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Etiopathogenesis behind Semicircular Canal Dehiscence Syndrome: Review Article

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Abstract

Semicircular canal dehiscence remains one of the rare and least touched chronic vestibular diseases worldwide. Even though microbial, congenital, and genetically determined, as well as mixed characteristics are known to be involved in the pathogenesis of superior semicircular canal dehiscence syndrome (SSCDS), many facets of the pathogenesis of semicircular canal dehiscence syndrome still need to be simplified. Management plan related to pathogenesis has not yet been established. The primary objective of this review is to present and evaluate the etiopathogenesis behind SSCDS. This study is a systematic narrative review. A PubMed search (1970–2022) was performed for studies on epidemiology and pathogenesis of SSCDS. All included articles were categorized according to level of evidence. Five hundred and sixty papers were identified, of which 25 were found to be relevant for this review. SSCDS is a multifactorial disease. There is still no consensus as to what the specific etiology is behind the syndrome. No convincing evidence is available for most associated factors and pathogenesis. Important objectives in research of SSCDS should be achieving consensus about the definition of SSCDS and gaining more in-depth knowledge of the pathogenesis of SSCDS, especially the role of congenital and acquired causes. There is still a need for further well-designed studies on the various etiopathogenesis and management of SSCDS.

Keywords

- superior semicircular canal
- superior semicircular canal dehiscence
- vertigo
- middle cranial fossa

Introduction

Any pathology causing fluid motion that can cause cupular deflection in the absence of head rotation and even that is read by the brain is head motion (vertigo) which is accompanied by eye movement (nystagmus). The association of vertigo induced by pressure change or loud sounds with dehiscence of the superior semicircular canal (SSC) was first laid out in 1998.[1] In 1949, Cawthorne[2] pointed out that a "third window" mechanism is responsible for the positive Hennebert sign of labyrinthine fistula. Dehiscence of bone over the SSC and consequential connection with the middle cranial fossa allows a third mobile window into the inner ear in superior semicircular canal dehiscence syndrome (SSCDS). In a normal person where the semicircular canal is covered with bone, sound coming from the stapes and oval window is dissipated across the cochlear partition. In the third labyrinthine window, there is an additional pathway for acoustic energy dissipation that goes to semicircular canal, which leads to fluid movement causing cupula deflection. Increased intracranial pressure can also trigger semicircular canal (Urban Schwarzenberg).[3] Tragal compression, nose-blowing, or other also results in pressure gradient between the inner ear and middle fossa. In addition, SCDS may be characterized by autophony, conductive hearing

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loss, which is not due to middle ear pathology, and/or pulsatile tinnitus.[4] High-resolution computed tomography scans with reconstructions in the plane of the superior canal and orthogonal to that plane are done to establish the diagnosis.

Materials and Methods

A PubMed search (1970–2022) was performed for studies on epidemiology and pathogenesis of SSCDS. All included articles were categorized according to the level of evidence. Articles were sorted based on keywords "superior," "canal," "dehiscence," "vertigo" in all age group irrespective of gender
Design: Systematic narrative review.

Etiopathogenesis

The true cause of canal dehiscence syndrome is still unknown. The dehiscence may, at least in part, be congenital and also may have occurred during the development of the inner ear. It can as well be caused from certain infections and head trauma. The prevalence of SCD is 0.5 to 0.6% as seen in histological studies in cadaver temporal bone.[5,6] The prevalence of SCD from reviewing high resolution CT scans accounts to 4 to 8%.[7,8] Dehiscence alone cannot cause the syndrome since it needs some sort of trigger to initiate it, and therefore many patients might have dehiscence which remains asymptomatic. SCD has been found to be[1] congenital,[2] acquired, or[3] mixed where a genetic predisposition followed by a secondary event trigger SSCD. Predominantly, it has a middle-aged onset and there is a statistically significant increase in prevalence with increasing age,[9] makes a pure congenital cause less likely. However, SSCDS has also been reported in children and affected siblings[10] indicating genetic associations.

Congenital Anomalies

The SSC roof was indicated to show continuous thickening during the early years of life, with reports proposing that dehiscence ensues from a defect in postnatal development.[11] Due to protrusion of the membranous labyrinth into the middle cranial fossa in fetuses, adhesion with the overlying dura has been postulated to prevent entire bone coverage of the SSC in some individuals resulting into thinning of the roof of canal and thereby causing SSCD.[12] Ossification of the petrous bone over the labyrinth occurs from birth to the age of three.[4,5,13] If the bone is insufficient, the resulting morphological abnormality may prone certain individuals to SSCD.

Secondary Triggers

Shearing force arising from a head injury or trauma to the temporal bone causes slow progressive erosion of thin bone and increased dural elasticity with time. A sudden increase in intracranial pressure is also a suggested secondary trigger.[14] A cadaveric study concluded that, when only an endosteal layer is present over the SSC, this is accountable to secondary events that could cause dehiscence.[15] Nager explained single case of a defect of the superior canal in the middle cranial fossa. He attributed the bony defect to senile osteoporosis of the petrous bone.[13] Changes seen in osteoporosis, thinning of trabeculae, and relative increase in the fatty marrow can be related to semicircular canal. A study also accounted for an association between aging and SSCD prevalence. Decreased semicircular canal roof height with age indicates that SSC dehiscence might be an acquired phenomenon, linked somehow to aging of the base of the skull.[16] Changes in cranial sphenoid angle with age demonstrate skeletal aging of the skull base. Skull base wear has been seen as a general progressive process, denoting the frequency of bilateral involvement.[17] SCD may also be hugely associated with a reduction in contralateral temporal bone thickness (< 0.5 mm). This theory of acquired etiology and skull base wear was carried out by other temporal bone pathologies such as spontaneous oto-liquorrhea due to osseous-dural fistula of the tegmen tympani.[18]

Combination of Causes

A continuous negative balance of labyrinthine osseous metabolism occurring with a congenital thin layer of bone over the SSC has been suggested as casual mechanism of SCD.[19] Syphilis has shown its effect on the otic capsule, especially the semicircular canals. Lesions range from a proliferative periostitis and fibrosis of the canals to bony obliteration of the semicircular canals. Specimens with clinical history of syphilis when examined for gummas as well as the nongummatous manifestations of otosyphilis resulted in periostitis, fibrosis, and deformation of the semicircular canals.[20] Multiple small intensity cranial trauma from combat sports or diving, for instance, has been associated with SCD. A defect in the floor of the middle cranial fossa at the level of the canal might also occur secondary to increments in the pressure of the cerebrospinal fluid.[21] Similarly, arachnoid granulations are known to cause tegmen erosion, encephaloceles, and ongoing cerebrospinal fluid leaks lead to dehiscence in wall of SSC.[22] Nakajima et al showed that an opening can change the impedance of the otic capsule to an adequate degree to cause a functional third mobile window.[23] Vestibular schwannoma-related erosion is noted in few subjects. Otosclerosis was found around the oval window on both sides. Ten percent of incidence of otosclerosis was seen in large samples of temporal bones.[24]. Microcavitations are found in periosteal and endosteal layers of temporal bone specimens of SCD. Microcavitation within the temporal bone is most likely due to osteoclastic activity, which is seen in both young and old patients, patients with and without otosclerosis, and in cases with SSCD. Pathologic changes of chronic otitis media also result in thinning of semicircular canal followed by its dehiscence. Bone remodeling within the otic capsule has been reported to be inhibited especially at or near the cochlea, except under some pathological conditions such as otosclerosis, Paget's disease, or mastoiditis, when bone remodeling can occur.[25].

Final Comments

Due to the absence of a single theory, it is suggested that multiple genetic, congenital, and acquired causes are responsible for SCD. Elevated intracranial pressure over the years may be an

etiologic factor in bilateral thinning of the skull base. Our study reveals symptoms and signs of SCD first develop in adulthood. We suggest, therefore, that a secondary event may occur, which fractures the thin bone or destabilizes dura over a pre-existing dehiscence. This second event may be a head injury or sudden change in intracranial pressure. The increased radiologic prevalence of SCD among older age groups suggests that this is more commonly an acquired rather than congenital condition. There is still a need for further well-designed studies on the various etiopathogenesis and management of SSCDS.

Conflict of Interest
None declared.

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