli without any verbal response (Glasgow Coma Score: 8). Pupils were anisocoric (L>R) and light reflexes were positive but sluggish. Laboratory investigations showed that she had pneumonia and severe metabolic acidosis. A CT examination was performed due to depressed level of consciousness and anisocoria. It revealed bilateral separated lip schizencephaly and chronic subdural fluid collection overlying left parieto-occipital region (Figure 1). She was intubated and appropriate therapy was started. Despite treatment, patient's

clinical status got worse, her GCS score progressed and she died a day after her admission due to sepsis. Patient's clinical condition was attributed to metabolic coma and any intervention was not performed for the treatment of chronic subdural collection. Also mass effect of subdural fuid collection wasn't significant and there was no midline shift or ventricular compression.

DISCUSSION

Schizencephaly is a rare congenital malformation and occurs as a result of abnormal neuronal migration of cerebral tissue during the embryonic development [1,5,6]. This pathology is of two types, seperated-lip and fused-lip [4,5]. It can be seen unilaterally and bilaterally, and both are encountered equally [5]. Clefts are lined with gray matter and covered by pia. Other developmental CNS anomalies like white matter diminution, septal and optic pathway anomalies, callosal anomalies and hippocampal anomalies are also seen in association with schizencephaly [4-7,9]. Schizencephalic patients generally present in infancy with seizure disorders and neurological deficits like cognitive and mental problems, hemiparesis, quadriparesis related to clefts [4]. Psychotic disorders have been also reported frequently in schizencephalic patients [1]. However, they can also show signs of increased intracranial pressure and hydrocephalus can be associated with schizencephaly. One third of schizencephaly patients require placement of ventriculoperitoneal shunts. Cause of hydrocephalus is thought to be disturbance of CSF circulation due to absence of cerebral mantle [7]. Presentation with subdural fluid collection is a rare phenomenon, and a neonatal case has been reported in the literature [6]. The patient was treated initially with surgical drainage and ventriculoperitonal shunting. Shunt system was removed later due to CNS infection but patient didn't need shunting later. Our adult patient had also subdural fluid collection and any adult case with subdural fluid collection has not been reported in the literature so far. Subdural fluid collection in our patient was detected incidentally when she was in coma as a result of septicemia. Subdural fluid collection didn't cause any symptom in our comatose patient but possibly it would be mildly symptomatic in a conscious patient. It is also possible that symptoms of schizophrenia might mask symptoms due to chronic subdural fluid collection. Careful follow up of these patients is needed to find out if there are new symptoms. Radiological investigations should also be performed in case of suspicion. Treatment of this type subdural fluid collection is burr hole drainage in a symptomatic patient. Due to small size of subdural fluid collection, clinical situation of our patient was attributed to metabolic coma, and no treatment of subdural fluid collection was planned.

Arachnoid cysts are commonly associated with schizencephaly [6-8]. It is postulated that the mechanism which causes schizencephaly may lead to some kind of a traction effect and splitting of the leptomeninges, resulting in the formation of an arachnoid cyst next to schizencephaly cleft [8]. There may be adhesions left between clefts and meninges due to this developmental pathology. If there is any adhesion present between schizencephalic cleft and arachnoid matter, a head trauma can cause traction on arachnoid at birth or in the postnatal life. Thus, traction can tear the arachnoid membrane and cause the subdural space to be filled with fluid. Resultant subdural fluid collection become symptomatic when enlarged. Inoue et al [6] reported cause of subdural fluid collection as minor trauma at birth. Trauma is a possible cause of subdural collections but in the case of schizencephaly, there is an anomaly and a passage between the ventricle and cortical subarachnoid space. Risk for the occurrence of subdural fluid collection may increase with altered cerebrospinal fluid dynamics. Also, developmental structural anomalies can promote collection of subdural fluid secondary to traction by

adhesions. Our patient's subdural collection is a bit hyperdense than reported for neonatal cases, possibly because of its higher protein content.

Instead of common symptoms, presentation with subdural fluid collection and its mass effect is possible in schizencephaly patients. That's why, careful follow up of these cases is needed. The mechanisms responsible for the development of subdural fluid collection may be altered CSF dynamics, traction and tearing of the arachnoid membrane due to adhesions left from embryologic development.

REFERENCES

1. **Alexander RC, Patkar AA, Lapointe JS, Flynn SW, Honer WG.** Schizencephaly associated with psychosis. *J Neurol Neurosurg Psychiatyr* 1997;63:373-5. http://dx.doi.org/10.1136/jnnp.63.3.373
2. **Denis D, Chateil JF, Brun M, Brissaud O, Lacombe D, Fontan D, et al.** Scizencephaly: clinical and imaging features in 30 infantile cases. *Brain and Development* 2000;22:475-83. http://dx.doi.org/10.1016/S0387-7604(00)00173-X
3. **Dubey AK, Grupta RK, Sharma P, Sharma RK.** Schizencephaly Type-I. *Indian Pediatr* 2001;38:1049-52.
4. **Guerrini R, Carrozzo R.** Epileptogenic brain malformations: clinical presentation, malformative patterns and indications for genetic testing. *Seizure* 2001;10: 532-47. http://dx.doi.org/10.1053/seiz.2001.0650
5. **Hayashi N, Tsutsumi Y, Barkovitch AJ.** Morphological features and associated anomalies of schizencephaly in the clinical population: detailed analysis of MR images. *Neuroradiology* 2002;44:418-27. http://dx.doi.org/10.1007/s00234-001-0719-1
6. **Inoue R, Isono M, Kamida T, Izumi T, Kobayashi H.** A case of schizencephaly with subdural fluid collection in a neonate. *Child's Nerv Syst* 2002;18:348-50. http://dx.doi.org/10.1007/s00381-002-0564-5
7. **Packard AM, Miller VS, Delgado MR.** Schizencephaly: correlations of clinical and radiologic features. *Neurology* 1997;48:1427-34. http://dx.doi.org/10.1212/WNL.48.5.1427
8. **Sener RN.** Coexistence of schizencephaly and middle fossa arachnoid cyst: Report of two patients. *Eur Radiol* 1997;7:409-11. http://dx.doi.org/10.1007/s003300050176
9. **Yakovlev PI, Wadsworth RC.** Schiz-encephalies: A study of congenital clefts in the cerebral mantle. I. Clefts with fused lips. *J Neuropathol Exp Neuro* 1946;5:116-30. http://dx.doi.org/10.1097/00005072-194604000-00003
10. **Yakovlev PI, Wadsworth RC.** Schiz-encephalies: A study of congenital clefts in the cerebral mantle. II. Clefts with hydro-cephalus and lips separated. *J Neuropathol Exp Neuro* 1946;5:169-206. http://dx.doi.org/10.1097/00005072-194607000-00001