Atypical Tumours of the Intratemporal Facial Nerve: A Review

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Abstract: *Objectives*: To describe the characteristic of tumours that affect the facial nerve and discuss the most prevalent signs and symptoms, clinical presentations, diagnostic features and therapeutic approach to such tumours.

Study Selection: Studies published between 2002 and 2013, referring to atypical tumours of the facial nerve. We excluded studies that contained only patients with schwannoma of the facial nerve, as well as those that did not report the number of patients treated.

Data Synthesis: 22 studies reported 99 patients with tumours of the facial nerve. The most prevalent symptoms were facial palsy (99%), tinnitus (12.1%), and ear pain (6.1%). Surgical excision of the tumour was performed in 99% of patients, and the most common approach was *via* the middle cranial fossa (30.3%). The degree of facial palsy after resection of the tumour remained unchanged in 23.2% of patients; 19.2% improved and 2% deteriorated.

Conclusions: The cardinal symptom for the diagnosis of tumours of the facial nerve was facial palsy. Treatment involves surgical removal of the tumour with or without the use of grafts for reconstruction of the facial nerve. Other adjuvant therapies such as radiosurgery and facial nerve decompression are not yet established and require further research.

Keywords: Tumours, facial nerve, schwannoma, haemangiomas, neurofibromas, paragangliomas.

INTRODUCTION

Primary tumours of the facial nerve are rare lesions. Most of these tumours (91%) are located in the temporal bone and only a few in the parotid gland [1]. They can arise from Schwann cells, fibroblasts (from endoneurial cells) and epithelial perineurium blood vessels [2]. Schwannomas are the most common tumour, although other lesions such as haemangiomas, neurofibromas, meningioma, dermoid cysts, glomus tumours and adenocarcinomas may affect the temporal bone [3].

Tumors of the facial nerve are a challenge for even the most experienced doctors. According to Salazat *et al*. [3], this type of injury tests the skills of the physician and requires sophisticated technology for diagnosis. Moreover, once detected, treatment is complex and multidisciplinary.

The aim of this study was to highlight the diversity of tumours affecting the facial nerve and discuss the signs and symptoms, clinical presentations, diagnostic and therapeutic approach to these tumours.

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METHODS

We performed a systematic review of three independent electronic databases (PubMed, SciELO and Cochrane Review) from March to June 2013, looking for studies about atypical tumours of the intratemporal portion of the facial nerve, published between the years 2002 and 2013. We included studies of patients of both sexes, with no restrictions on age, written in English, Portuguese or Spanish. The keywords used were "tumours" AND "intratemporal" AND "facial nerve" AND ("haemangioma" OR "meningioma" OR "paraganglioma" OR "neurofibroma") AND "facial paralysis. We excluded studies on exclusive schwannoma of the facial nerve, as well as those that did not report the number of patients treated.

RESULTS

The search identified 77 articles, and after reading the abstracts, 22 articles were selected for appraisal (Tables **1** to **3**). The selected studies report the cases of 99 patients with atypical facial nerve tumours. Forty-one (41.4%) patients were women and 51 (51.5%) were men. The gender of seven (7.1%) patients was not reported. The average age of the patients at the time of diagnosis was 45.7 years; the youngest patient was diagnosed at 9 months and the oldest at 70 years.

The time elapsed between first symptoms and the diagnosis of the tumour ranged from 3 months to 8

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Table 1: Results of the Database Search for Articles

Tumour	Number of articles	Number and percentage of patients
Haemangioma	05	27 (27.3%)
Neuroma and haemangioma	01	-*
Primary tumours of the facial nerve	03	-*
Neurofibromas	03	21 (21.2%)
Paragangliomas	03	4 (4%)
Traumatic neuroma	02	2 (2%)
Dermoid cyst	01	1 (1%)
Carcinoma	01	1 (1%)
Chondromyxoid fibroma	01	1 (1%)
Meningioma	01	4 (4%)
Atypical tumours	01	-*

*Tumours already classified in other categories.

years. Regarding the histology of tumours, 27 (27.3%) cases were haemangiomas, 21 (21.2%) neurofibromas, 4 (4%) meningiomas, 4 (4%) paragangliomas, 2 (2%) traumatic neuromas and 1 (1%) dermoid cyst, 1 (1%) a mixture of haemangioma and schwannoma, 1 (1%) chondromyxoid fibroma and 1 (1%) metastatic carcinoma.

Ninety-eight out of the 99 patients (99%) had peripheral facial paralysis at the time of diagnosis, and, according to the grading scale of House-Brackmann, grade VI was most frequent (27.3%). Twelve patients (15.3%) had facial paralysis grade I or II, 16.2% grade III, 9.1% grade IV and 15.1% grade V. In 20.2% of patients, the HB grade was not reported. Other less frequent symptoms were: tinnitus (14.1%), pain on the pinna (6.1%), dizziness (5%), unilateral headache (2%), epiphora (1%) and otorrhagia (1%).

Surgical treatment was performed in 99% of patients. In one case (a patient with haemangioma) the treatment was not reported. A complete tumour resection was performed in 97 patients (98%) and partial resection in one case (1%). Radiotherapy associated with complete resection of the tumour was performed in 1% of the patients (one case of metastatic carcinoma).

The most common surgical approach was *via* the middle cranial fossa (30.3%), followed by the transmastoid (26.3%). In 23.2% of patients a combined approach was necessary (transtemporal craniotomy with mastoidectomy, middle fossa and transmastoid; transmastoid and cervical). The translabyrinthic route was used in 7% of cases, followed by mastoidectomy (4%), retrosigmoid (3%) and transtemporal craniotomy (2%). In 4% of patients, the type of approach used was not reported.

Grafts for reconstruction of the facial nerve were used in 45.4% of patients. The auricular nerve was used in 18 (18.2%) patients and the sural nerve in three (3%) patients. In 24.2% of patients the type of graft used was not specified and some authors (1,2,7,11-17,20-23) did not report whether they used grafts or not.

The pre- or postoperative degree of facial paralysis according to the HB scale was not reported in 55.5% of patients. In 23.2% of cases the degree of facial paralysis did not improve in comparison with the preoperative grade, while 19.2% achieved some degree of improvement and 2% deteriorated.

DISCUSSION

Schwannomas are the most common tumour found in the facial nerve [3]. Although they usually affect the labyrinthine and tympanic portions, they may affect any segment of the nerve [1]. Due to the high incidence of schwannomas of the facial nerve in the literature compared to other kinds of tumours (classified as atypical by the literature), we chose to remove from this review articles containing only cases of schwannomas.

According to Ross *et al*. [3], haemangiomas are tumours that most commonly occur in people between the third and sixth decades of life and have a predilection for the geniculate ganglion. In this review,

Table 2: Summary of Characteristics of Patients and Atypical Tumours of Intratemporal Facial Nerve

Author / Year	Patients	Age/ Sex	Tumour	Signs/Symptoms	ENT examination	Time to diagnosis	Portion of the facial nerve affected	Treatment	Lesion appearance	Histopathology	Change in House-Brackmann postoperatively
Collin *et al.*, [5] 2013	01	48 y/F	Meningioma	FP progressive; R vestibular areflexia	HB III, blepharospasm; tearing	18 m	GG	Surgical resection by MF	Comp. com Schwannoma	Tu with calcifications spiral cells and pseudo inclusion IN	NI
Dai *et al.*, [12] 2013	11	25.5 ± 4.4 y / 36.4% F and 63.6% M	Neurofibroma	FP progressive (54.5%); sudden (45.5%); otalgia (36.4%); vertigo (9%); mild conductive HL (27.3%);	HB IV (9%); HB V (18.2%); HB VI (72.7%)	NI	MP 27.3%; PS 9%, Only GG 9% and 2 or +segm. in 54.5%	By TM	NI	Plexiform pattern or wheel fusiform cells elongated core and varying amounts of collagen S100 is usually positive.	0 (63.6%); - 2 (27.3%); - 4 (9%);
Ross *et al.*, [10] 2013	04	X=53.7 y; / 50% F, 50% M	Paraganglioma, schwannoma, meningioma, haemangioma	FP (75%), F (25%), vertigo (50%)	HB III in paraganglioma; HB VI in schwannoma; HB I-II in haemangioma	21 m average	VP (50%) - paraganglioma and schwannoma; only GG (25%) - meningioma; GG and LP (25%) - haemangioma;	Surgical resection: TT craniotomy with mastoidectomy and decompression of FN in paraganglioma; TT craniotomy in schwannoma; craniotomy by MF and TM in meningioma; craniotomy by MF in haemangioma	NI	NI	0 (50%); -5 (25%)
Takahashi *et al.*, [20] 2013	01	23 y/M	Paraganglioma	FP progressive, R mixed HL	IAC red mass nonpulsatile ;HB VI	1 y 8 m	FC	Surgery (by TM)	Granular tumour with significant bleeding	Paraganglioma	NI
Kunzel *et al.*, [6] 2012	01	39 y/M	Paraganglioma	FP progressive	HB VI	NI	FC	Total surgical resection	Bright red tumour	Medium-sized tumor cells with cytoplasm surrounded by vessels	PO =HB II
Nwojo *et al.*, [7] 2011	01	9 y/M	Dermoid cyst	FP progressive	HB VI	3 m	2^0 K and VP	Surgery = mastoidectomy with decompression and monitoring of FN	Fleshy portion extending out to the facial recess and Tm; scaly debris with hair in the posterior and medial FC	Dermoid cyst ruptured	Changes in HB: PO = HB III

(Table 2). Continued.

Author / Year	Patients	Age/ Sex	Tumour	Signs/Symptoms	ENT examination	Time to diagnosis	Portion of the facial nerve affected	Treatment	Lesion appearance	Histopathology	Change in House-Brackmann postoperatively
Benoit et al., [15] 2010	07	X=45 y/NI	Haemangioma	FP (100%), vertigo (14.3%), facial spasms (14.3%), HL (28.57%)	NI	10 m (14.3%); 15 m (14.3%), 18 m (14.3%); 3 y (42.8%); 8 y (14.3%)	GG	Surgery by MF	Vascular	Dilated vessels, irregularly shaped blade, SM scarce in all cases, the absence of IEL	0 (28.57%) + 1 (14.28%) - 2 (42.85%) - 3 (14.28%)
Clark et al., [18] 2010	01	16 y/M	Traumatic neuroma	FP progressive, with progressive reduction of the force to close the right eye	HB III	2 y	FP	Surgery by FM	NI	Dense layer of fusiform cells. Overall architecture and rich content axons compatible with traumatic neuroma	PO = In 1y, excellent facial symmetry at rest and symmetrical smile
Moberly & Fritsch, [13] 2009	01	28 y/F	Neurofibroma	L conductive HL and L COM	L tympanic membrane intact, opaque and retracted; HB I	NI	MP, TP and TC	Surgical resection	Mass of fibrous tissue in the midface of uncus	Benign tumuor of nerve tissue = neurofibroma	PO = HB I
Saliba & Fayad, [11] 2009	01	39 y/F	Haemangioma	R FP; R conductive HL	Otoscopy = bluish-red vascular mass behind the TM that filled the posterosuperior part of the TC right	2 y	2^0knee	Surgery by TM/TT	NI	Haemangioma	-2
Thopson et al., [8] 2009	01	33 y/F	Chondro-myxoid fibroma	FP	NI	22 m	FC-MP	Surgical resection = mastoidectomy	Cystic lesion irregular	Myxoid lesion with mild stromal cells mixed with bone spurs and cartilage – chondromyxoid fibroma	NI
Towfigh et al., [21] 2008	01	28 y/M	Neuroma and haemangioma	L FP progressive	HB VI	12 m	GG	Surgery by TM + MF	Tissue fragments reddish white	Abundant vascular spaces. Bundles of spindle cells with disorganized collagen = compatible with neuroma	NI

(Table 2). Continued.

Author / Year	Patients	Age/ Sex	Tumour	Signs/Symptoms	ENT examination	Time to diagnosis	Portion of the facial nerve affected	Treatment	Lesion appearance	Histopathology	Change in House-Brackmann postoperatively
Liu et al., [2] 2007	22	9 to 70 y/ 41% M and 59% F	14 neurinomas 6 neurofibromas 2 haemangiomas	Disf. of FN (63.6%); HL (45.4%); tinnitus (18.2%); EF (13.6%); FP progressive (81.8%); FP sudden (1 neuroma)	HB III (18.2%); IV (18.2%); V (27.3%) and VI (18.2%); swollen mass in IAC (22.7%)	NI	CPA (13.6%); LP+GG (18.2%); HP (4.5%); HP-VP (31.8%); VP (18.2%); VP-EC (9.1%); 2°j-PP (9.1%)	RS (9.1%); MF (13.6%); MF+TM (22.7%); TM (45.45%); TM and cervical (9.1%)	NI	NI	NI
Saliba & Fayad, 2009	Um	39,	Hemangioma	PF à D DA condutiva leve à D	Oto = massa vascular vermelho-azulada atrás da MT enchendo a parte PS da CT D						
Myashita et al., [19] 2007	01	47 y/ M	Haemangioma	L progressive FP L moderate SN H	L FP HBVI	14m	GG	Surgery by MF	Dark red mass	Haemangioma cavernous type	-1
Hopkins et al., [17] 2007	01	60 y/M	Haemangioma	R FP and facial spasms; chronic infection of the middle ear	R FP HB II	1y	Vertical	NI	NI	Intratumoral ossification and irregular projections	NI
Rainsbury et al., [16] 2007	01	43 y/ F	Traumatic neuroma	FP; vertigo; conductive HL	HB III	NI	2°knee	Surgery = mastoidectomy	Firm tumour, gray-white, non-encapsulated	Interrupted axons with demyelination and Wallerian degeneration distal, a tangle of Schwann cells and fibroblasts, all in a dense matrix of collagen	HB remained III
McMonagle et al., [22] 2006	01	27 y/F	Neurofibroma	FP	HB V	NI	LP and TP	Surgical resection by TL	NI	Proliferation of cells. Spindle corrugated IN; axons were preexisting +, distributed evenly throughout the typical neurofibroma	NI
Isaacson et al., [14] 2005	06	X=38.7 y/ 83.3% M and 16.7% F	Hemangioma	FP (100%); SN HL (33.3%); mixed HL (16.7%); unilateral tinnitus (16.7%); unilateral headache (16.7%); epiphora (16.7%); blepharospasm (16.7%).	HB III (33.33%); V (50%) and VI (16.67%).	2 y (66.7%), 3 y (16.7%), 11 y (16.7%)	GG (1= GG+ LP+ TP and 1 = GG +TP+ AT)	Surgery (83.3% MF and 16.7% MF and TM)	Purple vascular mass (33.3%); with bone fragments (16.7%); vasc. (33.3%) and NI in 16.7%	Haemangioma within the fascicles of the facial nerve (50%), cavernous haemangioma (33.3%) and not described in 16.7%	0 (16.67%); +1 (16.67%); -2 (33.33%0; -1(33.33%)

(Table 2). Continued.

Author / Year	Patients	Age/ Sex	Tumour	Signs/Symptoms	ENT examination	Time to diagnosis	Portion of the facial nerve affected	Treatment	Lesion appearance	Histopathology	Change in House-Brackmann postoperatively
Suryanarayanan et al., [9] 2005	01	64 y/F	Metastatic carcinoma of breast	Pulsatile tinnitus; imbalance; FP progressive; otorrhagia; L invasive lobular carcinoma of the breast 12 years ago; anacusis	HB V	NI	LP until GG and TP until 2^0 knee	Surgery by TL; 8 Rt sessions followed by hormonal therapy	NI	Lobular carcinoma metastatic breast	PO = HB IV
Salazar et al., [3] 2004	'06	16–46 y/ 50% F and 50% M	66.7% schwannoma and 33.3% haemangioma	FP (100%); SN HL (33.3%); anacusia (33.3%); tinnitus (50%); otalgia (33.3%) e headache (16.7%)	NI	3–7 y (average 4.5 y)	2^0k 50%; GG (33.3%) and IC (16.7%)	RS (16.7%) and MF (83.3%)	NI	NI	HB PO = III in 33.3% and IV in 66.7%
Wippold et al., [23] 2004	01	37 y/F	Paraganglioma	FP	IAC mass	NI	FC	Surgery	Soft tissue mass that violated the posterior wall of the TC and IAT	NI	NI
Falcione et al., [4] 2003	28	X= 40.3 y/ 64.3% M and 35.7% F	64.3% schwannoma, 21.4% haemangioma, 7.1% meningioma and 7.1% neurofibroma	FP (100%); HL (46.4%), conductive (53.8%); SN (23.1%) and anacusia (23.1%); tinnitus (17.8%); hemifacial spasm (14.3%)	HB I (17.8%); II (10.7%); III (21.4%); IV (14.3%); V (7.1%) and VI (28.6%); polyp occluding IAC (10.7%) and retrotympanic mass (21.4%)	X=31.2 m	CPA 10.7%; IAC 25%; GG 75%; 2k 46.4%; ET 10.7 %; 82.1% +1 seg	RT 96.4% and partial in 3.4% MF 25%; MF + TM 21.4%; TM 14.3%; TL 17.8%; TC 10.7%	NI	Haemangioma = bone spurs intratumour	HB PO: I (7.1%); III (28.6%); IV (32.1%); V (10.7%), VI (10.7%) and fora (10.7%)

Legend. X (average); y (years); m (months); M (male); F (female); HB (House-Brackman); change in the House-Brackmann postoperatively: 0 = no change; + = worsening of the House Brackman and - = improvement in House Brackman; FP (facial paralysis); R (right); L (left); HL (hearing loss); F (fullness); SN (sensorineural); COM (chronic otitis media); EAC (external auditory canal); NI (not reported); GG (geniculate ganglion); CPA (cerebellopontine angle); MP (mastoid portion); PS (pyramidal segment); LP (labyrinthine portion); FC (fallopian canal); VP (vertical portion); PP (parotid portion); TP (tympanic portion); HT (hearing tube); MF (middle fossa); TM (transmastoid); TT (trans temporal); RS (retrosigmoid); TM (tympanic membrane); IN (intra neural); SM (smooth muscle); IEL (internal elastic lamina); PO (postoperative); Rt (radiotherapy); vasc. (vascular); IAM (internal auditory meatus); IC (intra cranial); ET (extra temporal).

Table 3: Authors who used Intraoperative Graft for Reconstruction of the Facial Nerve

Author / Year	N° Pat	Type of tumour + Location in NF	Type of graft
Collin *et al.*, [5] 2013	01	Meningioma, GG	Auricular nerve
Dai *et al.*, [12] 2013	11	Neurofibroma, MP 27.3%; PS 9%, GG 9% and two or more segments in 54.5%	Four, did not specify where
Kunzel *et al.*, [6] 2012	01	Paraganglioma, FC	Auricular nerve
Nwojo *et al.*, [7] 2011	01	Dermoid cyst	Sural nerve
Saliba & Fayad, [11] 2009	01	Haemangioma, 2^0 knee	Sural nerve
Thompson *et al.*, [8] 2009	01	Chondromyxoid fibroma	Sural nerve
Liu *et al.*, [2] 2007	22	14 neurinomas, 6 neurofibromas, 2 haemangiomas; CPA; LP; GG; MP; 2^0 K-PP	Auricular nerve in ten cases
Isaacson *et al.*, [14] 2005	06	Haemangioma, GG	Auricular nerve
Falcione *et al.*, [4] 2003	28	64.3% schwannoma, 21.4% haemangioma, 7.1% meningioma, 7.1% neurofibroma	In 24 cases, did not specify where
		CPA10.7%; IAM 25%; GG 75%; 2K 46.4%; ET 10.7% (82.1% +1)	

Legend. GG (geniculate ganglion); CPA (cerebellopontine angle); IAM (internal auditory meatus); 2K (second knee); ET (extra temporal); LP (labyrinthine portion); MP (mastoid portion); PP (parotid portion); FC (fallopian canal); PS (pyramid segment).

the geniculate ganglion was affected in 48.1% of patients and the mean age at the time of diagnosis was 44 years. The literature reports no differences in distribution according to gender [3].

Falcioni *et al*. [4] considered intracranial meningiomas as frequent injuries, being the second most common tumour in the cerebellopontine angle; however, they are infrequent in the geniculate ganglion, with only four cases described in the literature [5, 4, 3]. Although the aetiology is not clear, associations with progesterone, breast cancer and radiation therapy have been described [3]. It has been suggested that they originate in the arachnoid villi along the acoustic porus [3]. According to Colin *et al*. [3], the mean age at presentation is 25 years, ranging between 5 and 56 years, information that contrasts with that found in the present review, in which the average age of patients was 47.3 years. The same authors observed a predominance of such tumours in females [5].

Paragangliomas of the head and neck areas are highly vascularized, and in most cases turn out to be benign. They mostly originate in paraganglionic tissue in the area of the carotid bifurcation, and less frequent sites of origin are the jugular foramen, the vagus nerve and the tympanic plexus. They represent 0.6% of all tumours of the head and neck [6]. Paraganglioma of the facial nerve is a rare condition (4% of the tumours in this review), but should be considered as a differential diagnosis of peripheral facial paralysis caused by tumours.

Dermoid cysts are rare tumours (only one case described in the literature, equivalent to 1% of cases included in this review) that are derived from a variety of types of parenchyma cells (representative of all three germ cell layers) [7]; they are cystic tumours that contain skin tissue, hair, dental structures and sebaceous glands. The chondromyxoid fibroma is a rare benign tumour characterized by a lesion of cartilaginous origin. Thompson *et al*. [8] described four cases located in the mastoid portion of the temporal bone. The patients had facial paralysis associated with hearing loss and headache. The authors reported a slight predilection for males and a peak incidence in the second and third decades of life.

Suryanarayanan *et al*. [9] reported the case of one patient with metastatic breast carcinoma in the intratemporal portion of the facial nerve. The authors found that the primary tumour sites, in descending order of frequency, were: breast, lung, kidney, prostate, stomach, and thyroid. Their patient had progressive facial paralysis, classified as HB V at diagnosis; she had a personal history of breast infiltrating lobular

carcinoma 12 years previously, treated with radiotherapy, mastectomy and tamoxifen. The metastasis was removed *via* translabyrinthic surgery, and additional treatment with radiation therapy and hormone therapy was performed. The degree of facial palsy after treatment was HB IV. These tumours can cause erosion of the temporal bone and their clinical presentation depends on the extent of the destruction. Hearing loss, tinnitus, dizziness, instability and progressive facial paralysis may occur as unique or combined symptoms [9]. Metastatic lesions can also be diagnosed years after treatment and cure of the primary tumour [9].

Some studies [10, 11] compared the evolution of idiopathic facial paralysis (Bell's palsy) and paralysis secondary to tumours of the facial nerve. Most (85%) patients with Bell's palsy recovered from facial paralysis to HB grade I or II in approximately 8–12 weeks, while patients with facial nerve tumours experience a more indolent course, with slower recovery of facial motor function or even permanent sequelae.

Primary tumours of the facial nerve can be located in any portion of the facial nerve, and tumours that involve more than one segment of the nerve are the most common [3]. In this review, the tumours were located in more than one portion of the nerve in 53.5% of patients, while 46.5% involved only one segment.

Approximately 97% of patients with atypical facial nerve tumours present with some degree of facial paralysis [12]. Morbely and Fritch [13] described a case of neurofibroma in the facial nerve that did not cause facial paralysis. The tumour extended along the chorda tympani nerve and fused to the facial nerve superiorly, preserving the facial recess and other portions of the nerve. However, ten cases of neurofibroma described by Dai *et al*. [12] showed progressive facial paralysis (54.5%), 45.5% with sudden facial palsy, ear pain (36.4%), dizziness (9%) and mild conductive hearing loss (27.3%).

Falcione *et al*. [4] and Isaacson *et al*. [14] reported that hearing loss is a common symptom in different kinds of tumours of the facial nerve. This could be conductive when the horizontal segment of the facial nerve is involved, or sensorineural when it affects the labyrinthine or geniculate ganglion portions.

Histologically, plexiform neurofibromas present in a fusiform and elongated shape with different amounts of collagen, which are separated by small amounts of mucoid material without a capsule [12]. Schwannomas also have a well-defined capsule [12]. Haemangiomas could be considered as solid tumours or vascular malformations. They have dilated vessels in an unusual formation and very few muscle fibres, associated with no internal elastic lamina [15]; these tumours also contain fragments of bone [4]. Similarly to meningiomas, these tumours appear as calcifications with sparse cells and spirals with pseudo intranuclear inclusion [5].

Traumatic neuromas (2% of the tumours in this review) are not true neoplasms, since they are a hyperplastic response to nerve injury, such as direct or indirect trauma, or even chronic inflammation. They are firm, have a gray-white colouration, are not encapsuled and present as a single lesion [16]. In contrast, schwannomas are encapsulated and may be multiple, though they often present individually [16].

The definitive diagnosis of tumour of the facial nerve can only be made intraoperatively by histopathological examination of tissue. However, magnetic resonance imaging (MRI) and computed tomography (CT) are useful to assess tumour location, evaluate its characteristics and plan the approach. Table **4** describes the main differences between radiological schwannomas, meningiomas and haemangiomas of the geniculate ganglion. Electromyography was used in 27.3% of the studies in this review [6-22], and even though it does not identify tumours, it is a useful tool for evaluating the degree of nerve dysfunction.

Haemangiomas in the temporal bone appear on CT as a soft tissue mass with bone erosion. Intratumoural bone spurs are responsible for their characteristic honeycomb appearance [17]. On MRI, they are typically bright on T2 and impregnate with gadolinium. Their intensity on T1 can be variable [17]. Compared with schwannomas, haemangiomas tend to have well-defined margins, a more fusiform configuration and an increased signal intensity [17].

In MRI, paragangliomas usually appear as a hyperintense signal on T2 and without contrast catchment in T1. The "salt-and-pepper" appearance on T1-weighted images is a typical diagnostic sign [6]. However, digital subtraction angiography is not only considered the gold standard for the diagnosis of paraganglioma, but also allows preoperative embolization [6].

Table 4: Main Differences between Radiological Schwannomas, Meningiomas and Hemangiomas of the Geniculate Ganglion

Imaging modality		Meningioma	Schwannoma	Hemangioma
High-resolution CT Scan		Bony marginspoorly defined contrast enhancement ++	Sharp margins larger than meningiomas and hemangionas	Intratumoral bone spicules 'Salt and pepper aspect
MRI	T1	Hypo-to-isointense	Hypo-to-isointense 1st and 2nd portion of the facial nerveinvolved Iso-to-hypertense	Isointense
	T2	Iso-to-hyperintense	Iso-to- hyperintense	Isointense
	Gadolinium enhancement	+	++	Heterogeneous

The diagnosis of atypical tumours of the facial nerve is usually made long after the first symptoms. A review of studies showed an average of 2 years from onset of the symptoms to diagnosis [5, 4, 7, 8, 10, 11, 20, 17, 18, 19, 20, 21]. According to Falcione *et al*. [6], the likelihood of recovering satisfactory function of the facial nerve decreases if surgery is not performed in the first year after the onset of clinical deficit. In this review, the degree of facial paralysis remained equal to the preoperative index in 23.2% of patients, 19.2% showed some improvement and 2% deteriorated. However, in 55.5% of patients there was no description of the evolution of facial paralysis with treatment.

Many authors did not report the use of grafts or their type [3, 5, 7, 8, 10, 13, 16-23]. Among the patients who received graft reconstruction of the facial nerve, a graft from the greater auricular nerve was the most commonly used (18.2% of cases). According to Dai *et al*. [12], the great auricular nerve is most commonly used because it has a diameter similar to that of the facial nerve. When a longer graft is needed, the sural nerve or the lateral femoral cutaneous nerve are good choices, even though their diameter is smaller than that of the facial nerve.

Approximately 82% of patients with facial nerve neuromas that underwent grafting because of injury to the facial nerve recovered to HB III in 10.1 ± 1.3 years of follow-up after surgery (range 2–19 years) [12]. Dai *et al*. [12] reported 11 patients with neurofibromas of the temporal bone with facial paralysis HB IV who underwent surgical excision of the tumour. Four of the 11 individuals (36.36%) required grafting to repair the surgical lesion of the facial nerve. Among the seven patients who did not require grafts, four (57.1%) showed improvement, although only one of those who underwent grafting (25%) achieved improvements in the motor function of the nerve, while in the other three (75%) the degree of facial paralysis worsened. This study concluded that patients with facial nerve integrity preserved intraoperatively achieved better recovery of facial motor function postoperatively than patients who required grafts.

CONCLUSION

The average time between onset of symptoms and diagnosis of the tumour in the facial nerve was 2 years. More than 25% of the tumours were haemangiomas. The cardinal symptom for diagnosis was peripheral facial palsy, and the degree of facial paralysis most commonly found at diagnosis was the House-Brackmann IV.

The absence of pre- and post-treatment ratings of facial palsy as well as the lack of information on the degree of improvement in facial paralysis in most studies made it difficult to obtain accurate information on the best therapeutic approach.

Other adjuvant therapies such as radiosurgery and facial nerve decompression are not yet established and require further research.

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