Case Report

A rare case of arteriovenous malformation in the internal acoustic meatus

Ruth Ng, Roslenda Abdul Rahman, Azizi Abu Bakar, Asma Abdullah

Department of Otorhinolaryngology Head and Neck Surgery, Universiti Kebangsaan Malaysia Medical Centre (UKMMC), Kuala Lumpur, Malaysia

Arteriovenous malformation (AVM) is a high flow vascular malformation, in which arteriovenous shunting occurs in a central nidus. AVM in the internal acoustic meatus is rare. We report a case of 53 year-old lady presenting with right hearing associated with hemifacial hypoaesthesia and facial asymmetry for 2 years. Preoperatively working diagnosis was acoustic neuroma and excision of tumor via translabyrithine approach was performed. Histophathological result revealed as AVM of internal acoustic meatus (IAM). Post-operatively, the hemifacial hypoaesthesia had complete resolution and there was improvement of the facial asymmetry. (Rawal Med J 201;41:250-252).

Keywords: Arteriovenous malformation, internal acoustic meatus, translabyrithine approach.

INTRODUCTION

Arteriovenous malformation (AVM) is tangled blood vessels in which multiple feeding arteries converge and enlarged veins drain with the arteriovenous shunting occurs in a central nidus. It is a high flow vascular lesion.[1] The chronic high flow condition results in endothelial thickening and intima hyperplasia of the feeding arteries and dilated draining veins.[1] AVM in the internal acoustic meatus is a rare condition. In English literature, less than 10 cases have been reported.[2] Here, we report another case of AVM in the internal acoustic meatus.

CASE PRESENTATION

This 53 year-old lady with underlying hypertension, dyslipidemia and bronchial asthma presented with history of worsening right hearing for 2 years associated with intermittent right tinnitus. She developed right facial hypoaesthesia for the past year, which is persistent. Six months later, she had right facial asymmetry. She denied any vertigo, otalgia and otorrhea. She had no history of ear trauma or ear infection. There were no symptoms of raised intracranial pressure symptoms such headache, nausea, vomiting and giddiness.

On clinical examination, sensations were reduced at the right trigeminal nerve over V2/V3 distribution, however right corneal reflex was present. There was right facial nerve palsy, with House-Brackmann grade V with Bell's phenomenon. No right eye redness was seen. Other cranial nerves examination appeared grossly intact and other ENT examinations were normal. The pure tone audiogram (PTA) revealed right profound hearing loss and left normal hearing with both tympanogram showed type A.

MRI brain and IAM showed an avidly enhancing homogenous lobulated lesion in the right intracanalicular portion of the internal auditory meatus extending to the cerebellopontine angle, measuring about 1.1x1.0x1.2 cm, with normal flow void signal (Fig. 1). T1 weighted and T2 weighted FLAIR MRI scan revealed hypointense and hyperintense lesion, respectively (Fig. 2 and 3). There was no enhancement noted at the labyrinthine segment of facial nerve, (unlikely facial schwannoma); no dural tail was seen,(unlikely meningioma). Radiological impression was acoustic neuroma.

Subsequently, Otorhinolaryngology and Neurosurgery teams performed excision of the tumor via translabyrinthine approach. Introperatively, the mass was noted to be arising from superior vestibular nerve and extending to cerebellopontine angle. It appeared reddish and vascular (Fig. 4). Upon removal of the mass, massive bleeding occurred with estimated blood loss of 1500cc, which required intraoperative blood transfusion. Most of the tumor (95%) was removed

and the remaining mass was adherent to the facial nerve and was not removed.

The Histopathology examination showed fragments of vascular spaces lined by flattened endothelial cells (CD31 and CD34 positive). The vascular spaces were separated by fibrous septa. There were some thick walled blood vessels seen. The underlying stroma is composed of spindle smooth muscle cells (SMA positive). Immunohistochemical stain for S100 was negative. There was no evidence of malignancy.

Fig. 1. Post-gadolinium MRI in axial view showed enhancing homogenous lobulated lesion in the right intracanalicular portion of the internal auditory meatus extending to the cerebellopontie angle (arrow).

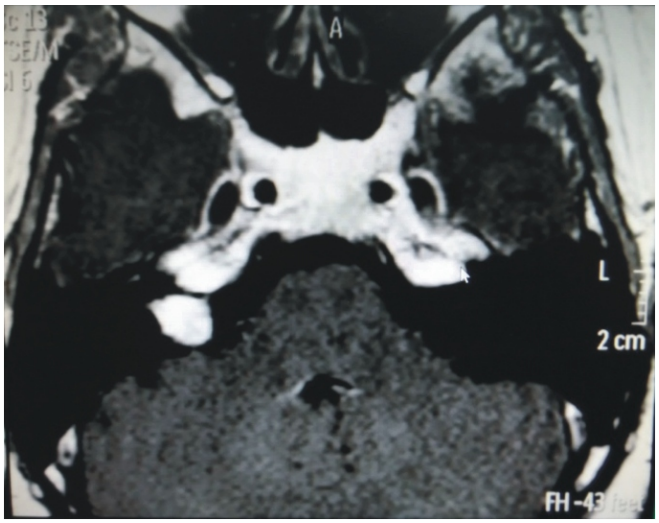

Fig. 2. T1 weighted MRI in axial view showed hypointense lesion in the right intracanalicular portion of the internal auditory meatus extending to the cerebellopontie angle (arrow).

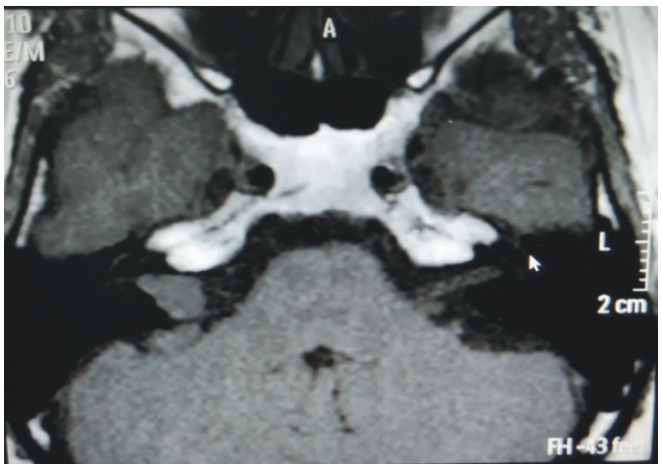

Fig. 3. T2 weighted FLAIR MRI in coronal view showed hyperintense lesion in the right intracanalicular portion of the internal auditory meatus extending to the cerebellopontie angle (arrow).

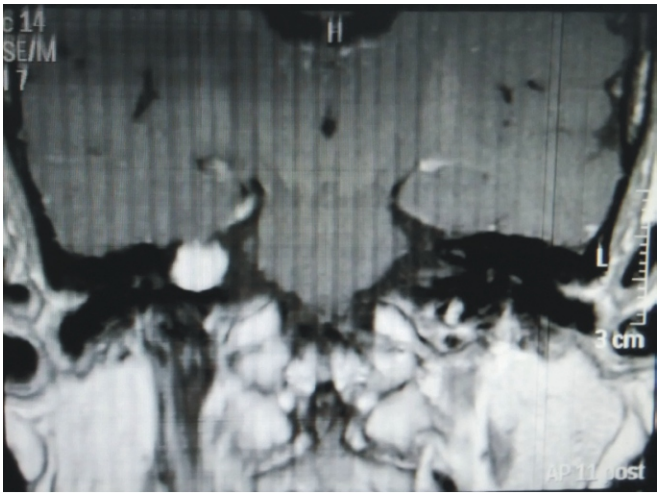

Fig. 4. The mass (arrow) appeared reddish and vascular.

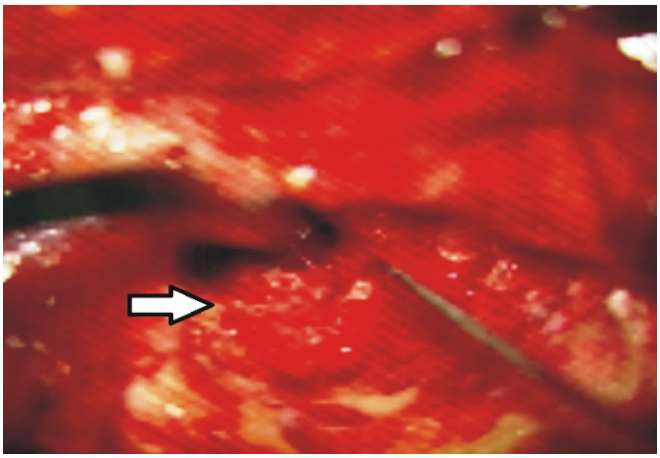

Fig. 5. Right facial nerve palsy 5 months after surgery. A, House Brackmann grade IV of the right eye closure with Bell's phenomenon. B, House Brackmann grade III for the right nasolabial movement.

Five months post-surgery, right facial hypoaesthesia was fully recovered. Right facial nerve palsy improved to House brackmann grade IV for the eye closure and grade III for the nasolabial fold movement (Fig. 5).

DISCUSSION

Tumor in the IAM can present with unilateral hearing loss, hemifacial hypoasthesia, facial weakness, tinnitus and vertigo.[2] Trigeminal neuralgia can be also the only presenting symptom and resolv after radiosurgery.[3] Kim et al reported a case with hemifacial spasm with AVM in the cerebellopontine angle and the symptom was completely resolved after excision.[4] The presenting symptoms of AVM in IAM and cerebellopontine angle are different from the presentation of cerebral AVM. The presentation of cerebral AVM include hemorrhage, stroke, seizure and headache.[1] In our case, complete recovery of the hypoaesthesia in the trigeminal distribution and the improvement of the facial nerve palsy was observed after the surgery.

The typical MR imaging of AVM finding is flow voids signal lesion, which is a result of the turbulent flow of the lesion. Angiography has been suggested before any intervention.[5] However, in our case the MRI findings of the lesion appeared like acoustic neuroma, which were hypointense in T1, hyperintense in T2 FLAIR and enhanced in post-gadolinium scan without flow void signal.[6] Despite the discrepancy between the MRI findings and histopathological findings, MRI scan is still considered a useful imaging modality to detect small internal acoustic meatus tumor.[7]

Literatures had reported the recurrence of cerebral AVM despite complete resection. The recurrence interval widely ranged from 3 months to 10 years.7-9 These patients were followed up and monitored by angiography, either by Digital Subtraction Angiography (DSA) or CT angiogram. However, routine surveillance imaging to look for recurrence is still debatable.8-10 Unfortunately, there is inadequate data in literature regarding the recurrence of the AVM in IAM and cerebellopontine angle. Nevertheless, follow up of patient up to 10 years is needed to assess any recurrence clinically and radiological imaging needs to be requested if necessary. In summary, AVM in the internal acoustic meatus is a rare condition. The presentation of this lesion can be similar with any tumorlike acoustic neuroma.

Author contributions:
Conception and design: Asma Abdullah
Collection and assembly of data: Ruth Ng, Roslenda Abdul Rahman
Analysis and interpretation of the data: Ruth Ng, Azizi Abu Bakar
Drafting of the article: Ruth Ng
Critical revision of the article for important intellectual content: Roslenda Abdul Rahman
Final approval and guarantor of the article: Asma Abdullah
Corresponding author email: ruth_ng_2511@yahoo.com
Conflict of Interest: None declared
Rec. Date: Dec 9, 2015 Accept Date: Mar 1, 2016

REFERENCES

1. Al-Shahi R, Warlow C. A systematic review of the frequency and prognosis of arteriovenous malformations of the brain in adults. Brain 2001;124:1900-26.
2. Patel PN, Connor S, Brew S, Gleeson MJ. An arteriovenous malformation within the internal acoustic meatus and cerebellopontine angle cistern. J Laryngol Otol 2011;125:1275-8.
3. Anderson WS, Wang PP, Rigamonti D. Case of microarteriovenous malformation-induced trigeminal neuralgia treated with radiosurgery. J Headache Pain 2006;7:217-21.
4. Kim Y, Tanaka A, Kimura M, Yoshinaga S, Tomonaga M. Arteriovenous malformation in the cerebellopontine angle presenting as hemifacial spasm. Neurologia medico-chirurgica 1991;31:109-12.
5. Lowe LH, Marchant TC, Rivard DC, Scherbel AJ. Vascular malformations: classification and terminology the radiologist needs to know. Sem Roentgenol 2012;47:106-117.
6. Haque S, Hossain A, Quddus MA, Jahan MU. Role of MRI in the evaluation of acoustic schwannoma and its comparison to histopathological findings. Bangladesh Med Res Council Bull 2011;37:92-6.
7. Freudenstein D, Duffner F, Ernemann U, Rachinger J, Grote EH. Recurrence of a cerebral arteriovenous malformation after surgical excision. Cerebrovasc Dis 2001;11:59-64.
8. Wostrack M, Meyer B, Stoffel M. Asymptomatic arteriovenous malformation recurrence in an adult. Acta Neurochirurgica 2011;15:1821-3.
9. McCarthy C, Kaliaperumal C, O'Sullivan M. Recurrence of a paediatric arteriovenous malformation 9 years postcomplete excision: case report and review of literature. BMJ Case Rep 2012;2012:bcr2012006826.
10. Iżycka-Świeszewska E, Szurowska E, Kloc W, Rzepko R, Dubaniewicz-Wybieralska M, Skorek A, et al. Cerebellopontine angle tumours: radiologic-pathologic correlation and diagnostic difficulties. Folia Neuropathol 2006;44:274-81.