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Review

"Wait and scan" management of patients with vestibular schwannoma and the relevance of non-contrast MRI in the follow-up

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Abstract

Vestibular schwannoma (VS) is a slow-growing benign neoplasm. There has been an evolution in the management of VS from active treatments (microsurgery and stereotactic radiotherapy) to conservative management (wait and scan). Regular MRI scanning is necessary to monitor tumor progression. Conservative management causes significantly less complications and offers a higher quality of life compared with active treatments. The mean growth rate of VS varies from 0.4 to 2.9 mm/year, and spontaneous shrinkage is observed in 3.8 percent of tumors during observation. If significant growth occurs, active treatment is considered. Significant growth is defined as an increase of at least 3 mm in the largest extrameatal diameter in any plane between the first and last available scans. The vestibulocochlear nerve is surrounded by cerebrospinal fluid, which provides natural contrast for MRI; thus, gadolinium may not be needed to detect VS. Specific sequences have high sensitivity, specificity, and accuracy for detection of progression. Hypointense signal in the ipsilateral inner ear fluid might be a useful sign to distinguish VS from meningioma. In this paper, we summarize the current status of research on conservative management and non-contrast MRI for the detection of VS.

Keywords: Vestibular schwannomas; Treatment; Magnetic resonance imaging; Contrast; Quality of life

Contents

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1. Introduction

Vestibular schwannomas (VSs) are benign neoplasms of the 8th cranial nerve originating from the myelin-forming Schwann cells of either the superior (SVN) or inferior vestibular nerve (IVN), and the latter accounts for more than 90% of cases (Khrais et al., 2008). Patients with VSs commonly complain of asymmetric hearing loss that may be accompanied by tinnitus, dizziness, facial numbness or weakness, or a combination of these. The VS is the most common cerebellopontine angle tumor with a proportion of approximately 80–90% and it accounts for 8–10% of all intracranial neoplasms (Tallan et al., 1993; Valvassori, 1988). The VS has an estimated annual incidence of 1.09 per 100,000 population in USA and 2.55–3.32 per 100,000 population in Netherlands (Kleijwegt et al., 2016; Kshettry et al., 2015). However, this number is significantly higher in Denmark, where the annual incidence has risen from 78 per 100,000 in 1976 to 230 per 100,000 in 2004 likely due to improved detection with magnetic resonance imaging (MRI) (Stangerup and Caye-Thomasen, 2012). In addition, tumor size at diagnosis has decreased significantly over time, probably due to same reason. The current trend in treatment is from traditional microsurgical resection or stereotactic radiotherapy to conservative management (it is also called wait and scan) (Fayad et al., 2014; Jufas et al., 2015; Lee et al., 2014; Patnaik et al., 2015; Rosenberg, 2000; Stangerup and Caye-Thomasen, 2012; Stangerup et al., 2008). For the "wait and scan" strategy, non-contrast MRI has the advantage of avoiding exposure to the contrast agent of gadolinium chelate in either screening or follow-up of VS, but still providing reliable results with constant improvements in technique (Oh et al., 2013; Ozgen et al., 2009; Schulze et al., 2016). This is important for the patients who need serial imaging. This review will present the recent progress in conservative management and non-contrast MRI in the treatment of VS.

2. The biological behavior of vestibular schwannoma

VSs can be either bilateral (neurofibromatosis 2, NF2) or unilateral (sporadic VSs), in which the NF2-associated VSs tend to be more aggressive than the sporadic VSs (Linthicum and Brackmann, 1980) although the latter one may also sparsely exhibit aggressive behavior (Feghali and Kantrowitz, 1995). Inactivation of the NF2 gene, that leads to a loss of merlin (schwannomin, a putative tumor suppressor) expression, is believed to be paramount to the pathogenesis of both NF2 and sporadic VSs (Zwarthoff, 1996). The involvement of merlin depletion in the schwannoma tumorigenesis induces deregulation of ErbB receptor signaling, promoting a dedifferentiated state, and increasing Schwann cell proliferation (Ahmad et al., 2010). Expressed merlin may also be phosphorylated at the site of S518 by members of the PAK family of kinase and thus failed to enter the nucleus of Schwann cells of VS at S phase that is opposite to the wild type merlin (Lu et al., 2008). The ratio of merlin and S518 phosphorylated merlin may contribute to the biological behavior of VS with respect to the growth rate.

In general, VSs grow very slowly but variably with the average annual tumor growth rate varying from 0.4 to 2.9 mm/y (Yoshimoto, 2005). Even spontaneous shrinkage was observed in 3.8 percent of tumors during the "wait and scan" period (Huang et al., 2013). The variability may be associated with the clinical parameters selected, limited number of patients, different observation periods, location of the tumor, and gene status. Intratumoral hemorrhage, vessel density, the inflammatory reaction, and M2-polarized macrophages associated angiogenesis contribute to volume increase of sporadic vestibular schwannomas (Caye-Thomasen et al., 2005; de Vries et al., 2012; de Vries et al., 2013; Koutsimpelas et al., 2007). The VSs usually cause expansion of the internal auditory meatus within the temporal bone due to the growth. It is generally believed that the size and location of the tumor are the prime determinants of the extent of internal auditory meatus bone remodeling and expansion. Tumors that arise laterally along the course of the vestibular nerve tend to cause more expansion of the internal auditory meatus than those that arise more medially which can grow towards the cerebellopontine angle cistern. Due to this, large VSs may compress the brainstem, and an active intervention is needed.