

Spontaneous Regression of a Growing Vestibular Schwannoma

—Case Report—

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Abstract

A 75-year-old woman presented with a small right vestibular schwannoma which enlarged and then spontaneously regressed during conservative management. The tumor increased in size from 5.2 to 16.7 mm over 7 years with worsening symptoms of tinnitus, dizziness, and headache. The tumor then regressed to 8.2 mm and the symptoms improved for 4 years without further treatment. Changes in tumor growth should be carefully monitored during conservative management of patients with vestibular schwannoma.

Key words: vestibular schwannoma, magnetic resonance imaging, spontaneous regression, conservative management

Introduction

The natural history of vestibular schwannomas has been retrospectively studied in patients managed conservatively.^{4,6-8,12-18} These benign tumors grow slowly at less than 2 mm/year in most patients.^{4,8,12,14-18} Significant tumor growth occurred in 38.9–82% of the patients, no or insignificant growth in 12–47%, and negative growth in 2.8–19.4%.^{4,6,7,10,14-18} An extensive review of 26 studies including 1340 patients found that the overall frequency of growth was 46% and that of regression was 8%.¹⁸ Investigation of tumor growth patterns during the observation period indicated variable growth in vestibular schwannomas: continuous growth in 38%, no measurable growth 12%, no measurable growth and then continuous growth in 24%, negative growth in 6%, and various positive tumor growth in 20%.⁷ However, spontaneous regression is rare.^{4,6-8,10,14-18}

We report a case of spontaneous regression of a growing vestibular schwannoma during an 11-year follow-up period.

Case Report

A 75-year-old woman presented with worsening tinnitus on the right and dizziness on walking. She had a 35-year-old history of right hearing loss and tinnitus.

Neurootological examinations found no nystagmus both without and with Frenzel eyeglasses. However, pure tone audiometry found severe right sensorineural hearing loss and the eye tracking test showed saccadic pursuit. The caloric test showed good bilateral responses. Magnetic resonance (MR) imaging showed a small enhanced tumor occupying the right internal auditory canal (IAC) and slightly protruding into the cerebellopontine angle (CPA) cistern (Fig. 1A). The neuroimaging diagnosis was small vestibular schwannoma.

The tumor was conservatively managed because of her advanced age, mild symptoms, and small tumor size. The patient was followed up for 11 years at 6-month or 1-year intervals using serial axial T₁-weighted MR imaging with gadolinium to assess tumor growth. The tumor size was determined using the 1995 guidelines of the American Academy of Otolaryngology-Head and Neck Surgery.¹ The

Received March 22, 2006; Accepted July 27, 2006

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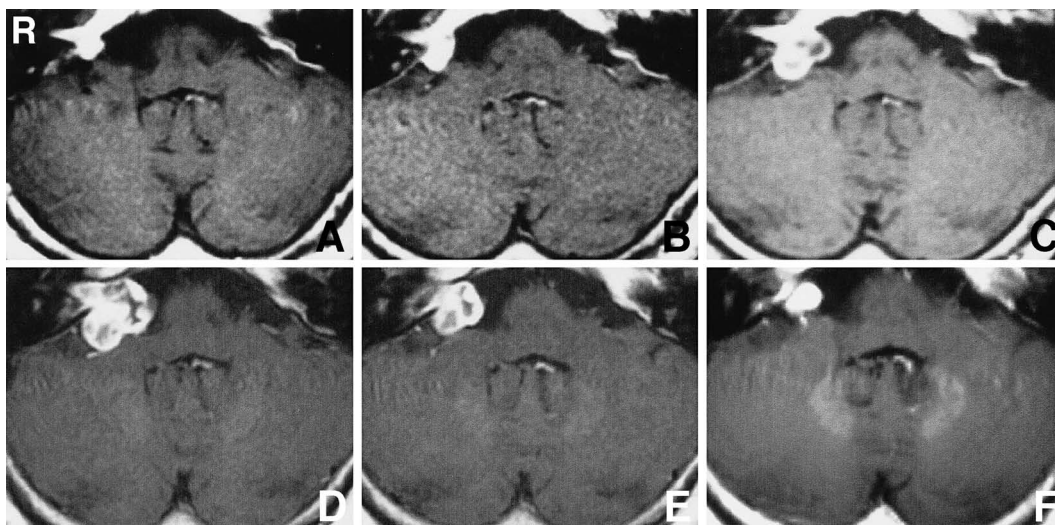


Fig. 1 Axial T₁-weighted magnetic resonance images with gadolinium showing the increase and decrease in size of the vestibular schwannoma during an 11-year follow-up period. The small enhanced tumor occupied the right internal auditory canal and slightly protruded into the cerebellopontine angle cistern. The tumor size was 5.2 mm at diagnosis (A), 8.4 mm at 1 year (B), 12.1 mm at 2 years (C), 16.7 mm at 7 years (D), 11.7 mm at 9 years (E), and 8.2 mm at 11 years (F).

extracanalicular component of the CPA tumors was determined as follows: the axial image with the largest extracanalicular tumor diameter was selected and the maximum antero-posterior and medial-lateral tumor diameters were calculated with a micrometer. The antero-posterior diameter was measured parallel and the medial-lateral diameter was measured perpendicular to the posterior surface of the petrous bone. The size of the tumor was calculated as the square root of the product of these two diameters. The size of the tumor limited to the IAC was not calculated because of the varying MR imaging presentation.

The patient frequently complained of headache, right-sided tinnitus, and dizziness during the observation period. However, the tinnitus disappeared, and the headache and dizziness improved over the 8 years after diagnosis. Follow-up MR imaging showed increased tumor size in the right CPA cistern (Fig. 1B–D). Tumor growth rate was 3.2 mm/year for the first 2 years and then 0.8 mm/year for the next 5 years, reaching 16.7 mm at 7 years from the initial size of 5.2 mm (Fig. 2). During tumor growth, a part of the solid portion became cystic. The tumor size thereafter decreased to 8.2 mm at 11 years after diagnosis without treatment (Fig. 1E, F). The tumor regression rate was 2.0 mm/year (Fig. 2). The cystic portion disappeared during regression.

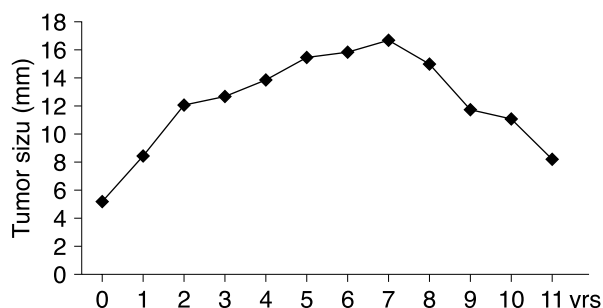


Fig. 2 Tumor size versus time. The tumor grew from an initial size of 5.2 mm up to 16.7 mm at 7 years. Tumor growth rate was 3.2 mm/year for the first 2 years and then 0.8 mm/year for the next 5 years. The tumor decreased in size to 8.2 mm without treatment. Tumor regression rate was 2.0 mm/year.

Discussion

The present case of spontaneous regression of a growing vestibular schwannoma is rare among the tumor growth patterns. One among 70 conservatively-treated patients had slight tumor growth during the 1st year followed by continuous tumor regression during the next 5 years.⁴⁾ A 64-year-old female with an intracanalicular tumor initially showed increase in tumor size but computed tomo-

graphic scan indicated shrinkage of tumor during a 18-year follow-up period.²⁾ Except for these patients, no similar case was described in the literature on spontaneous regression.^{6,7,10-12,14-18)}

The clinical features in patients managed conservatively were investigated to identify the tumor growth-promoting factors. Although no predisposing factor to predict tumor growth was identified,^{12,14,16)} patient age was correlated with tumor growth.¹³⁾ An increase in growth rate during the 1st year of observation may be predictive of growth in the following year.^{4,8)} Cystic formation, as observed during tumor enlargement in our patient, may also contribute to significant tumor growth.⁵⁾ The mechanisms of cyst formation in vestibular schwannomas are unknown. However, histological study of three cases with vestibular schwannoma mainly consisting of cysts suggested that repeated small hemorrhages from abundant abnormal sinusoid or telangiectasis-like vessels were the most probable cause of cyst formation in vestibular schwannoma.³⁾

The mechanisms of spontaneous regression of vestibular schwannoma are speculative. Ischemic necrosis secondary to intratumoral thrombosis and subsequent necrosis following fibrosis may be related to tumor regression.¹⁰⁾ Spontaneous regression of human neoplasms triggered by immunologically-mediated apoptosis or programmed cell death may also occur.⁹⁾ Prediction of spontaneous regression of vestibular schwannomas is unreliable based on age and tumor size at diagnosis.¹⁰⁾ Our patient showed disappearance of the cystic portion during regression. This phenomenon has not been described in cases of spontaneous regression.^{2,4,6,10-12)} Prospective studies to identify the prognostic factors underlying spontaneous regression are needed for better conservative management of vestibular schwannoma.

Surgery was necessary in 13% to 32% of patients managed conservatively due to neurological deterioration and rapid tumor growth.^{2,4,6,8,14,16)} Tumor growth in patients with failed conservative management was 3.0–4.2 mm/year.^{8,14,16)} Our patient showed rapid tumor growth of 3.2 mm/year in the 1st and 2nd years of follow-up and she complained of worsening symptoms. Our patient could have been a candidate for surgery or radiotherapy if the tumor had not regressed spontaneously, as any delay leads to unfavorable outcome. No difference in postoperative neurological outcome has been reported between patients who initially underwent surgery and those who initially received conservative treatment but subsequently required surgery.^{4,14,16)} However, we should closely monitor patients with

rapidly growing vestibular schwannomas. In particular, hearing deterioration may occur during conservative management regardless of tumor growth.^{6,7,14,16)} Microsurgery should be considered to preserve the hearing.

The various growth patterns of vestibular schwannomas suggest that spontaneous regression is not so rare. Because even a rapidly growing tumor can spontaneously shrink as shown in this case, changes in tumor growth should be carefully monitored at regular intervals by MR imaging during conservative management of patients with vestibular schwannomas.

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