Geniculo-Temporo-Parotideal Neurofibroma of the Facial Nerve
A Case Report

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Key words: facial nerve, neurofibroma, parotid gland

SUMMARY – *Preoperative diagnosis of facial nerve neurofibroma is difficult when it presents as an asymptomatic parotideal mass. Facial nerve tumor suspicion arises during parotid surgery, histopathologic evaluation confirms diagnosis and postoperative imaging demonstrates a nerve tumor and its extension. We present the multimodality imaging findings of a 43-year-old man with a solitary neurofibroma involving the whole facial nerve continuously from geniculate ganglion to parotideal segment as the first case in the literature.*

Introduction

Neurogenic neoplasm of the facial nerve is uncommon and those involving the intraparotid portion of the facial nerve is far less common. The estimated frequency of parotid tumors originating in the facial nerve ranges from 0.2% to 1.5% [1]. For this reason these tumors are unlikely to be included in the differential diagnosis of a parotid mass when facial nerve function is normal and in the absence of neurofibromatosis [2,3]. Neurogenic neoplasms of the facial nerve are predominantly schwannoma whereas neurofibroma is exceedingly rare [2]. Up to now about 70 cases of schwannoma [4] and 11 cases of neurofibroma of the facial nerve involving the intraparotideal portion have been reported in the literature [2,4]. We describe a case of geniculo-temporo-parotideal solitary facial nerve neurofibroma, initially presenting as an asymptomatic parotid mass. To our knowledge, this is the first case of neurofibroma involving all facial nerve segments from geniculate ganglion to parotid.

Case Report

A 43-year-old man was admitted for evaluation and management of a gradually enlarging right parotid mass of two years duration. There was no sign of facial weakness, no family history of neurofibromatosis and the patient had no café-au-lait spots. On physical examination, a 3×4 cm firm mobile mass was palpable slightly superior to the right angle of the mandible. There was no associated lymphadenopathy. A complete blood count, biochemical parameters and peripheral smear were within normal limits. Ultrasound examination showed that there was a well circumscribed hypoechoic mass in the tail of the right parotid with minimal posterior acoustic enhancement (figure 1 A). There was no vascularity within the tumor in color Doppler ultrasound. Preoperatively fine needle aspiration biopsy (FNA) was performed for histologic diagnosis but it was inconclusive. Superficial parotidectomy had been planned. However because of being unable to locate the facial nerve during surgery, suspicion of a nerve

tumor arose and a wedge resection was done. A 2.5×2.5×2 cm solid mass was excised and the histopathological diagnosis was a neurofibroma (figure 1 B). After surgery right partial facial paralysis developed but regressed after administration of oral corticosteroids. High resolution temporal bone computed tomography (HRCT) and temporal bone magnetic resonance imaging (MRI) were performed to determine the extension of the tumor. HRCT images showed an ill-defined curvilinear hyperdense band indicating the impression of the enlarged right geniculate ganglion (figure 2 A), while contrast enhancement was detected on MR images (figure 3 A). The intratympanic part of the nerve was diffusely thickened along its course (figure 2 B-D). The vertical and extratemporal segments of the nerve were thickened as well causing expansion of the canal and stylomastoid foramen (figure 2 E, F). There was peripherally increased signal intensity on T2-weighted image (figure 3 B) and peripheral enhancement of the thickened nerve segment on pre- and post gadolinium T1-weighted images (figure 3 C, D). Thickened nerve extending from the stylomastoid foramen to the parotid was hypointense on T2-weighted image (figure 4 A) and showed heterogenous enhancement on post gadolinium T1-weighted image (figure 4 B) The residual part of the tumor in the parotid gland was isointense compared to muscle on T1- weighted image (figure 4 C) and displayed both peripheral and central 'dot-like' hyperintensity on T2-weighted image (figure 4 D). The patient declined radical surgery which was quite risky due to involvement of the geniculate ganglion and a long segment of the facial nerve by the tumor. There was no significant difference in size and length of the tumor on the first year control in temporal bone MRI. Localized radiotherapy is planned to decrease the mass effect of the tumor.

Discussion

Facial nerve tumors arising within the course of parotid gland are rare and are unlikely to be included in the differential diagnosis of a parotid mass [2]. The rate of involvement of facial nerve segments by nerve tumor in decreasing order are geniculate ganglion (75%), the first portion (46.4%), second genu (46.4%), third portion (32.1%), internal auditory canal (25%), cerebellopontine angle (10.7%) and extracranial portion (10.7%) [5]. The segments of the nerve running into the cerebellopontine angle or in the parotid gland are usually affected with adjacent portions of nerve [5]. We describe a case of a solitary neurofibroma involving the four contiguous segments (geniculo-tympano-mastoideo-parotideal) of the facial nerve presenting with a parotid mass without paresis or paralysis. Clinical symptoms depend on the affected segment of the facial nerve. Conductive hearing loss and facial palsy are the most common symptoms due to frequently affected geniculate ganglion and tympanic portion [5]. Due to firmly attached cells in tumors of neurogenic origin obtaining positive cytology is quite challenging [3]. The rarity of the tumor, the low specificity of ultrasound examination and inconclusive FNA almost always lead to preoperative misdiagnosis in these patients [3,4]. In our case, FNA failed to provide the correct diagnosis and sonographic features were indistinguishable from a pleomorphic adenoma. Neurogenic neoplasms of the facial nerve are usually diagnosed intraoperatively by tissue biopsy as in our case. Therefore in case of a parotideal mass and concomitant facial disorder, both radiologists and clinicians must be aware of the probability of a facial nerve tumor involving the intraparotideal segment even if the mass sonographically looks like a pleomorphic adenoma. The favorable imaging modality for facial nerve diseases depends on topographical localization and clinical indication. MRI is preferred for imaging of the brainstem nuclei, the cisternal segment or the intracanalicular portion of the nerve; high resolution CT is preferred for the tympanic portion. The normal facial nerve shows low or slightly intermediate intensity on T1-weighted image, while physiological enhancement is seen in the intratemporal part due to surrounding circumneural venous plexus. Enhancement in the distal intracanalicular is never present while it may be rarely seen in the labyrinthine portion [6]. Neurofibromas have low attenuation on unenhanced CT which might be due to the fat content of Schwann cells, water content of myxoid tissue and cystic areas of hemorrhage and necrosis. The tumor is isointense with muscle on T1-weighted images. On T2-weighted images the lesion is homogenously hyperintense or may have characteristic target sign resulting from the location of collagen centrally and more myxoid tissue peripherally [7]. The target sign was described as high signal intensity surrounding a central region of lower signal intensity [1]. The target sign on axial T2-weighted images is not specific for neurofibromas, as it is also seen in other neurogenic neoplasms such as schwan-

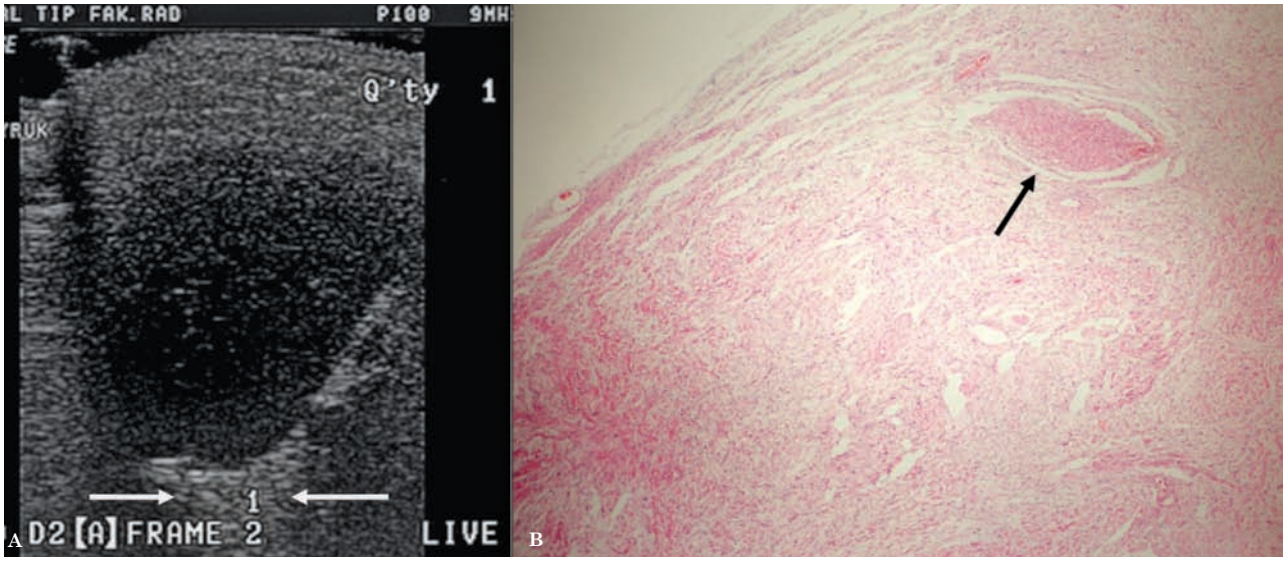

Figure 1 A gray scale ultrasound image in sagittal plane (A). Neurofibroma is seen as a well circumcribed hypoechoic mass in the tail of the right parotid gland with posterior acoustic enhancement (arrows) resembling a pleomorphic adenoma. The microphotograph shows a typical neurofibroma characterized by spindle cells within the mixoid background (B). A small neurite (arrow) is seen within the lesion (H&E, ×40).

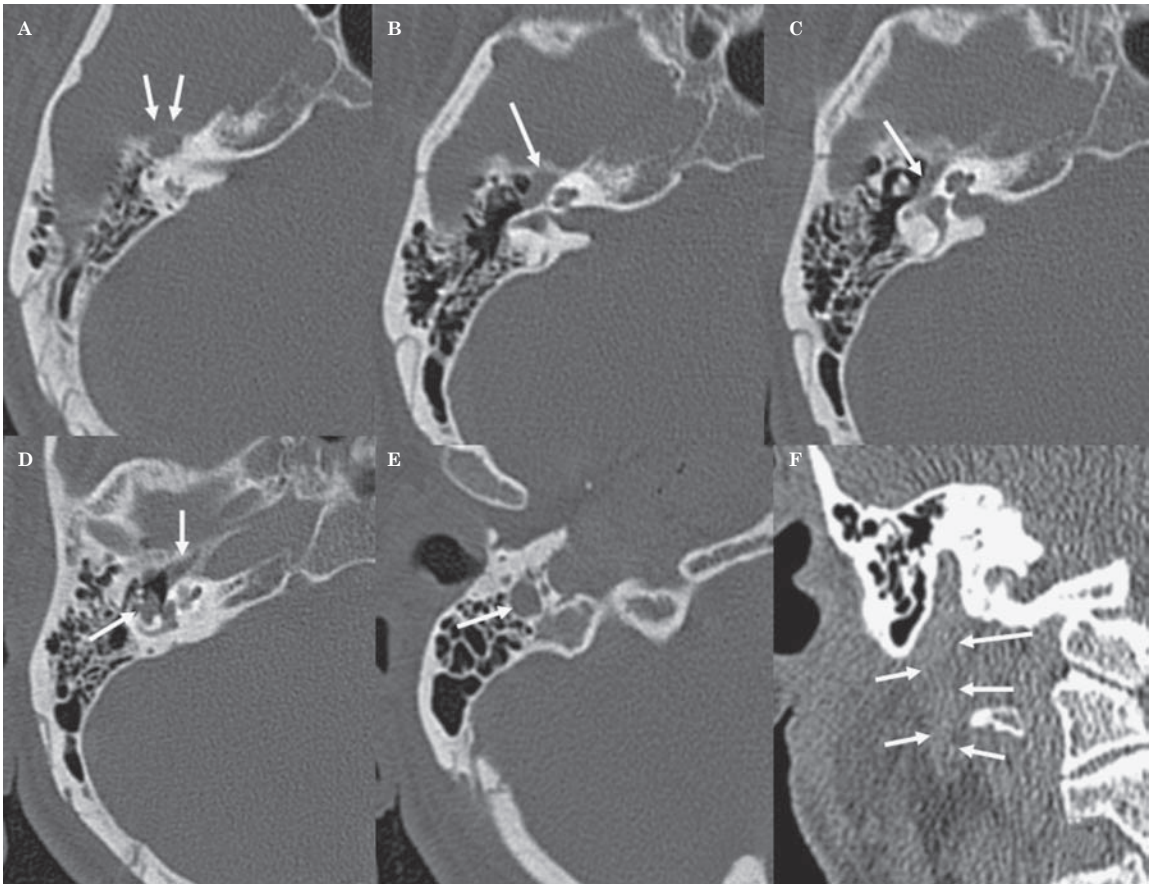

Figure 2 Axial HRCT image (A) shows an ill-defined curvilinear hyperdense band (arrow) indicating the impression of enlarged right geniculate ganglion on the bone. Thickened intratemporal horizontal and vertical segments of the facial nerve (arrows) (B-E) are demonstrated om sequentially. The coronal HRCT image (F) obtained at the level of stylomastoid foramina shows foraminal expansion and thickening of the nerve (arrows) along the extratemporal portion through the parotid gland.

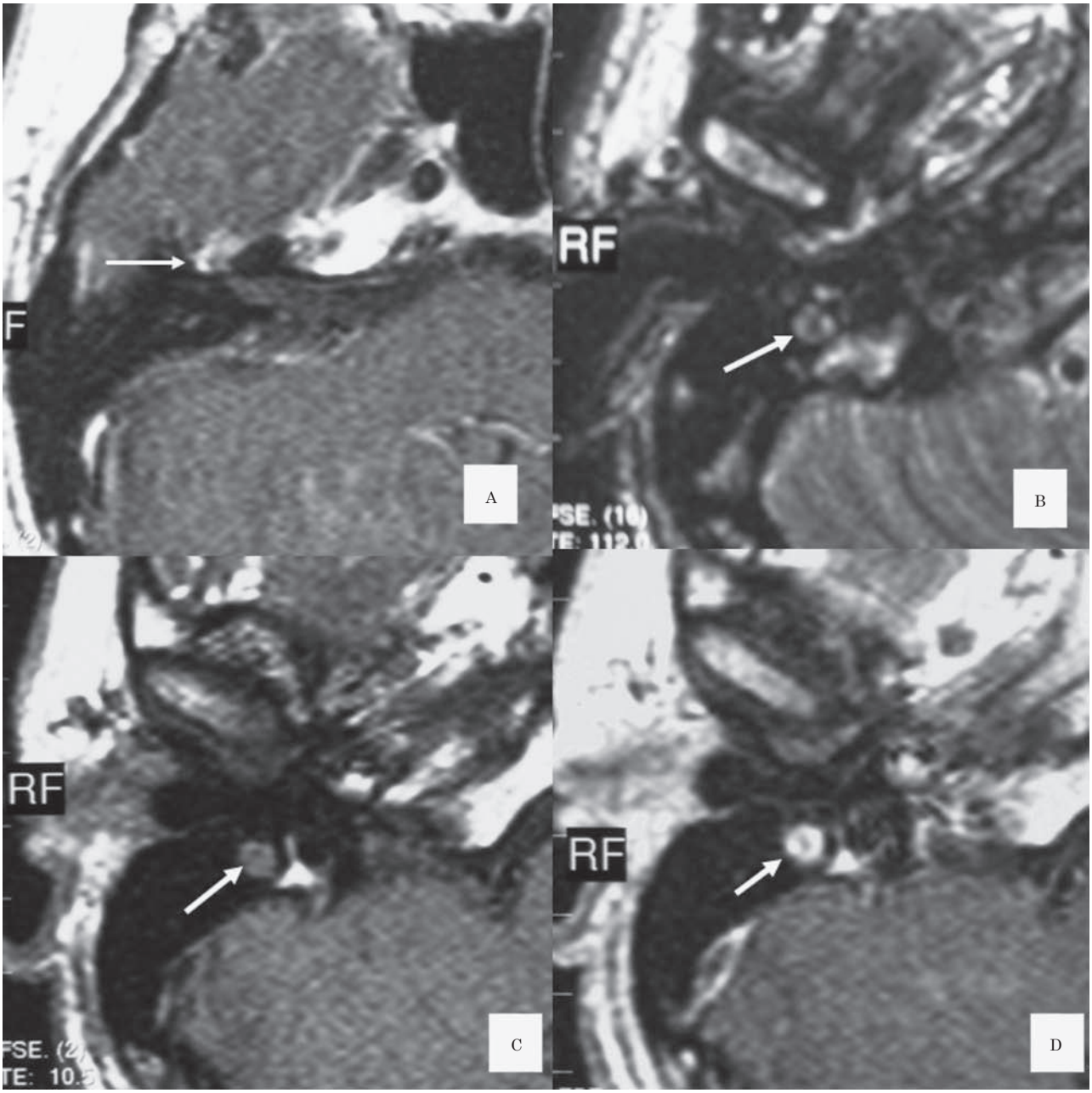

Figure 3 Axial postgadolinium T1-weighted image (A) illustrates heterogenous peripheral contrast enhancement (arrow) at the right geniculate ganglion. At the level of intratemporal vertical portion. On the axial T2-weighted image the central hypointensity and peripheral hyperintensity (target sign) (arrow) are illustrated in the thickened nerve (B). Axial pre- (C) and post-gadolinium (D) T1-weighted images show the thickened nerve (arrow) with brisk peripheral contrast enhancement (arrow).

nomas and malignant peripheral nerve sheath tumors. In our case, there was a typical target sign at the intramastoid segment and both peripheral and central 'dot-like' hyperintensity at the extratemporal segment on T2-weighted images.

Differential diagnosis of facial nerve neurofibromas from schwannomas is extremely difficult by currently available imaging methods. Both of them appear as well-circumscribed fusiform enhancing masses along the intratemporal course on post-gadolinium T1-weighted images and cause sharply defined bony canal enlargement on CT images. Other imaging features of

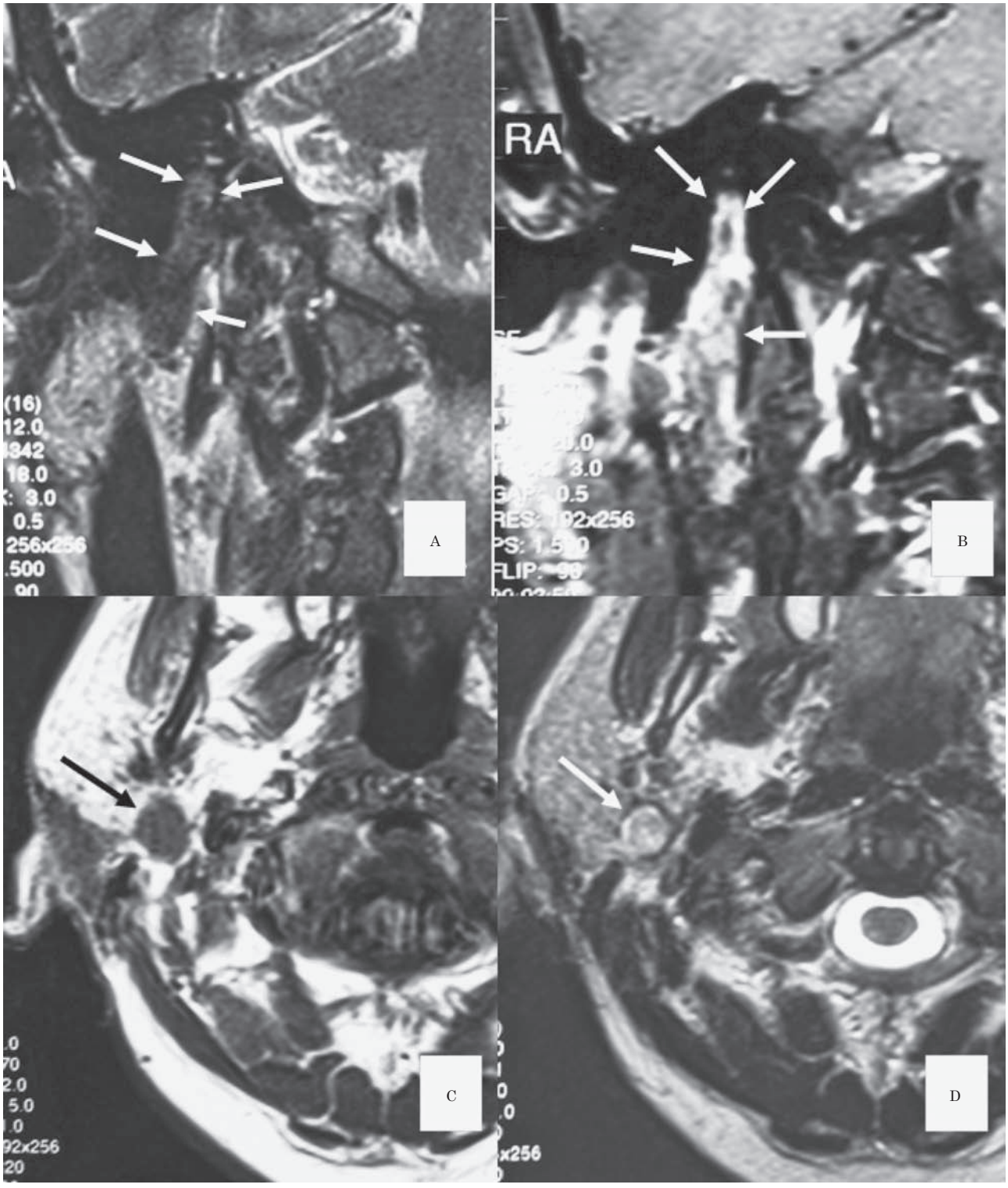

Figure 4 Coronal T2-weighted (A), and coronal postgadolinium T1-weighted (B) images obtained at the level of the stylomastoid foramina show foraminal expansion and thickening of the nerve (arrows) along the extratemporal portion through the parotid gland. The postgadolinium T1-weighted image (B) shows heterogenous contrast enhancement (arrows) of the thickened nerve. The pre-gadolinium axial T1-weighted image (C) shows the distal part of the neurofibroma in the right parotid gland (arrow) adjacent to the resection border. The axial T2-weighted image (D) depicts peripheral 'ring-like' and central 'dot-like' hyperintensity.

schwannomas include a mass mimicking acoustic schwannoma at the cerebellopontine angle-internal auditory canal; a tubular-round mass at the geniculate ganglion enlarging the geniculate fossa; a round extra-axial mass of the middle cranial fossa involving the greater superficial petrosal nerve; a lobulated mass involving the tympanic segment, and a mass at mastoid segment causing disruption of adjacent air cells [8]. These features can also be seen in neurofibromas. Both schwannomas and neurofibromas are nerve sheath tumors developing from proliferating perineural fibroblasts and Schwann cells. Schwannomas, histopathologically, do not contain axons and can occasionally be separated from the adjacent nerve fibers surgically. By contrast, neurofibromas contain axons penetrating directly into the tumor mass making surgical dissection challenging [2]. The other differential considerations for the radiologist evaluating intratemporal facial nerve lesions are cholesteatoma, facial nerve hemangioma, and perineural parotid malignancy. All of them enlarge the facial nerve canal. Cholesteatomas involving the facial nerve canal do not enhance on postgadolinium T1-weighted images. Facial nerve hemangioma involving the canal displays more aggressive bony changes seen as irregular margins and/or a 'moth-eaten' appearance. When an internal calcified honeycomb matrix is present (50%), ossifying hemangioma is quite distinctive [8]. The contrast enhancement pattern of neurofibroma on CT or MRI is variable [7]. In our case, the lesion was isointense on T1-weighted images and slightly hyperintense on T2-weighted images compared to muscle. Contrast enhancement at the geniculate ganglion was peripheral in CT and heterogeneous in MRI. The extratemporal part had no apparent enhancement on CT but displayed peripherally dominant heterogeneous enhancement on MRI. These tumors are primarily benign and might be quiescent for a long time, therefore therapy might not even be necessary. In general, management of facial nerve neurofibromas should be conservative and based on facial nerve functional status [2]. However, functional deficit, extensive growth, and/or cosmetic disfigurement might require therapeutic intervention. On the other hand, it has to be considered that up to 10% of plexiform neurofibromas undergo malignant differentiation and those have an extremely poor prognosis despite all therapeutic efforts. In the surgical treatment of neurofibromas, total excision and cable graft is recommended. There have been few alternatives to surgical excision suggested in the literature to date. Radiotherapy is often not feasible for the control of tumor spread, but may have a role for solitary plexiform neurofibroma of the head and neck [9]. In our patient surgery was not performed because the geniculate ganglion and a long segment of the facial nerve were involved by the tumor and the patient refused surgery due to its risks. He has been in follow-up for more than one year and no gross difference in size and length of the tumor was detected on the first year control in temporal MRI. Localized radiotherapy was planned to decrease the volume effect of the tumor. We present the first case of a neurofibroma involving whole segments of the facial nerve from geniculate ganglion to parotid in the literature with demonstrative US, CT and MRI images.

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