Hypoglossal Schwannoma of Parapharyngeal Space: An Unusual Case Report

Hari Ram · S. P. Agrawal · Nuzhat Husain ·
Swagnik Chakrabarti

Received: 6 November 2010 / Accepted: 5 October 2011 / Published online: 22 October 2011
© Association of Oral and Maxillofacial Surgeons of India 2011

Abstract Schwannomas are benign tumor of the schwann cells. These are common in eighth, tenth cranial nerves and parasympathetic chain. Hypoglossal schwannomas are rare. Among the hypoglossal schwannomas the intracranial and dumb-bell shaped schwannomas are frequent but extra cranial hypoglossal schwannomas are rare. We describe a parapharyngeal hypoglossal schwannoma in a 25 years female presenting as slow growing mass in right side of the neck.

Keywords Parapharyngeal space · Hypoglossal nerve · Schwannoma

Introduction

Schwannomas are tumors arising from Schwann cells of the peripheral sensory nervous system. Most schwannomas of the head and neck region are intracranial in origin [1]. Clinical manifestations are variable depending on location and can be associated with neurofibromatosis [2]. Schwannomas of the hypoglossal nerve usually develop in the intracranial portion, but sometimes may develop in both

the intra and the extra-cranial components forming a dumb-bell shaped lesion [3]. Schwannomas usually arise from the sensory divisions of cranial nerves, most commonly the eighth and tenth cranial nerves. Hypoglossal schwannoma is rare as the nerve has only a motor component [4].

Here we describe a rare case of extracranial schwannoma of hypoglossal nerve presenting as a mass in parapharyngeal space.

Case Report

A 25 years female presented with complaint of a slowly increasing swelling over right side of soft palate, fever and difficulty in swallowing of both solids and liquids since 5 months. Pain over swelling and fullness over retro-mandibular region was also noted since 4 months.

There was no history of difficulty in breathing, change in voice and bleeding from nose. On inspection a smooth swelling was present over soft palate measuring 6×4 cm in size extending from right last molar to base of tongue. Margins were ill defined and no pulsation was felt. On palpation it was firm with mild tenderness. No regional lymph nodes were palpable and no neural deficit was observed.

CT scan revealed a large mildly enhancing oval shaped soft tissue mass in right parapharyngeal space about $4.2 \times 4.5 \times 6$ cm in size. It extended from skull base to C3 medial to vertical ramus in pre-styloid space (Fig. 1). Non enhancing hypodense areas of necrosis were seen. Lesion was seen pushing the oropharynx towards left side and internal carotid artery medially and slightly anteriorly (Fig. 2). No evidence of bony erosion was noted.

Fine needle aspiration cytology (FNAC) revealed schwannoma of the parapharyngeal space. Whole body MRI was done to see any other lesion and to rule out

H. Ram (✉)
Department of Oral and Maxillofacial Surgery, CSM Medical
University (Erstwhile King George's Medical University),
Lucknow, India
e-mail: hariram81@yahoo.com

S. P. Agrawal · S. Chakrabarti
Department of Otolaryngology, CSM Medical University
(Erstwhile King George's Medical University), Lucknow, India

N. Husain
Pathology, CSM Medical University (Erstwhile King George's
Medical University), Lucknow, India

Fig. 1 Coronal CT showing tumor in para pharyngeal space

Fig. 2 Axial CT showing tumor in para pharyngeal space

schwannomatosis. A combined surgical approach (Trans cervical and paramedian mandibulotomy) was used to expose the tumor. A well capsulated mass was seen lying deep in parapharyngeal space (Fig. 3). The hypoglossal nerve was identified, entering in the proximal pole of tumor and running out distally. The capsule of the tumor was gently dissected to preserve the hypoglossal nerve (Fig. 4). The post-operative course was uneventful and no neural deficit was observed.

Histopathological examination showed presence of both Antoni A and B type of tissues, confirming diagnosis of schwannoma (Fig. 5).

Discussion

Schwannomas are uncommon benign tumors of nerve that are usually slow growing and asymptomatic [5, 6]. Approximately 25–45% of the cases occur in the head and neck region and are sometimes (8–19%) associated with Neurofibromatosis type-1 (NF-1) [7, 8]. They are found on the face, scalp, intracranial cavity, orbit, oral and nasal

Fig. 3 Exposed capsulated tumor

Fig. 4 Post operative site after excision of tumor

cavities, parapharyngeal space, middle ear, larynx, and medial and lateral regions of the neck. 70–80% tumors in the parapharyngeal space are benign and schwannomas account for 18% of the tumors. In the parapharyngeal space, they mostly arise from the vagus nerve (12%) and cervical sympathetic chain (6%) [9, 10]. Reported case was present in hypoglossal nerve, situated in parapharyngeal space which is very rare (Fig. 6).

Schwannomas of the parapharyngeal space usually occur between the ages of 30 and 70 years [6] with greatest incidence in third to fifth decades [8]. Hoshi et al. [11] described 85 cases of hypoglossal schwannomas in 16–85 years of age (mean 47.8). Male–female ratio for schwannoma of head and neck is almost equal or has a

Fig. 5 Excised tumor specimen

Fig. 6 Histopathology of tumor showing Antoni A and B type of tissues

slight preponderance for males (61.9%) than females (38.1%). Incidence of cervical sympathetic chain schwannomas is either equal or in ratio of 3:1 [5, 12]. In case of vagus nerve schwannomas there is no sex related predisposition [6, 13]. Hoshi et al. [11] reported slight preponderance of females (67.1%) for hypoglossal schwannoma as compared to males (32.9%). Study of Smith et al. [1] also showed schwannomas of the hypoglossal nerve being 2.5 times more common in women than men. We also report the lesion in a female which supports the findings of the above studies.

Out of 85 cases of hypoglossal schwannoma reported by Hoshi et al. [11] 39/71 (54.9%) were reported on left side and 32/71 (45.1%) were found on right side, in 14 cases side was not reported. Reported tumor was situated on the less common right side.

Hypoglossal nerve may be divided into five parts: the medullary, cisternal, skull base, pharyngeal, and sublingual

parts [14]. According to Hoshi et al. [11] dumb-bell shaped hypoglossal schwannomas are most common 27/54 (50%), followed by intra cranial 17/54 (31.5%). Extracranial was reported in 10/54 (18.5%) of cases. In contrast Karpoti et al. [15] reported 49 cases of hypoglossal schwannoma of which 28 (57.14%) were intracranial, 13 (26.53%) transcranial and 8 (16.3%) extracranial in origin. Out of 8 extracranial, 4 (8.16%) were located in the parapharyngeal space and 4 in the submandibular space.

Para pharyngeal schwannoma presents as a firm, painless slow growing mass in the lateral side of neck. Vagal schwannoma is typically characterized by dysphagia, cough, sore throat and hoarseness. Sympathetic schwannoma generally presents as Horner's syndrome [6, 9, 13]. Most common symptoms of intracranial and dumb-bell shaped schwannoma of hypoglossal nerve are hemi-atrophy with weakness of the tongue, deviation of tongue towards affected side, numbness of the lip and paresis of hypoglossal nerve. Head ache, nuchal pain, hemifacial spasm on affected side and dysphagia may also be present. Enlargement of right hypoglossal canal is seen in majority of cases [16]. However, many patients may not complain of hypoglossal deficits because dysfunction of one hypoglossal nerve does not cause disruption of speech or dysphasia [17].

Imaging studies play an important role in diagnosis of parapharyngeal space neoplasms [10]. Schwannomas appears hypodense as compared with muscle on non-contrast CT. Administration of contrast results in enhancement of lesion, which may be homogeneously solid or heterogeneous and patchy [12, 18]. It can be difficult to differentiate the hypervascular schwannoma from a paraganglioma on CT. Schwannomas are well-circumscribed homogenous masses that exhibit high-signal intensity on T2-weighted images and relatively homogeneous low signal intensity on T1-weighted images on MRI [9, 12]. In contrast with paragangliomas, no vascular flow voids are seen in schwannomas [12, 18].

Schwannoma appears as a single, well circumscribed and encapsulated lesion. Microscopically, they are well-encapsulated tumors with presence of distinctive cylindrical structures (Antoni type A tissue), which are set into a non distinctive, loose stroma of fibers and cells (Antoni type B tissue). Classical features include necrosis, haemorrhage, and cystic degeneration. Nuclear palisading is also seen in some areas [3].

The differential diagnosis of the parapharyngeal space lesions include tumors of the deep lobe of the parotid gland, tumors of minor salivary gland, metastatic cervical nodes, paragangliomas, branchial cysts, lymphomas, neurofibromas, and aneurysms of the internal carotid artery [5, 6].

Complete surgical excision is the treatment of choice as these tumors are radio resistant and chemotherapy has

limited effectiveness [6, 8, 13]. The surgical planning is influenced by the size of lesion, location, vascularity, and relation to adjacent structures like sympathetic chain, vagus nerve, carotid artery and jugular vein and other vital structures [19]. The transcervical surgical approach to the parapharyngeal space is the preferred operating procedure. If the schwannoma is completely removed, recurrence rates are extremely low [5]. Recently, intracapsular enucleation has been introduced which preserve the nerve fibres, and there by the neurological functions [19].

Though schwannoma has been described as a benign tumor but local recurrence and malignant schwannoma has been described by Langner et al. [8] in one (14.2%), out of 21 cases. Approximately 4% of malignant schwannomas has also been reported in cervical nerves [7].

Conclusion

Extra cranial hypoglossal schwannoma is very rare and few cases have been described in the literature. They are easily ignored in the diagnosis of parapharyngeal mass. We emphasise that during the examination of lateral neck masses, schwannomas should be included in the differential diagnosis.

References

- Smith PG, Backer RJ, Kletzer GR, Misler ET, Loosmore JL et al (1995) Surgical management of transcranial hypoglossal schwannomas. *Am J Otol* 16:451–456
- Kabatas S, Cansever T, Yilmaz C, Demiralay E, Celebi S, Caner H (2010) Giant craniocervical junction schwannoma involving the hypoglossal nerve: case report. *Turk Neurosurg* 20(1):73–76
- Drevelengas A, Kalaitzoglou I, Lazaridis N (1998) Sublingual hypoglossal neurilemmoma. Case report. *Aust Dent J* 43:311–314
- Kawakami R, Kaneko T, Kadoya M, Matsushita T, Fujinaga Y, Oguchi K, Kurashina K (2004) Schwannoma in the sublingual space. *Dentomaxillofac Radiol* 33:259–261
- Mikaelian DO, Holmes WF, Simonian SK (1981) Parapharyngeal schwannomas. *Otolaryngol Head Neck Surg* 89(1):77–81
- Chang SC, Schi YM (1984) Neurilemmoma of the vagus nerve: a case report and brief literature review. *Laryngoscope* 94(7):946–949
- Valentino J, Boggess MA, Ellis JL, Hester TO, Jones RO (1998) Expected neurologic outcomes for surgical treatment of cervical neurilemmomas. *Laryngoscope* 108(7):1009–1013
- Langner E, Del Negro A, Akashi HK, Araujo PP, Tincani AJ, Martins AS (2007) Schwannomas in the head and neck: retrospective analysis of 21 patients and review of the literature. *Sao Paulo Med J* 125(4):220–222
- Miller FR, Wanamaker JR, Lavertu P, Wood BG (1996) Magnetic resonance imaging and the management of parapharyngeal space tumors. *Head Neck* 18(1):67–77
- Saito DM, Glastonbury CM, EL-Sayad IH, Eisele DW (2007) Parapharyngeal space schwannomas preoperative imaging determination of the nerve of origin. *Arch Otolaryngol Head Neck Surg* 133(7):662–667
- Hoshi M, Yoshida K, Ogawa K, Kawase T (2000) Hypoglossal neurinoma—two case reports. *Neurol med Chir* 40(9):489–493
- Wax MK, Shiley SG, Robinson JL, Weissman JL (2004) Cervical sympathetic chain schwannoma. *Laryngoscope* 114(12):2210–2213
- Gilmer-Hill HS, Kline DG (2000) Neurogenic tumors of the cervical vagus nerve: report of four cases and review of the literature. *Neurosurgery* 46(6):1498–1503
- Thompson EO, Smoker WR (1994) Hypoglossal nerve palsy: a segmental approach. *RadioGraphics* 14:939–958
- Karapati RL, Loefer LA, Cunnig DM, Yousem DM, Li S, Weber RS (1998) Synchronous schwannomas of the hypoglossal nerve and cervical sympathetic chain. *AJR Am J Roentgenol* 171(6):1505–1507
- Lee HH, Liu CH, Hsu YH, Jung SM, Chen MT, Huang CC (2007) Hypoglossal schwannoma presenting as hemi-atrophy of the tongue. *Acta Neurol Taiwan* 16(1):37–40
- Raney RB, Raney AA, Bayless AE (1954) Tumors of the hypoglossal nerve: report of a case. *Bull Los Angel Neurol Soc* 19(3):160–165
- Som PM, Sacher M, Stollman AL, Biller HF, Lawson W (1988) Common tumors of the parapharyngeal space: refined imaging diagnosis. *Radiology* 169(1):81–85
- Kim SH, Kim NH, Kim KR, Lee JH, Choi HS (2010) Schwannoma in head and neck: preoperative imaging study and intracapsular enucleation for functional nerve preservation. *Yonsei Med J* 51(6):938–942