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# Distinct spontaneous shrinkage of a sporadic vestibular schwannoma

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ABSTRACT

We present a case with outspoken spontaneous vestibular schwannoma shrinkage and review the related literature. The patient was initially diagnosed with a left-sided, intrameatal vestibular schwannoma, which subsequently grew into the cerebello-pontine angle (CPA), followed by total shrinkage of the CPA component without any intervention over a 12-year observation period. The literature on spontaneous tumor shrinkage was retrieved by searching the subject terms "vestibular schwannoma, conservative management" in PubMed/MEDLINE database, without a time limit. Of the published data, the articles on "shrinkage" or "negative growth" or "regression" or "involution" of the tumor were selected, and the contents on the rate, extent and mechanism of spontaneous tumor shrinkage were extracted and reviewed. The reported rate of spontaneous shrinkage of vestibular schwannoma is 5–10% of patients managed conservatively. Extreme shrinkage of the tumor may occur spontaneously.



## 1. Introduction

Vestibular schwannomas (VS) are intracranial, benign nerve sheath tumors, which originate in the transition zone between central and peripheral myelin on the vestibular branches of cranial nerve VIII [1]. Most frequently, VSs are sporadic, unilateral tumors, whereas bilateral lesions are pathognomonic for NF2 [2]. From epidemiological studies we know that only about 30% of the tumors grow after diagnosis, in most cases within the first five years after diagnosis [3].

During the past 30 years, the number of diagnosed VS in Denmark has increased from 15 in 1976 to 110 in 2009 (5.5 million inhabitants). During the same period, the size of the tumors has decreased from a mean extrameatal diameter of 30 mm to 10 mm and the mean age at the time of diagnosis has increased from 50 years in 1976 to 60 years in 2009 [3]. Interestingly, the smallest tumors are found in the oldest age group. This may indicate: (1) that small VSs develop as a degenerative phenomenon in old age, (2) that small tumors with mild symptoms are found incidentally in old age, when symptoms such as hearing impairment and dizziness are not uncommon, or (3) it may reflect that some tumors shrink spontaneously after initial growth.

The aim of this paper is to focus on spontaneous tumor shrinkage. Thus, we present a patient initially diagnosed with a left-sided, intrameatal vestibular schwannoma, which subsequently grew into the cerebellopontine angle (CPA), followed by total shrinkage of the CPA component without any intervention over a 12-year observation period. Finally, we review the relevant literature of shrinking VS.

## 2. Case report

A 67-year-old woman was referred to our tertiary referral center in June 1998 by a private ENT doctor, because of sensorineural hearing impairment and tinnitus in her left ear. She had no dizziness/unsteadiness at any time and the tinnitus later disappeared. The patient had no history of previous ear problems or any systemic disease. Otomicroscopy showed a normal ear drum. Audiometry revealed the pure-tone average (PTA) on the right ear to be 33.8 dB, and on the left ear 42.5 dB. The discrimination was normal on the right ear, whereas the left ear had a 4% loss. An auditory brainstem response (ABR) was abnormal on the left side and a following CT showed a normal result. In April 1999, the hearing of the left ear deteriorated further, to a PTA of 32.5/67.5 dB, discrimination loss 0/100%. A subsequent gadolinium-contrasted MR imaging in June 1999 demonstrated an elongated, uniformly enhanced tumor with a largest diameter of 10 mm within the left internal auditory canal (IAC), resembling a purely intrameatal vestibular schwannoma. The diagnosis of a vestibular schwannoma was made from the very characteristic

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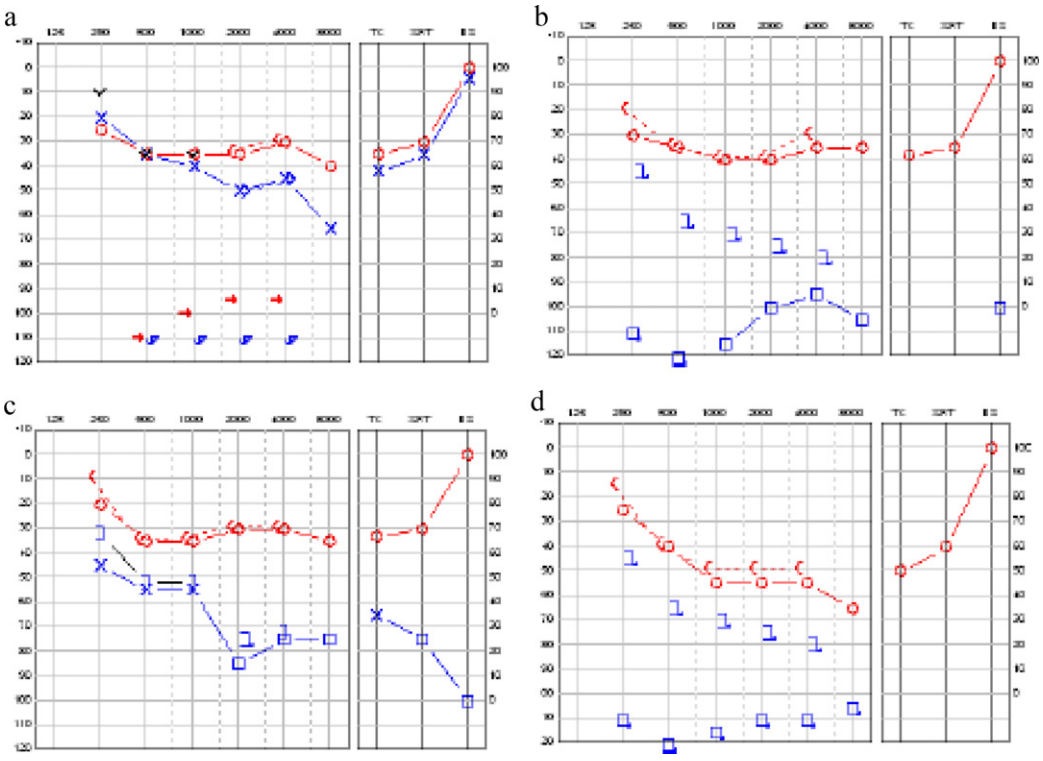


**Fig. 1.** (a) The initial audiometry in 1998 showed slight pure tone asymmetry, with left-sided hearing loss at the high frequencies and 4% discrimination loss (DS). (b) The hearing loss progressed rapidly and in 1999, all discrimination was lost. (c) In 2001, the patient had become practically deaf, also for pure tones, on the left (tumor) side. For that reason, the hearing on the left side was not recorded subsequently. (d) The 2009 audiometry show that the hearing on the right (non-tumor) side has also worsened over the years, due to presbyacusis (patient age 78 years).

shape, the tumor density in T1 and T2W images, and the localization of the tumor, which makes another diagnosis highly unlikely. MRI equipment used was a 1.5 T Philips Achiva, T2W images using 0.8 mm slicing in FIESTA-C. Because of a small tumor size and no serviceable hearing on the tumor ear, the patient was allocated to "wait and scan" management. At the next MRI in July 2000 the tumor had grown into the CPA, with a largest extrameatal diameter of 7 mm. The hearing on her left ear presented no change. Between 2000 and 2001, however, the patient became completely deaf on the left side (Fig. 1). The patient was reluctant to surgical intervention, so "wait and scan" conservative management continued. Followingly, the size of the tumor peaked on the MRI of 2002 and 2004, with a largest extrameatal diameter of 13–14 mm (Fig. 2a and b), without progressing or additional symptoms. During the following three years, the tumor displayed slow, spontaneous shrinkage (Fig. 2c and d). Then the tumor shrinkage accelerated with a rate of mean 3 mm per year from 2007. The MRI in February 2011 showed that the extrameatal part of the tumor had disappeared completely (Fig. 2e and f). The MRI results are resumed in Table 1.

## 3. Discussion

There has been a more than 6-times increase in the number of diagnosed VS over past four decades in Denmark [4]. In the very nature of things, this increase in the incidence has been argued to be the result of improved diagnostic techniques and enhanced awareness of the disease among patients, physicians and otologists. The progress in image diagnostic techniques, especially gadolinium-contrasted MRI, has led to an increase in the number of VSs diagnosed, especially small tumors with mild symptoms. MRI is now widespread available, such that the elder patients and individuals with minimal symptoms more readily are referred to determine whether a VS is exist or not.

Conservative clinical observation is one of current treatment options, which also include microsurgical resection and radiation. As more and more small VSs are diagnosed, the conservative management strategy, or "wait and scan" policy, for these patients is becoming increasingly relevant. During conservative observation, the various growth patterns of VS have shown, that only a minor part of the tumors grow following diagnosis [3]. Although considerably less common, the tumor may even shrink.

Data from 11 studies on conservative management [5–16] have demonstrated, that 2.9–22% of patients display tumor regression, spanning follow-up periods from one to 12.5 years (see Table 2). Among these reports, only Luetje [7] and Battaglia et al. [14] described the extent of tumor regression, Luetje's study having the longest observation period, of up to 12.5 years. The patients of Hoistad et al. [10] presented the lowest tumor shrinkage rate of 2.9%, whereas Hajioff et al. [16] demonstrated 22% with tumor shrinkage in a series of 72 patients. Summarizing these reports on tumor regression, 83 (7.8%) of 1062 VS patients managed

**Table 1**
The MRI findings of the patient at regular intervals.

| Date | Tumor localization | Tumor largest diameter (mm) |
|---|---|---|
| June 1999 | Intrameatal | 10 |
| July 2000 | Extrameatal | 7 |
| July 2001 | Extrameatal | 10 |
| October 2002 | Extrameatal | 14 |
| March 2004 | Extrameatal | 13 |
| April 2006 | Extrameatal | 11 |
| December 2007 | Extrameatal | 12 |
| February 2008 | Extrameatal | 9 |
| February 2011 | Intrameatal | 10 |