Cranial nerve schwannoma - A pictorial essay

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Abstract

Schwannomas are peripheral nerve sheath tumours arising from cranial, spinal or peripheral nerves. Most of the schwannomas are benign with the rare possibility of malignant transformation. Cranial nerve schwannomas can be seen along the course of any cranial nerve in the intracranial region or head and neck location. Although a majority are solitary sporadic lesions, multiple schwannomas can be seen in syndromes like neurofibromatosis type 2 and rarely in type 1. Since intracranial schwannomas are slow-growing, clinical presentation varies between no symptoms to cranial nerve palsy. Most of the times, the symptoms are due to mass effect over the adjacent structures, foraminal widening, compression of other cranial nerves, denervation injury or hydrocephalus. Familiarity with the course of the cranial nerves, imaging appearances and clinical presentation of schwannomas helps in accurate diagnosis and possible differential diagnosis, especially in uncommon clinical and radiological appearances. In this pictorial review, we illustrate relevant anatomy of cranial nerves, imaging features of schwannomas of most of the cranial nerves, clinical presentation and differential diagnosis.

Key words: Anatomy; clinical presentation; cranial nerve; differential diagnosis; magnetic resonance imaging; schwannoma

Introduction

Schwannoma, also known as neurilemmoma and neurofibroma, are two major types of benign peripheral nerve sheath tumours. Schwannoma arises from the myelinating cells of the cranial or peripheral nervous system, composed of Schwann cells that normally produce myelin sheath covering the nerve.[1] Schwannoma grows eccentrically within a capsule of parent nerve. By contrast, neurofibroma contains cellular elements of peripheral nerves such as axons, perineural cells, fibroblasts and Schwann cells. Neurofibroma grows diffusely within and along the involved nerves, expanding radially entrapping the neural elements. Schwannoma has a typical histological pattern, namely, Antony Type A and B.[1] Type A is highly cellular and demonstrates nuclear palisading and Verocay bodies. Type B is loosely organised microcystic tissue which may represent degenerated Antoni A tissue. Some authors found that heterogeneous signal intensity of larger lesions on T2-weighted imaging is more commonly associated with a higher ratio of type B to type A tissue.[2] Ancient schwannoma is a rare subtype of schwannoma with degeneration and hypocellular matrix.[3] Histopathological features include relative reduction of hypercellular Antoni type A tissue, cystic necrosis, calcifications, hyalinisation and degenerative

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nuclei which may be misdiagnosed as pleomorphism of sarcoma.\textsuperscript{[4]}

Since schwannoma is a slow-growing tumour, the clinical presentation of intracranial schwannomas varies between no symptoms to cranial nerve palsy. Symptoms may also be due to mass effect over the adjacent structures, denervation injury or hydrocephalus. Herein, we present schwannomas of most of the cranial nerves, relevant anatomy of cranial nerves, clinical presentation and differential diagnoses.

**Imaging Features of Cranial Nerve Schwannoma**

Among intracranial schwannomas, vestibular schwannoma constitutes 90%, followed by trigeminal, facial and other lower cranial nerve schwannomas. Schwannoma is typically iso-hypointense on T1-weighted images, hyperintense on T2 and FLAIR images and shows variable enhancement on contrast administration. Heterogeneous appearance and enhancement pattern are due to necrosis, cystic degeneration and haemorrhage. Haemorrhage is seen as a loss of signal on gradient imaging. 3D heavily T2-weighted fast spin-echo and fast gradient echo techniques are useful for assessing the fine anatomic details of the cranial nerve and its relationship with the lesion. Target appearance on T2-weighted image with central hypointensity and peripheral hyperintensity which is typically described in neurofibroma can also be observed in schwannoma.\textsuperscript{[5]} On computed tomography (CT), associated bony changes like bone remodelling and widening of neural foramen are better visualised.

**Olfactory Schwannoma**

Schwannoma can arise from almost all cranial nerves with exception of olfactory and optic nerves which lacks Schwann cell layer and only have oligodendrocytes. Developmental and non-developmental hypothesis explain the origin of olfactory schwannoma. The developmental hypothesis states that ectodermal Schwann cells are transformed from mesenchymal pial cells while non-developmental hypothesis states that olfactory schwannoma arises from the adjoining perivascular nerve plexus, meningeal branches of 5\textsuperscript{th} cranial nerve and ethmoidal nerves in the anterior cranial fossa and olfactory groove.\textsuperscript{[6]}

Murakami et al. state that the fila olfactoria has Schwann cell sheath at approximately 0.5 mm beyond the level of the olfactory bulb and can thus give rise to a schwannoma.\textsuperscript{[7]}

Sensory cells for smell are placed in the olfactory epithelium in the superior nasal cavity, penetrating axons pass through the cribriform plate, the expanded portion of the nerve which is the olfactory bulb is placed in the olfactory groove and the olfactory nerve courses between the gyrus rectus and medial orbital gyrus. Therefore, olfactory nerve schwannomas may involve the superior nasal cavity, olfactory groove and anterior cranial fossa. Clinical presentation is nasal block, anosmia and congestion. If there is an orbital or intracranial extension of the mass, the clinical presentation varies between proptosis, visual disturbances and raised intracranial pressure. Differential diagnoses to be considered are schwannoma from maxillary branches of the trigeminal nerve [Figure 1], squamous cell carcinoma, lymphoma, esthesioneuroblastoma which usually show aggressive imaging features.

**Orbital Schwannomas**

Optic nerve schwannomas are rare as it is a white matter tract, lacks Schwann cells like olfactory nerve and myelinated by oligodendrocytes. Schwannomas may arise from the small sympathetic fibres of perivascular nerve plexus that innervate the vasculature around the optic nerve and sheath\textsuperscript{[8]} [Figure 2]. Orbital schwannoma can arise from the cranial nerves 3, 4 and ophthalmic division of 5\textsuperscript{th} cranial nerve which course from the cavernous sinus to the superior orbital fissure and orbit. Clinical presentation varies from headache, retro-orbital pain, proptosis to visual field defects. Differential diagnoses are haemangioma, neurofibroma, venovascular malformations, meningioma, lymphoma, glioma and metastases.

**Oculomotor Nerve**

Oculomotor nerve arises from two nuclei in midbrain namely oculomotor nucleus and Edinger-Westpal nucleus (accessory parasympathetic) which are placed anterior to the aqueduct, the nerve traverses the midbrain...
and emerges into the interpeduncular cistern. The nerve travels the prepontine cistern between the superior cerebellar artery and posterior cerebral artery and enters the cavernous sinus, runs along the superolateral aspect and enters the orbit via superior orbital fissure. Third cranial nerve innervates the extraocular muscles, namely, superior, medial and inferior recti and inferior oblique muscles and levator palpebrae superioris. Clinical presentation of patient’s with schwannoma in the third cranial nerve [Figures 3 and 4] includes diplopia, unable to elevate the upper eyelid resulting in ptosis and lack of coordination of extraocular muscles for vision tracking and fixation. Autonomic parasymptathetic component of the third cranial nerve innervates the ciliary muscle and sphincter papillae, lesions in the nerve may result in pupillary dilatation and unresponsiveness. Differential diagnoses of the third cranial nerve schwannoma are meningioma, the perineural spread of malignancy, aneurysm and lymphoma.

**Trochlear Nerve**

The trochlear nerve is the only cranial nerve that exits from the dorsal midbrain and it has the longest intracranial course. After exiting from the midbrain, the nerve passes anteriorly, along with the ambient cistern and runs alongside the 3rd cranial nerve. The nerve pierces the dura between the attached and free borders of the tentorium. The nerve runs in the lateral wall of the cavernous sinus with third, sixth, ophthalmic and maxillary division of 5th nerves before entering the orbit via superior orbital fissure. Fibres from the nucleus cross the midline before exiting the midbrain, so the nerve fibres innervate the opposite side superior oblique muscle, which is the only muscle innervated by trochlear nerve. Schwannoma in trochlear nerve [Figure 5] clinically present as vertical diplopia, exacerbated by looking down and in activities such as climbing the stairs and reading.

**Figure 2 (A and B):** Optic nerve schwannoma. Coronal T2 fat-suppressed (A) and axial post-contrast T1-weighted (B) images show a well-defined heterointense mass in the intraconal compartment of the left orbit involving the left optic nerve with inhomogeneous enhancement. This patient is a case of NF2 with multiple intracranial schwannomas. No biopsy was done. Symptomatic lesions were treated with radiosurgery.

**Figure 3 (A-D):** Oculomotor nerve schwannoma. Axial T1-weighted (A) and T2-weighted (B) images show a heterointense lesion in interpeduncular cistern arising from the right oculomotor nerve. Axial (C) and coronal (D) post-contrast T1-weighted images show heterogeneous enhancement with non-enhancing foci of necrosis. No surgery was done. The lesion was treated with cyberknife therapy.

**Figure 4 (A and B):** Another oculomotor nerve schwannoma. Axial B-FFE (A) and (B) post-contrast T1-weighted images show a well-defined nodular lesion (arrow) arising from the left oculomotor nerve with significant enhancement. This 61-year-old woman presented with left eye ptosis and underwent cyberknife surgery. No surgery was done.

**Figure 5 (A and B):** Trochlear nerve schwannoma in a 53-year-old man with neurofibromatosis 2. Axial B-FFE (A) and post-contrast T1-weighted (B) images show well-defined nodule (arrow) arising from a trochlear nerve in the right ambient cistern. The nodule shows homogeneous enhancement on contrast imaging. Enhancing lesions are also seen in the right cerebellopontine cistern and para sellar regions, are part of vestibular and trigeminal schwannomas. No biopsy of trochlear nerve schwannoma was done. The lesion has been included in cyberknife therapy of vestibular schwannoma.
Differential diagnosis includes aneurysm and perineural spread of neoplasm.

**Trigeminal Nerve**

The trigeminal nerve is the largest intracranial nerve.\(^{[12]}\) The nerve arises from the lateral surface of mid pons. It has a larger sensory root which is laterally placed to the thinner
The nerve traverses the prepontine cistern anterolaterally and pierces the dura to enter the Meckel’s cave, which is a CSF containing space, placed posterolaterally to the cavernous sinus. In an anterior aspect of the Meckel’s cave, the trigeminal ganglion is enclosed which then divides into 3 subdivisions, namely, ophthalmic (V1), maxillary (V2) and mandibular (V3) branches. Ophthalmic and maxillary divisions contain sensory fibres which enter the lateral aspect of the brainstem and nucleus tractus solitarius. The mandibular division contains motor fibres which innervate the muscles of mastication.

As this patient is a recurrent biopsy-proven case of glossopharyngeal schwannoma, further radiotherapy was given.
of the cavernous sinus and exit the skull via the superior orbital fissure and foramen rotundum, respectively. The mandibular division which has motor fibres for muscles of mastication exits inferiorly via the foramen of ovale. Trigeminal schwannomas are the second most common intracranial schwannomas. Schwannoma of the trigeminal nerve can involve the preganglionic segment which is cisternal, the ganglionic segment which is in Meckel’s cave [Figure 6] or postganglionic which is in cavernous sinus [Figure 7] or foraminal [Figure 8]. Schwannoma involving both preganglionic and ganglionic segment has a typical dumb-bell appearance. Clinical presentations include facial numbness, pain and tingling sensation. Denervation changes can be seen in muscles of mastication if the lesion involves the mandibular division. Differential diagnoses include cerebellopontine angle meningoima, large vestibular schwannoma, ependymoma, the perineural spread of metastasis, atypical pituitary macroadenoma, internal carotid artery aneurysm and skull base lesions such as chondrosarcoma and chordoma.

Abducens Nerve

Abducens nerve arises from the abducens nucleus which is placed anterior to the fourth ventricle in dorsal pons. The nerve courses anteriorly and exits from the pontomedullary junction then traverses the preopticine cistern from posterior to the anterior direction and enters the Dorello’s canal by piercing the dura near the posterior aspect of the clivus. Successively the nerve runs along the central portion of the cavernous sinus and enters the orbit through the superior orbital fissure to innervate the lateral rectus muscle. Schwannoma of abducens nerve can be seen anywhere along its course in preopticine cistern [Figure 9], Dorello’s canal, cavernous sinus or superior orbital fissure. The clinical presentation would be diplopia due to lateral rectus palsy. Secondary denervation injury of lateral rectus will be seen as the increased signal intensity of muscle on T2-weighted images or atrophy with fatty change. Differential diagnosis includes meningioma, the perineural spread of malignancy, lymphoma and cavernous haemangioma.

Facial Nerve

The facial nerve has one motor, one secretomotor and one sensory nucleus in dorsal pons. The nerve emerges from the posterolateral aspect of the pons and the cisternal segment runs anterolaterally towards the porus acusticus. A canalicular segment of the facial nerve is placed anterosuperior. A labyrinthine segment of the nerve extends from the internal auditory canal to geniculate ganglion. The tympanic segment starts from the geniculate ganglion runs to the pyramidal eminence. In the tympanic cavity, the facial nerve runs superolateral to the oval window. The mastoid segment runs from the pyramidal eminence to stylomastoid foramen. An extratemporal segment of facial nerve enters the parotid gland and divides into five main branches namely frontal, zygomatic, buccal, mandibular and cervical. Facial nerve schwannoma [Figure 10] can involve any of these segments among which geniculate ganglion is the most common location. Clinical presentation is via facial nerve paresis. Differential diagnoses include cerebellopontine angle meningoima, schwannoma of other lower cranial nerves, epidermoid and the perineural spread of metastases. The differential diagnosis for a temporal segment of facial nerve schwannoma includes...
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Figure 18 (A-F): Another patient with neurofibromatosis 2: Post-contrast administration axial T1 (A-F) and Axial B-FFE (d) images show left optic nerve (A), bilateral vestibular (B) and trigeminal (B) schwannomas. Left trochlear nerve (C and D), bilateral jugular foramen schwannomas (F) and meningioma in left cerebello medullary cistern (E) are also seen. No biopsy was done.

Veins. Inferiorly the nerve traverses the thorax and reaches the abdominal cavity via oesophageal hiatus. Vagus nerve schwannomas can involve the cerebello-medullary cistern, jugular foramen, suprathyroid [Figure 13] or infrathyroid carotid spaces and splay the carotid bifurcation, carotid arteries and internal jugular vein [Figure 14]. Clinical presentation varies between no symptoms to vocal cord paresis and clinical features of other lower cranial nerve paresis may present due to compression. Differential diagnoses are similar to glossohypopharyngeal nerve schwannoma which include schwannoma of 9, 11th cranial nerves and paragangliomas of the jugular foramen, supra and infra hyoid carotid spaces.

**Spinal Accessory Nerve**

Spinal accessory nerve has cranial and spinal rootlets. Cranial rootlets arise from the lateral aspect of medulla below the vagus nerve. Spinal rootlets arise from ventral horn cells of the upper (C1–C5) segment of the cervical spinal cord. These rootlets join together to form a single trunk and pass towards the foramen magnum. In cerebello-medullary cistern, spinal and cranial rootlets join together and traverse towards the pars vascularis of the jugular foramen. The nerve passes posterior to the glossohypopharyngeal and vagus nerves in the jugular foramen and exits the skull. Spinal accessory nerve schwannomas can involve the cerebello-medullary cistern [Figure 15], jugular foramen and suprathyroid carotid space. Clinical presentation includes difficulty in shrugging the shoulder and inability to rotate the neck secondary to denervation injury of sternomastoid and trapezius muscles, respectively. Differential diagnosis includes schwannoma of 9, 10th cranial nerves and paragangliomas of jugular foramen and suprathyroid carotid space.

**Hypoglossal Nerve**

The hypoglossal nerve is a pure motor nerve which supplies the intrinsic and extrinsic muscles of the tongue. The nucleus is located in front of the fourth ventricle, the nerve fibres traverse the medulla in paramedian location and exits as multiple rootlets in the ventrolateral aspect between the pyramid and olivary nucleus. The nerve traverses the cerebello medullary cistern anterolaterally and enters the bony hypoglossal canal. While passing through the cerebello medullary cistern the nerve is posterior to the vertebral artery and anterior to the posterior inferior cerebellar artery. In the neck, the nerve traverses medial to 9th to 11th cranial nerves, then lies deep to dia
gastic muscle and supply the muscles of the tongue. Clinical presentation includes dysarthria, difficulty in moving the tongue, deviation of the tongue to one side and hemiatrophy. Hypoglossal schwannoma [Figure 16] can involve cerebello medullary cistern, hypoglossal canal, suprathyroid neck and floor of the mouth. Hemiatrophy and fat intensity of one half of the tongue on MRI [Figure 16D] will clinch the diagnosis of hypoglossal schwannoma. Differential diagnoses are paraganglioma of carotid space, squamous cell carcinoma and salivary gland tumours involving lingual space and floor of the mouth.

**Conclusion**

Knowledge about the anatomy of cranial nerves, clinical presentation, characteristic clinching imaging features of each one of the cranial nerve schwannomas like ‘ice cream cone’ appearance of vestibular schwannoma, an extension of enhancement to the geniculate ganglion in facial nerve schwannomas, the involvement of foramen ovale and extracranial extension of the mandibular division of trigeminal schwannomas, infrathyroid carotid space extension of vagal schwannomas , syndromic association of multiple cranio- spinal schwannomas like neurofibromatosis 2 [Figures 17-19] and secondary imaging features like denervation injury of end organs will aid the radiologist in establishing the diagnosis and thereby treatment planning of intracranial schwannomas.

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**Conflicts of interest**

There are no conflicts of interest.
Figure 19 (A and B): (A) One another patient of NF2: Bilateral vestibular schwannomas (a and b), bilateral trigeminal nerve schwannomas extending to cavernous sinuses (a and b), right hypoglossal nerve (d) and left upper cervical (c) schwannomas are seen. No biopsy is done. Symptomatic lesions treated with radiosurgery. (B) Same patient with NF2: Numerous spinal nerve root schwannomas are seen in the cervical, dorsal and lumbosacral spine. Schwannoma is also seen in the subcutaneous plane of the posterior lower chest wall (d). Intra medullary ependymoma (arrow in a) is also seen in the cervical cord at C1 level. No biopsy was done. Symptomatic lesions were treated with radiosurgery.

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