Trochlear Nerve Schwannoma: Case Report and Literature Review

Schwannoma do nervo troclear: revisão da literatura e relato de caso

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Abstract

Schwannomas arise from the Schwann cells of the peripheral and cranial nerves. They represent 8% of the primary cerebral neoplasms. Although schwannomas usually develop in sensory nerves, most often on the vestibular and trigeminal nerves, in very rare cases they can develop in motor nerves. We reported an unusual case of a 29-year-old woman with headache, nausea, vomiting, and blurred vision ongoing for 3 years. Magnetic resonance of the brain showed a solid-cystic expanded injury, heterogeneous, with limits partially defined and epicenter on the pineal gland. The lesion presented hypointensity in T1 and isointensity in T2. An intense enhancement of the solid part was observed after contrast injection. Foci of calcification and absence of diffusion restriction were also observed. The patient underwent microneurosurgery with supracerebellar infratentorial approach in a seated position. Subtotal resection was performed with maintenance of calcified tumor tissue adhered to the right Rosenthal basal vein. In the postoperative phase, the patient remained with diplopia when looking down; however, she reported improvement of headache and nausea.

Keywords
► neurilemmoma
► trochlear nerve
► diplopia

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Resumo

Os schwannomas surgem das células de Schwann, dos nervos periféricos e cranianos e representam 8% das neoplasias cerebrais primárias. Apesar de os schwannomas se desenvolverem comumente em nervos sensoriais, mais frequentemente no nervo vestibular e trigêmeo, em casos muito raros ele pode se desenvolver em nervos motores. Relatamos um caso raro, de uma paciente do sexo feminino, 29 anos, com quadro de cefaleia, náuseas, vômitos e turvação visual há três anos. Ressonância magnética de encéfalo demonstrou lesão expansiva sólido-cística, heterogênea, com limites parcialmente definidos e epicentro na glândula pineal. Apresentou hipossinal em T1 e isossinal em T2. Houve intensa reação da parte sólida após injeção de contraste. Foram observados focos de calcificação e ausência de restrição à difusão. A paciente foi submetida a tratamento microneurocirúrgico com acesso infratentorial supracerebelar em posição sentada. Houve ressecção subtotal com manutenção de tecido tumoral calcificado aderido à veia basal de Rosenthal direita. No pós-operatório, a paciente permaneceu com diplopia na mirada ocular para baixo, entretanto apresentou melhora de cefaleia e náuseas.

Keywords
► neurilemmoma
► nervo troclear
► diplopia

Palavras-chave
► neurilemmoma
► nervo troclear
► diplopia
Introduction

Schwannomas arise from the Schwann cells and represent 8% of the primary cerebral neoplasms. They usually develop in sensory nerves, most often in vestibular nerve. In very rare cases schwannomas can develop in motor nerves, including the trochlear nerve. Trochlear nerve schwannomas are extremely rare and, according to our review, there are 37 surgical cases related in the literature, including ours.

Case Report

The case described is that of a female patient, 29 years-old, with headache associated with nausea, vomiting and blurred vision for 3 years. Imaging studies performed in another service showed an expansive lesion, causing mass effect, resulting in hydrocephalus. The patient was submitted to ventriculoperitoneal shunt, with improvement of the clinical condition. After the surgery, clinical follow-up was realized.

The patient was admitted in our service 2 years after the ventriculoperitoneal shunt surgery. She had diplopia when looking down, headache and nausea refractory to the clinical treatment. Magnetic resonance imaging (MRI) of the brain showed a solid-cystic expanded injury, heterogeneous, with limits partially defined and epicenter on the pineal gland. The lesion presented hyposignal in T1 and isosignal in T2. An intense enhancement of the solid part was observed after contrast injection (Figs. 1, 2, 3). Foci of calcification and absence of diffusion restriction were observed as well.

Cerebrospinal fluid examination did not show tumor cells and the search for a-fetoprotein and human chorionic gonadotropin were negative. The patient underwent micro-neurosurgery with supracerebellar infratentorial approach in a seated position. The surgery was performed with ultrasonic aspiration and central enucleation of the tumor, allowing a better mobility of the tumor capsule and dissection of deep veins and mesencephalic roof. Subtotal resection was performed, with maintenance of calcified tumor tissue adhered to the right Rosenthal basal vein due to profuse bleeding when attempting resection of the tumor portion in this topography (Fig. 4).

The fourth nerve schwannomas typically show predominance of Antoni B tissue, have frequent clusters of xanthomatous macrophages and are poor in Verocay bodies (these are often more common in spinal tumors). Reticulin is abundant and distinctly pericellular in schwannomas.

Schwannomas are strongly and uniformly reactive for S-100 protein; they may be positive for epithelial membrane antigen (EMA), but not frequently, and do not exhibit the membrane-characteristic pattern of meningioma. Microscopic patterns and immunohistochemical findings in our case (Fig. 5). In the postoperative phase, the patient remained with diplopia when looking down, reporting improvement of headache and nausea.
Discussion

We did a literary review using the PubMed and COCHRANE platforms searching for the following terms: trochlear nerve schwannoma and schwannoma surgery.

We found the occurrence of 37 surgical cases of trochlear schwannoma. The average age at diagnosis was 44.8 years, with a standard deviation of 15.1. The female gender was predominant, with 21 cases (58.3%) (∆Table 1).

Trochlear nerve schwannoma presents as initial symptoms, in most cases, diplopia (60%), hemiplegia (43%), headache (40%) and cerebellar symptoms (37%), such as ataxia, dysmetria and nystagmus.29,33 In the present study, trochlear nerve palsy was evidenced in 17 (50%) of 34 cases (∆Table 1). In three case reports, intratumoral hemorrhage occurred as a complication.7,26,33

Macroscopically, schwannomas are encapsulated, well-delineated, lobulated, generally grayish masses, and may have areas of cystic alterations and xanthomatosis.36,37

Magnetic resonance imaging showed to be the most used imaging exam in the evaluation of schwannoma.24,29,31,32,38 Computed tomography (CT) may also be used for the same purpose.33 At CT, it tends to present as a single, well-circumscribed and solid-appearing mass, located mainly in the course of the IV cranial nerve.32,38 At MRI, schwannomas present with hyposignal in T1, hypersignal in T2 and show an intense enhancement after intravenous administration of gadolinium contrast.38

Schwannomas can be diagnosed with conventional techniques when they are small and located in the cerebellum-pontine cistern. However, when they reach very large sizes and the intracanalicular part is not obvious, it may be difficult to discriminate them from meningiomas and metastases.39 So, the use of advanced imaging techniques may be necessary. Studies with vestibular schwannomas by the cerebellar-angle have shown that, through diffusion-weighted imaging (DWI), it is possible to observe that the schwannomas solid component, usually isointense to brain parenchyma, presents an apparent diffusion coefficient (ADC) that ranges from 1.1 to 1.7 × 10⁻³ mm²/S, which is considered elevated when compared with the normal brain parenchyma (1.4 × 10⁻³–3 mm²/S).40,41 This increased coefficient may reflect the lower cell density of the Antoni B cells of schwannomas. However, the ADC alone cannot differentiate schwannoma from meningiomas. Even though schwannomas present a significantly higher average ADC value, there is considerable overlap among the values, which makes it difficult to differentiate meningiomas from schwannomas.41

Otherwise, it is possible to differentiate schwannomas from meningiomas through magnetic resonance (MR) perfusion. The schwannomas cerebral blood volume (CBV) is significantly smaller when compared with meningiomas.42 Even if there is an overlap in the reason between the CBV of both entities, a threshold of 4.4 is the highest found in the schwannomas, while the meningiomas CBV average ranges from 6 to 9.42–44

Another way to differentiate meningiomas from schwannomas is through MR proton spectroscopy, where the schwannomas show a myo-inositol peak of 3.55 pm and absence of alanine.45 Meningiomas are characterized by a high choline signal with very low signs of creatine and N-acetylaspartate, and alanine presence.46

The trochlear nerve schwannomas are classified in three subtypes: cisternal, cisternocavernous and cavernous.13 In our review, 34 of the 37 cases evaluated were cisternal (91.8%), two cases (5.4%) were cisternocavernous and one case (2.7%) was cavernous.

Contrary to what can be found in the literature regarding trochlear schwannomas centered in the cisternal portion, the present case reports that the trochlear schwannoma was found with epicenter in the pineal gland. In our review, this only occurred in one other occasion, generating a second reported case with this approach for the referred pathology. The
Table 1: Literary review of trochlear nerve schwannomas.

<table>
<thead>
<tr>
<th>Reference</th>
<th>Age/Sex</th>
<th>4th cranial nerve palsy</th>
<th>Location</th>
<th>Surgery approach</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>King⁴</td>
<td>55/F</td>
<td>No</td>
<td>Ambient Cistern</td>
<td>Subtemporal transtentorial to the right</td>
<td>Appearance of fourth cranial nerve palsy on the right side and paresis in the right leg</td>
</tr>
<tr>
<td>Boggan⁵</td>
<td>32/F</td>
<td>Yes</td>
<td>Ambient Cistern</td>
<td>Subtemporal transtentorial craniotomy to the right</td>
<td>Persistence of fourth cranial nerve palsy on the right side and absence of corneal reflex on the right side after 8 months.</td>
</tr>
<tr>
<td>Leunda⁶</td>
<td>54/M</td>
<td>Yes</td>
<td>Ambient Cistern</td>
<td>Subtemporal transtentorial craniotomy to the right</td>
<td>Persistence of fourth cranial nerve palsy on the right side after 1 year.</td>
</tr>
<tr>
<td>Leunda⁶</td>
<td>16/F</td>
<td>Yes</td>
<td>Cisternocavernous</td>
<td>Subtemporal transtentorial craniotomy to the left</td>
<td>Persistence of fourth cranial nerve palsy on the left side after 6 months.</td>
</tr>
<tr>
<td>Yamamoto⁷</td>
<td>37/F</td>
<td>Yes</td>
<td>Ambient Cistern</td>
<td>Subtemporal transtentorial craniotomy to the right</td>
<td>Persistence of fourth cranial nerve palsy on the right side after 1 year.</td>
</tr>
<tr>
<td>Garen⁸</td>
<td>18/F</td>
<td>No</td>
<td>Ambient Cistern</td>
<td>Subtemporal craniotomy to the left</td>
<td>Improvement of third cranial nerve palsy.</td>
</tr>
<tr>
<td>Tokuriki⁹</td>
<td>43/M</td>
<td>No</td>
<td>Ambient Cistern</td>
<td>Temporal craniotomy to the right</td>
<td>Appearance of fourth cranial nerve palsy on the right side after 1 month.</td>
</tr>
<tr>
<td>Murakawa¹⁰</td>
<td>23/F</td>
<td>No</td>
<td>Ambient Cistern</td>
<td>Pterional and suboccipital</td>
<td>Appearance of fourth cranial nerve palsy on the right side.</td>
</tr>
<tr>
<td>Maurice-Williams¹¹</td>
<td>56/M</td>
<td>No</td>
<td>Ambient Cistern</td>
<td>Lateral suboccipital craniotomy to the left</td>
<td>Appearance of fourth cranial nerve palsy on the left side after 2 years.</td>
</tr>
<tr>
<td>Samii¹²</td>
<td>53/F</td>
<td>Yes</td>
<td>Ambient Cistern</td>
<td>Pterional craniotomy to the left</td>
<td>Persistence of fourth cranial nerve palsy on the left side.</td>
</tr>
<tr>
<td>Celli¹³</td>
<td>51/M</td>
<td>Yes</td>
<td>Ambient Cistern</td>
<td>Subtemporal transtentorial craniotomy to the right</td>
<td>Persistence of fourth cranial nerve palsy on the right side after 5 years.</td>
</tr>
<tr>
<td>Lanotte¹⁴</td>
<td>NI</td>
<td>NI</td>
<td>Ambient Cistern</td>
<td>NI</td>
<td>NI</td>
</tr>
<tr>
<td>Jackowski¹⁵</td>
<td>26/F</td>
<td>Yes</td>
<td>Ambient Cistern</td>
<td>Transtemporal craniotomy to the left</td>
<td>Persistence of light fourth cranial nerve palsy on the left side.</td>
</tr>
<tr>
<td>Abe¹⁶</td>
<td>60/M</td>
<td>No</td>
<td>Ambient Cistern</td>
<td>Lateral suboccipital craniotomy</td>
<td>Appearance of paralysis in the IV pair.</td>
</tr>
<tr>
<td>Abe¹⁶</td>
<td>57/M</td>
<td>No</td>
<td>Ambient Cistern</td>
<td>Subtemporal transtentorial craniotomy to the right</td>
<td>Appearance of fourth cranial nerve palsy after 4 months.</td>
</tr>
<tr>
<td>Dolenc and Coscia¹⁷</td>
<td>68/M</td>
<td>No</td>
<td>Ambient Cistern</td>
<td>Lateral suboccipital craniotomy to the right</td>
<td>Appearance of postoperative fourth cranial nerve palsy, but with improvement in 2 months. Improvement of the hemiparesis to the left within 2 months.</td>
</tr>
<tr>
<td>Beppu¹⁸</td>
<td>66/M</td>
<td>No</td>
<td>Ambient Cistern</td>
<td>Lateral suboccipital craniotomy to the right</td>
<td>Appearance of paralysis of the abducent nerve.</td>
</tr>
<tr>
<td>Santoreneos¹⁹</td>
<td>35/F</td>
<td>No</td>
<td>Ambient Cistern</td>
<td>Subtemporal transtentorial craniotomy</td>
<td>Appearance of fourth cranial nerve palsy on the left side.</td>
</tr>
<tr>
<td>Nadkami and Goel²⁰</td>
<td>48/F</td>
<td>No</td>
<td>Ambient Cistern</td>
<td>Subtemporal transtentorial craniotomy</td>
<td>Superior oblique muscle weakness</td>
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</table>

(Continued)
<table>
<thead>
<tr>
<th>Reference</th>
<th>Age/Sex</th>
<th>4th cranial nerve palsy</th>
<th>Location</th>
<th>Surgery approach</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Matsui21</td>
<td>61/M</td>
<td>Yes</td>
<td>Ambient Cistern</td>
<td>Presigmoid transpetrosal craniotomy to the right</td>
<td>Persistence of fourth cranial nerve palsy on the right side.</td>
</tr>
<tr>
<td>Veshchev22</td>
<td>26/F</td>
<td>No</td>
<td>Cavernous Sinus</td>
<td>Pterional craniotomy to the left</td>
<td>Appearance of postoperative fourth cranial nerve palsy on the left side, with significant improvement in 4 months.</td>
</tr>
<tr>
<td>Türe23</td>
<td>31/M</td>
<td>Yes</td>
<td>Pineal Region</td>
<td>Infratentorial - Lateral supracerebellar</td>
<td>Persistence of fourth cranial nerve palsy on the left side.</td>
</tr>
<tr>
<td>Shenouda24</td>
<td>49/M</td>
<td>Yes</td>
<td>Cisternocavernous</td>
<td>Presigmoid combined transpetrosal craniotomy</td>
<td>Persistence of fourth cranial nerve palsy on the left side and deafness.</td>
</tr>
<tr>
<td>Du2</td>
<td>17/F</td>
<td>No</td>
<td>Ambient Cistern</td>
<td>Orbitozygomatic pterional</td>
<td>Asymptomatic</td>
</tr>
<tr>
<td>Shenoy and Raja25</td>
<td>54/F</td>
<td>No</td>
<td>Ambient Cistern</td>
<td>Subtemporal transtentorial</td>
<td>Appearance of hemianesthesia to the left after 1 year.</td>
</tr>
<tr>
<td>Ohba26</td>
<td>48/M</td>
<td>Yes</td>
<td>Ambient Cistern</td>
<td>Anterior transpetrosal</td>
<td>Permanence of the fourth cranial nerve palsy on the right side after 4 months.</td>
</tr>
<tr>
<td>Gerganov27</td>
<td>52/F</td>
<td>Yes</td>
<td>Ambient Cistern</td>
<td>Suboccipital</td>
<td>Permanence of dysfunction in the trochlear nerve.</td>
</tr>
<tr>
<td>Grigorian and Korobova28</td>
<td>47/F</td>
<td>NI</td>
<td>Ambient Cistern</td>
<td>Retromastoidal</td>
<td>NI</td>
</tr>
<tr>
<td>Grigorian and Korobova28</td>
<td>44/F</td>
<td>NI</td>
<td>Ambient Cistern</td>
<td>Paramedian subtentorial supracerebellar</td>
<td>NI</td>
</tr>
<tr>
<td>Kohama29</td>
<td>47/F</td>
<td>No</td>
<td>Ambient Cistern</td>
<td>Posterior transpetrosal craniotomy</td>
<td>Appearance of fourth cranial nerve palsy on the left side.</td>
</tr>
<tr>
<td>Bartalena30</td>
<td>50/F</td>
<td>Yes</td>
<td>Ambient Cistern</td>
<td>Subtemporal-transpetrosal</td>
<td>Persistence of diplopia with posterior surgical correction by myectomy.</td>
</tr>
<tr>
<td>Younes31</td>
<td>65/F</td>
<td>Yes</td>
<td>Ambient Cistern</td>
<td>Pterional</td>
<td>Improvement of fourth cranial nerve palsy after 2 years postoperative.</td>
</tr>
<tr>
<td>Boucher and Michael32</td>
<td>64/M</td>
<td>Yes</td>
<td>Ambient Cistern</td>
<td>Anterior transpetrosal</td>
<td>Persistence of fourth cranial nerve palsy on the right side.</td>
</tr>
<tr>
<td>Hataee33</td>
<td>44/M</td>
<td>Yes</td>
<td>Ambient Cistern</td>
<td>Zygomatic transpetrosal</td>
<td>Persistence of fourth cranial nerve palsy on the left side.</td>
</tr>
<tr>
<td>Samadian34</td>
<td>63/M</td>
<td>No</td>
<td>Ambient Cistern</td>
<td>Left retromastoid</td>
<td>Appearance of fourth cranial nerve palsy.</td>
</tr>
<tr>
<td>Chaudhry35</td>
<td>24/F</td>
<td>No</td>
<td>Ambient Cistern</td>
<td>Paracoccpital posterior interhemispheric transtentorial, from the right side</td>
<td>Appearance of postoperative fourth cranial nerve palsy without complete resolution after 6 months. Emergence of ataxia, with almost complete resolution after 6 months.</td>
</tr>
<tr>
<td>Cunha (present study)</td>
<td>29/F</td>
<td>Yes</td>
<td>Pineal Region</td>
<td>Supracerebellar – infratentorial</td>
<td>fourth cranial nerve palsy with 3 months postoperative follow-up.</td>
</tr>
</tbody>
</table>

Abbreviations: F, female; M, male; NI, not informed.
incidence of tumors in the pineal region ranges from 0.4 to 1% of all primary brain tumors, with studies showing a higher percentage of tumors in this region in children and adolescents, with rates ranging from 3 to 8%.\textsuperscript{47,51} As a differential diagnosis for the pineal region, we must consider the tumors divided in four categories: germ cell tumors, pineal parenchyma tumors, pineal gland support tissue tumors (for example, astrocytomas) and tumors originating from nearby structures (thalamic astrocytomas, tumors of Plexus chorioid).\textsuperscript{52}

The most common tumors of the pineal region are the germinative ones, responsible for 40 to 80% of the total number, with germinoma being the main representa-
tive.\textsuperscript{47,53} Tumors of the pineal parenchyma have a prevalence of 11 to 40% of pineal tumors, and can be divided into pineocytes, pineoblastomas, papillary tumors of the pineal region (PPTPs) and pineal parenchymal tumor of intermediate differentiation (PPTID).\textsuperscript{53,54}

Tumors derived from supporting tissues or structures around the pineal gland include astrocytomas, ependymomas, papillomas and carcinomas of the choroid plexus, lymphomas, gangliogliomas, dermoid and epidermoid cysts, meningiomas and metastases.\textsuperscript{55} According to our literature review, our study is the second case of trochlear schwannoma that has its origin in the pineal region.

The therapeutic options for non-vestibular schwannomas include clinical observation through serial image analysis, microsurgery and stereotactic radiosurgery.\textsuperscript{56} Usually, when treating asymptomatic tumors, clinical observation may be a reasonable alternative, especially in elderly patients or those with other comorbidities, since these tumors usually have slow growth.\textsuperscript{56}

According to our literature review, the most used surgical approach in the cases of trochlear schwannoma is the transtemporal subtemporal, performed in 13 of 36 cases, followed by transpetrosal (7 cases), lateral suboccipital (7 cases), peritonal (5 cases), supracerebellar infratentorial (2 cases) supracerebellar submentonial (1 case), and para-occipital transtentorial (1 case) (Table 1). In one case, the surgical approach was not described.\textsuperscript{54} In our case, we used the intratentorial supracerebellar approach, the same one used in the Ture study.\textsuperscript{23} The surgical approach used in the previous case of trochlear schwannoma in the pineal gland was the posterior transtentorial interhemispheric paraoccipital.\textsuperscript{35}

The postoperative of the previously reported cases demonstrated the difficulty and vulnerability in preserving the trochlear nerve during surgery (Table 1). Among the 17 cases that did not present preoperative paralysis, 12 developed a new trochlear paralysis in the postoperative period, and in only 2 cases there were significant improvements in trochlear paralysis in the postoperative follow-up. From the 17 cases in which the patients had IV paresis, in only 1 trochlear function improvement happened, with diplopia improvement between 4 weeks and paralysis improvement after 2 years postoperatively. However, in this same case, the MRI 3 months after surgery showed a small remnant tumor.\textsuperscript{31}

**Conclusion**

Trochlear nerve schwannomas are extremely rare; however, they generally have a good neurosurgical resectability. Our work presents an unusual trochlear nerve schwannoma, corresponding to the wide range of the possible pathological diagnoses when the region of the pineal gland is involved. In addition, our case also contrasts with the majority of the cases reported in the literature, which shows tumor epicenter furthest from nerve trajectory.

**References**

14. Lanotte M, Giordana MT, Forni C, Pagni CA. Schwannoma of the trochlear nerve during surgery (Table 1). Among the 17 cases that did not present preoperative paralysis, 12 developed a new trochlear paralysis in the postoperative period, and in only 2 cases there were significant improvements in trochlear paralysis in the postoperative follow-up. From the 17 cases in which the patients had IV paresis, in only 1 trochlear function improvement happened, with diplopia improvement between 4 weeks and paralysis improvement after 2 years postoperatively. However, in this same case, the MRI 3 months after surgery showed a small remnant tumor.\textsuperscript{31}

**Table 1**

<table>
<thead>
<tr>
<th>Case Report</th>
<th>Literature Review</th>
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</thead>
</table>
| 1 | Russell DS, Rubinstein LJ. Pathology of tumors of the nervous system. 5th ed. Baltimore: Williams and Wilkins; 1989
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28 Grigorian IuA, Korobova AN. [Neurinomas of the trochlear nerve]. Zh Vopr Neirokhir Im N N Burdenko 2008;01:50–52
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