

Case Report

Spontaneous shrinkage of vestibular schwannoma

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Abstract

Background: “Watch, wait, and rescans” (WWR) has an established place as a successful management option for a significant proportion of vestibular schwannomas (VS) as an alternative to microsurgical removal or stereotactic radiotherapy. VS may grow slowly and continuously, followed by stagnation or even shrinkage. We present two case reports of spontaneous shrinkage of VS along with a review of the literature.

Case Description: A 29-year-old female presented with a progressive history of visual blurring and intermittent diplopia over 2 months. A 29 mm of maximum intracranial diameter (ICD) VS with secondary obstructive hydrocephalus was diagnosed. The patient underwent a ventriculo-peritoneal shunt with resolution of her symptoms and opted for initial WWR management. Interval scanning between 2007 and 2014 showed progressive reduction in the maximum ICD together with reduction in the degree of central tumor enhancement. Maximum ICD at most recent follow up was 22 mm. A 28-year-old female was referred with right sensorineural deafness. A right VS of maximum ICD of 27 mm was diagnosed. Initial WWR management was planned after discussion. Serial imaging showed an initial increase in the size of the tumor followed by progressive reduction in size. The most recent follow up showed a maximum ICD of 20 mm.

Conclusion: Early WWR management can be associated with spontaneous shrinkage of VS over time. Prospective clinical study of larger numbers of such cases using the UK VS database may help to identify predictive factors for the spontaneous regression of VS.

Key Words: Conservative treatment, neurinoma, spontaneous shrinkage, vestibular schwannoma, watch, wait, and rescans

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Quick Response Code:**INTRODUCTION**

Vestibular schwannomas (VS) are benign, slow-growing tumors, which originate in the transition zone between the central and peripheral myelin of the vestibular branches of cranial nerve VIII.^[1,11] The incidence is 20 per million/year.^[23] Improvements in neuro-radiological techniques with gadolinium-enhanced magnetic resonance imaging (MRI) have led to an increase in the number of small VS detected with mild symptoms and without brainstem compression. Stangerup in 2006

defined VS as unpredictable tumors with respect to their patterns of growth. These tumors may grow slowly and

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continuously, followed by stagnation or even shrinkage. When growth is progressive or leads to compression of the brainstem or occlusion of the fourth ventricle, surgery may be indicated.^[23]

Non-surgical management with “watch, wait, and rescan” (WWR) policy is a frequent option, especially in asymptomatic patients,^[11] because a stable tumor size has been reported in almost 50% of cases with an average follow-up time of 3 years.^[6,8] We present two case reports of spontaneous shrinkage of VS out of 223 neurinoma referred to our Institution between December 2002 and May 2015. A review of the literature regarding spontaneous shrinkage of VS is also presented.

CASE DESCRIPTION

Case report 1

A 29-year-old female patient presented with a progressive history of visual blurring and double vision over 2 months in 2006. She was assessed by an optician who noted bilateral papilloedema. Clinical assessment performed in our hospital confirmed the presence of bilateral Grade 2 papilloedema. She had preserved speech discrimination on the right and double vision on the extreme left lateral downgaze. Audiometry in February 2007 showed slight loss of hearing on the right with no apparent high tone preponderance. MRI in 2007 disclosed a lesion in the right cerebellopontine angle (CPA) of maximum intracranial diameter (ICD) of 29 mm in continuity with the acoustic nerve, with associated secondary hydrocephalus [Figure 1a]. The patient had placement of a left ventriculo-peritoneal shunt.

Following surgery, the papilloedema and the diplopia resolved. Computed tomography (CT) scan showed

immediate improvement in the hydrocephalus. Following the complete resolution of her symptoms, the patient was managed with WWR. The tumor remained stable in ICD for 2 years with a maximum ICD of 29 mm [Figure 1b]. MRI at 4 years revealed a slight reduction in the lesion with an ICD of 26 mm [Figure 1c]. Further MRI performed at years 6 and 7 showed progressive reduction in the degree of central tumor enhancement and a reduction in the maximum ICD of, respectively, 24 and 22 mm [Figure 1d and e]. During clinical follow-up, the patient was in good neurological condition and serial audiometry showed stable hearing function. At year 10, she developed symptoms related to shunt dysfunction with new headache and evidence of ventricular enlargement. A revision of the shunt was required.

Case report 2

A 28-year-old female was referred to us by an ENT surgeon for right-sided hearing loss with preservation of speech discrimination. There was no impairment of coordination. A CT scan demonstrated the presence of a right VS with an extrameatal component in the CPA of maximum ICD of 25 mm [Figure 2a]. MRI at 1 year showed a 1 mm increase in the size of the lesion. A further MRI at 3 years showed the tumor maximum ICD of 25 mm [Figure 2b], and in 2013 the tumor maximum ICD was 24 mm [Figure 2c]. The most recent MRI investigation at 5 years showed a further reduction in the maximum ICD to 20 mm [Figure 2d].

DISCUSSION

Watch, wait, and rescan has an established place as a successful treatment option for a significant proportion of small VSs, as an alternative to microsurgical removal

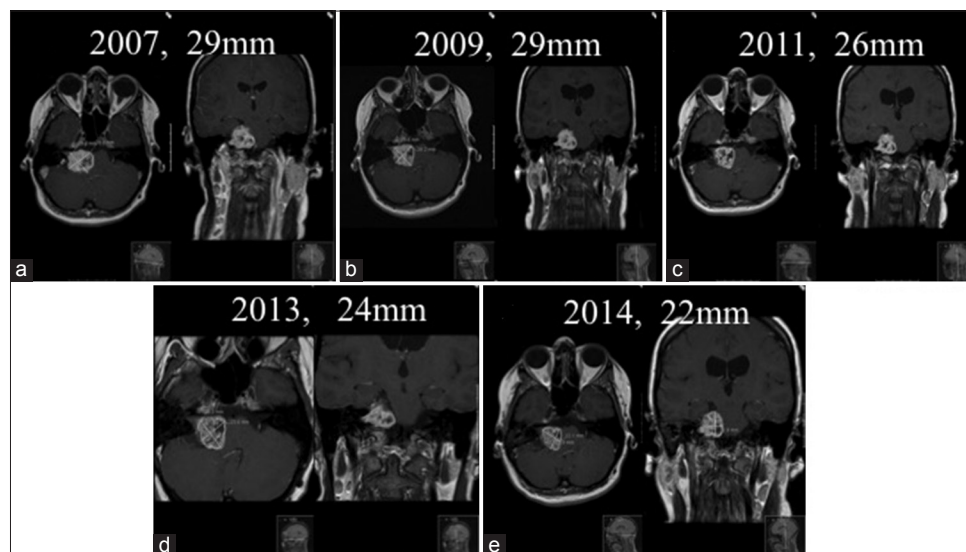


Figure 1: Right medium-sized (29 mm) vestibular schwannoma in 2007 (a) remaining stable in size in 2009 (b) and then showing a progressive reduction in the maximum intracranial diameter with conspicuous reduction in the degree of central tumor enhancement (c and d). Maximum intracranial diameter of 22 mm at most recent follow-up (e)

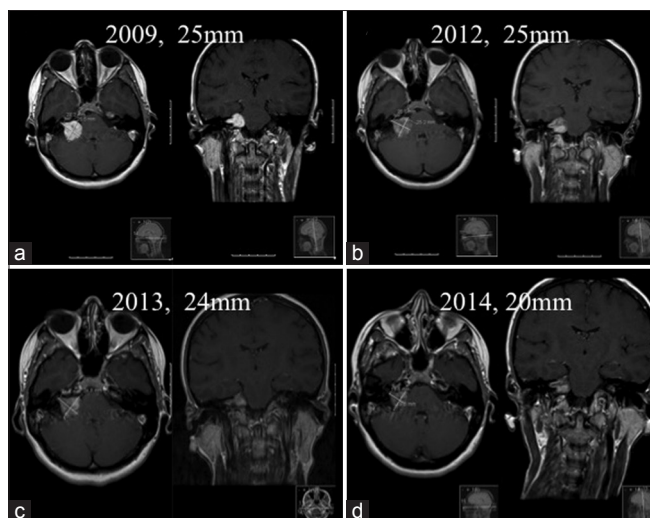


Figure 2: Right medium-sized (25 mm) vestibular schwannoma in 2009 (a) remaining stable in size in 2012 (b) and then showing a progressive reduction in size in 2013 (c) to a maximum intracranial diameter of 20 mm (d) at recent follow-up

or stereotactic radiotherapy.^[3,4,19,23] Treatment should be reserved for cases with tumor growth, in response to patient preference, or because of progressive symptoms.^[18] In published series of non-surgically managed VS, a small number of cases are known to demonstrate signs of regression.^[1] Table 1 presents a review of the previous literature describing spontaneous shrinkage in VS including the current study.

In 21 previous studies, between 1988 and 2013, the incidence of shrinkage of these tumors ranged between 1 and 29% during follow-up of 6 months to 27 years. The degree of tumor shrinkage identified ranged between 5.38 and 100% during the same follow-up period.^[1-4,7,9-12,14-16,18,19,21,22,24-27]

Tumor size can be classified as follows: Intracanalicular, small (<10 mm)-, medium (range, 11–25 mm)-, large (range, 25–40 mm)-, giant (>40 mm)-sized neurinoma.^[2]

In the present report, we assessed tumor size by measurement of the largest CPA tumor diameter, as reported by Rosenberg in 2000.^[20] A decrease of 2 mm or more in the largest extrameatal tumor diameter was defined as shrinkage.^[23]

Intracanalicular tumors tend to exhibit a slow growth rate, as reported by several authors.^[5,7,18,19,23] Complete regression of a VS in its extrameatal component from a 14.1 mm tumor was described by Huang *et al.*^[11] in 2013 during a follow-up period of 12 years. Yasumoto and Ito^[27] described a 75-year-old woman with a right small VS protruding into the CPA. This tumor was conservatively managed because of the advanced age of the patient. Initially, the tumor grew from 5.2 to 16.7 mm over 7 years with worsening of her tinnitus, dizziness, and headache. After the initial increase in size, the tumor

spontaneously shrank to 8.2 mm with improvement of the symptoms.^[27] The largest series of sporadic VS demonstrating shrinkage has been reported by Huang *et al.* in 2013^[12] who noted 48 cases in a series of 1261 patients. The mean age at diagnosis was 56.7 years and mean follow up was 9.5 years (range 1–27 years). Fourteen patients presented with initial growth of the tumor followed by shrinkage; 26 showed stable size followed by shrinkage, and eight showed evidence of shrinkage at first reimaging. The mean pre-shrinkage duration was 3.9 years. Our first patient had an initial period of stability in tumor size with shrinkage after four years, and our second patient showed initial growth for 1 year followed by shrinkage over the 3 subsequent years of follow up.

In patients affected by neurofibromatosis type 2 (NF2) with bilateral VSs, the gene responsible for NF2 is known to be localized on chromosome 22;^[17] however there is no known genetic correlation with spontaneous shrinkage. von Eckardstein^[26] described two patients with NF2 who underwent unilateral resection of VS and regression of the contralateral medium-sized VS during a follow-up period of 4 and 9 years. There are no large series describing shrinkage of VS managed with WWR in patients with NF2. A multicentre study of 56 NF2 patients with 84 VSs reported 16 tumors out of 84 with regression with a range of shrinkage between 1 and 7 mm during a follow-up of 51.3 months.^[22]

The mechanism of tumor shrinkage is not well understood. Environmental factors and/or genetic-molecular hypothesis should be considered. A reduction in vascular supply is hypothesized, which could lead to ischemic necrosis and fibrosis with associated shrinkage.^[7,13] However the absence of any age correlation with the incidence of shrinkage provides evidence to contradict this possible mechanism. Based on the existing literature, age, sex and tumor size have not been demonstrated as predictive factors for tumor shrinkage.^[1-4,7,9-12,14-16,18,19,21,22,24-27] In the present report, both patients presented at a young age and with no history of vascular disease.

CONCLUSIONS

We would endorse the value of a WWR policy as a valid current treatment option for VS. This should be always considered in patients with small- or medium-sized tumors, with minimum or absent symptoms. We have shown that significant spontaneous tumor shrinkage can occur even in medium-sized VS, and we recommend that patients should be followed up for a minimum of 4 years to seek evidence of this. Furthermore, initial growth of the tumor may not always mandate active treatment even in young patients or in patients with a significant CPA component.

Table 1: Spontaneous shrinkage of vestibular schwannoma: Review of the literature

Author, year	Number of patients with shrinkage/total patients studied (%)	Sex and age of patients	Size of tumor (mm)	mm or % of shrinkage	FU years (range)
Luetjje <i>et al.</i> , 1988 ^[11]	1/115	71, M	13	5, 38%	5
Bederson <i>et al.</i> , 1991 ^[3]	4/70 (8%)	N/A	N/A	5.7%	2.7
Tschudi <i>et al.</i> , 2000 ^[19]	8/74 (11%)	N/A	N/A	16.0%	3
Luetjje, 2000 ^[10]	6/47 (13%)	71, M	14	9, 64%	12.5
		67, F	11.6	5.9, 51%	9
		74, M	10	5, 50%	4.3
		59, M	46	15, 33%	12
		67, F	15	3.4, 23%	8
		63, F	15.9	5.9, 37%	5
Shin <i>et al.</i> , 2000 ^[16]	7/60 (12%)	63 year (mean age)	N/A	N/A	2.6
Hoistad <i>et al.</i> , 2001 ^[7]	3/102 (3%)	64, mean	N/A	N/A	2.4 (6 months-10 years)
Stipkovits <i>et al.</i> , 2001 ^[18]	3/44 (7%)	39, M	18	3, 16.7%	9
		67, F	19	3, 15.7%	10
		69, F	20	5, 20%	4
Mohyuddin <i>et al.</i> , 2003 ^[13]	10/50 (20%)	N/A	N/A	N/A	1.65
Slatteri WH III <i>et al.</i> , 2004 ^[17]	16/56 NF2 (29%)	N/A	N/A	1-7 mm	4.3
Bozorg Grayeli <i>et al.</i> , 2005 ^[4]	7/111 (6%)	N/A	N/A	N/A	2.75
Al Sanosi, 2006 ^[1]	6/205 (3%)	N/A	N/A	N/A	3.2 (2.2-5)
Battaglia <i>et al.</i> , 2006 ^[2]	6/111 (5%)	N/A	4: <10 mm 2: 11-25 mm	0.74 mm/year	3.1
Yasumoto and Ito, 2006 ^[21]	1	75, F	16.7	8.5, 51%	4
De Oliveira Penido <i>et al.</i> , 2007 ^[14]	2	48, M	10 (intracanalicular)	N/A	4
		60, F	7 (intracanalicular)	1, 14%	2
Mick <i>et al.</i> , 2008 ^[12]	2	60.1 years	N/A	N/A	2.1
Ferri <i>et al.</i> , 2008 ^[5]	6/123 (5%)	N/A	N/A	N/A	4, 8
Hajioff <i>et al.</i> , 2008 ^[6]	16/72 (22%)	61, median	11 CPA tumors 4 intracanalicular	N/A N/A	10
von Eckardstein <i>et al.</i> , 2010 ^[20]	2 (NF2)	49, F	30.1 mm	11.5, 38%	4
		61, F	27 mm	11, 41%	8
Pennings <i>et al.</i> , 2011 ^[15]	4/47 (9%)	N/A	Intracanalicular	N/A	5.1
Huang <i>et al.</i> , 2013 ^[9]	48/1261 (4%)	25 pts M 23 pts F 56.7 years (mean age)	12.04 mm (mean)	6.25 (mean) (52%)	9.5 (1-27)
Huang <i>et al.</i> , 2013 ^[8]	1	67, F	14 mm	14, 100% (extrameatal component)	12
Present report	2/223 (1%)	29, F 28, F	29 mm 25 mm	7, 24% 5, 20%	7 5

Prospective multicentre studies with a large number of patients such as the UK vestibular schwannoma database will be a valuable future aid in identifying predictive factors for tumor shrinkage. In young patients with an anticipated long-term follow-up, there may be merit in considering biopsy in selected cases for a better understanding of the mechanism of shrinkage.

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Conflicts of interest

There are no conflicts of interest.

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