Superior semicircular canal dehiscence syndrome: A rare cause for dizziness

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ABSTRACT

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CASE REPORT

A 35-year-old female presented with a 18-month history of intermittent dizziness triggered by hearing loud sound and symptoms resolve as soon as sound stops. She saw her general practitioner 12 months ago and was diagnosed with benign paroxysmal vertigo. However, patient's symptom gradually worsened. She described that, in recent months, her dizziness symptom could be triggered just by putting a mug on a table or talking too loudly. She also described hearing her eyeball movements at night when the surroundings were quiet. Her worsening symptoms severely affected her quality of life. She was otherwise healthy and had no medical or surgical history.

The patient was referred to an ENT specialist for further investigations. A high resolution CT scan of the left temporal bone was performed and it showed 1 mm dehiscence of the superior semicircular canal antero-superiorly; consistent with superior semicircular canal dehiscence (Figure 1). Patient subsequently underwent an elective surgery to repair the superior semicircular canal dehiscence. The surgery was done by middle cranial fossa approach and the area of dehiscence was occluded with bone pâté and covered with fascia. Postoperatively, patient was admitted to intensive care unit for overnight observation and was transferred to ENT ward on day-1. Initially, her cranial examination revealed nystagmus on lateral gazes but it resolved on day-3. Patient continued

Figure 1: 1 mm dehiscence of the superior semicircular canal antero-superiorly.

to make a good recovery and was discharged on day-4 postoperatively. At fourth week follow-up, she had complete resolution of the symptoms and she was able to perform any activities of daily living without problems.

DISCUSSION

Superior semicircular canal dehiscence syndrome (SSCDS) is a rare condition which is characterized by sound or pressure induced vestibular symptoms. It is caused by dehiscence of the bone overlying the superior canal and the prevalence is estimated to be 0.5–0.6% [1]. It is postulated that the dehiscence creates a third mobile window into the inner ear. As a result, external sound and pressure can cause changes in the middle ear pressure, resulting in vertigo and oscillopsia [2]. The evoked eye movements in this syndrome align with the plane of the dehiscence superior canal. Patients may also experience hyperacusis to bone-conducted sounds.

The diagnosis is best established upon clinical findings and imaging, include the following [2]:

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- (i) Vertical-torsional eye movements evoked by sound or pressure stimuli noted on examination performed with Frenzel goggles.
- (ii) Lowered thresholds for responses to vestibular-evoked myogenic potentials
- (iii) CT scan of the temporal bones.

In terms of management, SSCDS can be treated by surgically occluding the dehiscence and it can effectively alleviate symptoms [3]. In our case, the patient had full resolution of her symptoms after receiving the surgery. On review of literature, only a small number of SSCDS cases have been reported. The majority of the patients experienced similar sound or pressure induced vestibular symptoms as observed in this case. Fortunately, all of the patients had good symptom control if not full resolution after surgical treatment.

CONCLUSION

Superior semicircular canal dehiscence syndrome (SSCDS) is characterised by vestibular symptoms induced by sound or pressure. Although uncommon, this condition can significantly affect quality of life. Awareness of this rare cause of dizziness is essential for timely diagnosis and intervention.

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Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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