Dalla ricerca

Intraorbital schwannomas: case report and systematic review of the literature through the history in the last 20 years

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Abstract*:* Intraorbital schwannomas account for 1 to 4% of all orbital tumors, they can remain subclinical for years before usually manifesting with diplopia and progressive proptosis. We present a rare case of schwannoma of the right infraorbital nerve. The patient underwent biopsy and gross total resection of the tumor in two different surgical times. At follow-up he experienced complete relief of preoperative excruciating pain. In order to discuss our case, we performed a literature review of the reported cases of intraorbital schwannomas in the last 20 years. 17 cases of sporadic intraorbital schwannoma have been previously reported. Although orbital schwannomas are rare lesions, they should be always included in the differential diagnosis of intraorbital tumors. Excisional surgery is a valid option in symptomatic patients radiological suspicion of schwannoma.

Keywords: orbit; intraorbital, schwannoma, neurinoma.

Schwannomi intraorbitali: case report e revisione sistematica della letteratura.

Riassunto: Gli schwannomi intraorbitali rappresentano dall'1 al 4% di tutti i tumori orbitali, possono rimanere subclinici per anni prima di manifestarsi solitamente con diplopia e proptosi progressiva. Presentiamo un raro caso di schwannoma del nervo infraorbitario destro. Il paziente è stato sottoposto a biopsia e resezione totale macroscopica del tumore in due diversi tempi chirurgici. Al follow-up ha sperimentato un completo sollievo dal dolore lancinante preoperatorio. Per discutere il nostro caso, abbiamo effettuato una revisione della letteratura dei casi segnalati di schwannomi intraorbitali negli ultimi 20 anni. In precedenza sono stati segnalati 17 casi di schwannoma intraorbitale sporadico. Sebbene gli schwannomi orbitali siano lesioni rare, dovrebbero essere sempre inclusi nella diagnosi differenziale dei tumori intraorbitali. La chirurgia escissionale è una valida opzione nei pazienti sintomatici con sospetto radiologico di schwannoma.

Parole chiave: orbita; intraorbitale, schwannoma, neurinoma.

Schwannomas intraorbitarios: reporte de un caso y revisión sistemática de la literatura.

Resumen: Los schwannomas intraorbitarios representan del 1 al 4% de todos los tumores orbitarios, pueden permanecer subclínicos durante años antes de manifestarse habitualmente con diplopía y proptosis progresiva.

Presentamos un caso raro de schwannoma del nervio infraorbitario derecho. El paciente fue sometido a biopsia y resección total macroscópica del tumor en dos tiempos quirúrgicos diferentes. En el seguimiento experimentó un alivio completo del dolor insoportable preoperatorio. Para discutir nuestro caso, realizamos una revisión de la literatura de los casos reportados de schwannomas intraorbitarios en los últimos 20 años. Anteriormente se han informado 17 casos de schwannoma intraorbitario esporádico. Aunque los schwannomas orbitarios son lesiones raras, siempre deben incluirse en el diagnóstico diferencial de los tumores intraorbitarios. La cirugía escisional es una opción válida en pacientes sintomáticos con sospecha radiológica de schwannoma.

Palabras llave: órbita; intraorbitario, schwannoma, neurinoma.

Introduction

Intraorbital schwannomas (IS) are rare lesions, unilateral, account for 1-4% of all orbital tumors (1–3). Sporadic lesions are usually unilateral. IS originate from sensory fibers of the ophthalmic branch of the trigeminal nerve. The supraorbital and the supratrochlear nerves are most commonly affected, the superior orbital compartment is most frequently involved(1,4). More rarely, IS may arise from the infraorbital or ciliary nerves, or alternatively from the optic, oculomotor, trochlear or abducens nerves. These slow-growing tumors often remain subclinical for years before they manifest usually with diplopia and progressive proptosis. Due to their low incidence and variability in site and presentation, preoperative diagnosis is often difficult(5–7). We present a case of a patient operated for a schwannoma of right infraorbital nerve. To further describe the most appropriate management of these rare intraorbital lesions, we performed performing a systematic literature review.

Methods

A systematic review according to PRISMA-P (Preferred Reporting Items for Systematic review and Meta-Analysis Protocols) guidelines was performed (Figure 1). An online database search (Medline/Pubmed) was performed applying the following terms used as free terms, keywords, or MeSH terms: “schwannoma”, “neurinoma”, “orbit” and combining them with AND, OR, NOT operators (Appendix 1). Additional articles were identified by crossing references. All English language papers published over the past 20 years (January 2000 to December 2019) were considered.

The above-mentioned criteria led to identification of 44 articles that were independently screened by three authors. Disagreements were resolved through consensus by discussion. Each paper’s full text was read and critically assessed by three authors. Articles concerning tumors different from intraorbital schwannomas or intracranial schwannomas were excluded. Only papers reporting management of sporadic IS were considered, case reports and series involving patients affected by neurofibromatosis or without histological confirmation of Schwannoma were excluded.

Finally, we identified 15 articles published between 2000 and 2019(8,9,18–22,10–17), that reported

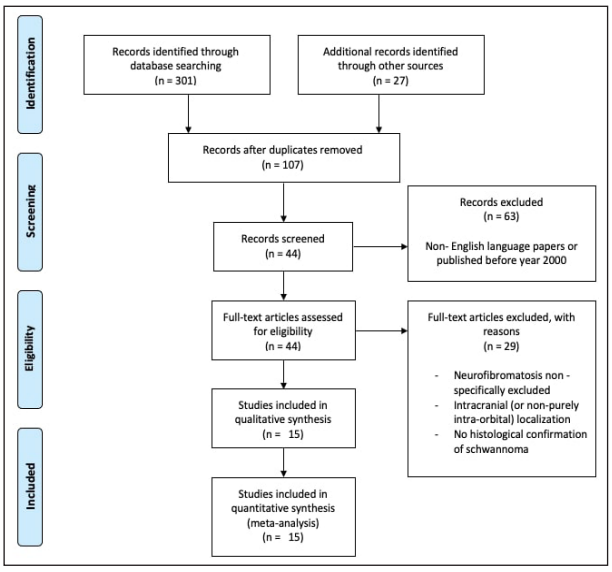

Figure 1. PRISMA flow diagram for systematic literature review.

clinical and surgical data of 16 patients with intraorbital schwannomas. For each publication, we reviewed all data provided by the authors regarding demographic informations, nerve involvement, symptoms and their duration, volumes of the lesions, clinical outcomes and complications.

Results

Case report

A 76-year-old man with Parkinson's disease presented at our outpatients clinic with a 2-years history of intermittent diplopia and stabbing, electric shock-like pain in the right orbital-maxillary area. Brain computed tomography (CT) showed revealed an intraorbital lesion, localized at the posterior third of the right orbital floor. Orbital CT and magnetic resonance imaging (MRI) confirmed the presence of an intraorbital mass displacing the inferior rectus muscle and extending into the maxillary sinus through an erosion of the orbital floor. The mass showed hypodense appearance on T1-weighted images, with inhomogeneous areas on T2-weighted images that enhanced contrast. It fusiform-shaped and measured 15 mm of maximal diameter. Neurological exam at admission revealed diplopia in horizontal and downward gaze, the patient reported visual worsening in the latest weeks. The referred stabbing pain was likely related to direct compression of the first and second right branches of the trigeminal nerve. Since radiological diagnosis was not clear, incisional biopsy was performed, in order to define the nature of the lesion. Intraoperatively, the lesion appeared to be in continuity with the infraorbital nerve. Histological examination allowed diagnosis of schwannoma (WHO grade: I). 1 month post-operatively, an orbital MRI was performed, showing further growth of the lesion. At this point, basing on histological diagnosis and on the progressive growth of the tumor, even if slow-growing in nature, surgical resection was indicated. Through an inferior infra-palpebral approach neurotomy, complete tumor removal and orbital floor reconstruction were performed. Histological examination confirmed diagnosis of schwannoma (Ki-67: 2.88%). The patient experienced complete pain relief but, unfortunately, diplopia did not improve.

Literature review

The literature search retrieved 16 cases of intraorbital schwannoma (Table 1). Mean age at presentation was 50.3 years (standard deviation (SD): ± 16.9, range: 22-76, median: 53). The male-to-female ratio was 0.6. The most frequently involved nerves were the infraorbital nerve (4 cases) and the oculomotor nerve (4 cases), followed by the abducens and optic nerve (3 cases), the supraorbital nerve (2 cases), and the frontal nerve (1 case). The range in largest diameter at presentation was 15-45 mm. The most commonly reported symptoms were proptosis and diplopia; whereas decrease in vision was reported in 5 patients. The duration of symptoms in average was 19,5 months. Unprovoked pain was reported in 22.2 % of patients. All patients underwent gross total resection. Transient surgery-related complications developed in 1 patient. Preoperative symptoms resolved completely in 5 patients and partially in 9.

Discussion

IS are slow-growing benign tumors usually presenting with progressive proptosis, diplopia, and neuralgia. Since symptoms are often non-specific, neuroimaging studies can be helpful to achieve preoperative diagnosis. These lesions frequently show T1-weighted isointense signal and T2-weighted homogeneous, hyperintense signal and are characterized by markedly homogeneous contrast-enhancement, and sometimes bone erosion associated(6,7). Differential diagnosis includes primary or secondary tumor, nerve entrapment, vascular malformations and infections(8, 23, 24). Histological findings are essential for establishing a diagnosis. Surgical management is the only treatment modality described in literature.

In our literature review, regarding treatment, biopsy was performed before complete surgical removal, as in our case, in 2 patients(9,18). Since it is not always possible to spare the nerve, surgery of orbital schwannomas is associated with a high neurological morbidity(24). Involvement of ocular motor nerves warrants less aggressive treatment, with close follow-up or incisional biopsy(18,24). Only few cases of optic nerve schwannomas have been described, and

Table 1. Literature review.

Author (year)	Age, sex	Involved nerve	Symptoms	Duration of symptoms (months)	Dimensions	Post-surgical morbidity	Follow-up
Mahore et al (2019) [8]	22, M	Optic	Proptosis, vision impairment	2	>20 mm	No	Recovery
Mahore et al (2019) [8]	39, M	Optic	Proptosis, vision impairment, diplopia	4	>20 mm	No	Partial recovery
Young et al (2018) [9]	71, M	Medial rectus motor nerve	Proptosis, vision impairment, eye pain	12	45 mm	No	Partial recovery
Mortuza et al (2014) [10]	56, M	Optic	Eye pain	NA	23 × 20 mm	NA	NA
Scheller et al (2013) [11]	42, F	Oculomotor	Fixed dilatated pupil, vision impairment	1	17 x 10 mm	No	Partial recovery
Feichtinger et al (2013) [12]	53, F	Abducens	Proptosis, diplopia	24	45 x 20 mm	No	Partial recovery
Rato et al (2012) [13]	42, M	Abducens	Diplopia	12	>20 mm	No	Recovery
De Jong (2010) [14]	44, M	Supraorbital	Forehead numbness	NA	NA	Forehead anesthesia	NA
Clarençon et al (2009) [15]	45, F	Infraorbital	Proptosis	NA	>20 mm	Cheek hypoesthesia	Recovery
Irace et al (2008) [16]	55, M	Abducens	Proptosis	120	>20 mm	Diplopia	Partial recovery
Garg et al (2008) [17]	35, F	Infraorbital	Proptosis	24	20 x 15 mm	No	Recovery
Colapinto et al (2007) [18]	68, M	Right Inferior Motor Nerve	Diplopia	8	18 x 12 mm	No	Partial recovery
Tezer et al (2006) [19]	16, F	Infraorbital	Eye pain, proptosis	6	30 x 40 mm	Cheek hypoesthesia	Recovery
Barbagallo et al (2004) [20]	65, M	Supraorbital	Proptosis	24	NA	NA	NA
Tokugawa et al (2003) [21]	64, F	Frontal	Proptosis, forehead numbness	6	30 x 25 mm	No	Partial recovery
Tsuzuki et al (2000) [22]	62, F	Oculomotor	Proptosis, vision impairment, diplopia	6	>20 mm	No	Recovery
Present case	76, M	Infraorbital	Diplopia, facial pain	24	10 x 15 mm	No	Partial recovery

M: Male; F: Female; NA: Not Available.

their management, as before-mentioned, is associated with high morbidity. The present systematic review identified 3 cases of histologically-confirmed optic nerve schwannoma. Surgery was universally identified as the first-line treatment, even though surgical management is challenging because not only aims at complete resection of the tumor but also aims at optic nerve functional preservation. Conversely, in cases of sensory nerve involvement the nerve, it may be acceptable to sacrifice the nerve in order to achieve oncological completeness.

The surgical approach should be chosen on the basis of tumor location in respect to the optic nerve. Superior orbitotomy is therefore indicated for lesions localized superiorly to the optic nerve, whereas, considering tumor's location, inferior, lateral or medial orbitotomy should be respectively considered, in order to minimize manipulation and mobilization of the nerve. The use of bipolar forceps should be minimized to what is strictly necessary, in order to avoid thermal injury to the nerves. Adjuvant radiotherapy or stereotactic radiosurgery may be considered for residual

tumor in close relationship with the nerves. In these cases, further surgical resection may be considered too hazardous(8).

The reported literature reports low morbidity rates after surgery, that may be explained by the most frequent involvement of sensory nerves, which may be more easily sacrificed than motor nerves. In only one case diplopia was present also postoperatively, even if improved(16). Neurological symptoms, including diplopia, completely recovered in the remaining cases without additional postsurgical deficits(13,22,25).

Conclusions

Orbital schwannoma should be included in the differential diagnosis of intraorbital tumors. Variability in tumor site and clinical presentation of peripheral nerve lesions of the orbit, associated with non-specific findings on imaging and potential surgical relapse, constitute important variables that surgeons must take into account when planning the removal of an intraconal nerve lesion. Excisional surgery is a valid option in symptomatic patients with radiological suspicion of schwannoma, given the low postoperative morbidity rate and the optimal diagnosis, control of symptoms, and long-term control of disease.

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