Case Report / Olgu Sunumu

Cerebrospinal fluid otorrhea and recurrent bacterial meningitis in a pediatric case with Mondini dysplasia

Mondini displazisi olan pediatrik bir olguda beyin-omurilik sıvısı otoresi ve tekrarlayan bakteriyel menenjit

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Mondini dysplasia is a congenital malformation of the inner ear, which is characterized by a short and large cochlear canal of 1.5 turn rather than 2.5 turns and an apical region with cystic dilatation. Patients present with congenital deafness, when both cochlea are affected. Unilateral disease may cause recurrent meningitis, otorrhea or rhinorrhea. In this article, we report a three-year-old pediatric case with a history of meningitis and cerebrospinal fluid otorrhea following tympanostomy tube placement for serous otitis media.

Key Words: Cerebrospinal fluid; Mondini dysplasia; otorrhea.

Mondini displazisi, 2.5 dönüşe kıyasla, 1.5 dönüş gösteren kısa ve büyük koklear kanalı ve kistik dilatasyonlu apikal bölge ile karakterize, iç kulağa ait doğuştan yapısal bir bozukluktur. Her iki kokleanın da etkilendiği hastalarda doğuştan sağırlık görülür. Tek taraflı hastalık ise, tekrarlayan menenjit, otore ve rinoreye neden olabilir. Bu makalede seröz otitis media için tamponostomi tüpü yerleştirilmesinin ardından menenjit ve beyin-omurilik sıvısı otoresi öyküsü olan üç yaşında pediatrik bir olgu sunuldu.

Anahtar Sözcükler: Beyin-omurilik sıvısı; Mondini displazisi; otore.

Mondini dysplasia, an abnormal development of otic capsule in the seventh week of gestation, was first described by Carlo Mondini in 1791. The incidence in children is 1:1000 - 1:2000.[1] Mondini dysplasia is a common inner ear malformation related to recurrent meningitis and hearing loss.[2] Mondini dysplasia usually presents with hearing loss, rhinorrhea, otorrhea, and recurrent meningitis in the first 5-10 years of life and is one of the most common causes of spontaneous cerebrospinal fluid (CSF) otorrhea.

In this report, a male child with a history of two meningitis attacks and CSF otorrhea following tympanostomy tube placement and treated with exploratory tympanotomy via postaricular incision is presented.

CASE REPORT

A three-year-old boy presented with nasal obstruction unresponsive to medical treatment, and hearing loss of the right ear. The patient had a history of two pneumococcal meningitis attacks.

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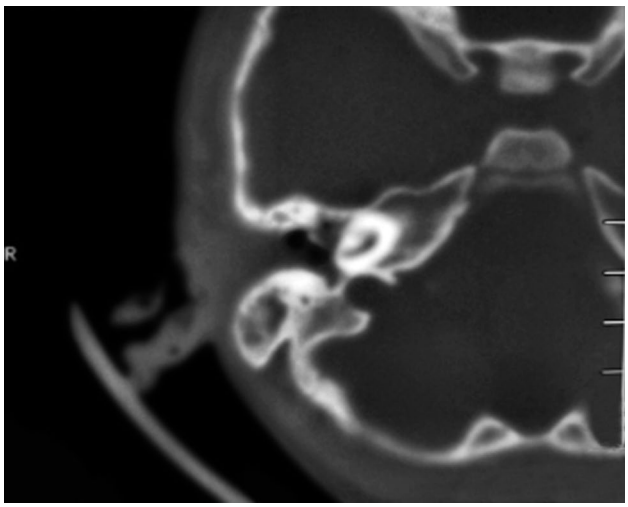

Figure 1. *High resolution computed tomography of the temporal bone showing the cochlea and vestibule replaced by a cystic cavity.*

Nasopharyngeal endoscopy revealed grade 3 adenoid vegetation. There was a type B pattern on the right side and type A pattern on the left side on impedance audiometry. Adenoidectomy and right tympanostomy tube placement were performed. After tube placement, a serous and pulsatile fluid discharge was observed immediately. Fluid sampling from otorrhea was done and high beta transferrin level consistent with CSF was detected. Brain evoked response auditory (BERA) test was performed with no response to 90 dB click stimulation on the right ear while the left ear was normal. High resolution computed tomography (HRCT) of the temporal bone showed the cochlea and vestibule replaced by a cystic cavity related to the enlarged internal auditory canal (Figure 1). Temporal bone magnetic resonance imaging (MRI) revealed high signal within right-sided mastoid cells and middle ear cavity, and a cystic dilatation with an incomplete configuration in the right vestibule on T_2-weighted images (Figure 2). The patient was diagnosed with Mondini dysplasia. Because of non-regressing CSF otorrhea, a repair operation was planned. Ceftriaxone was given preoperatively for meningitis prophylaxis, and lumbar drainage was done. The operation was performed via post-auricular approach. A piece of temporal muscle fascia and muscle were harvested. The tympanostomy tube was removed. The middle ear was visualized after tympanomeatal flap elevation. A defective stapes footplate and oval window were identified (Figure 3). There was CSF leakage from the defect. The stapes was removed and the oval window defect was plugged with harvested fascia and muscle, supported with tissue adhesive. Lumbar drainage was removed on the second day. There were no postoperative complications and no recurrence within the three-year follow-up.

DISCUSSION

Carlo Mondini first described Mondini dysplasia after the postmortem examination of the temporal bone of a patient with congenital deafness.[1] The cochlea makes 1.5 turns instead of 2.5, and the abnormal basal turn forms a short and large cochlear canal with cystic dilatation of the apical region. This malformation is characterized by an immature organ of Corti with decreased number of spiral ganglion cells and lack of intercalar septum (scala communis). There is a large and abnormal relationship between cochlear sac and vestibule. Semicircular canals, especially lateral, are dilated at various degrees.[3] Thalidomide use or infections like rubella can cause

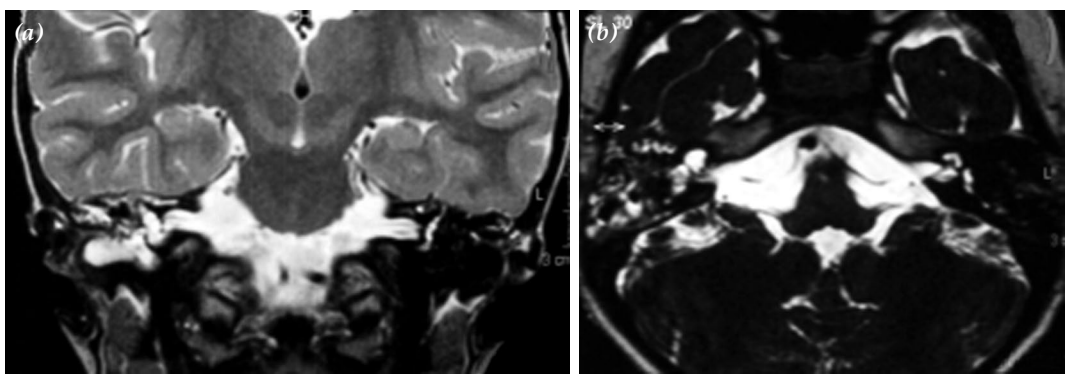

Figure 2. *High signal within right sided mastoid cells and middle ear cavity, and a cystic dilatation with an incomplete configuration at right vestibule;* ***(a)*** *T_2-weighted coronal MRI,* ***(b)*** *T_2-weighted axial magnetic resonance imaging.*

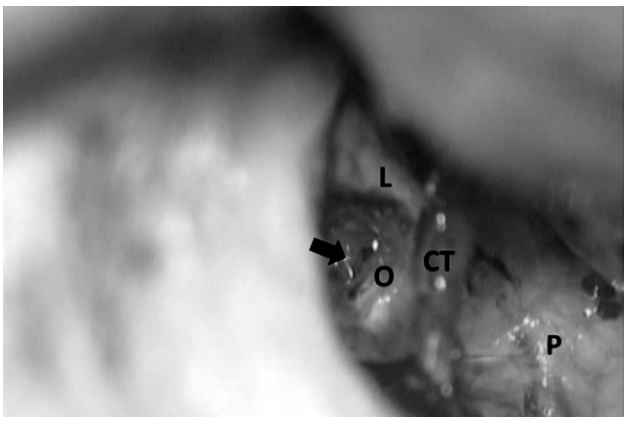

Figure 3. *Intraoperative view of the defective stapes footplate. CT: Chorda tympani; L: Long process of the incus; O: Oval window; P: Promontory; Black arrow defective stapes footplate.*

Mondini dysplasia which is mostly encountered sporadically. Patients present with congenital deafness when both canals are affected while unilateral disease may go undetected for long periods. There is a sensorineural type hearing loss due to corti organ abnormality and pressure changes at the perilymphatic sac.[4] Sensorineural hearing loss can also accompany conductive hearing loss due to CSF otorrhea and ossicular chain defects. An enlarged cochlear aqueduct, abnormal relationship between internal auditory canal and membranous labyrinth, and absent or defective stapes result in CSF leakage from the fistula at the oval window.[5] Cerebrospinal fluid leakage to the middle ear can cause recurrent meningitis attacks when bacterial contamination occurs or rhinorrhea if the tympanic membrane is intact.[6] The source of the effusion to the middle ear can be CSF leakage because of ear abnormalities like Mondini dysplasia instead of serous secretion of middle ear mucosa. Therefore, in these cases tympanostomy tube placement can cause unwanted complications like CSF otorrhea. Mondini dysplasia should be included in the differential diagnosis in patients with serous otitis media and recurrent meningitis attacks. Cerebrospinal fluid leakage is best demonstrated by magnetic resonance cisternography while characteristic temporal bone abnormalities are recognized with temporal bone HRCT.[7] Various methods to repair CSF leakage into the middle ear have been described in the literature. These include obliteration of vestibule with fascia, fat or muscle after stapedectomy, covering the oval window with bony elements, changing CSF flow direction with lumbar drainage, and subtotal petrosectomy and total obliteration of the ear. In our case, we used temporal fascia and muscle with additional tissue adhesive for plugging. Three-year follow-up was eventless.

In conclusion, in patients presenting with sensorineural hearing loss who have meningitis attacks in their history, abnormalities like Mondini dysplasia should be considered in the differential diagnosis. High resolution computed tomography of the temporal bone is extremely helpful for the correct diagnosis.

Declaration of conflicting interests

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