ABSTRACT

Trigeminal schwannoma is a rare cause of cerebellopontine angle tumour. This case report of a 65-year-old lady presented with ipsilateral facial numbness and instability. She was finally diagnosed to have trigeminal schwannoma after seeking multiple medical consultations with her doctors. This case report highlights a rare cause of cerebellopontine angle tumour.

Keywords: cerebellopontine angle tumour, trigeminal schwannoma, schwannoma

INTRODUCTION

Trigeminal schwannoma is a rare cause of cerebellopontine angle tumour as it only constitutes 0.2 – 0.4% of all intracranial tumours. This tumour is slightly more common in female than male and usually occurs in the age group of 30 to 40s. The tumour grows slow and may cause compression to the surrounding structure. Due to the slow growing nature, patient with trigeminal schwannoma may experience symptoms for a long time before seeking medical attention and rarely presented with features of raised intracranial pressure as there is sufficient time to compensate for the increase pressure. With the advancement of imaging techniques and refinement of neurosurgical methods, trigeminal schwannoma is highly treatable with a good prognosis if detected and acted upon timely.

CASE PRESENTATION

A 65-year-old lady with dyslipidaemia on diet control under health clinic follow-up. She was referred to the medical outpatient clinic with the complaint of left-sided facial numbness for a year. She initially noticed the symptom of numbness describing as ‘pins and needles’ over the left side of her face occurring intermittently. However, for the past 3 months, the facial numbness was consistently associated with instability when walking. She felt that her body swayed to the left side when she walks and there was fullness felt in her left ear. She had sought multiple medical attentions at various private general practitioners without any symptomatic relief. There was no associated headache, blurring of vision or vomiting to suggest raised intracranial pressure. Apart from instability, her four limbs were of normal strength without any numbness or pain sensation. Prior to the facial numbness, there was no preceding fall or injury to her face. The numbness was not associated with lancinating facial pain suggestive of trigeminal neuralgia. She had no difficulty in chewing and swallowing food or drink and no drooling of saliva. She has no history of hypertension or diabetes. Her family history was otherwise unremarkable for any brain tumour or neurocutaneous disorder like neurofibromatosis.

Assessment revealed a full Glasgow Coma Scale with normal speech. Her face appeared symmetrical without any involuntary twitching of the facial musculature. Both eyelids were symmetrical without any obvious ptosis or proptosis; pupils were equal and reactive. There was no obvious scar or skin lesions like neurofibroma appreciated on her face and any other part of body. Local palpation of the temporomandibular joints was non-tender, no crepitus felt on passive closure of her mouth. Oro-buccal cavity assessment revealed good dental hygiene without any dental carries. There
was reduced pin prick and light touch sensation on the left side of her face in the V<sub>1</sub> to V<sub>3</sub> distribution of the trigeminal nerve. The corneal reflex was impaired on both sides. Muscle bulk and power of mastication muscle was intact both sides. Auditory assessment revealed Rinne’s test of air conduction better than bone condition for both ears; Weber’s test was centralised. Formal audiometry assessments were done (Table 1). Otherwise other cranial nerves assessments were unremarkable.

All four limbs showed normal tone, power and sensation with intact normal tendon reflexes. Coordination was intact with negative Romberg sign.

The differentials for ipsilateral facial numbness includes trauma to the face and its underlying nerve branches, dental pathology with referred pain to the ipsilateral face, lesions involving the trigeminal nerve along its route from the brainstem to the cerebello-pontine space, cavernous sinus and lastly skull exit foramina. There was neither associated crossed long tract sign nor ophthalmoplegia to suggest brainstem and cavernous sinus involvement respectively.

As she had ipsilateral reduced sensation of V<sub>1</sub> – V<sub>3</sub> distribution of trigeminal nerve, with absence of corneal reflex on both sides and subjective ipsilateral instability over a year duration; cerebello-pontine angle tumour need to be considered.

We proceeded with Computed Tomography of the Brain for her case which revealed a suspicious cerebro-pontine angle mass lesion. A Magnetic Resonance Imaging of the Brain was performed to further delineate the nature and extent of the mass in relation to the surrounding structures (Figure 1).

Figure 1 (A) MRI axial view showing tumour occupying the left cerebello-pontine angle; (B) A more rostral axial view showing the same tumour compressing the adjacent pons; (C) MRI coronal view showing tumour compressive effect on the brainstem. The tumour measured at 2.8 cm (W) × 2.7 cm (H) × 3.5 cm (AP). The tumour lies superior and medial to the left internal auditory canal without any extension into the canal. The pons, left middle cerebellar peduncle and the 4th ventricle are compressed by the tumour without any hydrocephalus or significant midline shift.

Table 1 Formal Audiometry Assessment

<table>
<thead>
<tr>
<th></th>
<th>Left Ear</th>
<th>Right Ear</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tympanometry</td>
<td>Type As bilateral suggestive of middle ear stiffness</td>
<td>Normal bilateral</td>
</tr>
<tr>
<td>Pure Tone Audiometry</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Once diagnosed with cerebello-pontine angle tumour, she was referred promptly to neurosurgical team for evaluation. She underwent neurosurgical operation for excision of the tumour, followed by Gamma Knife Surgical Intervention to remove the residual tumour. The intraoperative specimen was sent for histopathological examination which was later reported as trigeminal schwannoma. Six months post-operation, she still had left-sided reduced pin prick and light touch sensation over V₁ – V₂ distribution with intact corneal reflex. Other aspects of neurological assessment were unremarkable. She was able to ambulate without feeling unstable unlike prior to neurosurgical intervention. Figure 2 shows residual tumour after neurosurgical intervention.

DISCUSSION

Cerebellopontine (CP) angle is a shallow triangle space bounded by the undersurface of cerebellar hemisphere, lateral aspect of pons and the superior surface of the inner third petrous ridge. This space spans cranially from the cranial nerve V<sup>th</sup> to the cranial nerve IX<sup>th</sup> rostrally. In view of the close proximity of the structures, a lesion in CP angle (mostly tumours) causes a constellation of signs and symptoms. The extent of involvement depends on the location and size of the lesion. CP angle tumours consist of 5 – 10% of all intracranial tumours. Of all these CP angle tumours, 80 – 90% comprise of vestibular schwannoma or better known as acoustic neuroma. The remainder 10 – 20% consists of trigeminal schwannoma, epidermoid cysts, dermoid cysts, meningioma, arachnoid cysts, lipomas and secondary tumours. Trigeminal schwannoma as in our case constitutes only 0.2 to 0.4% of all intracranial tumours. Hence it is a very rare cause of CP angle tumours.

Trigeminal schwannoma is a benign tumour of the Schwann cells which surrounds the trigeminal nerve. This tumour can arise from the trigeminal nerve root entry zone, the Gasserian ganglion, or any of the three branches of trigeminal nerves, namely ophthalmic, maxillary and mandibular branches. Patients with trigeminal schwannoma commonly present with facial numbness, pain, hypoesthesia like this case. Other clinical features include diplopia, gaze abnormalities, nystagmus, wasting of mastication muscles. Less frequently, they may have hearing loss, ataxia, dysarthria or pathological crying due to brainstem compression. The symptomatology varies largely depending on the size and location of the lesion. In view of the benign nature, most trigeminal schwannoma grows slowly and are treatable with neurosurgical intervention. Magnetic Resonance Imaging of the brain is the best diagnostic imaging modality to guide neurosurgical intervention given its high soft tissue resolution compare to computer tomography of the brain. The outlook and prognosis have gotten better given the improvement in diagnostic imaging and operative techniques.

We highlight the challenges in the diagnosis of cerebellopontine (CP) angle tumour. The patient’s symptom had been there for over a year and the only reason that prompted her to seek medical consultation was her facial numbness that had been more frequent.
of late. Her symptom was overlooked as CP angle tumour, a rare diagnosis especially more so in our setting. The authors had only seen two cases of CP angle tumour in a year including the present case. During her initial visit to the doctors, there was no prompting to perform brain imaging given the rather subjective sign and symptom. Her presentation of ipsilateral facial numbness with subjective instability was typical for CP angle tumour. The finding of large tumour upon diagnosis was expected given that she had been experiencing the symptoms for a year before diagnosis. However, the absence of other neurological deficits was a surprise given that there was significant compression of the surrounding brain structures in particularly the brainstem. The absence of corneal reflex on the contralateral side when she initially presented could well represent a false localising sign yet there was no other features like abducens nerve palsy to support this postulation. Reduced facial sensation to light touch and pin prick persisted postoperatively as there was still residual tumour. Complete resection was not possible given the close proximity of tumour to the adjacent cranial nerves and brainstem.

CONCLUSION

Cerebellopontine angle tumour, although rare, need to be considered for any individual who presented with facial symptoms and subjective ipsilateral instability due to the good prognosis if detected early and treated. We have highlighted the challenges in diagnosing cerebellopontine angle tumour in our healthcare setting which could be due to unfamiliarity of this clinical entity.

ACKNOWLEDGEMENTS

The authors thank the Director General of Health, Ministry of Health Malaysia for his permission to publish this case report.

CONFLICT OF INTEREST

The authors declare that they have no competing interests.

CONSENTS

Written informed consent was obtained from the patient to publish the case with its related pictures. A copy of the written consent is available for review by the Chief Editor of this journal.

REFERENCES