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Original article

Post-surgical vestibular schwannoma remnant tumors: What to do?☆



Reliquat tumoral post-chirurgical de schwannome vestibulaire : que faire ?

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ABSTRACT

Background. – Vestibular schwannomas (VS) are benign tumors of the vestibular nerve's myelin sheath. The current trend in VS surgery is to preserve at the facial function, even if it means leaving a small vestibular schwannoma tumor remnant (VSTR) after the surgery. There is no defined therapeutic management VSTR. The aim of this study was to assess the evolution of the VSTR to define the best therapeutic management and identify predictive factors of VSTR progression.

Methods. – Among the 256 patients treated surgically for VS in the Department of Neurosurgery at Angers University Hospital, 33 patients with a post-surgical VSTR were included in this retrospective study. For all surgical patients, the data collected were age at diagnosis, the Koos classification, the surgical access, the existence of a type 2 neurofibromatosis (NF2), the TR location and size on control MRI-scans. Patients had a bi-annual follow-up with clinical status and VSTR size assessment with MRI-scan. Survival analyzes were performed to determine the time and rate of VSTR progression, and identify factors of progression.

Results. – The mean follow-up of the population was 51 months. All VS remnant progression occurred between 38 and 58 months after surgery. In non-NF2 patients with first follow-up MRI-scan three months after surgery, 43% presented a spontaneous regression, 50% a stability and 7% a progression of the VSTR. In the same population with the 1-year MR-scan after surgery as baseline, 25% presented a spontaneous regression, 62.5% a stability and 12.5% a VSTR progression. These data are consistent with the data reported in the literature. The post-operative facial function impairment and an initial remnant $\geq 1.5 \text{ cm}^3$ were found to be significant risk factors of VS remnant progression in non-NF2 population in univariate analysis ($P=0.048$ and 0.031) but not in multivariate analysis.

Conclusion. – In our experience, the best therapeutic management of the post-surgical VSTR in non-NF2 patients with no risk factor of progression is a simple clinical radiological follow-up otherwise complementary radiosurgery should be considered.

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R É S U M É

Introduction. – Les schwannomes vestibulaires (SV) sont des tumeurs bénignes de la gaine du nerf vestibulaire. La tendance actuelle dans la prise en charge des SV est de conserver la fonction faciale, même si cela signifie laisser un reliquat tumoral (RT) en place lors de la chirurgie. Il n'existe pas de prise en charge thérapeutique définie de ce RT. Le but de cette étude est d'étudier l'évolution du RT afin de déterminer la meilleure prise en charge thérapeutique, ainsi que les facteurs pronostiques de progression du RT.

Patients et méthodes. – Nous avons réalisé une étude rétrospective portant sur 256 patients traités chirurgicalement pour SV dans le département de neurochirurgie du CHU d'Angers. Trente-trois patients

Mots clés :

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Abbreviations: GTR, gross total resection; IAC, internal auditory canal; NF2, type 2 neurofibromatosis; Retrosig, retrosigmoidian surgical approach; Translab, translabyrinthine approach; VS, vestibular schwannomas; VSTR, vestibular schwannomas tumor remnants.

☆ The preliminary results of this study was the subject of an oral communication at: Congress of the French Neurosurgical Society, Bordeaux, France, 22 Mars 2013; Congress of the French Speaking Association of Neuro-Oncologists, Bordeaux, France, 23 Mars 2013.

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porteurs d'un RT confirmé sur l'IRM de contrôle ont été inclus. Pour tous les patients chirurgicaux, les données recueillies étaient l'âge au moment du diagnostic, la classification de Koos de la lésion, la voie d'abord chirurgicale, l'existence d'une neurofibromatose de type 2 (NF2), l'emplacement du RT et la taille du RT sur les scanners de contrôle. Les patients ont eu un suivi bi-annuel avec bilan clinique et suivi de la taille du RT sur l'IRM de contrôle. Des analyses de survie ont été réalisées afin de déterminer le délai et le taux de reprise évolutive du RT, ainsi que les facteurs prédictifs de cette reprise évolutive.

Résultats. – La durée de suivi moyen de la population était de 51 mois. Toutes les reprises évolutives de RT de schwannome vestibulaire sont survenues entre 38 et 58 mois après la chirurgie. Chez les patients non NF2 avec comme IRM de référence celle faite 3 mois après la chirurgie, 43 % présentaient une régression spontanée, 50 % une stabilité et 7 % une progression du RT. Dans la même population comme IRM de référence celle faite 1 an après la chirurgie, 25 % présentaient une régression spontanée, 62,5 % une stabilité et 12,5 % une progression du RT. Ces résultats sont cohérents avec les données de la littérature. L'altération de la fonction faciale post-opératoire et un volume tumoral initial $\geq 1,5 \text{ cm}^3$ sont significativement associés à un risque de progression du RT de VS dans la population non NF2 en analyse univariée ($p = 0,048$ et $0,031$) mais pas dans l'analyse multivariée.

Conclusion. – La prise en charge thérapeutique des RT post-chirurgicaux de SV la plus adaptée chez les patients non-NF2 sans facteur de risque de progression semble être un suivi clinico-radiologique simple, tandis qu'une radiochirurgie complémentaire nous semble être nécessaire dans les autres cas.

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1. Introduction

Vestibular schwannoma (VS) is an histologically benign tumor arising from the schwannoma cell sheath of the vestibular portion of the VIII cranial nerve. VS is the most common tumor of the cerebellopontine angle, with an incidence of 2/100 000 per year, currently increasing as the initial size at diagnosis has decreased. These changes in the epidemiology of the VS have been reported as a consequence of the multiplication of brain MRI-scan investigations [1,2]. Typically, VS clinical symptoms begin with otologic symptoms characterized by unilateral perception hearing loss. If there is no subsequent treatment neurologic signs i.e. ataxia, gait disturbance, facial function impairment or hydrocephalus may then appear.

There are three main possibilities in the management of VS: a “wait-and-see” policy, consisting of a simple clinical follow-up with regular MRI-scan, primary radiosurgery or surgery. Decision management of VS is complex and will depend on a multitude of factors influencing the therapeutic approach.

VS localization in the cerebellopontine angle makes the surgery a technical challenge. The previous trend in therapeutic management was to treat VS surgically with a complete resection even at the cost of an impairment of the facial function. However, currently facial palsy is regarded as a severe handicap by the patient and is no longer considered an acceptable post-operative result. This attitude has led to a change of approach with the preservation of facial function as the first aim of surgery, before the complete resection of the VS [3–5].

Surgeons now prefer to leave a small remnant instead of risking the facial function of the patient by performing a complete resection [6], and this change of attitude in VS surgery is leading to an increase in the prevalence of post-operative vestibular schwannoma tumor remnants (VSTR), which makes the problematics of VSTR management a more frequent question.

There is no defined therapeutic approach towards this VS remnant. Some surgical teams recommend routine radiosurgery [7–12] while other teams have adopted a “wait-and-see” attitude [13–15], keeping radiosurgery for regrowth of the VS remnant [16,17].

Despite being safe compared to surgery, radiosurgery is not without potential side effects and complications especially in a post-surgical cerebellopontine angle, and its place as a routine treatment of a VS post-surgical remnant is widely discussed.

2. Aims of the study

The objectives of this original study were to define the best therapeutic attitude towards post-surgical VS remnant tumors and to identify the factors of VS remnant progression.

3. Methods

3.1. Patient population

All consecutive patients who underwent surgical treatment of VS in the Department of Neurosurgery at Angers University Hospital between 1977 and 1st May 2013 were included in the study. They all underwent a planned gross total resection (GTR). The surgical indication was the appearance of neurological symptoms or hydrocephalus. Most of the VS were stage III or IV in the Koos classification [18,19], and no longer had a useful hearing capacity before surgery, with a Tokyo score of C or worse. The diagnosis was confirmed histologically in all cases.

3.2. Surgical technique

A multidisciplinary team, made up of a neuro-otologist and a neurosurgeon performed the surgical procedures. The primary surgical objective was the GTR and internal auditory canal decompression, with preservation of the facial function. The translabyrinthine approach, allowing a more effective exposition of the internal auditory canal, was favored, except in patients with an ipsilateral partial hearing preservation, where the retrosigmoid approach was preferred in an attempt to preserve hearing. Facial nerve function was continuously checked during the intervention by facial nerve monitoring. The surgical procedure was halted when the facial nerve stopped responding to neurostimulation during surgery or when the surgeon estimated that the benefit/risk ratio regarding facial function preservation was weighed against the GTR of the tumor, and chose to leave a small tumor remnant to avoid any facial nerve lesions.

3.3. Clinico-radiological follow-up

All patients benefited from a multidisciplinary follow-up by their neuro-otologist and neurosurgeon with regular consultations and control MR-scans when available, or with CT-scan by default.

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