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Spontaneous Tegmen Defect and Semicircular Canal Dehiscence: Same Etiopathogenic Entity?

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Objectives: To describe the clinical and radiologic features of associated spontaneous tegmen defects (STDs) with semicircular canal dehiscences (SCCDs) and to postulate a novel etiopathogenic hypothesis of these pathologic conditions.
Methods: Medical records of all patients with surgically confirmed STD between 2001 and May 2010 were reviewed. We excluded all secondary tegmen defects. Clinical, audiological, and radiologic data were analyzed.
Results: Twenty-three patients matched the inclusion criteria. Semicircular canal dehiscence was associated to STD in 13 patients. Of these patients, 12 (95%) had protruding superior semicircular canals in the middle cranial fossa versus only 3 (30%) of 10 patients for the nondehiscent cases. Twenty-two patients complained of hearing loss. Cerebrospinal fluid leak was found in 13 patients. Four patients had history of meningitis. Vestibular symptoms were present in 8 patients.
Conclusion: This is the largest series of reported coexistence of STD and SCCD in the literature. Protrusion of the superior semicircular canal in the middle cranial fossa is probably an additional factor underlying STD and SCCD etiopathogeny. Semicircular canal dehiscence should always be looked for when STD is present. **Key Words:** Cerebrospinal fluid leak—Semicircular canal dehiscence—Spontaneous tegmen defect.
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Temporal bone defects may be secondary or primary in nature. Acquired tegmen defects are most commonly due to chronic otitis media, with or without cholesteatoma, or to middle ear and cranial base surgery. Other causes include temporal bone trauma, neoplasia, and radiotherapy (1). Spontaneous temporal bone defects present in 2 major distinct categories based on the age of onset. Congenital temporal bone abnormalities in children include defects in the Hyrtl fissure, the wide fallopian canal, the Mondini dysplasia, and the patent cochlear acqueduct (2). Spontaneous temporal bone defects in adults are less common and generally present without previous history of petrous pyramid disease. They occur predominantly in the floor of the middle cranial fossa (MCF) at the level of the tegmen (1).

Spontaneous tegmen defects (STDs) and semicircular canal dehiscences (SCCDs) are 2 varieties of spontaneous temporal bone defects that have been widely explored and analyzed in the literature both anatomoradiologically and clinicosurgically, but in a separate way. A recent histologic study, performed on 79 fetal temporal bones at different gestational ages, demonstrated that the medial part of the tegmental process develops from the cartilaginous otic capsule during fetal development (1).

Considering their common anatomic location and embryological origin, the tegmen and the semicircular canals are theoretically exposed to the same potential etiopathogenic factors leading to the development of spontaneous tegmental and SCC defects. This fact implies that the coexistence of STD and SCCD should be more frequent than reported in the literature (3–6). We think that this discordance is probably because STD and SCCD are considered as 2 distinct pathologic conditions and therefore usually explored separately. In fact, a recent anatomoradiologic study found an incidence of 36.4% of associated STD in a series of 22 SCCDs confirmed with high-resolution computed tomography (HRCT) (2).

This study focused mainly on the aspect of the coexistence of STD with SCCD as an indicator of a unique disease entity. Moreover, we have shed light on potential anatomic factors probably involved in the etiopathogeny of temporal bone defects. We have postulated a novel hypothesis that may help understand the mysterious underlying mechanisms of STD and SCCD. The secondary purpose of the present article was to describe the clinical presentation of such a combination as well as its radiologic

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features in a consecutive series of 23 patients diagnosed for symptomatic STD in our otology and neurotology department during a period of 9 years.

MATERIALS AND METHODS

A retrospective chart review of all patients scheduled for surgical repair of STD at our otology and neurotology department from January 2001 to June 2010 was undertaken. Patients with a history of head injury, chronic otitis media, middle ear and cranial base surgery, temporal bone neoplasia, and osteoradionecrosis were excluded from the analysis. Demographic pattern and clinical presentation data were collected. High-resolution computed tomography was performed in all patients. Collimated axial and coronal images of 0.5 or 1 mm were obtained. Multiplanar reconstructions in the Stenvers (plane of the superior canal) and Pöschl (plane of the posterior canal) planes were made when SCCD was suspected. The following features were determined in every computed tomographic scan: degree of mastoid pneumatization, integrity and thickness of tegmen tympani and antri, presence of dehiscence of the superior or posterior SCC, presence of supralabyrinthine mastoid cells over the superior SCC and behind the posterior SCC, and protrusive SCC in the middle or posterior cranial fossa (Table 1). Magnetic resonance imaging was performed in patients who had suspicion of meningocele or encephalocele from the computed tomographic findings. We looked mainly for magnetic resonance imaging stigmats of high intracranial pressure.

RESULTS

In the database, 23 patients were found to match the inclusion criteria (Table 2). Sex ratio was 12:11. The mean age was 54.4 years, ranging from 30 to 72 years. Fifteen patients (65.2%) were misdiagnosed for a serous otitis media; 7 of them (30.5%) had previously benefited from tympanostomy tube placement. Cerebrospinal fluid (CSF) leak was the main clinical symptom in 13 patients (56.5%), with 10 clear otorrhea and 3 rhinorrhea (β_2 transferrin test was not performed in these patients). One-third of these misdiagnosed cases (4 patients) were complicated by meningitis. Vestibular symptoms were present in 8 (61.5%) of the 13 SCCD cases. Tullio and Hennebert signs were found in 7 (53.8%) of these patients. All but 1 patient complained of hearing loss (95.6%). This patient had asymptomatic unilateral STD with ipsilateral superior SCCD manifested by an authentic Minor syndrome.

All patients had HRCT performed (Figs. 1–3). Table 3 summarizes the overall findings. Ten patients (43.5%) had STD alone, which was unilateral in 8 patients and bilateral in the remaining 2 patients. Thirteen patients (56.5%) had SCCD combined to STD. Of these patients, 7 (53.8%) presented both bilateral STD and bilateral superior SCCD. Of them, 2 (8.7%) had additional bilateral posterior SCCD. Bilateral superior SCCD associated with unilateral STD appeared in 2 patients. In 5 patients (21.7%), the STD and the superior SCCD were both unilateral and on the same side. The tegmen was found to be excessively thin in

TABLE 1. *Adopted criteria for computed tomography features*

(A) Degree of mastoid pneumatization:
1. Very pneumatized mastoid: air cells in the mastoid tip, in the petrous apex, and/or around the carotid canal
2. Fairly pneumatized mastoid: partial pneumatization sparing the mastoid tip, the petrous apex, and the carotid canal
3. Poorly pneumatized mastoid: air cells limited to the antrum and periantral region
(B) Tegmen tympani and/or antri thickness:
1. Absent: tegmen absent in 1 or more coronal cuts
2. Very thin: tegmen barely visible in most coronal cuts
3. Thick: tegmen clearly visible in all coronal cuts
(C) Semicircular canals:
1. Superior SCC dehiscence: absence of bone overlying the canal in the Pöschl plane
2. Posterior SCC dehiscence: absence of bone overlying the canal in the Stenvers plane
3. Thin bony coverage of superior or posterior SCC: bone barely visible in the coronal cuts for the superior SCC and axial cuts for the posterior SCC
(D) Perilabyrinthine air cells:
1. Present (Fig. 1): air cells between the superior SCC and the tegmen and between the posterior SCC and the posterior fossa plate
2. Absent (Fig. 2): SCC directly in contact with middle and/or posterior cranial fossa
(E) Protrusion of SCC in the cranial fossa:
1. Protruding superior SCC (Fig. 3): superior canal overpassing the horizontal line of the tegmen in coronal cuts
2. Protruding posterior SCC: posterior canal overpassing the oblique line of the posterior fossa plate in axial cuts

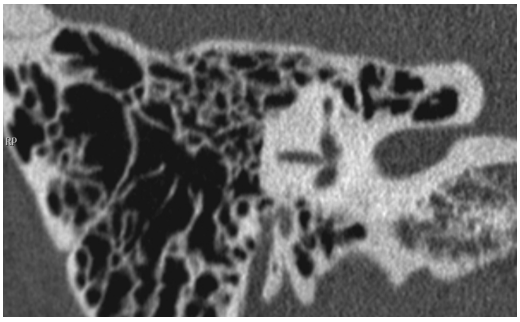

FIG. 1. Coronal computed tomographic scan showing an example of a nondehiscent superior SCC, which is covered by supralabyrinthine air cells.

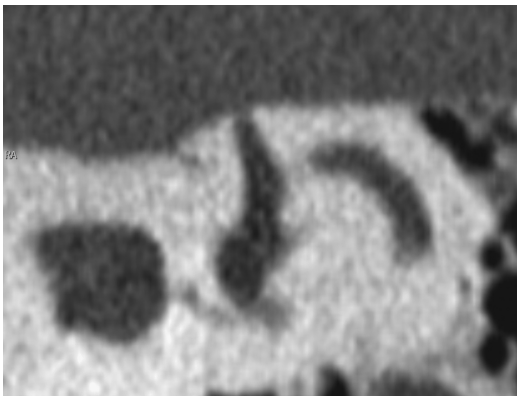

FIG. 2. Coronal computed tomographic scan showing an example of dehiscent superior SCC, which is at the same level of the tegmen tympani.

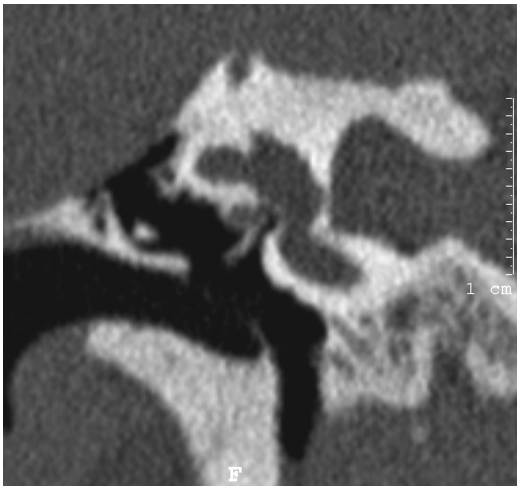

FIG. 3. Coronal computed tomographic scan showing an example of dehiscent superior SCC, which overpasses the level of tegmen tympani (protruding canal).

82.6% of the cases. Of these patients, 78.2% had hyperpneumatized mastoid according to our adopted criteria. The superior SCC was protruding into the MCF in 92.3% of the patients who had confirmed superior SCCD in the Pöschl plane. Only 30% of the patients who had isolated STD exhibited protruding superior SCC in the MCF. The bone covering the superior SCC in this category of patients was not dehiscent but excessively thin. The remaining 70% of the patients with isolated STD had the superior SCC protected either by supralabyrinthine cells (40%) or by a thick layer of bone (30%).

The posterior SCC was protruding bilaterally in the posterior cranial fossa in only 4 patients (17.4%), 2 of whom had posterior SCCD. In most cases, the posterior SCC was covered by mastoid air cells (47.8%). Of the posterior canals, 34.8% were neither protruding nor protected by mastoid cells but had rather a thick bony coverage. Regarding the group of patients who presented with unilateral superior SCCD, the contralateral superior SCCs were protrusive with thick overlying bone in 2 patients, at the same level of the tegmen in 2 patients, and covered by supralabyrinthine cells in the last patient.

The only patient who had poorly pneumatized mastoids in our series had bilateral STD and bilateral superior and posterior SCCD. This patient had confirmed high intracranial pressure with protruding superior and posterior SCC in the MCF and the posterior cranial fossa, respectively.

Bilateral otosclerosis foci were present in a female patient who had unilateral STD and bilateral superior SCCD. Her twin sister also had bilateral STD with bilateral superior SCCD.

Magnetic resonance imaging was performed in 18 patients (78.3%), showing meningoencephalocele in 4 patients. Hydrocephalous was found in 2 patients. It was a consequence of Sylvius aqueduct stenosis in 1 patient and supracerebellar cyst with Chiari malformation in the other. Bilateral diffuse meningiomatosis was noticed in 1 patient mainly in the frontotemporal and parieto-occipital areas.

DISCUSSION

Spontaneous tegmen defects and SCCDs have been increasingly drawing the interest of many neurotologists during the last decade. Carey et al. (7) had largely studied the incidence of superior SCCD and found a rate of 0.5% in a survey of 1000 temporal bones. A similar incidence value (0.6%) was reported by Crovetto et al. (4) in their original anatomoradiologic study on 604 ears. Regarding STD, Åhren and Thulin (8) found an incidence of 21% in a series of 94 temporal bones, which is very similar to the findings of Lang (9) (20%) in his 70 temporal bone series.

These studies addressed STD and SCCD as 2 different pathologic conditions. In fact, very few articles reported the association of STD to SCCD (3–6). Fewer articles analyzed the anatomic and radiologic features accompanying this kind of association (3,4). Brantberg et al. (3) reported in a series of 8 unilateral superior SCCD the association of ipsilateral STD in 7 patients. Crovetto et al. (4), in their 604 ears series, found that the incidence of STD is statistically greater in ears with SCCD (36.4%, i.e., 8 of 22 ears with SCCD) than ears without SCCD (10% of 582 ears without SCCD) with a $p < 0.001$. This is, to date, the largest published series of associated STD and SCCD.

Our study provides further evidence supporting the original findings of Crovetto et al. (4). In fact, the major finding in our series is that SCCD was associated to STD in 56.5% of the cases. It is also noteworthy to mention that more than half (53.8%) of the combined cases had a bilateral pattern and that nearly 85% of the unilateral combined cases had STD and SCCD on the same side, which is similar to the findings of Brantberg et al. (3). We may conclude at this stage that STD and SCCD may have the same etiopathogenic factors based on the following facts: their occurrence is concomitant in more than half of the cases, they are mostly bilateral, and they occur on the same side in case they are unilateral.

Tóth et al. (10) performed an anatomic and histologic study on 79 human temporal bones of different ages ranging from the 14th fetal week to adult age. They found that the medial part of tegmen tympani develops from the

TABLE 2. *Epidemiological data and clinical patterns*

Sex, n (%)	
Male	12 (52.2)
Female	11 (47.8)
Age (yr)	
Mean	54.4
Range	30–72
Hearing loss, n (%)	22 (95.7)
Conductive	10 (43.5)
Mixed	10 (43.5)
Sensorineural	2 (8.7)
Normal, n (%)	1 (4.3)
Otitis media effusion, n (%)	15 (65.2)
CSF leak, n (%)	13 (56.5)
Otoliquorrhea	10 (43.5)
Rhinoliquorrhea	3 (13)
Meningitis, n (%)	4 (17.4)
Vertigo, n (%)	8 (34.8)

TABLE 3. *Findings from the HRCT*

	n (%)	Unilateral	Bilateral	SCC protrusion	Perilabyrinthine cells	SCC thick bony coverage
STD alone	10 (43.5%)	8/10 (80%)	2/10 (20%)	3/10 (30%)	4/10 (40%)	3/10 (30%)
STD + SCCD	13 (56.5%)	6/13 (46.2%)	7/13 (53.8%)	12/13 (92.3%)	1/13 (7.7%)	0/13 (0%)
After SCCD	2/23 (8.6%)	0/2 (0%)	2/2 (100%)	4/23 (17.4%)	11/23 (47.8%)	8/23 (34.8%)

Patient	**STD**	**SCCD**	**Mastoid**	**Tegmen**	**SSCC protrusion**	**PSCC protrusion**	**Perilabyrinthine cells**
1	L	—	+++	Thick	—	—	Sup and Post ×2
2	L	—	+++	Thin	×2	—	Post ×2
3	L	—	+++	Thin	×2	—	Post ×2
4	L	—	+	Thin	×2	—	Post ×2
5	R	—	+++	Thin	—	—	Sup and Post ×2
6	R	—	+	Thin	—	—	Post ×2
7	×2	—	+++	Thin	—	—	Sup and Post ×2
8	×2	—	+++	Thin	—	—	Sup and Post ×2
9	L	—	+	Thin	—	—	Post ×2
10	L	—	+++	Thin	—	—	—
11	×2	Sup ×2	+++	Thin	×2	—	—
12	×2	Sup ×2	+++	Thin	—	—	—
13	×2	Sup ×2	+	Thick	×2	—	Post ×2
14	×2	Sup ×2	+++	Thin	×2	×2	—
15	×2	Sup and Post ×2	+++	Thick	×2	×2	—
16	×2	Sup and Post ×2	—	Thin	×2	×2	—
17	L	Sup ×2	+++	Thin	×2	—	—
18	R	Sup ×2	+++	Thin	×2	×2	—
19	R	Right Sup	+++	Thin	Right	—	—
20	R	Right Sup	+++	Thin	×2	—	Post ×2
21	R	Right Sup	+++	Thick	Right	—	Left Sup
22	R	Right Sup	+++	Thin	Right	Right	—
23	L	Left Sup	+++	Thin	×2	—	—

(×2) indicates bilateral; (+), fairly pneumatized mastoid; (+++), hyperpneumatized mastoid; L, left; PSCC, posterior semicircular canal; R, right; SCCD, semicircular canal dehiscence; SSCC, superior semicircular canal; Sup, superior.

otic capsule during chondral ossification forming the tegmental process. The lateral part of the tegmen tympani develops from the squamous part of the temporal bone. They demonstrated that the medial anlage of the tegmen tympani fuses with the cochlear part of otic capsule after the 25th fetal week. They concluded that the congenital defect of the tegmen may be due to an incomplete development of the tegmental process of the otic capsule and, thus, can be defined as an inner ear defect.

Therefore, in view of their common embryological origin, SCCD and STD may be considered as a unique etiopathogenic entity owing to a developmental deficiency in the inner ear. In fact, Chen et al. (11) found an incidence of 4% and 11% posterior and superior SCCDs, respectively, in a series of 131 temporal bone HRCT in the pediatric population. None of the patients displayed vestibular symptoms; nevertheless, all of them had hearing loss, which was the main indication for HRCT (11). This interesting finding points toward a developmental cause, although the asymptomatic feature of these patients remains enigmatic.

Although the developmental theory seems to be most plausible, the sequence of development of semicircular canals does not support it. Development of the semicircular canals starts with the budding of the membranous labyrinth from the otocyst. The first canal to develop is the superior canal followed by the posterior and then the horizontal canal. When the membranous labyrinth nears adult size, ossification starts at the basal turn of the cochlea and then progresses toward the semicircular canals in the same order previously listed (12,13). If an interruption or delay were to occur during ossification of the superior semicircular canal, the posterior and lateral canals would also be defective because their ossification develops later. Therefore, a developmental mechanism does not fully explain the predilection of dehiscences to the superior and posterior SCC, specifically sparing the lateral SCC. We think that other factors, mainly anatomic, probably intervene during the development of semicircular canals as well.

Actually, there are 3 additional observations from the present study, which are of particular significance. The first is that 92.3% of the surgically confirmed SCCDs were protruding in the MCF versus only 30% for the nondehiscent cases. These had a particularly thin bone overlying the superior SCC. The second remarkable observation is that 28.6% of the nonprotruding canals in the group of nondehiscent superior canals were covered by supralabyrinthine mastoid air cells whereas the remaining 71.4% were at the same level of the tegmen and had a thick bony coverage. The third interesting finding is that posterior SCCD was present in 8.6% of the cases in our series, whereas no case of lateral SCCD was observed. We postulate that anatomic variations are probably involved in the etiopathogeny of SCCD during fetal development in the sense that protrusion of the superior and posterior canals into the middle and posterior cranial fossa, respectively, exposes them to temporal lobe pressure, thus preventing appropriate ossification or thickening of the bony labyrinth.

Other factors may contribute to the development of STD and SCCD. Twin sisters in our series had both STD and SCCD. This finding raises the number of reported cases of SCCD in the same family members to 6 cases in the English literature (3,14). It suggests that some patients may be genetically predisposed to tegmen defects and SCCDs. Mutation in the *COCH* gene (chromosome 14q12), which encodes cochlin, has been incriminated in a recently published article (15).

Some authors consider the congenital, developmental, and genetic factors as the ''first event'' leading to STD and SCCD, whereas the ''second event'' would be a sudden change in middle ear pressure (excessive straining, head trauma, etc.), arachnoid granulations, or pulsatile high intracranial pressure during adulthood (16). In fact, in the present series, all patients with STD alone who had protrusion of the superior SCC into the MCF had a very thin bony coverage, exposing them to the risk of canal dehiscence in case the second event happens in the future.

Increased intracranial pressure, especially in obese patients, may be an additional etiologic factor responsible for both STD and SCCD during adulthood (17). The pressure of CSF pulsating directly against the bone results in bony erosions, which, over time, can culminate in obvious bony defects (18). The etiology for increased CSF pressure in obese patients is thought to be caused by persistent increased intra-abdominal pressure. This results in decreased venous return leading to increased intracranial pressure (19). Unfortunately, neither body mass index nor intracranial pressure values were found in our database. A prospective study is being conducted in this regard.

No sex predominance was found in our study. Mean age was 54.4 years, which is very similar to the reported data in the literature. However, it is important to emphasize that an incidence of 11% of SCCD has been recently reported in children by Chen et al. (11). Thus, many questions remain unanswered regarding the asymptomatic pattern of this condition in the pediatric population.

Cerebrospinal fluid leak and recurrent serous otitis media in adults are 2 misleading symptoms of tegmen defects that must be thoroughly explored by otolaryngologists. In the present study, 65.2% of STD were manifested by serous otitis media and 17.4% were complicated by meningitis, which is indeed a high rate. It seems that the risk for cophosis is higher when SCCD is associated to CSF leak especially when the membranous labyrinth is disrupted, allowing direct spread of the infection into the inner ear. High-resolution computed tomography is the key diagnostic tool for both STD and SCCD.

CONCLUSION

We report the largest series of associated STD and SCCD in the literature. We postulate that such a coexistence is an indicator of a unique disease entity. Many factors are involved in the etiopathogeny of both conditions. Anatomic variations seem to play a fundamental role during the embryologic development of the inner ear. Other factors may intervene in the continuous pathologic process during adulthood. High clinical suspicion is required from the physician, who should always look for SCCD in case STD is present by means of HRCT with multiplanar reconstructions.

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