Schwannomas of Head and Neck and Review of Literature

Dinesh Kumar Sharma · Barjinder Singh Sohal ·
T. L. Parmar · Harvinder Arora

Received: 24 March 2010 / Accepted: 13 June 2010 / Published online: 11 April 2011
© Association of Otolaryngologists of India 2011

Abstract Benign nerve cell tumours have been given various names like schwannoma, neurilemmoma, neurinoma, neurofibroma, spindle cell tumours etc. Extra cranial head and neck schwannomas usually present as solitary and well-demarcated lesions. The lesion can cause secondary symptoms, such as nasal obstruction, dysphasia, and hoarseness, depending upon the location of the lesion. Fine needle aspiration cytology, CT scans, and MRI may be of limited help in the diagnosis of schwannomas. The treatment is complete surgical excision of the benign tumour and postoperative histopathological examination establishes the final diagnosis.

Keywords Schwannoma · Neurilemmoma · Benign nerve cell tumours

Benign nerve cell tumours have been given various names like schwannoma, neurilemmoma, neurinoma, neurofibroma, spindle cell tumours etc. Schwannomas of the head and neck region are not uncommon but they are rarely reported. About 25–45% of all the extracranial schwannomas have been reported in the head and neck region

[4, 15]. Calcaterra et al. [7] reported that head and neck region is the site of origin in more than one third of all solitary neurilemmomas and they occur most often in the lateral part of the neck. These are reported in the parapharyngeal space, retropharyngeal space, posterior pharyngeal wall, para-nasal sinuses, nasal cavity, scalp, sub-mandibular region, larynx, epiglottis, tongue, infratemporal fossa, cheek, oral cavity etc [2, 6, 19]. Three cases of extracranial schwannomas presented in the Dept. of ENT, Govt. Medical College & Rajendra Hospital Patiala during the period 1999–2006. These presented in the lateral pharyngeal wall, submandibular region and cheek and are being reported and discussed. Two of them were male and one female. The incidence of age ranged between 25 and 51 years.

Case No 1

A 51 year-old male patient from Haryana presented with dysphagia of 2 years duration. There was foreign body sensation in the throat. Since 2–3 months he was having a change of voice. There was no history of loss of appetite, loss of weight or trauma. In the past he had taken antitubercular treatment twice. He had been a smoker for 40 years taking 20 beedies daily.

On examination of the neck there was no lymphadenopathy. Oral examination showed a medial bulging on the left posterior tonsillar pillar. The overlying mucosa was smooth. Indirect laryngoscopy showed the mass extending downwards into the hypopharynx overhanging above left vocal cord making it invisible. On needle aspiration, few drops of blood came out. Computed tomography revealed a heterogenous mass (dV 10–40 HV) present in relation to left pharyngeal wall pushing the parapharyngeal fat planes laterally. The mass contained a few dense areas and a speck

D. K. Sharma (✉)
56, New Majithia Enclave, Patiala, India
e-mail: drdinesh11@gmail.com

B. S. Sohal · T. L. Parmar
Department of ENT Rajindra Hospital, Govt. Medical College
Patiala, Patiala, India
e-mail: drsohal52@gmail.com

H. Arora
Agroha Hissar Maharaja Agrasen Medical College, Agroha,
Hissar, India

of calcification. The mass extended from the left pterygoid plates to the level of the pyriform sinus obliterating the pyriform sinus. The mass was pushing, distorting and compressing the airway. The mass was well defined with a smooth border. Epiglottis had been pushed anteriorly by the mass, distorting the valleculae. Contrast study showed enhancement of hypodense area to dV 40–50 HV.

Surgical excision of the mass was planned under general anaesthesia through the per-oral route. Blood transfusion was arranged for any emergency need. A vertical incision was given behind the posterior tonsillar pillar. A yellowish growth was defined. It was encapsulated and separated from the surrounding structures by blunt dissection using a finger. With a slight push from the lateral side the globulated mass came out completely. No significant bleeding was seen. Postoperative period was uneventful. Histopathological report showed interlacing bands of delicate spindle cells and palisading of nuclei at places—suggestive of neurilemmoma.

Case No 2

A 25 year-old female patient presented with history of a persistent painless swelling in the right submandibular region since one and half year. The swelling had no relation with the meals. She gave the history of an operation 3 months back and showed a histopathological report of chronic sialadenitis.

On examination the swelling was firm, mobile, non-tender, irregular and well-defined with the size of 3.5 and 3 cm not fixed to the underlying structures or the overlying skin. There was a scar of the previous surgery over the swelling (Fig. 1). Bi-manual palpation showed a firm swelling opposite the right 2nd and 3rd molar teeth. The mucosa over the swelling area was intact. Ductal path was smooth and normal. Radiological examination showed a soft tissue shadow in the right submandibular area. FNAC showed changes of benign mesenchymal tumour.

Under general anaesthesia excision biopsy was done. Incision was given just by the side of the first incision. There was some difficulty during exploration due to the previous surgery, but the mass was completely taken out.

Postoperative period was uneventful. Biopsy of the specimen showed features of schwannoma i.e. spindle shaped cells with fibrous tissue stroma. No recurrence of the mass has been seen after 6 years.

Case No 3

A male patient aged 25 years belonging to a rural area presented with a swelling over the left cheek since 2 years.

It was slowly increasing in size. There was no history of pain or anaesthesia over the cheek. The patient gave a history of trauma over the cheek in the past (Fig. 2).

On examination the swelling was ovoid 2 cm by 2.5 cm, firm, margins not well defined and mobile. Radiological examination showed a soft tissue density mass outside the antero-lateral wall of the maxilla. Surgical excision was done under local anaesthesia through a sub-labial incision. The mass was removed completely (Fig. 3).

Histopathological report showed the presence of interlacing bundles of elongated cells having wavy dark staining nuclei. The cells were intimately associated with wire like strands of collagen suggesting a benign nerve sheath tumour.

Discussion

The study of literature showed that the neurogenic tumours arise from the neural crest which differentiates into the schwann cells and the sympatheticoblasts. The schwann cells give rise to neurofibroma and neurilemmoma (Schwannoma) (AGD) [13]. A schwannoma is a slow-growing solitary and encapsulated tumour attached to a nerve. A schwannoma shows degenerative changes such as cystic alterations and haemorrhagic necrosis whereas such changes are not seen in neurofibroma [4]. Schwannoma may arise from any cranial or spinal nerve that has a sheath i.e. any motor or sensory nerve other than the optic and the olfactory nerves which do not have the schwann cell sheath. Schwannoma was first established as a pathological entity by Verocay in 1908 who later called it neurinoma in 1910. Later the term neurilemmoma was coined by Stout in 1935 [15].

Approximately 25–45% of all the reported neurilemmomas occur in the head and neck and most of them are in the eighth nerve [3]. The most common site of the extracranial schwannomas in the head and neck region is the parapharyngeal space [5, 14]. The first case of neurilemmoma within the parapharyngeal space was reported in 1933 by Figi. Other sites in the head and neck like submandibular space, para-nasal sinuses, cheek, oral cavity etc. are rare [10].

The size of the tumour may vary from few mm to over 24 cms. The clinical signs and symptoms may vary according to the anatomic site of the tumour in the head and neck. Majority of the patients present with a painless mass and some may present with pain and tenderness. Other symptoms may be difficulty in breathing (Nasal), dysphagia (pharynx), epistaxis (PNS), hoarseness (larynx) or only a swelling in the neck (parapharyngeal space). The swelling is most often freely mobile in soft tissues, but when it is connected to a large nerve or trunk there is

Fig. 1 Swelling Right submandibular region

Fig. 2 Swelling Cheek (Intra-operative)

Fig. 3 Mass after removal (en bloc)

restriction of the movements. The primary nerve of origin may be entirely encompassed within the tumour. [7, 15, 17].

The schwannoma may arise at any age and there is no gender and race predilection but Putney et al. [15] in their study quoted a sex difference of approximately six females to four males. Leu and Chang [12] reported the predilection for males in their study of 52 cases. Though there are no known predisposing factors but Hatziotis and Aspiudes [9] believe that the trauma may be the cause. Calcaterra et al. [7] noted that the nerve of origin is usually not identifiable. Batsakis [4] reported that in the nose and paranasal sinuses, these tumours arise from intranasal nerves, ophthalmic and maxillary branches of trigeminal nerve and branches of autonomic nervous system. In the neck he divided the schwannomas into medial and lateral groups on the basis of nerve of origin. The medial group arises from the last four cranial nerves and the cervical sympathetic chain; the lateral group arises from the cervical neck trunk, cervical plexus and the brachial plexus. Rajan and Rajan [16] reported that in the parapharyngeal space the schwannoma may arise from the vagus nerve, the cervical sympathetic chain or the glossopharyngeal nerve. Ahad [1] has reported a case of lingual nerve schwannoma in the submandibular region.

The preoperative diagnosis of schwannomas in the head neck region is difficult. Abdul Ahad [1] has recommended the FNAC. But most of the investigations, like FNAC, may help to reveal diagnosis, but they are inadequate. FNAC is very effective in differentiating benign and malignant tumours of soft tissue. Although FNAC is very useful in most neck masses, it has a low accuracy in the diagnosis of neural tumours [8].

Computerised Tomography with contrast enhancement should be routinely done as some of the schwannomas are very vascular [4, 17]. Shoss et al. [18] has also recommended high resolution CT to determine the size and extent of the tumour, to demonstrate degree of tumour vascularity and to differentiate between benign and malignant lesions. On CT and MRI studies the schwannoma usually appear as spherical or ovoid soft tissue mass and may show moderate to marked often inhomogenous contrast enhancement, cystic component may be apparent and fatty degeneration is sometimes seen.

The differential diagnosis of these tumours must include metastatic or reactive lymphadenopathy, soft tissue neoplasms like fibroma, leiomyoma and lipoma, paraganglioma, carotid artery aneurysm, branchial cleft cyst, angioma, and other neurogenic tumours.

The treatment is complete surgical excision of the benign tumour. Recurrence after successful en bloc removal of the tumour is very rare (Khanna et al. [11]). Histo-pathological examination reveal two types of schwannomas Antoni type A and Antoni type B. Type A consists of spindle shaped cells with long slender fibres forming parallel arrangement of nuclei, known as the palisading of the nuclei around a

central mass of cytoplasm (Verocay bodies). Antoni type B tumours are degenerative in nature. The tissue is loosely arranged stroma in which the fibres and cells form no distinctive pattern. The two types may also be mixed (Maran AGD) [13]. The tumour is radioresistant and the possibility of the malignant degeneration of the benign tumour is extremely rare. Radiotherapy should be reserved for palliation when the surgical management is impossible [7].

In our study the male:female ratio has been 2:1 but this may not be significant as the number of cases has been very less. The nerve of origin could not be precisely identified. In one of the cases there was history of trauma prior to the appearance of the lesion which supports the belief of Hatzioitis and Aspiudes. We used the oral approach for the removal of the lateral pharyngeal wall tumour because there was no swelling seen from outside, but generally the trans-cervical approach is recommended for the parapharyngeal tumours. Vaid et al. [20] said that a transoral approach offers direct route to tumours presenting in the oropharynx but provides no control of the great vessels. This approach is only for small lesions not extending to the styloid process. It may be added that though the CT scan has made the diagnosis of the schwannoma easy but there are problems faced due to the economic status of most of the patients. We faced this problem in two of our patients.

Conclusion

Extracranial schwannomas which most often present as asymptomatic solitary masses, are rare tumours. The pre-operative, diagnosis may be difficult and it is often made after the surgery. The definitive diagnosis relies on clinical suspicion and histopathological confirmation. Complete surgical excision with appropriate approaches is the treatment of head and neck schwannomas. The possibility of nerve injury should be kept in mind. Local recurrence is extremely rare.

References

- Ahad A (1997) Schwannoma of submandibular region. *Indian J Otolaryngol Head Neck Surg* 49(4):341–343
- Ahmed S, Ahmed R, Lateef M, Ahmed M, Wani NA (2002) Neurilemmoma (Schwannoma) of the Maxillary sinus. *Indian J Otolaryngol Head Neck Surg* 54(3):234–235
- Balle VH, Greissen O (1984) Neurilemmoma of the facial nerve presenting as parotid tumour. *Ann Otol Rhinol Laryngol* 93:70–72
- Batsakis JG (1979) Tumours of the peripheral nervous system, 2nd edn. Williams and Wilkins, Baltimore, pp 313–333
- Batsakis JG, Sneige N (1989) Parapharyngeal and retropharyngeal space diseases. *Ann Otol Rhinol Laryngol* 98:320–321
- Bhandari A, Mishra P, Jain S, Sankhya N, Bapna AS (2000) Neurofibroma of the posterior pharyngeal wall. *Indian J Otolaryngol Head Neck Surg* 52(2):161
- Calcaterra TC, Rich JR, Ward PW (1974) Neurilemmoma of the sphenoid sinus. *Arch Otolaryngol* 100:383–385
- Dey P, Mallik MK, Gupta SK, Yasishtha RK (2004) Role of fine needle aspiration cytology in the diagnosis of soft tissue tumours and tumour-like lesions. *Cytopathology* 15(1):32–37(s)
- Hatzioitis JCh, Aspiudes H (1967) Neurilemmoma (Schwannoma) of oral cavity. *Oral Surg Oral Med Oral Pathol* 24(4):510–526
- Hazarika P, Nayak DR, Pujary K, Rao L (2003) Schwannoma of the nose and paranasal sinuses. *Indian J Otolaryngol Head Neck Surg* 55(1):34–38
- Khanna S, Gupta SC, Sinh PA (2003) Schwannoma of the maxillary sinus. *Indian J Otolaryngol Head Neck Surg* 55(2):132–135
- Leu YS, Chang KC (2002) Extracranial head and neck schwannoma—a review of 8 years experience. *Acta Otolaryngol* 122(4):435–437
- Maran AGD (1987) Benign diseases of the neck. *Scott Brown's Otolaryngol* 5:283–300
- Muranjan SN, Jagasia V, Pusalkar A (2001) Schwannoma of the cheek. *Indian J Otolaryngol Head Neck Surg* 53(2):140–141
- Putney FJ, Moran JJ, Thomas GK (1964) Neurogenic tumours of the head and neck. *Laryngoscope* 74:1037–1059
- Rajan R, Rajan R (1994) Tumours of the parapharyngeal space our experience. *Indian J Otolaryngol Head Neck Surg* 3(2):88–91
- Robitaille Y, Seemayer TAEI, Deiry A (1975) Peripheral nerve tumours involving paranasal sinuses. A case report and review of literature. *Cancer* 35:1254–1258
- Shoss SM, Donovan DT, Alford BR (1985) Tumours of parapharyngeal space. *Arch Otolaryngol* 111:753–757
- Sonkhya N, Luckwani A, Mishra P (2001) Neurofibroma of the Infratemporal Fossa. *Indian J Otolaryngol Head Neck Surg* 53(2):142–144
- Vaid N, Puntambekar S, Bora M, Kothadia A (2001) A transpalatal approach (palatal split) to the Parapharyngeal space. *Indian J Otolaryngol Head Neck Surg* 53(4):307–308