A large vestibular schwannoma that did not grow for 18 years

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Abstract. A large vestibular schwannoma that did not grow for 18 years. Introduction: Treatment strategies for vestibular schwannoma include microsurgery, stereotactic radiotherapy and conservative management (wait and scan). To avoid neurological complications or even death, surgery is the preferred treatment for large tumours with an extrameatal diameter > 3.0 cm.

Objective/methods: We present the case history of a man with a large vestibular schwannoma who had refused treatment and was seen again eighteen years later.

Results: This patient had not developed symptoms other than the initial hearing loss. Repeated imaging showed that the tumour had not grown and the brainstem compression had not progressed.

Conclusion: This case history illustrates the unpredictable growth pattern of vestibular schwannomas. Apparently, even large tumours in close proximity to the brainstem may remain stable for many years. However, there still are no valid arguments to refrain from therapy in patients with a large vestibular schwannoma, since reliable growth predictors are not available.

Introduction

Treatment strategies for vestibular schwannoma (VS) include microsurgery, stereotactic radiotherapy and conservative management (wait and scan). To prevent brainstem compression, microsurgery is usually the preferred treatment for large tumours. We discuss the history of a man with a large vestibular schwannoma who initially refused treatment and was seen again after eighteen years.

Case report

A 65-year-old male was seen in 1989 for hearing loss and tinnitus in his right ear. He reported some vertigo in the past but, at that point in time, he was suffering only from a mild loss of balance. Physical examination revealed a normal aspect of the eardrum on both sides.

The function of the cranial nerves V and VII was normal. Audiometry showed profound deafness on the right side and subnormal hearing in his left ear (Figure 1). There was a spontaneous rotatory nystagmus to the left side.

Electronystagmographic examinations revealed a complete right vestibular paresis.

Because of this audiovestibular asymmetry, it was decided to conduct a magnetic resonance imaging (MRI) scan. This scan revealed a large cerebellopontine angle mass on the right side. The maximal extrameatal diameter was 3.0 cm. The internal auditory canal was enlarged. The tumour was compressing the cerebellum and the brainstem, causing a minor shift of the brainstem to the left (Figure 2). Obviously, the presence of a large VS on the right side was appropriately documented.

When counselling the patient, we tried to convince him that his lesion was potentially life-threatening and strongly advised microsurgery. However, the patient persisted in refusing any type of treatment, particularly surgery. He expressed a preference for herbal and magnetic therapy and deliberately disappeared from follow-up. In 2007, we were able to get in contact with him again, and on that occasion he was willing to give us some relevant information. By then he was 83 years old and said he was in excellent condition. While the deafness in his right ear had persisted, he had got used to his handicap, as his hearing on the left was subjectively normal.

The patient refused our new offer of clinical examination and audiology but did agree to undergo MR imaging. Figure 3 shows the results. Apparently the tumour is still there. The size of the VS...
has not changed, although part of it has developed a cystic consistency.

Recently (in December 2010) we contacted him again. He said he was in good health, without any complaints related to his VS, increasing his “survival without treatment” to almost 22 years.

Discussion

This patient had survived at least 22 years with a large VS. His symptoms did not change during that period, nor did the tumour change in size during the first 18 years after the diagnosis. This case history illustrates the unpredictable growth pattern of vestibular schwannomas. It also puts into perspective our effort to convince the patient to have surgery eighteen years ago.

Conservative management has gained popularity since the introduction of MR imaging. Many authors have published their “wait and scan” results. On average, 50 percent of the vestibular schwannomas are found to be growing after three years of follow-up. Multiple growth patterns have been identified, but so far it is impossible to predict future tumour growth. In this patient, growth had stopped at an advanced stage.

In the decision-making process about the preferred treatment modality for VS, only small lesions used to be eligible for conservative management. The general consensus is that large tumours should be treated. Moreover, it is widely accepted that these large lesions should be treated with microsurgery. The localisation of this lesion in the posterior fossa close to the brainstem implies that progression is life-threatening. Mortality can ensue from the compression of the fourth ventricle and/or the brainstem, which results in hydrocephalus and, at a later stage, brainstem herniation. This possibility is mentioned in a few studies of the “wait and scan” approach. In these studies, mortality rates range from zero to six percent. However, given the selection bias and the limited follow-up period, these data are hardly applicable to the natural history and mortality of VS.

The literature on the treatment of VS is vast and includes extensive reports about the probabilities of success and failure. Presumably, all large tumours will be treated once the diagnosis has been made;
refraining from offering therapy would be considered unethical. Only rarely do large vestibular schwannomas go undiagnosed until death by tumour progression is inevitable.

The patient described above did not develop progressive symptomatology. Indeed, his subjectively perceived condition is now at least as good as it would have been if he had been operated on eighteen years ago. However, he has been under constant threat of progressive neurological deficits, and the risk of a deteriorating course has remained over the years. In view of these considerations, while the advice we gave to the patient eighteen years ago is still justifiable, it is hard to explain to him with hindsight. This case history demonstrates that even large tumours in close proximity to the brainstem may remain stable for many years. It also suggests that counselling patients with large vestibular schwannomas may sometimes be a problematic and unrewarding task.

Conclusion

This case history illustrates the unpredictable growth pattern of vestibular schwannomas. Apparently, even large tumours in close proximity to the brainstem may remain stable for many years. However, there still are no valid arguments to refrain from therapy in patients with a large vestibular schwannoma, since reliable predictors of tumour growth are not available.

References


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