

Treatment for head and neck neurilemoma: an integrative review

Tratamento para neurilemoma de cabeça e pescoço: uma revisão integrativa

Tratamiento del neurilemoma de cabeza y cuello: una revisión integrativa

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Abstract

Neurilemmoma or Schwannoma is a benign neoplasm of neural origin originating in the Schwann cell, which can be considered relatively uncommon, and which affects soft tissues in the head and neck region, being noted in these regions in 25% to 48% of cases. The diagnosis is made by histopathological examination, and the most indicated treatment is conservative surgical excision, with a favorable prognosis, and the lesion should not recur or present malignant transformations. Thus, the aim of the study is to discuss the main treatments available for Neurilemmoma, through an integrative literature review. This study was carried out from February 2023 to April 2023, through a literature review of scientific articles obtained from the Scielo, PubMed and Google academic databases, in which articles directly related to the theme of the work were included. There are several treatment methods for Neurilemmoma, with surgical excision being the most commonly used method. There are also innovative options, such as the Gamma Knife, and Five-Fraction Stereotactic Radiotherapy with Linear Accelerator, which can be used together to ensure greater treatment efficiency. Surgical excision is still the most used treatment method for Neurilemmomas. However, other methods have been used, with a view to ensuring greater benefit in the treatment.

Keywords: Neurilemmoma; Plexiform Schwannomatosis; Therapeutics; Treatment.

Resumo

O Neurilemoma ou Schwannoma é uma neoplasia benigna de proveniência neural originado na célula de Schwann, que pode ser considerado relativamente incomum, e que atinge tecidos moles da região de cabeça e pescoço, sendo notados nessas regiões em 25 % a 48% dos casos. O diagnóstico é realizado pelo exame histopatológico, e o tratamento mais indicado é a excisão cirúrgica conservadora, com prognóstico favorável, não devendo a lesão recidivar ou apresentar transformações malignas. Assim, o objetivo do estudo é discutir sobre os principais tratamentos disponíveis para o Neurilemoma, através de uma revisão integrativa da literatura. Este estudo foi realizado no período de Fevereiro de 2023 até Abril de 2023, por meio de revisão da literatura de artigos científicos obtidos nas bases de dados Scielo, PubMed e Google acadêmico em que foram incluídos artigos diretamente relacionados à temática do trabalho. Vários são os métodos de tratamento para o Neurilemoma, sendo a excisão cirúrgica o método mais comumente utilizado. Também existem opções inovadoras, como a Faca Gama, e Radioterapia Estereotáxica Hipofracionada de Cinco Frações com Acelerador Linear, podendo ser utilizadas em conjunto a fim de garantir maior eficiência no tratamento. A excisão cirúrgica ainda é o método de tratamento mais utilizado para Neurilemmomas. No entanto, outros métodos têm sido utilizados, com vistas a garantir um maior benefício no tratamento.

Palavras-chave: Neurilemoma; *Schwannomatose* Plexiforme; Terapêutica; Tratamento.

Resumen

El neurilemoma o schwannoma es una neoplasia benigna de origen neural con origen en la célula de Schwann, que puede considerarse relativamente poco frecuente, y que afecta a los tejidos blandos de la región de cabeza y cuello, observándose en estas regiones en un 25% a 48% de los casos. El diagnóstico se realiza por examen histopatológico, y el tratamiento más indicado es la extirpación quirúrgica conservadora, con pronóstico favorable, y la lesión no debe recidivar ni presentar transformaciones malignas. Así, el objetivo del estudio es discutir los principales tratamientos disponibles para el Neurilemoma, a través de una revisión integradora de la literatura. Este estudio se realizó desde febrero de 2023 hasta abril de 2023, a través de una revisión bibliográfica de artículos científicos obtenidos de las bases de datos académicas Scielo, PubMed y Google, en las que se incluyeron artículos relacionados directamente con la temática del trabajo. Existen varios métodos de tratamiento para el neurilemoma, siendo la escisión quirúrgica el método más utilizado. También hay opciones innovadoras, como el bisturí de rayos gamma y la radioterapia estereotáctica de cinco fracciones con acelerador lineal, que se pueden usar juntas para garantizar una mayor eficacia del tratamiento. La escisión quirúrgica sigue siendo el método de tratamiento más utilizado para los neurilemmomas. Sin embargo, se han utilizado otros métodos, con el fin de garantizar un mayor beneficio en el tratamiento.

Palabras clave: Neurilemoma; Schwannomatosis plexiforme; Terapéutica; Tratamiento.

1. Introduction

Neurilemmoma or Schwannoma is a benign neoplasm of neural origin and unknown origin, which affects the soft tissues of the head and neck region and develops from a disorderly proliferation of Schwann cells in the myelin sheath, which function as electrical insulators and enhance nerve conduction. There is no gender predilection, it is more prevalent among young and middle-aged adults. In the oral cavity, in addition to the tongue, other regions can be affected, such as the base of the tongue, floor of the mouth, palate, lips and vestibular mucosa (Líbera et al., 2021; Neville, 2009; Santana et al., 2022). Microscopically, there are two patterns, the Antoni A and Antoni B patterns, which can be analyzed in different ways, with the possibility of demonstrating the two patterns in the same lesion in variable shapes and quantities, in addition to finding a pattern in isolation. The variation of the histopathological pattern is not clearly defined in the literature, no data were found that show differences in clinical behavior, treatment and recurrence. (Líbera et al., 2021; Santana et al., 2022, Vieira de Miranda et al., 2017).

The diagnosis is made by histopathological examination and the most indicated treatment is conservative surgical removal, with a favorable prognosis. The inconstancy of the histopathological pattern can help the pathologist in the diagnosis of this lesion, which reinforces the importance of associating clinical characteristics with the histological pattern found (Líbera et al., 2021; Santana et al., 2022, Vieira de Miranda et al., 2017). Usually, the increase in volume is asymptomatic, but tenderness or pain may occur, varying in size from a few millimeters to many centimeters (Neville, 2009). When there are great clinical similarities between lesions, reaching the final diagnosis can only be concluded through complementary exams, previous differential clinical diagnoses, as these are essential for adequate treatment. In the case of benign mesenchymal neoplasms, such as Neurofibroma and Neurilemmoma, the differential clinical diagnoses can be considered, because they usually present as submucosal nodules covered by intact mucosa and are asymptomatic with unknown evolution. In addition, the characteristic microscopic findings of each lesion guide the final diagnosis (Pires, 2022).

Thus, histopathological examination is the main method for diagnosis, and can sometimes be inconclusive. Therefore, it is necessary to aim for other techniques, such as immunohistochemistry, to reach a conclusive diagnosis. It is believed that the histopathological pattern related to the location is relevant in helping the pathologist to make a correct diagnosis and indicate treatment (Pires, 2022).

Based on the above, the objective of the study was to discuss the main treatments available for Neurilemmoma, through an integrative literature review.

2. Methodology

The study is an integrative review, an investigation that consists of finding relevant themes in a given topic that allow

exploration, as gaps in the literature are identified by recognizing others (Vieira de Miranda et al., 2017). Thus, the study was prepared according to the structure of the Integrative Systematic Review Manual (Cunha, 2014).

The guiding question determined for this integrative literature review was: "Is there a consensus on the best treatment for Neurilemmoma?".

The access portals to the Scientific Electronic Library Online (SciELO), PubMed, BIREME and SCOPUS electronic databases were consulted from February to March 2023, using the following descriptors: "Neurilemoma", "Schwannoma", "Schwannomatosis, Plexiform", "Therapeutics" and "Treatment"; according to the search strategy using the Boolean operators "OR" and "AND" in the association of descriptors, with the languages of the filters in English, Portuguese and Spanish, and publications between 2019 and 2023, that is, in the last 5 years. Duplicate evidence was located with Mendeley's help and excluded. In addition, a search was carried out in the gray literature and manually in the references of the selected articles.

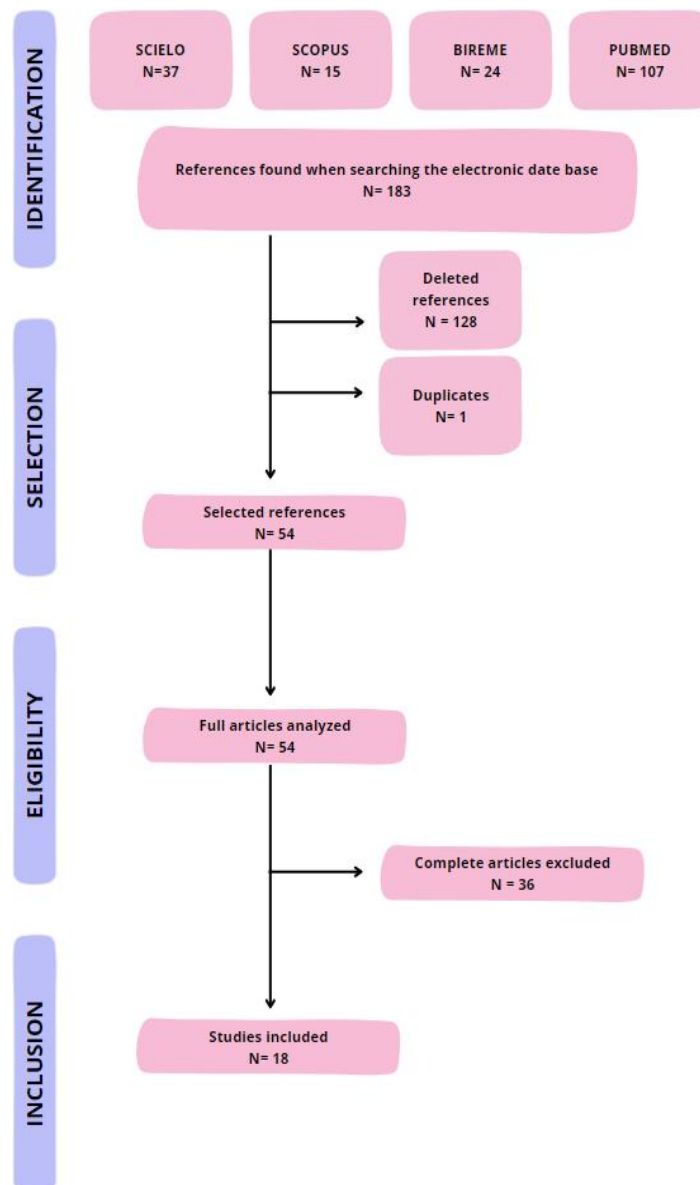
The results found through the integrative literature review were submitted to content analysis, carried out in 03 phases: a) pre-analysis; b) exploration of the material; and c) treatment of results.

Inclusion criteria were: systematic reviews, meta-analyses and clinical studies; and as exclusion criteria: narrative reviews, integrative reviews and animal model studies.

Papers that met the eligibility criteria were summarized according to year of publication, journals, authors, titles, objectives, results and conclusions.

Figure 1 shows the summary of the search and selection of papers - 183 articles were identified and 54 were selected. Thus, 128 were excluded and 18 were chosen in this integrative literature review.

Figure 1 - Work search and selection flowchart.



Source: Authors.

3. Results and Discussion

The survey in the databases located 183 productions, of which 129 were excluded after reading the titles and abstracts, as they did not meet the established criteria. Of the 54 works selected for reading in full, only 18 were compatible with the research inclusion criteria.

One author of this study was selected to perform a content analysis. In Table 1 it is possible to observe the summary of the articles included according to the year of publication, journals, authors, titles, objectives, results and conclusions.

Table 1 - Summary of articles included according to year of publication, journals, authors, title, objectives, results and conclusions.

| Publication year | Journal | Authors | Title | Goals | Results | Conclusion |
|------------------|---------|---|---|---|--|---|
| 2019 | science | Dokania, Vivek; Rajguru, Anagha; Mayashankar, Vishwakarma; Mukherjee, Indranil; Jaipuria, Bhagyashree; Shere, Devika; (Dokania et al., 2019) | Palatal Schwannoma: An Analysis of 45 Literature Reports and of an Illustrative Case | To review previously reported cases of palatal schwannoma along with an illustrative case and provide better insight into the clinicopathological and radiological features of this neural tumor in a rare intraoral location | We present a case of palatal schwannoma in a 16-year-old girl. Another 45 cases were identified in 2 searches in medical databases (PubMed and Google Scholar) published from 1985 onwards, and from 13 countries on 5 continents. The age of the patients ranged from 3 to 84 years. Palatal schwannoma showed a slight predilection for females, with a male/female ratio of ~ 1:1.81. Hard palate involvement is almost twice as high as soft palate involvement. Surgical excision was employed in almost all cases, and recurrence was reported only once. | Palatal schwannomas are rare, but have been reported in both the hard and soft palate. They present as a painless, firm, well-encapsulated, slow-growing lesion on the lateral palatal surface. The diagnosis is confirmed by histopathological examination. Fine needle aspiration cytology (FNAC) is almost always inconclusive. Complete surgical excision is the treatment of choice, and recurrence or malignant transformation is rare. |
| 2022 | Scopus | Priya Kumthekar, Sean Aaron Grimm, Roxanne T Aleman, Marc C Chamberlain, David Schiff, Patrick Y Wen, Fabio Massaiti Iwamoto, Demirkan Besim Gursel, David A Reardon, Benjamin Purow, Masha Kocherginski, Irene Helenowski, Jeffrey J Raizer (Kumthekar et al., 2022) | A multi-institutional phase II trial of bevacizumab for recurrent and refractory meningioma | To better evaluate the activity of bevacizumab (BEV) in patients with meningiomas where definitive surgery and radiotherapy (RT) have been considered impossible or something that has already been attempted. | Fifty patients, 42 with progressive meningioma were treated: 10 G1M, 20 G2M and 12 G3M. Previous treatments include surgical resection, radiosurgery, external beam radiation therapy, and chemotherapy. Median infusions administered were 16. Response was graded using Macdonald's criteria. PFS-6, medium PFS and mOS were 87%, 22 months, 35 months for G1M; 77%, 23 months, 41 months for G2M; and 46%, 8 months, 12 months for G3M. The best radiographic responses include stable disease; partial response and progressive disease. The most common toxicities were hypertension, proteinuria, and fatigue. | Bevacizumab (BEV) is well tolerated and appears to be a promising systemic treatment option for patients with recurrent and refractory meningiomas. |
| 2022 | PubMed | Yang, Siyuan; Wang, Jiahe; Li, Xiang; Li, Hang; Zhong, Yi; Zhou, Xinmin; Chen, Gang; (Yang et al., 2022) | Gamma knife radiosurgery for trigeminal schwannomas: A systematic review and meta-analysis | To systematically evaluate the efficacy, safety, and complications of gamma knife radiosurgery for trigeminal schwannomas. | The search reached 351 studies, of which 35 were evaluated for full-text eligibility. 19 studies were included in the meta-analysis. 456 of 504 patients (0.94, 95% CI 0.91–0.96, I2 = 3.02%, p < 0.01) from 18 studies had local control and 278 of 489 patients (0.63, CI 95% 0.48–0.78, I2 = 88.75%, p < 0.01) of 16 studies showed tumor regression or disappearance. 231 of 499 patients (0.50, 95% CI 0.37–0.62; I2 = 83.89%, P < 0.01) from 17 studies had improved clinical symptoms. There was no | GKRS is an effective primary and adjuvant method in the treatment of trigeminal schwannomas, with reliable rates of tumor control. Randomized clinical trials are needed to more broadly and comprehensively assess the benefit-risk balance of gamma knife radiosurgery. |

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| | | | | | significant difference in tumor control between those treated with GKRS as a primary treatment or adjunctive to surgery (p = 0.390). | |
| 2021 | PubMed | (Balossier et al., 2021) | Repeat stereotactic radiosurgery for progressive vestibular schwannomas after previous radiosurgery: a systematic review and meta-analysis | To perform a systematic review and meta-analysis of the literature on repeat stereotactic radiosurgery (SRS) for SV. Using PRISMA guidelines, we reviewed manuscripts published between January 1990 and October 2020 and referenced in PubMed. Tumor control and cranial nerve outcomes were assessed with separate meta-analyses. | Cranial nerve results remain favorable, particularly for the facial nerve. The rate of hearing loss seems similar to that related to the first SRS. New facial nerve paralysis or worsening of the previous one was reported in 8/183 patients, ie a rate of 4.3% (range 1.4–7.2%, p = 0.004). Hearing loss was reported in 12/22 patients, ie a rate of 54.3% (range 24.8–83.8%, I2 = 70.7%, p < 0.001). Repeat SRS after previous failure of SRS to VS is associated with high rates of tumor control. | Repeat SRS after previous failure of SRS to VS is associated with high rates of tumor control. Tumor progression was reported at an overall rate of 16%, while only a few series reported additional need for surgical treatment. So, in the event of such progression, an ETT must be excluded. Cranial nerve outcomes remain favourable, particularly in relation to the facial nerve. Hearing loss rates are similar to the first SRS. |
| 2021 | PubMed | (Ignacio et al., 2021) | Efficacy of aspirin for sporadic vestibular schwannoma: a meta-analysis | we aimed to systematically review the evidence on the effect of ASA ingestion on tumor growth in patients with SVs. Methods: Pubmed, Cochrane, Scopus, Embase, ClinicalTrials.gov, and Web of Science were searched for studies comparing VS tumor growth in patients with aspirin intake and those without. | This result is valid for the analysis of linear tumor growth (OR 1.23; 95% CI 0.49, 3.10), volumetric tumor growth (OR 1.41; 95% CI 0.36, 5.59) and both combined (OR 1.02; 95% CI 0.56, 1.86). Conclusions: Our meta-analysis suggests that there is insufficient evidence to recommend ASA therapy in patients with SVs. | |
| 2021 | PubMed | (Peciu-Florianu et al., 2021) | Trigeminal Neuralgia Secondary to Meningiomas and Vestibular Schwannoma Is Improved after Stereotactic Radiosurgery: A Systematic Review and Meta-Analysis | We sought to review the medical literature on TN treated with SRS for meningiomas and SV and to investigate rates of improvement in TN symptoms. Methods: We reviewed articles published between January 1990 and December 2019 in PubMed. | There was no significant difference in the series that discussed the results for tumor targeting versus tumor and nerve targeting. Recurrences were described in 34.7% (range 21.7–47.6; tumor target). Maintenance of BNI I was reported in 36.4% (range 20.1–52.7) and BNI I-IIIb in 41.2% (range 29.8–52.7; tumor targeting series). When nerve and tumor were targeted, only 1 series reported 86.7% with BNI I-IIIb at last follow-up. Complications were found in 12.6% (range 6.3–18.8; tumor targeting series) of patients; however, they were much higher, reaching 26.7%, in the only study that reported them after reaching both the nerve and the tumor. | Future clinical trials should focus on standard reporting of clinical outcomes and randomization of targeting methods. heterogeneity between reports and segmentation approaches. Although nerve and tumor targeting seemed to achieve better long-term results, the complication rate was much higher and the number of patients treated was limited. |

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| 2022 | PubMed | (Thai et al., 2022) | Treatment for vestibular schwannoma: Systematic review and single arm meta-analysis | systematically review and single-arm meta-analysis the different techniques of treatment of vestibular schwannoma. Methods: A comprehensive literature search was performed using thirteen databases, including PubMed, Scopus, and Web of Science. | A total of 35 clinical trials were included in the final analysis. The pooled proportion of stable hearing ability in patients undergoing gamma knife radiosurgery (GKRS) was 64% (95% CI: 52%–74%). The GKRS favored an increase in hearing ability by 10% (95% CI: 7%–16%). Regarding tumor size, GKRS is the most protective method 53% (95% CI: 37%–69%). Complications occurred most commonly in single fractionated linac stereotaxic radiosurgery (SFSRT) 37% (95% CI: 12%–72%). | Our analysis suggests that gamma knife radiosurgery may be the most optimal treatment for vestibular schwannoma based on stabilization of hearing ability, increase in hearing ability, decrease in tumor size and complications. |
| 2021 | PubMed | Peciu-Florianu (Peciu-Florianu et al., 2021) | Tumor control and trigeminal dysfunction improvement after stereotactic radiosurgery for trigeminal schwannomas: a systematic review and meta-analysis | review the medical literature on TS treated with SRS to investigate tumor control rates and symptomatic improvement. We reviewed manuscripts published between January 1990 and December 2019 in PubMed. Rates of tumor control and symptomatic improvement were assessed with separate meta-analyses. | Tumor control rates after SRS were 92.3% (range 90.1–94.5; $p < 0.001$) and tumor shrinkage rates were 62.7% (range 54.3–71, $p < 0.001$). Tumor progression rates were 9.4% (range 6.8–11.9, $p < 0.001$). Clinical improvement rates for trigeminal neuralgia were 63.5% (52.9–74.1, $p < 0.001$) and oculomotor nerves were 48.2% (range 36–60.5, $p < 0.001$). The rate of clinical worsening was 10.7% (range 7.6 to 13.8, $p < 0.001$). | Stereotactic radiosurgery for TS is associated with high rates of tumor control and favorable clinical outcomes, especially for trigeminal neuralgia and oculomotor nerves. However, patients must be correctly counseled about the risk of tumor progression and potential clinical worsening. Future clinical trials should focus on standard reporting of clinical outcomes. |
| 2020 | PubMed | (Söderlund Diaz et al., 2020) | LINAC-based stereotactic radiosurgery versus hypofractionated stereotactic radiotherapy delivered in 3 or 5 fractions for vestibular schwannomas: comparative assessment from a single institution | to evaluate outcomes after LINAC-based radiosurgery (SRS) and hypofractionated radiotherapy (hypo-FSRT) and to identify possible differences in outcomes between hypo-FSRT administered in 3 or 5 fractions. | The median follow-up was 57 months. Eight patients had progression requiring surgery, corresponding to an overall local control rate of 93.4%, with no significant difference between fractionation schemes. A borderline significant correlation ($p = 0.052$) was detected between cystic tumors and local failure. A trend towards a higher incidence of local failure was observed after 2015, when SRS treatment increased to include slightly larger tumours. Hearing preservation was seen in 35% of patients and 36% of patients experienced acute side effects, but persistent facial or trigeminal nerve toxicity was rare. | SRS and hypo-FSRT with 3 or 5 fractions provided a high rate of local control with no significant differences between treatment regimens. SRS is a well-documented radiation technique for VS and is the recommendation for small to medium-sized tumors. This report demonstrates excellent long-term results after hypo-FSRT; |
| 2020 | PubMed | (Ding et al., 2020) | Meta-analysis of tumor control rates in patients undergoing stereotactic radiosurgery for cystic vestibular | This review serves to introduce a renewed understanding of the effectiveness of SRS as a viable treatment modality for cystic SV. | A total of 246 patients underwent SRS for cystic VS, with reported mean or median follow-up ranging from 49.7 to 150 months, and an overall range of 6 to 201 months. After SRS treatment for cystic SV in all studies, 92% of patients had | Despite the paucity of pertinent data, the results of our meta-analysis suggest that SRS has effective rates of tumor control in patients with cystic VS. Therefore, SRS can be considered a viable treatment |

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| | | | schwannomas | | tumor control at follow-up (95% CI: 88–95%). The tumor control rate specifically for patients undergoing the GammaKnife was 93% (95-CI: 88%–95%). | modality when choosing among interventions for cystic VS. |
| 2018 | PubMed | (Starnoni et al., 2018) | Systematic review and meta-analysis of the technique of subtotal resection and stereotactic radiosurgery for large vestibular schwannomas: A "nerve-centered" approach | The aim of this study was to provide a systematic review and meta-analysis of the available literature on the combined strategy of subtotal resection (STR) followed by stereotactic radiosurgery (SRS) for large SVs. | The authors included 9 studies (248 patients). With a weighted mean follow-up of 46 months (range 28-68.8 months), the pooled rate of overall tumor control was 93.9% (95% CI 91.0%-96.8%). Salvage treatment (according to STR and/or SRS) was required in only 13 (5.24%) of the 18 patients who failed initial treatment. According to the House-Brackmann (HB) grading scale, functional preservation of the facial nerve (HB grade I-II) was achieved in 96.1% of patients (95% CI 93.7%-98.5%). Serviceable hearing after the combined approach was preserved in 59.9% (95% CI 36.5%-83.2%). | A combined approach of STR followed by SRS has been shown to have excellent clinical and functional results, while achieving a tumor control rate comparable to that obtained with a total resection. Longer-term follow-up and larger cohorts of patients are needed to fully assess the rate of tumor control achieved with this approach. |
| 2019 | PubMed | (ROMIYO ET AL., 2019) | Radiosurgery treatment is associated with improved facial nerve preservation versus repeat resection in recurrent vestibular schwannomas | PubMed, Scopus, Embase, Cochrane, and Web of Science databases were searched for studies reporting patients undergoing radiosurgery or repeated surgical resection after resection. | Our results reveal that tumor control rates are comparable between adjuvant radiosurgery and secondary resection. However, adjuvant radiosurgery has been shown to better preserve good facial nerve function compared to secondary surgical resection. | With comparable rates of tumor control and better preservation of good facial nerve function, this study suggests that secondary radiosurgery for recurrent VS is associated with optimal tumor control and preservation of good facial nerve function. |
| 2018 | PubMed | (Comps et al., 2018) | Upfront gamma knife surgery for facial nerve schwannomas: retrospective case series analysis and systematic review. | We reviewed our series of four consecutive cases treated with Gamma Knife Surgery (GKS) between July 2010 and July 2017 at the University Hospital of Lausanne, Switzerland. Radiosurgery was performed using Leksell Gamma Knife Perfexion. | The mean age at the time of the GKS was 44.25 years. Two cases had facial paralysis and two had hemifacial spasm. Pretherapeutically, the House-Brackmann (HB) grade was II for one case, III for two, and VI for one. All patients maintained stable Gardner-Robertson class 1 at the last follow-up. | The results of our systematic review are also encouraging, with satisfactory rates of stabilization and/or clinical improvement and a high rate of tumor control. Complications are infrequent and generally transient. |
| 2018 | PubMed | (Papatsoutsos et al., 2018) | Self-Evaluated Quality of Life and Functional Outcomes after Microsurgery, Stereotactic Radiation or Observation-Only for Vestibular | To assess the impact of different treatment options on health-related quality of life (HRQoL) in patients with vestibular schwannoma. | Ten prospective and 29 retrospective studies were identified: microsurgery initially had a negative effect on HRQoL, but this tended to improve with follow-up. Radiotherapy had a less negative effect, but with minimal changes over the follow-up. | The heterogeneity and methodological weaknesses of the included studies constitute the main limitation of this review. |

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| | | | Schwannoma of the Adult Patient: A Systematic Review | | | |
| 2018 | PubMed | (Nguyen et al., 2018) | Hypo-fractionated stereotactic radiotherapy of five fractions with linear accelerator for vestibular schwannomas: A systematic review and meta-analysis | In this systematic review, PubMed, Scopus, Web of Science, Embase, and Cochrane databases were selected to identify relevant studies for five-fraction hypo-FSRT for VS patients. | Five retrospective studies spanning 1999-2014 were identified. We identified 228 patients with a mean age ranging from 43 to 63.4 years and a mean follow-up of 12.5 to 43.1 months. The total tumor dose was 25Gy for three studies and 20Gy for one study, all distributed in 5 fractions. | Hypo-FSRT shows clinical results comparable to SRS regarding tumor control, with slightly superior facial nerve sparing. However, studies with longer follow-up periods must be performed. |
| 2020 | PubMed | (Starnoni et al., 2018) | Surgical management for large vestibular schwannomas: a systematic review on behalf of the EANS skull base section | Get recommendations for managing Schwannomas from a European perspective. | There is a lack of consensus to support the superiority of any surgical strategy with regard to the extent of resection and use of adjuvant radiosurgery. Intraoperative neuromonitoring needs to be used routinely to preserve neural function. | Quality of life for the patient; functional preservation of the facial/cochlear nerve, ensuring optimal oncological control. |
| 2022 | PubMed | (Bal <i>et al.</i> , 2022) | Management of non-vestibular schwannomas in adult patients: a systematic review and consensus statement on behalf of the EANS skull base section. Part I: oculomotor and other rare non-vestibular schwannomas (I, II, III, IV, VI) | Provide information on the main principles that define surgical management and strategy, in order to formulate a series of recommendations. | A summary of the literature evidence was proposed after discussion in the EANS skull base section. The task force constituted dealt with existing standards of practice regarding preoperative radiologic investigations, ophthalmologic evaluations, optimal surgical and radiotherapy strategies, and follow-up management. | This article represents the consensus opinion of the task force regarding the treatment of non-vestibular schwannomas. For each of these tumors, the management of these patients is complex, and for those who are symptomatic tumors |
| 2018 | PubMed | (Torun <i>et al.</i> , 2018) | Schwannoma of the trochlear nerve—an illustrated case series and a systematic review of management | Illustrate a case series and suggest a rational treatment algorithm for trochlear schwannomas and a pertinent literature review. | Of those reported, less than half (40%) of the patients underwent surgical resection, while the rest were treated conservatively or with SRS. Seventy-six percent (65/85) of all less than half (40%) of patients underwent surgical resection, while the remainder were treated conservatively or with SRS. Seventy-six percent (65/85) of all less than half (40%) of patients underwent surgical resection, while the remainder were | Both open microsurgical removal and SRS can achieve good long-term local control. Consequently, a personalized multidisciplinary treatment algorithm based on individual presentation and tumor configuration is proposed. |

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| | | | | | <p>treated conservatively or with SRS. Seventy-six percent (65/85) of the entire cohort had diplopia, which was the lone symptom in more than half of the cases (n = 39). All patients who had symptoms other than diplopia or headache as the only symptoms underwent surgical resection. Patients in the non-surgical group were mostly male (M/F = 3.5:1), older and with a smaller mean diameter (4.6 vs. 30.4 mm, $p < 0.0001$) when compared to the surgical group. Twelve patients in the entire cohort were treated with SRS, none of whom underwent surgical resection before or after radiation treatment. Patients with trochlear schwannoma without systemic neurofibromatosis are rare and rarely reported in the literature. Of these, patients with symptomatic trochlear schwannomas do not form a single homogeneous group, but fall into two quite distinct subgroups in terms of demographic and clinical characteristics. Among patients who require intervention, there are open microsurgical resections, as well as less invasive treatment options aimed at safely relieving symptoms and preventing progression.</p> | |
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Source: Authors.

Regarding the treatment of Neurilemma, it was possible to observe a significant agreement of the consulted authors in the use of surgical excision. However, there was a scarcity in the literature consulted on this discussion, which denotes the need for further studies.

For Torun et al. (2018), microsurgical removal and single-fraction radiosurgery (SRS) can be efficient and control the lesion in the long term. Furthermore, a multidisciplinary and personalized approach for each patient and tumor condition must be proposed.

It is also important to highlight that fine needle aspiration cytology (FNAC) is almost always inconclusive. Furthermore, as far as treatment is concerned, opting for complete surgical excision of the lesion is usually the best choice (Dokania, 2019). Still according to the literature, both the recurrence of Neurilemma after surgical removal and its evolution into a malignant lesion are extremely rare events (Dokania, 2019).

In some situations, in the case of small or medium-sized vestibular schwannomas, stereotactic radiosurgery may be a viable option. The technique consists of administering high doses of radiation to stop the growth of the lesion, and it can also be performed a second time in case of initial failure. In addition, the technique can also be associated with microsurgical resection (Balossier et al., 2021). Furthermore, the use of the technique with the aim of improving symptomatic conditions associated with neurilemoma, such as trigeminal neuralgia, presents favorable clinical results (Peciu-Florianu et al., 2021).

It is worth noting that the use of some medications such as aspirin does not represent a safe option for the treatment of Neurilemmoma, even if conservative, as there is still not enough evidence to support this recommendation (Ignacio et al., 2021). Gamma knife radiosurgery (GKRS) treatment represents a less invasive form of therapy. In addition, it demonstrates good results in relation to hearing capacity, when this is compromised due to vestibular schwannoma, reducing the size of the tumor and decreasing complication rates, when compared to other non-invasive treatment options (Thai et al., 2022).

Furthermore, bevacizumab has been shown to be a positive alternative to systemic treatment for patients with recurrent and refractory meningiomas. Similarly, the Basic Technical Manual of Oncology-SIA/SUS- Ambulatory Information Systems describes bevacizumab as a medicine that continuously multiplies in many therapeutic areas of benign and malignant diseases (Dos Sistemas, 2022).

GKRS is an effective primary and adjuvant method in the treatment of trigeminal schwannomas, with reliable rates of tumor control. However, the study by Thai et al. (2022), states that this method can be considered a more suitable treatment for vestibular Schwannoma based on the stabilization of the auditory capacity, increase of auditory capacity, decrease of the tumor size and complications.

Furthermore, it is worth noting the benefits of five-fraction hypofractionated stereotactic radiotherapy with a linear accelerator in the treatment of Schwannomas. This form of treatment, according to the data obtained, demonstrates results such as control of the size of tumors and preservation of the facial nerve that are superior to other forms of treatment. During this type of treatment, the millimetric positioning of the instruments and radiation beams that reach only the necessary structures, that is, the tumor, is taken into account. However, more studies need to be carried out, with a longer follow-up period for patients with Vestibular Schwannomas (Nguyen et al., 2018).

In addition, it is possible to observe that the combination of different forms of treatment, during the same procedure, shows a greater benefit to the patient, such as the union of surgical excision with the subsequent use of Hypofractionated Stereotactic Radiotherapy of Five Fractions with Linear Accelerator. This combination of procedures, excision with the subsequent use of radiotherapy promotes the removal of cell remnants that could promote a recurrence (Nguyen et al., 2018).

By combining different strategies, it is feasible to find an effective treatment for Neurilemmoma. According to (Söderlund Diaz et al., 2020), the application of radiosurgery techniques based on LINAC (SRS) and hypofractionated radiotherapy (hypo-FSRT) in 3 or 5 fractions resulted in a high rate of tumor control, with only 36% of patients experiencing

acute side effects and a rare persistent facial or trigeminal nerve toxicity. It is important to mention that the SRS technique is well documented for the treatment of small and medium sized Neurilemmomas, while a hypo-FSRT has been shown to be effective in the long term.

Still under this perspective, Starnoni et al. (2018) presents a technique that has been shown to be safe and adequate for the treatment of large Neurilemmomas. Combining subtotal resection (STR) with stereotactic radiosurgery (SRS), it is possible to achieve excellent clinical results and a next evolution tumor control rate that can be achieved with a total type resection. Also requiring a higher incidence of studies with control groups, with a greater number of patients and for a longer period, as this way, relapses and consequently sequelae can also be controlled.

4. Conclusion

Therefore, it is possible to point out that currently surgical excision is still the most used method for the treatment of Neurilemmomas. However, other methods have been used and guaranteeing patients a greater benefit in the treatment. Due to these benefits, many studies have addressed the combination of techniques in order to add excision, such as the use of radiotherapy, so that there are no recurrences, and a lower rate of sequelae. However, few reports were found in the literature on aspects of the treatment of Neurilemmomas that affect the oral cavity and its annexes. In this sense, it is recommended to carry out more research on the subject in question, with a view to contributing even more to the scientific debate on this pathology.

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