Bilateral Sigmoid Sinus Thrombosis and Otitis Media

Case Report

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Abstract

Sigmoid sinus thrombosis is a rare complication of otitis media and is known to be unilateral. In this report, we present a case of bilateral sigmoid sinus thrombosis secondary to bilateral otitis. To the best of our knowledge, there are no bilateral-simultaneous cases reported in literature to date. Here, the diagno-

sis of and treatment for the disease as well as predisposing factors are discussed.

Keywords: Complications, mastoiditis, sigmoid sinus thrombosis, otitis media

Introduction

Sigmoid sinus thrombosis is rarely seen (1). Its incidence varies between 3 and 4 per million and 75% of adult patients are female. In 70% of sigmoid sinus thrombosis cases, the reason can be explained and these reasons include infection, inflammatory diseases, trauma, neoplasm, autoimmune diseases, or oral contraceptive usage (2). Thrombosis can be associated with antithrombin 3, protein C or S deficiency, factor 2 or 5 mutation, active protein C resistance and hyperhomocysteinaemia, and genetic prothrombotic states (2). In cases, the etiology of which is infection, acute or chronic disease of the middle ear is seen frequently.

Lateral sinus and sigmoid sinus thrombosis are rare, but they are serious intracranial complications of otitis media, which can be fatal, and they are often associated with mastoiditis (1). Thrombosis typically begins from the region of sinus wall adjacent to the inflammation area and extends to the sigmoid sinus. Thrombus can spread superiorly to other neighbor sinuses or inferiorly internal jugular vein. Vascular occlusion developing due to thrombosis leads to increased intracranial pressure by impairing the drainage of the cerebrospinal fluid and causes hydrocephalus (1).

Clinically, patient can have behavioral symptoms like impaired consciousness, amnesia, severe headache, seizure, delirium, and focal lateralized findings. With common and efficient use of antibiotics, its incidence and also mortality rate have been reported to be dramatically decreased. The use of antibiotics decreased the incidence rate of complications from 17% to 1% (3).

Sigmoid sinus thrombosis, which is seen as a rare otitis complication, is presented in this study in the way that it was not reported in literature previously. Before preparing the study, the written informed consent was obtained from the patient with bilateral sigmoid sinus thrombosis.

Case Presentation

A 12-year-old male patient, who had been hospitalized in the Pediatric Clinic with the diagnosis of left mastoiditis, was consulted to our department because his computed tomography (CT) and magnetic resonance imaging (MRI) revealed bilateral mastoiditis and thrombus in the left sigmoid sinus (Figure 1). It was learned from the history of the patient that he had had headache for 1.5 month and he had been given antibiotherapy due to the pre-diagnosis of acute mastoiditis in another health center. No feature was observed in his medical history and his family history. In his physical examination, subfebrile fever was found and other findings were normal. In the initial ear examination of the patient, the right tympanum was pale and there was cambering in the posterior, but the left tympanum was in natural appearance. The patient had no neurological deficit. In the tympanometry test, tympanogram curve was obtained as type B in the right ear and as type A in the left ear. Audiometric examination revealed conductive hearing loss of 20 dB in the right ear and the mean pure tone was normal in the left ear. The patient was transferred from the Pediatrics Clinic and MRI venography was performed. Left lateral sinus thrombosis was detected in MRI venography Figure 2.

The patient was initiated enoxaparin sodium 2x0.3 mg in addition to ceftazidime 100 mg/kg/

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Figure 1. a, b. Computed tomography of the temporal bone (CT): bilateral otomastoiditis (a) CT of the temporal bone: (CT): Delta sign (the area shown by a black arrow) (b)

Figure 2. Magnetic resonance imaging (MRI) venography; Decreased flow associated with thrombosis is observed in the left sigmoid sinus, lateral and transverse sinus, and jugular vein at extracranial level

day and vancomycin 60 mg/kg/day. With the suspicion of left lateral sinus thrombosis due to acute-subacute otitis complication, the tympanum of the right ear was applied paracentesis

and seromucoid discharge was observed. Mastoidectomy was performed in the left ear. Mastoid cells were opened and inflamed tissues were removed. Biopsy was taken from inflamed tissues. Mastoid cavity was expanded superiorly until dural plate and posteriorly until sigmoid sinus (Figure 3). Sigmoid sinus was entered with an injector and it was observed that there was no pus and a little bleeding occurred due to the movement of the needle. Therefore, it was thought that there was re-canalizing thrombus in the sigmoid sinus and it was not infected. The result of biopsy was reported as chronic active non-specific inflammation. Because of the reproduction of coagulase-positive staphylococci in the fluid culture taken in paracentesis performed in the right side, the patient was initiated linezolid 2x20mg/kg, ceftazidime 100mg/kg/ day, and metronidazole 2x15mg/kg.

Five days after the operation, MRI and MRI venography were repeated because the headache of the patient aggravated rather than healing. The results of these examinations revealed decreased flow in the sigmoid, transverse, and confluence sinuses in the right ear (Figure 3). Thrombosis detected in the right side was evaluated as a complication of otitis media with effusion and right mastoidectomy was performed to the patient. Intense inflamed tissues in the mastoid cavity were cleaned. Bony layer over the sigmoid sinus was opened. Sinus was entered with an injector and controlled. It was seen that no pus was available. It was thought that there was thrombus in it, but it was not infected. When it was controlled with the needle, decreased flow was observed in the sinus.

MRI venography was performed again one week after the operation. It was seen that recanalization began in the right

Figure 3. a,b. Magnetic resonance (MRI) venography (filling defect in the right transverse sinus proximal, superior sagittal sinus, and confluence sinus) (a), Magnetic resonance (MRI) venography filling defect in the right transverse sinus proximal, superior sagittal sinus, and confluence sinus) (b)

side, but the dominance of thrombosis still continued in the venous system. The brain provided venous blood supply from the collateral regions (Figure 4). Because bilateral diffuse thrombosis was found in the patient, possible non-infectious reasons were investigated. Factor5 51(50-150), factor8 53(50-150), protein C 83(70-130), protein S 136 (60-140), Von Willebrand factor (VWF) 99 (60-150), homocysteine 12.9 (0-12), antithrombin 3 113 (80-120), and lipid a 48.9 (0-30) were detected. C3, C4 antiphospholipid antibodies, antinuclear antibodies, anti-ds dna lupus anticoagulants, and immunoglobulin negative were found and lymphocyte panel was observed to be within normal limits. Thrombophilia gene analysis was performed for the patient. Methylenetetrahydrofolate reductase (MTHFR) C677T heterozygote, Plasminogen activator inhibitor-1 (PAI-1) homozygote and angiotensin- converting enzyme (ACE) Ins/del heterozygote mutations were found. The results of other tests for genetic analysis were normal. MTHFR C677T heterozygote gene mutation was present in the patient and his homocysteine level was high at the limit. However, it was decided that existing mutation and homocysteine level did not have enough efficiency for making thrombosis.

After it was learned that the patient had recurrent aphthous lesions, pathergy test was carried out for Behçet's disease and the result was found as negative. No pathological finding was found in the peripheral smear of the patient. Coombs test was positive, but hemolysis was not detected and hemoglobin levels were within normal intervals.

After 14-day antibiotic treatment, it was discontinued. His anticoagulant therapy was switched to enoxaparin sodium

1x0.3 mg and 1x3\4 tablet of warfarin 5mg. The patient who was fully recovered symptomatically was discharged from the hospital with oral warfarin therapy. In his control examination done 2 weeks after discharge, the tympana of both ears were normal and the mean pure tone in both ears was within normal intervals in audiometric examination. In his MRI venography taken in the postoperative 4th month, partial recanalization was detected (Figure 5). Oral warfarin therapy of the patient has been continuing.

Discussion

Sigmoid sinus thrombosis was seen in this case as acute-subacute otitis media for the left ear and as complication of otitis media with effusion for the right ear. The finding of acute infection in mastoid cells and sigmoid sinuses of both sides and the absence of purulent material were attributed to his previous antibiotic therapy about 1.5 month ago.

The diagnosis and treatment of intracranial otitis complications are carried out considering clinical feature, neurological finding, and examination results. Ear-derived intracranial complications should be treated with otitis media which is a primary lesion. Treatment also includes surgical interventions such as mastoidectomy and removal of cholesteatoma as well as antibiotic therapy. It has been reported that 9% of patients undergoing mastoidectomy have intracranial complications (4). In our case, antibiotic therapy was used with mastoidectomy. Besides that, because antibiotics are used more commonly, intracranial complications are seen less in otitis media (4).

Barbara et al. (5) showed a genetic thrombotic disorder (prothrombin G20210A allele mutation) in deep cerebral sinus

Figure 4. Control magnetic resonance (MRI) venography taken after right and left mastoidectomy; a finding consistent with venous thrombosis, which involves superior sagittal sinus distal, right transverse sinus, and right sigmoid sinus in addition to sinus rectus, left transverse sinus, and left sigmoid sinus

Figure 5. Control magnetic resonance (MRI) venography taken after right and left mastoidectomy; a finding consistent with venous thrombosis, which involves superior sagittal sinus distal, right transverse sinus, and right sigmoid sinus in addition to sinus rectus, left transverse sinus, and left sigmoid sinus

thrombosis occurring in a child with non-coalescent mastoiditis as a predisposing factor. Venous thromboembolism is an important medical problem and it affects 1-5 per thousand individuals every year (6, 7). As a result of developments in medicine, coagulation and anti-coagulant mechanisms of blood have been explained at molecular level and various genetic risk factors have been defined for venous thrombosis. Genetic risk factors cause an increase in coagulation due to the impaired

balance between anticoagulant mechanisms and predisposition to coagulation. Thrombophilic diseases are multi-factorial disorders with environmental and genetic pathogenesis (6-8). Three most common genetic disorders leading to thrombophilia are factor V Leiden mutation, MTHFR mutation, and prothrombin gene mutation (FII). Moreover, thrombophilia can be seen in autoimmune diseases such as antiphospholipid syndrome and Behçet's disease (9). In studies conducted with healthy individuals, familial Protein S deficiency was found to vary between 0.03% and 2% (10, 11). It was detected between 1% and 13% in patients with thrombosis (12). In this case that we presented, the level of protein S was within normal limits.

Another factor causing predisposition to thrombosis is MTHFR gene mutation. Tuğ et al. (6) revealed the rate of MTHFR gene mutation as 30.4% in the Western Black Sea Region. In our case, there was MTHFRC677T heterozygote gene mutation and the level of homocysteine was high at the limit. However, it was thought that existing mutation and homocysteine level was not efficient enough to cause thrombosis.

In chronic otitis media cases with sigmoid sinus thrombosis, MRI is more sensitive than CT for diagnosis and it demonstrates blood flow better. Sinus congestion, lesion margins, and adjacent structures can be shown better through MRI. However, final diagnosis is demonstrated with MRI angiography or arteriography (13). In CT, the sign of delta appearing as an empty triangle covered with dura contrast in the sigmoid sinus area is typical for sigmoid sinus thrombosis (13, 14) (Figure 1). Delta sign may not always be observed, but it was seen in our patient.

Monitoring with contrast-enhanced CT is important in surgical planning stage for the cases having chronic otitis media with complications and it should be performed.

In our case, following the suspicion of thrombosis in CT, MRI and MRI venography were taken. The images were consistent with subacute thrombosis in the left transverse sinus and left sigmoid sinus, left jugular bulb, and the beginning of the jugular vein, and with acute-subacute thrombosis in the superior sagittal sinus, confluence sinus, and right transverse sinus proximal (Figure 2, 3).

Angiography is the most sensitive method for the diagnosis of thrombosis. However, there are some hesitations because it is interventional and it is risky for allergy to contrast agent. MRI angiography is more sensitive than CT and less interventional than angiography (14-16).

In cases with thrombosis secondary to otitis media, the infection is intervened surgically, especially with mastoidectomy. Nevertheless, the approach to thrombus has not been clarified yet. Although there are some researchers who defend opening sinus through thrombectomy and jugular vein ligation (11, 14, 15), the most frequently used technique is

surgical cleaning of the surroundings of only sigmoid sinus with surgical drainage of infection (17). Surgical approach to thrombus creates some risks such as septic embolization, expansion towards subarachnoid space, and difficulty in spontaneous recanalization. Therefore, efficient use of antibiotics has become widespread (17).

Similar to the way of surgical therapy, the use of heparin or oral anticoagulant agents in sinus thrombosis is controversial. While some of researchers defend routine usage (11, 15, 18, 19), others think that this treatment way will not be efficient (14, 16, 17). Even some researchers defending the use of oral anticoagulants observed that desired anticoagulant level was not reached in most of patients treated with heparin (20). It was suggested that sigmoid sinus recanalization could be performed without use of oral anticoagulant (17). It is defended that oral anticoagulants should be used for preventing complications associated with resistant or diffuse thrombus (11, 13, 14). Successful neurological results have been obtained with the use of oral anticoagulant in more than 80% of patients. Besides that, the mortality rate has been reported between 5% and 30% (2). At present, there is no consensus on the use of thrombolytic agent.

Conclusion

While a notable pathology was not found in the air cells of the left mastoid in our case, moderate chronic inflamed tissues were observed in the mastoid cells of the right side, but no classical acute mastoiditis findings were seen. Therefore, it was suggested that bilateral sigmoid sinus thrombosis in our case could be associated with other factors as well as mastoiditis.

With radiological imaging, clinical suspicion is also important for differentiating sigmoid sinus thrombosis immediately and rapidly initiating its therapy. In the treatment of sigmoid sinus thrombosis, antibiotic therapy, myringotomy, placement of ventilation tube, simple mastoidectomy, and anticoagulant therapy play a role in the solution of neurological sequelae and restoration of hearing.

Informed Consent: Written informed consent was obtained from the patient.

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