Original Article

Extratemporal facial nerve schwannoma: are computed tomography findings predictive?

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Abstract: Objectives: To investigate the predictive value of computed tomography imaging in diagnosis of extratemporal facial nerve schwannoma. Materials and methods: Computed tomography records of eight patients with extratemporal facial nerve schwannoma managed from 2007~2015 were reviewed. Tumors of all cases were totally removed surgically. Intraoperative frozen section examination was employed. Results: computed tomography of one patient whose tumor derived from buccal branch demonstrated imaging of a round mass with clear margin; four out of five patients whose tumors derived from facial nerve trunk revealed a significant tail pointing to the stylomastoid foramen with one exception; one patient whose tumor located at the margin of parotid gland demonstrated round mass just outside the stylomastoid foramen deep to the mastoid process with a tail pointing to the parotid gland; one case of synchronous schwannomas took place in the parotid gland and parapharyngeal space presented obscure tail-like imaging. Diagnoses of schwannoma were inclined in all intraoperative frozen section examinations. There was no tumor relapse in all cases after one to three years follow-up while the tumor-involved facial nerve or nerve branches were disturbed in all cases. Conclusion: Computed tomography findings can provide some useful information for the diagnosis of extratemporal facial nerve schwannoma especially for those derived from nerve trunk.

Keywords: Facial nerve, schwannoma, computed tomography, diagnosis

Introduction

Extratemporal facial nerve schwannoma (ETFNS) is rarer than those take place intratemporally [1, 2]. Because most of ETFNS are quite similar to other primary parotid gland neoplasm such as mixed tumor, an early diagnosis is challenging. It is reported that both magnetic resonance imaging and computed tomography (CT) could offer some evidence for the diagnosis of intratemporal facial nerve schwannoma (ITFNS) in several papers, while a very few have talked about ETFNS and never reached an agreed criteria of diagnosis based on radiological findings [3-5]. Here we report some radiological findings which might favor a diagnosis of ETFNS.

Materials and methods

Eight patients with ETFNS managed from 2007~2015 in West China Hospital, were retrospectively studied. They are aged from 40 to 65 years old with the mean age of 48. There is only one male among them. All patients presented with a painless parotid mass with normal facial nerve function. CT of eight cases was evaluated. Complex facial nerve schwannoma involving both temporal bone and parotid gland were excluded. Tumors of all cases were totally removed surgically. Intraoperative frozen section examination was employed.

Results

CT of one patient whose tumor located at the margin of parotid gland, demonstrated round mass just outside the stylomastoid foramen deep to the mastoid process with a tail pointing to the parotid gland (Figure 1A); four of five patients with ETFNS derived from facial nerve trunk, revealed a significant tail pointing to the stylomastoid foramen (Figure 1B-D), except one with no tail-like imaging (Figure 2A); one patient of ETFNS derived from buccal branch...
demonstrated a round mass with clear margin; one patient of synchronous schwannomas took place in the parotid gland and parapharyngeal space, presented obscure tail-like imaging, the schwannoma was also derived from facial nerve trunk (Figure 3A). Pathological diagnoses of schwannomas were inclined in all intraoperative frozen section examinations based on the well-differentiated spindle cells accumulation. The final pathological diagnosis of schwannoma was also established by routine histology and immunohistochemistry.

Tumors of all cases were totally removed surgically. Superficial parotidectomy was performed in seven cases and total parotidectomy was performed in one case. Superficial parotidectomy and partial mastoidectomy was performed for the tumor deep to the mastoid process. Facial nerve trunk was spared in four cases including three cases of eccentric lesion of the trunk, and one case derived from the buccal branch, at the same time, Schwann membranous “sac” was also spared (Figure 2B-D). The facial nerve trunk could not spared in the other four cases because the tumor could not strip from a clear facial nerve trunk, or facial nerve trunk was found terminated, then the facial nerve defect was repaired with greater auricular nerve grafting (Figure 3B-D).
There was no tumor relapse in all cases after one to three years follow-up while the tumor-involved facial nerve or nerve branches were more or less disturbed in all cases.

**Discussion**

Parotid tumor is common in surgical practice. Of all parotid tumors, 82% are benign and, most of them are pleomorphic adenomas. Schwannoma is rare in the parotid gland, with approximately 160 cases described to date, and most of them are so-called intraparotid schwannoma as the facial nerve enters the parotid gland immediately upon leaving the stylomastoid foramen [2-4, 6]. In this case series, because only one case derived from facial nerve trunk was not embedded in the parotid gland, instead, it located just behind the parotid gland, so the topic of this paper was accordingly changed from intraparotid facial nerve schwannoma to ETFNS. Even though there was a clear boundary presenting between the tumor and parotid gland, surgical exploration has con-

**Figure 2.** ETFNS derived from facial nerve trunk CT demonstrating a round mass with clear margin (white arrow) but without a tail, enhanced outer layer of the tumor and obvious hypointense region inside (A), intraoperative view demonstrating the tumor at the bifurcation of the trunk (black arrow) and Schwann membranous sac (black triangle) (B); the yellowish content of the tumor accorded to the hypointense region of CT (C); the tapered-off facial nerve trunk eventually became into membranous sac which was still spared during surgery for good nerve function recovery (white arrow) (D).
CT of extratemporal facial nerve schwannoma

Figure 3. Synchronous schwannomas took place in parapharyngeal space (A) and in the parotid gland (B) Obscure tail-like imaging revealing between the upper belly of digastric muscle and anterior border of sternocleidomastoid muscle (white arrow) (a); intraoperative view demonstrating the nerve trunk terminating at the surface of the tumor (b); demonstration of tapered facial nerve trunk after tumor removal (c); demonstration of Schwann membranous sac of the proximal end of the facial nerve trunk (d).

firmed a contact relationship between parotid gland and the tumor. Schwannoma in the temporal bone with or without extracranial extension is more common than solely intraparotid ones [7].

Preoperative diagnosis of ETFNS is difficult. Fine needle aspiration biopsy is theoretically the most feasible method but practically inconclusive [8]. This may be explained by limited cell specimen or pathological complexity of salivary gland tumor. Such as spindle cell type of myoepidermoid tumor can be taken for schwannoma, or epithelialoid schwannoma can also be masqueraded as pleomorphic adenoma [9]. In addition, needle track implantation also make repeated fine needle biopsies controversial. Intraoperative frozen section examination proved to be more accurate than fine needle aspiration before surgery, and its routine application is recommended.

Facial nerve dysfunction occurred in only about 20% patients in a review of 53 ETFNS patients [10]. Whenever facial weakness presents, it mostly implies a primary or metastatic malignancy of parotid gland rather than a facial nerve schwannoma. For instance, adenoid cystic carcinoma and mucoepidermoid carcinoma are two kinds of primary parotid malignancies
that may easily invade facial nerve thus result in facial nerve weakness or paresis. The weakening process of facial nerve is slow and gradual, and the presence of slow and gradual facial nerve weakness could not tell different stories about adenoid cystic carcinoma or mucoepidermoid carcinoma from a facial nerve schwannoma. Facial electroneurography can perform little differential function likewise.

Medical imaging techniques such as ultrasonic, CT and MR are widely used for diagnosis of parotid tumor including ETFNS, among which MR is regarded as most valuable for facial nerve schwannoma. ETFNS can mimic a pleomorphic adenoma clinically, as well as on imaging [11, 12]. Though pleomorphic adenomas may extremely rarely show extension to the stylomastoid foramen, perineural spread of parotid malignancies should also be included in the differential diagnosis of an ETFNS showing extension along the stylomastoid foramen. In this case series report, a tail of imaging extending to the stylomastoid foramen favor the diagnosis of an ETFNS. It is supposed to be the result of sub-Schwann membraneous growth of the tumor. MR is thought to be the better technique in diagnosis of ITFNS, it presents a typically fusiform solid tumors with well-circumscribed smooth margins in the facial nerve canal, or may acquire a dumbbell-shape when it shows multisegmental involvement. They appear iso- to hypointense on T1-weighted imaging and hyperintense on T2-weighted imaging. Cystic degeneration and hemorrhage may result in heterogeneous enhancement, it may also present with typical or classical imaging in enhanced CT (Figure 2A). The tumor usually grows along the path of least resistance, but this property could not be demonstrated good enough outside the osseous tissue such as in the parotid gland. Presence of a target sign on T2-weighted images, if seen, excludes the possibility of a pleomorphic adenoma. Parotid malignancies often have infiltrating margins and heterogeneous appearance on T2-weighted images [13]. In this case series, we could conclude that ETFNS derived from nerve trunk is very likely to have a tail pointing to the stylomastoid foramen; those not derived from nerve trunk may reveal typical picture because of cystic degeneration and hemorrhage; solitary tumors locating deep to the mastoid process or upper sternocleido-mastoid muscle with clear margin shall also arouse suspicion of facial nerve schwannoma.

Owing to the rarity of facial nerve schwannoma and its benign and slow-growing nature, there is no universally accepted management. As to the treatment plan for a ITFNS, surgically total removal with facial nerve repair is usually the standard management for patients with poor facial function (House-Brackmann grade III or worse); In patients presenting with normal or near-normal facial nerve function, both of initial observation with periodic examination and early surgical intervention are recommended in different papers. Radiotherapy especially stereotactic radiotherapy can be also applied to tumors of small size to avoid surgery and get possible arrest of tumor growth. Meanwhile malignant transformation after radiotherapy has been reported in literature [4, 7, 14]. The diagnosis of ITFNS by medical imaging is easier than that of ETFNS especially via a long term radiological follow-up. For ETFNS with normal facial nerve function and recognizable nerve trunk, which is also named as eccentric, nerve-sparing approach can grant postoperative facial nerve function; in patients with unrecognizable nerve trunk, which is also named as concentric, nerve-resection and grafting approach can rarely keep the facial nerve function satisfactory after tumor removal. Inevitable sacrifice of facial nerve followed by reconstruction may gain some degree recovery, but the facial nerve function has never been better than House-Brackmann grade III [5-7].

In conclusion, computed tomography findings can provide some useful information for the diagnosis of ETFNS especially for those derived from nerve trunk. Early surgical intervention is recommended regardless the facial nerve function, in this manner, postoperative facial nerve function recovery may be easier to gain, and delayed management of primary parotid malignancy may be avoided.

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Disclosure of conflict of interest

None.
CT of extratemporal facial nerve schwannoma

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