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Paraganglioma of the facial nerve, a rare differential diagnosis for facial nerve paralysis: case report and review of the literature

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Abstract This report describes a rare case of histopathologically confirmed glomus faciale tumor. The role of imaging in the differential diagnosis is discussed and therapeutic options are evaluated, along with a review of the previous literature on glomus faciale tumors. A 39-year-old male patient presented with total peripheral facial nerve paralysis. He underwent radical tumor resection and facial nerve grafting for a histopathologically confirmed paraganglioma of the facial nerve. He is now tumor-free after a 4-year follow-up period, and the functional outcome after primary nerve grafting is satisfactory. Facial nerve paragangliomas are a rare cause of facial nerve paralysis. Early imaging using computed tomography and magnetic resonance imaging is essential to clarify the differential diagnosis and assess the location and extent of the tumor. Precise pathological diagnosis requires additional targeted immunohistochemical examinations. The treatment of choice in patients with preoperative facial nerve paralysis is radical tumor resection with nerve reconstruction.

Introduction

Paragangliomas in the head and neck region are highly vascularized tumors, which in most cases prove to be benign. They originate most often in paraganglionic tissue in the area of the carotid bifurcation (carotid body tumors). Other origins include the jugular foramen (glomus jugulare), vagus nerve (glomus vagale), and the tympanic plexus and promontory (glomus tympanicum). In general, paragangliomas appear in principle along the embryological migration path of paraganglionic nonchromaffin tissue from the skull base to the pelvic floor. They may occur either sporadically or in the context of a hereditary familial tumor syndrome such as multiple endocrine neoplasia type II (MEN II), von Hippel–Lindau syndrome, or neurofibromatosis type 1. Multilocular presentations of glomus tumors are observed in 10–20% of sporadic cases and up to 80% of hereditary cases. Overall, they represent 0.6% of all tumors in the head and neck region. Only some 10% of cases show histopathological criteria for malignancy or metastasis into nonendocrine tissue. The mean tumor doubling rate is 4.2 years [1].

To date, there have only been eight case reports of histologically confirmed primary glomus faciale tumors in the literature (Table 1) [2–7]. We report here on a further case of paraganglioma of the facial nerve with an intracanalicular location.

Case

A 39-year-old man presented at our University Outpatient Department in August 2006 with House-Brackmann index VI right-sided peripheral facial nerve paralysis, which had appeared acutely 6 months earlier and had since progressed (Fig. 1) [8]. There had been no trauma, and serological, microbiological, and neurological examinations, particularly of the other cranial nerves, were also unremarkable. The findings on ear, nose, and throat examinations and

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Table 1 Reported cases of histopathologically confirmed glomus faciale tumor

Reference	Patient's age (y), sex (F/M)	Symptoms	Tumor location	Therapy
Dutcher and Brackmann [2]	50, F	5 months' facial nerve paralysis	Descending facial nerve canal	Surgery and facial nerve grafting
Bartels et al. [3]	40, M	2 years' facial nerve paralysis, pulsatile tinnitus, conductive hearing loss	Descending facial nerve canal with retrotympanic soft tissue	Surgery and facial nerve grafting
Bartels et al. [3]	20, M	6 months' ear fullness	Descending facial nerve canal and mass in the external auditory canal with involvement of the mastoid	Surgery and facial nerve grafting
Petrus and Lo [4]	74, F	Pulsatile tinnitus	Descending facial nerve canal with breach into external auditory canal	Biopsy and radiotherapy
Petrus and Lo [4]	74, F	5 years' facial nerve paralysis and retrotympanic mass	Descending facial nerve canal with breach of external auditory canal	Biopsy and radiotherapy
Kania et al. [5]	63, F	9 months' facial nerve paralysis, pulsatile tinnitus, otalgia	Horizontal portion and descending facial nerve canal with retrotympanic soft tissue	Surgery and facial nerve grafting
Mafee et al. [6]	37, F	12 months' otalgia and recurrent facial nerve paralysis	Tympanic portion at the geniculate area extending to the middle ear and pyramidal turn	Surgery and facial nerve grafting
Connor et al. [7]	54, F	6 months' facial nerve paralysis, parotid mass, otalgia	Descending facial nerve canal extending to the parotid gland	Surgery and facial nerve grafting

otoscopy in particular were normal for the patient's age. Pure tone audiography showed bilateral normal hearing without tinnitus, and gustometry confirmed normal taste. Head and neck ultrasonography showed no evidence of parotid gland pathology. The differential diagnosis then included a possible middle ear process such as genuine cholesteatoma, vestibular schwannoma, or facial nerve neurinoma, as well as idiopathic or central facial nerve paralysis as a diagnosis of exclusion. For further diagnosis, we therefore requested computed tomography (CT) of the petrous bone and magnetic resonance imaging (MRI) of the skull. These showed an unclear tumorous process in the right petrous bone (Figs. 2, 3).

An exploratory mastoidectomy with posterior tympanotomy and decompression of the facial nerve was carried out to obtain a tissue sample. Intraoperatively, a reddish, shiny tumor was seen in the facial canal, which was already growing diffusely into the surrounding mastoid cells (Fig. 4). Histological and immunohistochemical examinations showed medium-sized tumor cells, with light-colored cytoplasm surrounded by partly prominent hyperemic capillary vessels (Fig. 5). A paraganglioma of the facial nerve in a partly intraosseous position was therefore diagnosed.

After the various treatment options had been explained to the patient, it was decided to carry out a radical tumor resection. This was done via a combined retroauricular and preauricular access route, with exposure of the nerve and facial nerve resection distal to the geniculate ganglion as far as the stylomastoid foramen, combined with a type III tympanoplasty. Primary facial nerve reconstruction was carried out with a 6-cm long interposition graft from the great auricular nerve. Inner ear function was intact immediately postoperatively.

Four years later, electromyography of the facial nerve showed a persistent defect after healing, with clinically incomplete peripheral facial nerve paralysis, House-Brackmann index III (Fig. 6). Pure tone audiometry showed residual conductive hearing loss of 20 dB.

Discussion

Paraganglioma of the facial nerve is an extremely rare condition—and precisely for that reason it should be included in the differential diagnosis in unclear cases of facial nerve paralysis after more frequent causes have been excluded. To date, eight other cases of histopathologically confirmed paraganglioma of the facial nerve have been described in the literature, in three men and six women, aged 20–74 years (mean age 51.1 years). Glomus faciale tumors originate in a small number of "glomus bodies" in the descending part of the facial nerve canal [7]. In almost

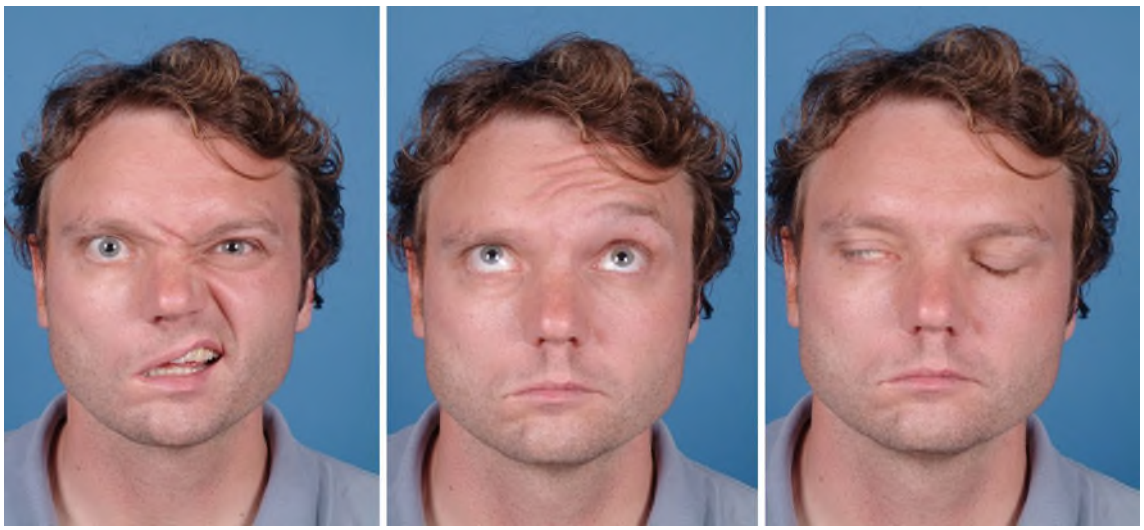

Fig. 1 Preoperative findings, with right-sided facial nerve paralysis (House-Brackmann index VI)

all cases, the clinical impression has been one of slowly progressive peripheral facial nerve paralysis of varying severity and duration [2, 4, 5, 7].

In one case, a young woman developed acute facial nerve paralysis with right-sided otalgia. She was initially treated with several injections of adrenocorticotropic hormone (ACTH), each of which led to almost complete symptomatic relief for a short period. The fact that recurrent facial nerve paralysis coincided with menstruation in the young woman is explained by the authors by an increased proportion of interstitial liquid and the resulting increase in volume in the hypervascular tumor, with compression of the nerve [6].

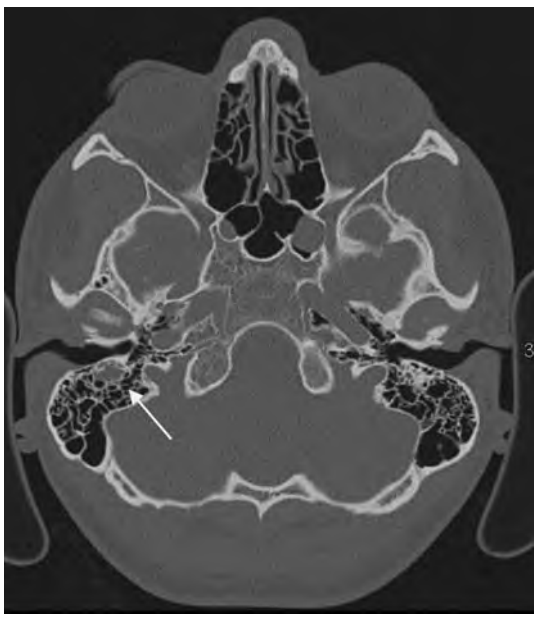

Fig. 2 Axial CT with an unclear osteodestructive process in the mastoid part of the facial nerve canal in the right petrous bone (*arrow*)

Other clinical symptoms described in the literature include pulsatile tinnitus, a feeling of pressure in the ear or otalgia, blepharospasm, and conductive hearing loss [2–7]. In two cases, a tumor mass was found in the external auditory canal on otoscopy [3, 4]. In the other cases, otoscopy was either unremarkable or a reddish space-occupying lesion was seen in a retrotympanic location [2, 5–7]. Ultrasonography of the parotid gland was unremarkable except in one case, in which Connor et al. [7] describe the only glomus faciale so far reported that had intraparotid and extracranial extension. The ultrasound examination showed a well-vascularized space-occupying lesion.

Contrast-enhanced CT of the petrous bone was carried out for further diagnosis in all of the reported cases. The tumor was located in the mastoid part of the facial nerve canal in 87.5% of cases ($n = 7$). Bone destruction in the neighboring mastoid cells was observed in all cases. The tumor had penetrated the external auditory canal or middle ear in three cases each. The extension of the tumors ranged from the geniculate ganglion to the parotid gland. MRI of the skull was also carried out in two cases, as in the present report. The tumor showed contrast uptake on T1-weighted imaging in both cases [5, 6]. Preoperative selective angiography was also carried out in a case described by Bartels et al. [3], with embolization of the afferent vessels.

In 75% of the reported cases ($n = 6$), the glomus faciale tumors were treated with radical tumor resection and facial nerve resection and reconstruction (e.g., with the great auricular nerve, sural nerve, or a hypoglossal jump anastomosis). In the two cases described by Petrus and Lo [4], treatment following biopsy confirmation of the diagnosis

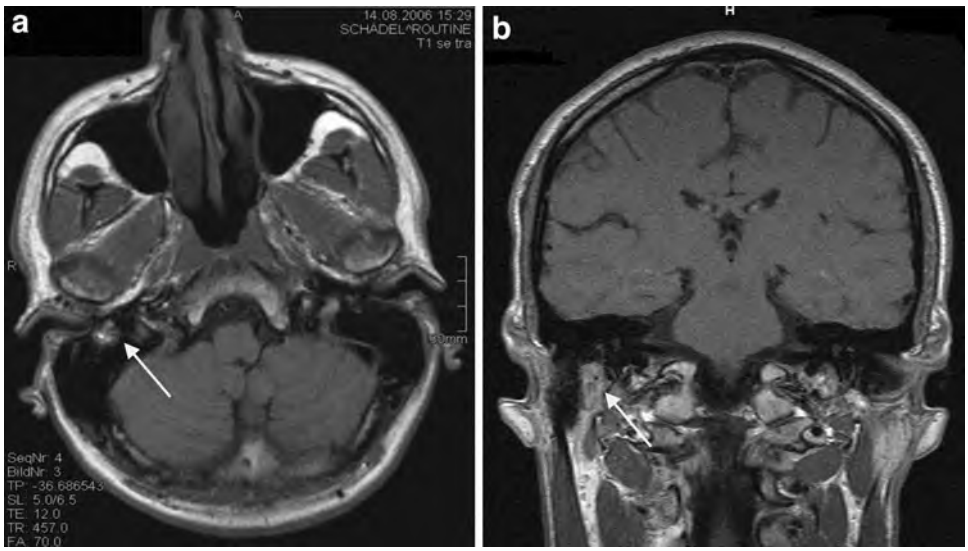

Fig. 3 Axial (**a**) and coronal (**b**) T1-weighted MRI with gadolinium enhancement, showing contrast uptake in the mastoid part of the facial nerve canal in the right petrous bone (*arrow*)

consisted of primary radiotherapy, partly due to the patients' advanced age. Facial nerve function recovered postoperatively within 12–24 months to a House-Brackmann index of III in the cases reported by Bartels et al. [3] and Kania et al. [5]

Involvement of the jugular foramen was not observed in any of the reported cases either using imaging morphology or intraoperatively. It can therefore be assumed that the paragangliomas described originated directly from non-chromaffin cells of the facial nerve and therefore represent a distinct tumor entity.

Early imaging with CT of the petrous bone and MRI of the skull is essential for localizing and assessing the extent of the tumor, as well as for precise planning of the treatment approach. On MRI, paragangliomas usually show a hyperintense signal on T2 weighting and clear contrast uptake on T1 weighting. A "salt-and-pepper" appearance on T1-weighted images is a typical morphological sign for paragangliomas. Due to their contrast uptake on MRI, paragangliomas can usually be distinguished from genuine cholesteatomas and other tumor entities in the differential diagnosis [6, 9].

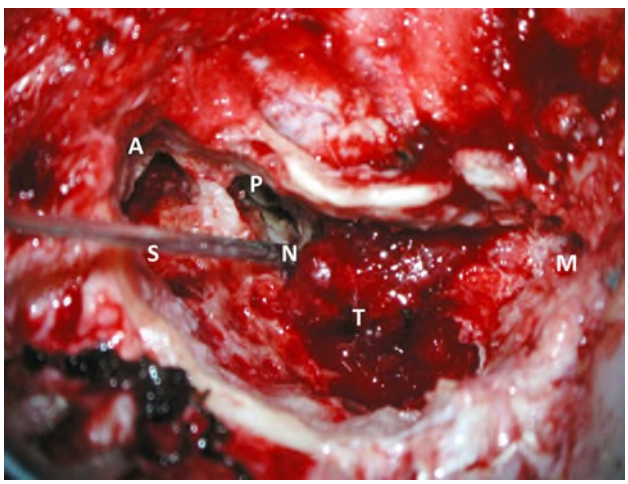

Fig. 4 The intraoperative site during exploratory mastoidectomy on the *right side*. *A* antrum, *M* tip of the mastoid, *N* facial nerve, *P* posterior tympanotomy, *S* stimulation electrode, *T* tumor

Digital subtraction angiography (DSA) is the ultimate gold standard for the diagnosis of paragangliomas. In addition to allowing preoperative embolization, DSA provides arterial "mapping" of the tumor's vascular supply. Somatostatin-receptor scintigraphy is not widely available as a diagnostic method and is reserved for specialized centers. However, ^{18}F-dihydroxyphenylalanine (DOPA) positron-emission tomography (PET) appears to be superior to MRI in very small tumors (<1 cm), and it may therefore become more important as a screening method in the future in patients with hereditary paragangliomas [1].

Precise pathological diagnosis requires additional targeted immunohistochemical examinations. These include the endocrine markers chromogranin and CD56. In addition, paragangliomas have protein S100-positive sustentacular cells and CD34-positive capillaries.

Depending on the location of the tumor (e.g., glomus tympanicum, glomus jugulare, glomus caroticum, glomus faciale) and its size (e.g., Fisch classification [10], cranial nerve involvement), the treatment of choice in patients with paraganglioma has generally shifted in recent years from radical resection to surgical tumor reduction with maintenance of function, as well as local control of any residual tumor tissue [11]. Depending on the individual case, local control may involve postoperative radiotherapy, or a "wait-and-scan" strategy can be used. In some cases—particularly in very large tumors—embolization of the afferent arteries should be carried out before surgery [12]. Depending on the individual situation (e.g., in relation to

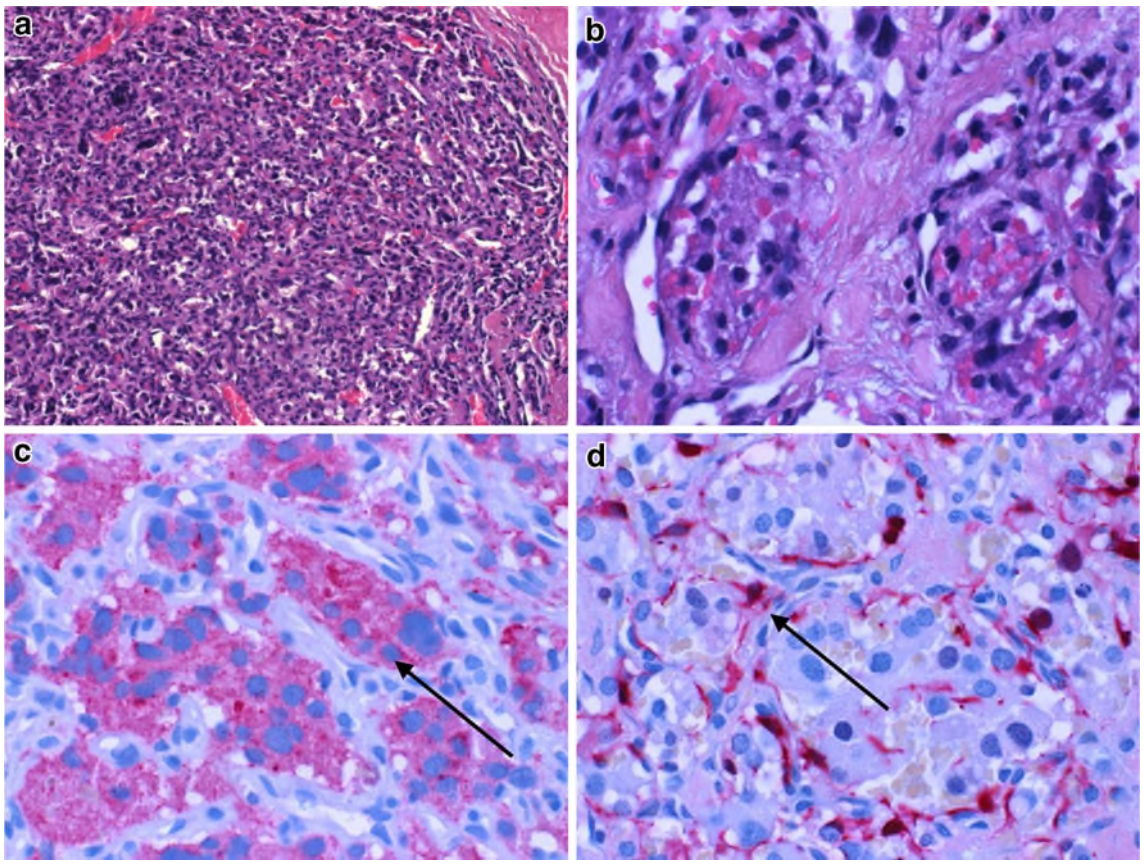

Fig. 5 Light microscopy showed small round and polygonal cells within highly vascularized and sclerotic stroma. **a** Hematoxylin–eosin (H&E), original magnification ×200. **b** H&E, original magnification ×631. **c** Protein S100-positive sustentacular cells (*black arrow*). **d** CD34-positive capillaries (*black arrow*)

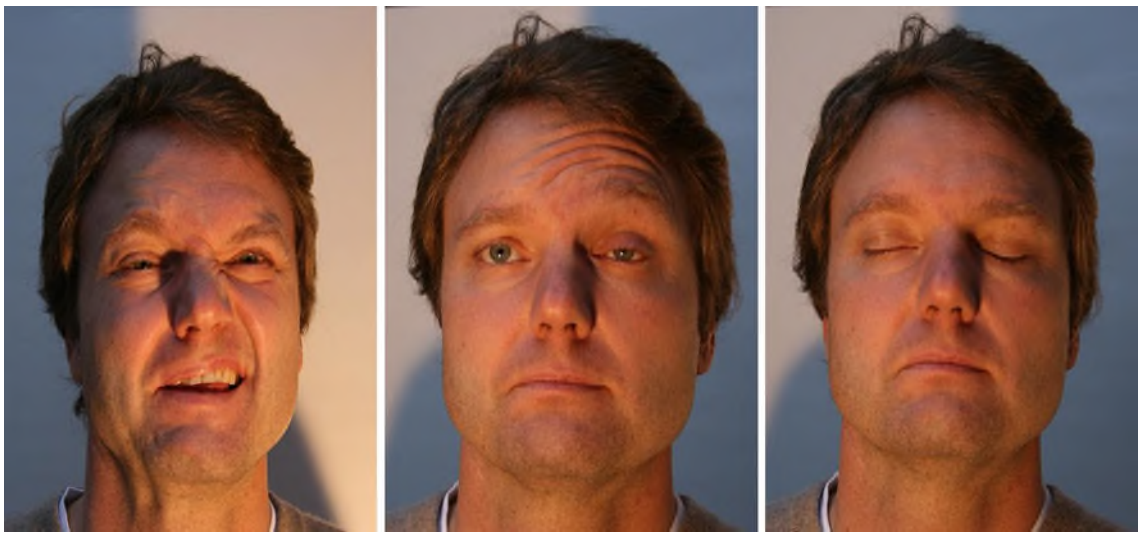

Fig. 6 The postoperative facial nerve results, showing House-Brackmann index III–IV conditions on the *right side* 4 years after primary facial nerve reconstruction using a great auricular nerve interposition graft

the patient's age, comorbid conditions, multifocal lesions, postradiotherapy recurrences, and the risk of injury to cranial nerves), alternative primary treatment options include conventional radiotherapy (45–50 Gy) or stereotactic radiosurgery (12–18 Gy) [13, 14]. Bianchi et al. [13] have reported on effective and safe use of CyberKnife

radiosurgery in the treatment of paragangliomas of the skull base. However, the follow-up period of approximately 20 months to date does not allow conclusive assessment of the long-term results of such treatment. A study by Cosetti et al. [15] reports on three patients with jugulotympanic paragangliomas in whom no tumor growth was seen over periods of up to 33 years without therapy. Bäck et al. [16] also report the case of a patient who initially received clinical checkups over a period of 20 months due to an intraparotid facial nerve schwannoma, with intact nerve function. A "wait-and-scan" strategy may therefore be justifiable, particularly in patients with a smaller glomus faciale and no evidence of facial nerve paralysis who are also unwilling to undergo therapy, or in patients at an advanced age.

Conclusion

In the case presented here, the patient already had complete peripheral facial nerve paralysis preoperatively, and radical facial nerve resection with subsequent reconstruction was therefore indicated. Early and appropriate rehabilitation measures are of course decisively important for achieving good postoperative results.

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