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Literature study: Superior semicircular canal syndrome (SSCS)

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Abstract

Superior Semicircular Canal Syndrome (SSCS) or Superior Canal Dehiscence Syndrome is a rare abnormality in the medical field in the form of balance disturbance or vertigo caused by loud noises or changes in pressure causing rupture of the roof of the superior semicircular canal. SSCS was first described by Lloyd Minor in 1998 after more than 70 years since Tullio and Hennebert discovered the Tullio phenomenon, a condition in which vertigo or nystagmus is triggered by loud noises, and the Hennebert phenomenon, a condition in which vertigo or nystagmus, is triggered by changes in pressure. The exact prevalence of superior semicircular canal syndrome is not known until now. Estimates of prevalence varied from 0.5–2% of cases in temporal bone studies, to 3–9% of cases in studies performed with CT scans. Cases of SSCS are more common in males than females. Clinical symptoms that appear in SSCS are caused by dehiscence so that patients will complain of autophonia, which the sound produced by their body sounds louder, hyperacusis, tinnitus, vertigo which is triggered by loud noises, and nystagmus. A Careful history, an examination of hearing and balance function, also high-resolution CT scan are needed to establish the diagnose of SSCS. Management can be done conservatively. If the complaints are severe, then operative options can be considered.

Keywords: Superior semicircular canal syndrome; Superior Canal Dehiscence Syndrome; Tullio phenomenon; Hennebert phenomenon

1. Introduction

Superior Semicircular Canal Syndrome (SSCS) or Superior Canal Dehiscence Syndrome is a rare abnormality in the medical field in the form of balance disturbances or vertigo caused by loud noises or changes in pressure causing rupture of the roof of the superior semicircular canal. Therefore, hearing and balance problems are one of the symptoms of SSCS. The two most common types of symptoms are those related to pressure and noise changes. SSCS was described firstly by Lloyd Minor in 1998 after more than 70 years since Tullio and Hennebert discovered the Tullio phenomenon, a condition in which vertigo or nystagmus is triggered by loud noises and the Hennebert phenomenon, a condition in which vertigo or nystagmus is triggered by changes in increased pressure, such as coughing, valsalva, and when lifting heavy objects [1–3].

The exact prevalence of superior semicircular canal syndrome is not known to date. Estimates of prevalence varied from 0.5–2% of cases in temporal bone studies, to 3–9% of cases in studies performed with CT scans. Cases of SSCS are more common in males than in females [2]. Clinical symptoms that appear in SSCS are caused by dehiscence so that patients will complain of autophonia, which the sound produced by their body sounds louder, hyperacusis, tinnitus, vertigo which is triggered by loud noises, and nystagmus. The diagnosis is based on a careful history, examination of hearing and balance function, and high-resolution of CT scan [1–4].

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2. Literature review

2.1. Embriology

During the development phase, the semicircular canals appear as budding in the membranous labyrinth of the otocyst. Development of the posterior and horizontal canals occur after the superior semicircular canals. Ossification occurs as the membranous labyrinth approaches its adult size. By the sixth month of pregnancy, the inner lining of the endosteal bone has formed. At birth, about 0.1 millimeter of bone separates the semicircular canals from the dura mater, which covers the temporal lobe. This bone grows thicker until the child is three years old [1,3].

The otic capsule will continue to thicken at the age of 4 months. The flap of superior semicircular canal is formed by 10 months of age. The bone layer of the superior semicircular canal reaches 1 mm thickness in the three years old. As a result, a CT scan cannot find the bone which covering the superior canal until the child is three years old. This layer of bone breaks easily due to trauma in about 1% untill 2% of the population and does not get thicker along with age. It will even thin out along with age. It is still not known why bone erosion occurs, or also known as slow erosion. Congenital anomalies can also cause this condition [1,3].

2.2. Etiology

The exact cause of superior semicircular canal syndrome is not known. There are several hypotheses such as congenital abnormalities, developmental abnormalities, especially one week postpartum, and genetic factors. The same study found that the temporal bone of children usually shows depletion of the bone layer over the superior semicircular canal, and begins to thicken around three years old. The researchers came to the conclusion that the thin covering bone that have been had by some adults caused by a defect in the development of this layer bone. This thin layer of bone can be damaged by trauma which will result in dehiscence [2,7,8].

2.3. Epidemiology

Superior semicircular canal syndrome is a fairly rare condition. Most of the cases counted so far only 65 cases were assessed over 9 years. A study of the temporal bone found a prevalence of SSCS in the amount of 0.5%, while the prevalence with a CT scan was 9%. Cases of SSCS are more common in men than women, with an age range from 13 to 78 years. There are no reports of children being infected. There are cases described of affected siblings, and this may lead us to consider some of the genetic factors associated with the development of SSCS [2,9].

Research conducted in 1999-2005, found that 97% of SSCS sufferers complained of vertigo due to loud noises, 82% of vertigo sufferers caused by changes in pressure in the middle ear, and 39% of hyperacusis sufferers. In 1999-2004, other researchers reported that 30 patients were diagnosed as SSCS. Out of 30 patients, 63% had chronic balance disorders, 41% had Tullio phenomenon, 44% had vertigo triggered by changes in pressure, 30% had hearing loss, and 7% had tinnitus [1,4].

2.4. Pathophysiology

There are two windows in the cocleovestibular system, namely the foramen rotundum and foramen ovale. The foramen ovale is connected to the base of the stapes, so that it can vibrate the perilymph of the vestibule scale if there is a flow of sound waves. The incoming vibrations will be transmitted through the Reissner membrane, so that it pushes the endolymph and basement membrane downwards, the perilymph of the tympanic scale will move and cause the foramen rotundum to be pushed outward. If dehiscence occurs in the superior semicircular canal, there will be a third window that plays a role, so that the endolymph in the labyrinth will continue the wave motion, causing activation of the vestibular system [1,2].

The cochlea is like a long chamber with two entrances and exits. Some people inside will be pushed out if the entrance is full and many people enter. These people are likened to voices. In contrast, the balance organ is a closed system, analogous to a race track. The movement of fluids in a confined space is detected as head movement, analogous to that of a person in a long room. The function of hearing and balance is independent if the balance organs are intact; However, if the balance organ has a third gate, fluid can move about and reduce the density of the cochlea, thereby reducing the sensation of sound. This condition will accelerate the discharge of fluid in an organ that has not been determined which causes complaints of vertigo [1,3].

2.5. Diagnosis

Clinical manifestations can occur over a long period of time or even appear suddenly. There is a tendency for a symptom to get worse after the first symptom appear. Symptoms that arise can occur up to 10-15 years before severe symptoms appear, so that sufferers will seek treatment relatively slowly, because it takes a long time to turn into severe symptoms. Symptoms that appear in semicircular canal syndrome can be auditory symptoms and vestibular symptoms. Auditory symptoms such as: autophony, hyperacoustics, tinnitus, and decreased hearing at low frequencies. Vestibular symptoms which are Tullio phenomenon (nystagmus and vertigo caused by loud noises), Hennebert's sign (nystagmus and vertigo caused by changes in pressure in the ear), oscillopsia (visual pressure disturbances, objects appear to sway due to changes in loud noises or changes in pressure) , and vertigo [10].

2.6. Physical Examination

On otoscopic assessment, no pathology or defects were found in the outer or middle ear. Lateralize to the affected ear or affected side using the 512 Hz tuning test. Nystagmus is evoked by low frequency or Valsalva movements [1]. Valsalva in a sitting position is not recommended. During induction of nystagmus with the Valsalva test in the sitting position, the duramater can effectively occlude the superior semicircular canal dehiscence, thereby masking the defect in the labyrinth bone and producing a false-negative test. In the supine Valsalva test, the patient is placed in a supine position while performing the usual Valsalva maneuver. Lying in the supine position prevents the duramater from accidentally closing the defect in the petrosal portion of the temporal bone. This hopefully exposes the defect in the labyrinth bone and allows visualization of the nystagmus [11]. Physical examination which is considered standard for examination of the vestibular apparatus such as electronystagmography and rotatory chair fails to show significant changes. Frenzel glasses should be worn in these patients to look for nystagmus to suppress the effects of ocular fixation [2].

2.7. Supporting Examination

Tympanometry examination usually gives normal results, unless accompanied by abnormalities in the middle ear and ossicles. Conductive deafness at low frequencies below 1000 Hz is usually the form of hearing loss that occurs in SSCS, with hearing threshold greater than or equal to 0 decibels (approximately -5 to -10 decibels). The third window of the superior semicircular canal raises the auditory threshold for air conduction, resulting in conductive deafness [1,12].

In the case of SSCS, Gait, Romberg, Stepping, Dix Hallpike, Head shake and Head thrust examination showed no abnormalities. However, examination with given of a 100-110 dB stimulation at a frequency of 500-2000 Hz will cause vertical and torsional nystagmus. Fistula assessment will get vertical and torsional nystagmus with a slow phase away from the affected side. But a valsalva assessment shows that nystagmus in the opposite direction. When tympanometry and valsalva are performed, nystagmus can be seen in SSCS patients, which can be diagnosed by electrostagmography (ENG). To establish the diagnosis of superior semicircular canal syndrome, a high-resolution of CT scan is essential [1].

On histopathological examination, the lamellae can be seen parallel to the surface of the superior petrosal sinus indicating that the ossification is stable at one point. This implies that the process leading to dehiscence can be a chronic and progressive condition. However, in 1000 temporal bone specimens, no local bone changes, osteoporotic changes or underlying bone disease were found in any of the specimens to explain the depletion. In addition, the prevalence of otosclerosis is not significantly different in cases of SSCS [10,13].

2.8. Differential Diagnosis

The differential diagnosis of semicircular canal syndrome is Meniere's disease, and Benign Paroxysmal Positional (BPPV). Attacks of vertigo, tinnitus, decreased in hearing that is fluctuating and a feeling of fullness in the ears are symptoms of Meniere's disease syndrome. Meniere's disease mostly occurs in adult men or women in the range of age 20-50 years. The cause of Meniere's disease is neurochemical and hormonal abnormalities in the blood flow to the labyrinth resulting in electrolyte disturbances in the labyrinth fluid, allergic and autoimmune reactions. Meniere's disease occurs when there is an imbalance of ear fluid caused by malabsorption in the endolymphatic saccus. Clinical symptoms in Meniere's disease are vertigo accompanied by nystagmus, nausea and vomiting which are periodic and subside in subsequent attacks. In each attack, it is usually accompanied by fluctuating sensorineural deafness. The hearing is felt to improve again, if there is an absence of an attack. Other symptoms accompanying the attack are tinnitus which sometimes persists even outside the attack, and a feeling of fullness in the ear. This disease can be cured without medication and the symptoms of the disease can disappear completely [16,17].

Benign Paroxysmal Positional (BPPV) is a vestibular disorder, with symptoms of sudden dizziness followed by nausea, vomiting and cold sweats, which is triggered by changes in the position of the head relative to gravity, accompanied by

torsional nystagmus, in which the upper part of the eye rotates towards the affected ear. BPPV usually affects women more often than men at the age of 50-70 years [18].

2.9. Management

Conservative treatment of SSCS can be done by avoiding trigger factors, such as lifting heavy loads, bending, changes in air pressure, for example during air travel, riding high-speed elevators, diving, blowing loudly, and loud noises. Ventilation pipes (grommets) can be used to minimize changes in air pressure in the middle ear. Most patients only experience mild complaints, so they do not require operative treatment. Complaints that do not interfere with the patient's activities, conservative therapy can be done. However, if it is very disturbing, operative therapy is carried out. Operative therapy for now is still being developed [1,9].

Lining the superior semicircular canal that is experiencing dehiscence (superior canal resurfacing) and occluding the superior semicircular canal (canal plugging) are two techniques used in operative management. The materials used to cover the dehiscence of the semicircular canals are soft tissue, bone, surgical materials, while bone powder and fascia can be used to block them [1]. The transmastoid approach to plugging involves separate labyrinthectomies, one proximal and one distal. Blockage dehiscence can use fascia, bone or bone powder that is inserted into the lumen of the superior semicircular canal. Thus, the dehiscence can be closed, thereby preventing the abnormal transmission of sound energy. Clinical symptoms that arise will slowly disappear. Because normal function of the semicircular canals is compromised, persistent imbalance often occurs, and movement of the head in the plane of the canal produces transient oscillopsia [2,3].

Very little case has been reported in the literature on the postoperative imaging appearance of SSCS. As the majority of patients are released symptom after surgery, there is usually no further imaging required. However, fear of complications, such as a fistula or pneumo-labyrinth, may recommend imaging evaluation. Dournes *et al*. reported a series of cases of postoperative CT scan of SSCS showing defect of the cover. Patients can have new bone formation at the site of closure with cartilage as early as 20 months post-operative. Some patients experience bone formation adjacent to dehiscence. The mechanism of bone formation is unknown. It is also not clear whether ossifying of the cartilage or growing of the new bone. MR imaging is very useful after surgery to evaluate the patency of the superior semicircular canal because post-operative healing may lead to canal fibrosis and secondary obstruction [3,12].

3. Conclusion

Superior Semicircular Canal Syndrome (SSCS) is a rare condition characterized with the vestibular symptoms which caused by intense sound stimulation or pressure changes due to rupture of the covering bone of the superior semicircular canal, accompanied by nystagmus, autophony, hyperacoustics, and decreased hearing. The characteristic signs and symptoms of SSCS are rarely seen on admission. Therefore, it is necessary to carry out a supporting examination approach to make a diagnosis, such as audiometric examinations, CT scans, and MRI. A Careful history and a careful examination are needed to establish the diagnosis of SSCS which are very rare. Management can be done conservatively. If the complaints are severe then consider operative options.

Compliance with ethical standards

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Disclosure of conflict of interest

The authors have declared that no competing interests exist in this study.

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