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Vestibular Schwannoma: An occasional diagnosis during an occasional examination

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Abstract

Introduction: Vestibular schwannoma (VS) is a rare and benign tumor originating from Schwann cells of the vestibular component of the eighth cranial nerve, which tends to give only late clinical manifestations. In general its diagnosis is late or occasional.

Case: We present the case of a 56-year-old woman who complained for difficult nasal breathing. It seemed to be a simple case of congestion of nasal turbinates, but during clinical interview, a spontaneous nystagmus beating to the right was noted. Continuing medical history, the patient reported an annoying right hearing loss and a slight dizziness which lasted from about 1 month. So we performed an audiometric examination and a CT scan. Finally MRI with gadolinium showed an expansive lesion of the right cerebellopontine angle with intra-extrameatal development, of 28 mm, and a large cystic intralesional component, compatible with an eighth cranial nerve VS. The VS was macroscopically removed with surgery.

Discussion: We explain an occasionally diagnosed case of VS. Although gold standard for its diagnosis is MRI, history and physical examination should always be as complete and thorough as possible, even when we are dealing with patients so-called “non-patients”. We say how we perform a vestibular visit and we make a review of recent literature, with the intention of providing basic information on how to treat a dizzy patient.

Conclusion: We want to emphasize the concept of never forget the VS, among the many causes of vertigo, and the importance for its diagnosis covered by history and physical examination.

Keywords Vestibular schwannoma; Cranial nerve; Rare tumor; Incidental diagnosis

Introduction

Vestibular Schwannoma (VS) is a rare (6%-8% of all newly diagnosed brain tumors; about 20 cases per million population diagnosed per year) and benign tumor typically originating from Schwann cells of the vestibular component of eighth cranial nerve. It tends to give only late clinical manifestations, like hearing loss, tinnitus, headache, vertigo and dizziness, and often diagnosis is obtained on an occasional basis, during a diagnostic imaging examination of head possibly performed for other reasons.

When an asymmetry of sensorineural hearing loss is ascertained, especially in presence of vestibular associated symptoms, it is always important to conduct a thorough investigation by brain imaging, with an emphasis on cerebellopontine angle. Even if the resort to techniques of neuro-imaging in such situation is mandatory, the importance of a good and complete clinical history and physical examination must not be neglected.

Explaining an occasionally diagnosed case of VS, we say how we perform a typical vestibular visit and we make a review of recent literature, with the intention of providing basic information on how to treat a dizzy patient.

Case

We present the case of a 56-year-old woman who comes to our attention, sent by the family physician because of a difficult nasal breathing: it seemed to be a typical case of “common symptoms with simple treatment”. At examination, nasal cavity, nasopharynx and oropharynx were evaluated, finding hypertrophy of inferior turbinates, a moderate serous nasal discharge and a picture of mild chronic pharyngitis.
During visit, the patient also reported an annoying right hearing loss, which lasted from about 1 month; otoscopy was normal, but at a more accurate evaluation, a nystagmus was shown. Using Frenzel’s glasses, we saw a spontaneous horizontal and second degree nystagmus beating to the right, which did not change with positioning maneuvers (sitting position; supine; left side; right side; Rose’s position, with hyperextended head) and which was not inhibited by fixation. Halmagyi Test (Head Impulsive Test, HIT) was difficult to interpret; Head Shaking Test (HST) did not change the spontaneous nystagmus. Romberg Test was indifferent, showing a good functional compensation of the patient. Facial nerves were working perfectly.

Investigating her history, patient reported a slight dizziness from about one month, without real vertiginous episodes. Vertigo is understood as a sensation of rotation of the person in the surrounding environment, or movement of the environment itself; it is present from 18% to 58% of patients with VS (2); this symptom is not as frequent as expected, as the VS grows slowly, and nervous system has time to adjust to the new situation, compensating for as long as possible. The patient denied tinnitus, that is conversely described in 53% to 70% of patients with VS (2). She was in good health, with exception of a mild hypercholesterolemia, treated with diet alone. She denied similar episodes in the past, recent head trauma or a history of migraine. She is not a smoker, as it was ascertained, remembering protective effect of cigarette smoking about the risk of VS, reported in literature (3,4,5).

Audiometric examination (Figure 1) showed a slight sensorineural hearing loss on the mid frequencies and a severe hearing loss on the acute frequencies at the right ear, with left normoacusia (compatible with patient’s age); left pure tone average (PTA), considering 500 Hz, 1000 Hz, 2000 Hz and 4000 Hz, was 32.5 dB; tympanograms were bilaterally normal and stapedial reflexes were absent. Wrongly, speech discrimination (SD) was not performed.

On suspicion of a central nervous system affection, a HRCT scan with enhancement was performed. A 33x23 mm mass, located at right cerebellopontine angle, with internal cystic-necrotic component was found; a suspected contralateral similar lesion of 17 mm was also described (Figure 2).

Consequently, a Magnetic Resonance Imaging with gadolinium was indicated: this exam showed an expansive lesion of right cerebellopontine angle, with intraextrameatal development (28 mm in largest extrameatal diameter), and a large cystic intrallesional component, compatible with an eighth cranial nerve neuroma; the tumor determined a moderate compression on right middle cerebellar peduncle, with minimal surrounding edema; all was normal on the left side (Figure 3-4-5).
Most of VS are solid; cystic VS compose 4%-20% of all VS and are commonly larger at the time of presentation (6).

After the imaging, patient was sent to a Neurosurgical evaluation, preparing the idea of surgery, due to the size of VS (28 mm), its cystic nature, that did not recommend gamma knife radiosurgery (1,7), despite the lack consensus in the literature (8), and considering her relatively young age (56 years) and her incoming symptoms (9-10). She was informed on the three principal possible routes of access to the surgical removal of VS (translabyrinthine, middle fossa and retrosigmoid approach) (11,12,13). She was told that retrosigmoid approach would have been the preferred, with more likely preservation of facial nerve’s function, and with minimal risk of persistence of residual disease (6,14,15). She agreed and she did not want to delay the possible date of intervention.

VS was macroscopically removed with surgery by retrosigmoid approach, ten days after diagnosis, with preservation of seventh cranial nerve (third grade of House-Brackmann, one week after surgery), but with ipsilateral hearing loss. After 3 months we saw only a slight motor deficit (first grade) of the lower branch of the facial nerve.

Discussion

VS is a rare benign tumor of eight cranial nerve. Generally its diagnosis is late or occasional. We were
surprised by this case, because the patient came to our attention complaining of very different symptoms: only a careful clinical history and a complete physical examination raised the suspicion of a serious disease, that induced to further investigate on the patient by imaging. When we found the spontaneous nystagmus associated with relatively poor symptoms, we initially thought an episode of vestibular neuritis. However, due the associated hearing loss, this hypothesis was discarded. So we thought of an attack of endolymphatic hydrops, but it is unlikely that the instability can last so long (1 month), with no associated tinnitus and vertigo. A thorough investigation with imaging techniques was then planned: CT first, and then MRI, that can not be carried out in emergency in our Country. These imaging methods are essential for the diagnosis, but we want to stress the concept that, among the many causes of vertigo, we must never forget the VS. We must always keep in mind it while preparing anamnesis and physical examination, even when dealing with so-called “non-patients”, because these are the prerequisite for the diagnosis of VS.

Conclusion

- Vestibular schwannoma (VS) is a rare and benign tumor, originating from Schwann cells of vestibular component of eighth cranial nerve
- Gold standard for diagnosis of VS is MRI
- Whereas diagnosis of VS is often occasional, you must always think about it, when examining a dizzy patient, especially in the case of asymmetric sensorineural hearing loss.
- Whereas VS tends to give only late clinical manifestations, you can visit a patient with VS, which consults you for a totally different problem.
- Don’t forget the importance of a good and complete clinical history and physical examination.

References

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