ORIGINAL ARTICLE

Arteriovenous malformation of the external ear: a clinical assessment with a scoping review of the literature

Shin Hye Kim a,b, Seung Hoon Han b, Yoonjae Song b, Chang Sik Park c, Jae-Jin Song b,*

a Korea University Medical Center, Korea University College of Medicine, Department of Otorhinolaryngology-Head and Neck Surgery, Seoul, Republic of Korea
b Seoul National University Bundang Hospital, Seoul National University College of Medicine, Department of Otorhinolaryngology-Head and Neck Surgery, Seongnam, Republic of Korea
c Seoul National University Bundang Hospital, Seoul National University College of Medicine, Department of Plastic Surgery, Seongnam, Republic of Korea

Received 20 May 2016; accepted 20 September 2016
Available online 17 October 2016

KEYWORDS
Ear; Tinnitus; Arteriovenous malformations; Embolization, therapeutic; Surgical procedures, operative

Abstract
Introduction: Auricular Arteriovenous Malformation of the external ear is a rarely encountered disease; in particular, arteriovenous malformation arising from the auricle, with spontaneous bleeding, has seldom been reported.
Objective: In the current study, we report an unusual case of late-onset auricular arteriovenous malformation originating from the posterior auricular artery that was confirmed by computed tomographic angiography. The case was successfully managed by pre-surgical intravascular embolization followed by total lesion excision. Prompted by this case, we also present a scoping review of the literature.
Methods: A case of a 60-year-old man with right auricular arteriovenous malformation treated in our tertiary care center, and 52 patients with auricular arteriovenous malformation described in 10 case reports and a retrospective review are presented. Auricular arteriovenous malformation can manifest as swelling of the ear, pulsatile tinnitus, pain, and/or bleeding. On physical examination, a pulsatile swelling and/or a tender mass is evident. When arteriovenous malformation is suspected, the lesions should be visualized using imaging modalities that optimally detect vascular lesions, and managed via embolization, mass excision, or auricular resection. Effectiveness of the various diagnostic methods used and the treatment outcomes were analyzed.

* Please cite this article as: Kim SH, Han SH, Song Y, Park CS, Song J-J. Arteriovenous malformation of the external ear: a clinical assessment with a scoping review of the literature. Braz J Otorhinolaryngol. 2017;83:683–90.
* Corresponding author.
E-mail: jjsong96@gmail.com (J. Song).
Peer Review under the responsibility of Associação Brasileira de Otorrinolaringologia e Cirurgia Cérvico-Facial.

http://dx.doi.org/10.1016/j.bjorl.2016.09.004
1808-8694/© 2016 Associação Brasileira de Otorrinolaringologia e Cirurgia Cérvico-Facial. Published by Elsevier Editora Ltda. This is an open access article under the CC BY license (http://creativecommons.org/licenses/by/4.0/).
Resultados: Várias modalidades incluindo Doppler sonografia, computado tomográfico angiografia, ressonância magnética angiografia, e/ou transfemoral cerebral angiografia foram usadas para diagnosticar 38 casos reportados na literatura. Em 15 casos, sem qualquer imagem foi realizada, tratamento foi determinado soamente pelo exame físico e auscultação. Do total de 53 casos, 12 não foram tratados (sua symptoms eram apenas observados) versus 20 que foram submetidos a embolização endovascular. Em total, 32 pacientes, incluindo 1 paciente que não foi tratado e 10 com persistente ou exacerbada malformação arteriovenosa após embolização, resultaram massa excisão ou auricular reseção dependendo da extensão da doença. Sem maiores complicações postoperatoriais foram registradas. O tempo médio de seguimento foi registrado. O tempo médio de seguimento foi de 1 mês a 19 anos, e apenas um caso de malformação arteriovenosa cervicocaudal irresecável, residual foi registrado.

Conclusão: A malformação arteriovenosa auricular é uma doença raramente encontrada, mas deve ser suspeitada se um paciente apresentar orelha inchada e zumbido pulsátil. A imagem apropriada é essencial para o diagnóstico e avaliação da extensão da doença. Como a embolização proporciona apenas um controle relativamente precário, a remoção cirúrgica total da lesão vascular é recomendada.

© 2016 Associação Brasileira de Otorrinolaringologia e Cirurgia Cérvico-Facial. Publicado por Elsevier Editora Ltda. Este é um artigo Open Access sob uma licença CC BY (http://creativecommons.org/licenses/by/4.0/).
Introduction

An Arteriovenous Malformation (AVM) is an abnormal connection between one or more arteries and veins, bypassing the capillary system. In most cases, the AVM arises from the intracranial area, but occasionally originates from extracranial vessels. In a retrospective review of 81 patients with AVM of the head-and-neck, the most common site was the cheek (31%), followed by an ear (16%). The AVM is almost always present at birth, but manifests later in life.

Here, we report an unusual case of late-onset auricular AVM originating from the posterior auricular artery. Also, a scoping review of the literature allows us to discuss the roles played by imaging in the diagnosis and management of AVM.

Materials and methods

Search criteria

We adhered to the PRISMA (Preferred Reporting Items for Systematic Reviews and Meta-Analyses) guideline when performing PubMed searches to identify all studies on AVM of the external ear. The keywords used were “arteriovenous malformation” and “ear”, and the search was limited to articles in the English language. AVMs originating from the external auditory canal, or the pre- or retro-auditory areas, were excluded.

Our case and literature review

We treated a 60-year-old man with right-side auricular AVM. The study was approved by the institutional review board of the Clinical Research Institute at our center (B-1601-329-002). The literature review identified a total of 52 AVM patients described in 11 reports (10 case reports and 1 retrospective review). The 10 case reports dealt with 11 cases of auricular AVM and the single review analyzed 41 cases. The chief complaints, duration of symptoms, events that aggravated the AVMs, and history of spontaneous AVM bleeding were analyzed.

Diagnostic process and differential diagnosis

The laterality, location, and extent of AVM were investigated. Various diagnostic imaging modalities including Doppler Sonography (DS), Temporal Bone Computed Tomographic Angiography (TBCTA), Magnetic Resonance Angiography (MRA), and/or Transfemoral Cerebral Angiography (TFCA) were employed in individual cases, and the findings of the key diagnostic modalities were reviewed. In patients who underwent diagnostic angiography, the main arteries and feeder vessels are summarized.

Management options and treatment outcomes

The AVM management options included observation, embolization, mass excision, or auricular resection. After the treatment, the clinical condition was described as controlled, improved, persistent, or aggravated. We also explored the application of reconstructive procedures such as Skin-Thickness Split Grafting (STSG) and total auricular reconstruction. Postoperative complications, follow-up durations, and final status were reviewed.

Results

Our experience with late-onset AVM

A 60-year-old man visited the emergency department with massive spontaneous bleeding from the right ear. He had a history of recurrent swelling of the right auricle, and pulsatile tinnitus. Three years prior, the patient had visited our outpatient clinic with recurrent auricular swelling, and was diagnosed with an otohematoma because of the cauliflower-like appearance of the auricle.

On physical examination, the right auricle exhibited excessive swelling and discoloration (Fig. 1A). The tympanic membrane and external auditory canal were normal. TBCTA revealed enlargement of the right helix, and an entangled vascular lesion (Fig. 1B). We suspected an auricular AVM and performed transarterial embolization using TFCA. The principal AVM-feeding artery originated from the posterior auricular artery, and we completely embolized the artery with glue (Fig. 2A). After embolization, bleeding was controlled, but ischemic necrosis of the auricular skin developed (Fig. 2B and C). Two weeks later, the necrotic skin boundary became distinct (Fig. 3A), and we thus planned total mass excision.

Under local anesthesia, the skin was incised along the vertical plane of the right auricle and the AVM mass was totally excised (Fig. 3B). The adjacent necrotic skin was excised and primary closure was performed (Fig. 3C). The patient recovered without any complications. At 6 months postoperatively, the auricle was well-healed without any evidence of recurrence.

Demographics and symptoms of AVM patients

Table 1 summarizes the findings of 12 studies on 53 cases of auricular AVM. Of 11 case reports, 7 were men and 4 were women, with a median age of 21 (15–45) years. In the retrospective review of 41 cases, the male-to-female patient ratio was not mentioned, and the mean age was 26 (1–55) years. Chief complaints were swelling of the ear, pulsatile tinnitus, intermittent pain, hearing loss, and/or profuse spontaneous bleeding. About two-thirds of patients reported that their symptoms had commenced at a very young age. Seven patients reported that their AVMs became aggravated during puberty, and 6 had histories of trauma.

Diagnosis

Table 1 shows key diagnostic study and their findings. In 38 of the 53 cases, DS, TBCTA, MRA, and/or TFCA were silent in terms of diagnosis. No imaging was performed in the other 15 cases; treatment was determined solely by physical examination and auscultation. On angiography, the most common feeding vessels were the posterior auricular, superficial temporal, and occipital arteries.
Figure 1  Gross findings and temporal bone computed tomographic angiography findings on the ear of a 60 year-old man, as recorded in the emergency room: (A) the patient presented with a swollen ear and spontaneous massive bleeding; (B) temporal bone computed tomographic angiography revealed a right-side, auricular vascular tangled lesion (white arrow).

Figure 2  Preoperative therapeutic embolization using transfemoral cerebral angiography and gross ear findings 3 days after embolization: (A) transfemoral cerebral angiography revealed large tortuous vessels and innumerable small vessels. The principal feeder vessel of the arteriovenous malformation originated from the posterior auricular artery (black arrow), and was completely occluded with glue; (B) and (C) after embolization, the bleeding stopped, but ischemic necrosis of the skin progressed.

Management and treatment outcomes

The flow chart of Fig. 4 shows the treatment options and outcomes. Twelve of 53 patients were initially followed-up without any treatment. Of these, AVMs persisted in 10, and became aggravated in 2. Of the latter 2, one underwent auricular resection but the other rejected an operation. Twenty patients underwent therapeutic embolization. Of these, the AVMs improved in 3, but became aggravated in 17. Of the latter 17, 10 underwent AVM mass excision or auricular resection.

A total of 21 patients underwent AVM mass excision or auricular resection as initial treatment. Of these patients, plus a further 11 who underwent excision or resection as salvage management, 22 (69%) were controlled and 9 (28%) improved, and only one had a persistent unresectable AVM in the adjacent region. In these 32 patients, wound closure was performed by simple linear closure (n = 18); STSG (n = 10); immediate reconstruction using salvaged auricular cartilaginous framework covered by temporoparietal flap (n = 1), expanded local flap (n = 1), advancement flap (n = 1), and free flap (n = 1). One patient underwent a two-staged operation: auricular resection and STSG initially and ear lobule creation and ear elevation secondarily. No major postoperative complication was noted, except for transient tinnitus in one patient and skin necrosis at the site of wound closure in two. The postoperative follow-up duration varied from 1 month to 19 years, and only one case had residual cervicofacial AVM.

Discussion

Schobinger classified AVM into four stages. The symptoms of stage I (quiescence) are warm and discolored skin; those of stage II (expansion) are bruit, pulsation, and swelling.
Stage III (destruction) is characterized by pain, ulceration, and bleeding; whereas stage IV (decompensation) features cardiac failure. The presenting signs and symptoms correlate with the stage of AVM. Although many AVMs are asymptomatic, they may alternatively trigger severe pain and/or bleeding. The most common symptoms are pulsation (51.2%), bleeding (41.5%), and pain (29.3%). Hearing can also deteriorate, presumably because the bruit is audible. There are 2 types of AVM with regard to the flow rate: fast-flowing and slow-flowing. Most of fast-flowing regions are arteriovenous fistulas whereas slow-flow AVMs are produced by venous, capillary, or lymphatic lesions. This flow rate-based classification may be of importance as different treatment options are needed for the 2 types.

Enlargement of an AVM may be triggered by trauma, infection, or hormonal influences. In the review paper of 41 AVM patients, expansion occurred during childhood in 7, adolescence in 14, pregnancy in 10, and adulthood in 10. Some AVMs may remain quiescent until adolescence and, even into adulthood. Our patient was rather older than those of other reports. Moreover, we had earlier misdiagnosed the lesion as an otohematoma. AVM is similar to cauliflower ear in that both diseases may present with a swollen deformed auricle. As simple drainage based on a misdiagnosis of otohematoma may trigger massive bleeding in a case of AVM, differential diagnosis of AVM from an otohematoma is important.

In two case reports, surgical intervention was planned under suspicion of AVM in the absence of imaging data. However, imaging modalities are required for accurate diagnosis of the vascular lesion and confirmation of the extent. MRA or TFCA revealed total auricular involvement in 24 (89%), although 56.1% of 27 patients were thought to exhibit only partial involvement upon physical examination. Although conventional TFCA is invasive, the technique is useful to identify the principal feeding vessel and an appropriate embolization portal. TFCA-guided embolization is a
<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>25Y</td>
<td>41Y</td>
<td>41Y (1Y-55Y)</td>
<td>16Y, 22Y</td>
<td>21Y</td>
</tr>
<tr>
<td>Sex</td>
<td>M</td>
<td>M</td>
<td>Not described</td>
<td>M, F</td>
<td>F</td>
</tr>
<tr>
<td>Nations</td>
<td>Bangladesh</td>
<td>USA</td>
<td>USA and France</td>
<td>India</td>
<td>India</td>
</tr>
<tr>
<td>Chief complaint</td>
<td>Ear swelling, profuse bleeding</td>
<td>Ear swelling, PT intermittent bleeding</td>
<td>PT (51.2%), bleeding (41.5%), pain (29.3%), bruit/thrill (24.4%)</td>
<td></td>
<td>Ear swelling, PT</td>
</tr>
<tr>
<td>Duration of symptom</td>
<td>7Y</td>
<td>6Y</td>
<td>6Y</td>
<td>6Y</td>
<td></td>
</tr>
<tr>
<td>Bleeding history</td>
<td>Several times</td>
<td>Several times</td>
<td>Auricle and extraauricular involvement (Retroauricular: 46.3%, Neck: 22%, None: 22%)</td>
<td>Pt.2: 2 times</td>
<td>None</td>
</tr>
<tr>
<td>Lesion site</td>
<td>R) auricle</td>
<td>L) auricle</td>
<td>A and/or MRI: 65.9%</td>
<td>L) auricle, R) auricle</td>
<td>R) auricle</td>
</tr>
<tr>
<td>Key diagnostic study/findings</td>
<td>A/enlarged and tortuous vessels</td>
<td>A/diffuse network of shunts</td>
<td>Pt.1: DS/multiple dilated anechoic areas, Pt.2: MRA/enlarged serpiginous structures</td>
<td>A/diffuse shunts with STA</td>
<td></td>
</tr>
<tr>
<td>Main feeding vessels</td>
<td>PAA, OA Excision and STSG, ear elevation after 4M</td>
<td>PAA, STA, OA Excision, and STSG after embolization</td>
<td>PAA, STA, OA Observation (n = 12, 29.3%), Embolization (n = 9, 21.9%), Auricular resection c/w embolization (n = 20, 48.8%)</td>
<td>STA</td>
<td>failed embolization, excision after feeding artery ligation</td>
</tr>
<tr>
<td>Treatment</td>
<td>Excision and STSG after embolization</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Follow-up duration/Final status</td>
<td>4M/follow-up lost after 2nd operation</td>
<td>2Y/no recurrence</td>
<td></td>
<td></td>
<td>3Y/no recurrence</td>
</tr>
<tr>
<td>No of patients</td>
<td>Age</td>
<td>Sex</td>
<td>Nations</td>
<td>Chief complaint</td>
<td>Duration of symptom</td>
</tr>
<tr>
<td>----------------</td>
<td>------</td>
<td>-----</td>
<td>---------</td>
<td>----------------</td>
<td>-------------------</td>
</tr>
<tr>
<td>Woo HJ (2008)</td>
<td>1</td>
<td>M</td>
<td>Korea</td>
<td>PT</td>
<td>7M</td>
</tr>
<tr>
<td>Whitty LA (2009)</td>
<td>1</td>
<td>M</td>
<td>USA</td>
<td>Ear swelling, intermittent pain</td>
<td>2Y</td>
</tr>
<tr>
<td>Prasad KC (2011)</td>
<td>1</td>
<td>M</td>
<td>India</td>
<td>Ear swelling</td>
<td>2-3Y</td>
</tr>
<tr>
<td>Goel A (2011)</td>
<td>1</td>
<td>F</td>
<td>India</td>
<td>Ear swelling, PT</td>
<td>4Y</td>
</tr>
<tr>
<td>Meena BK (2013)</td>
<td>1</td>
<td>F</td>
<td>India</td>
<td>Ear swelling, PT</td>
<td>1Y</td>
</tr>
<tr>
<td>Dixit SG (2013)</td>
<td>1</td>
<td>M</td>
<td>India</td>
<td>Ear swelling</td>
<td>Since birth</td>
</tr>
<tr>
<td>Kim SH (this study)</td>
<td>1</td>
<td>F</td>
<td>Korea</td>
<td>Ear swelling, massive bleeding, PT</td>
<td>3Y</td>
</tr>
</tbody>
</table>

Y, year; M, month; R, right; L, left; PT, pulsatile tinnitus; A, angiography; CTA, computed tomography angiography; MRA, magnetic resonance angiography; DS, Doppler sonography; PAA, posterior auricular artery; STA, superficial temporal artery; QA, occipital artery; AVM, arteriovenous malformation; STSG, Split-Thickness Skin Graft.
useful initial therapeutic step, but surgical excision is required in most cases to avoid recurrence.

Treatment is unnecessary (especially in children) if the AVM is small and asymptomatic. In an untreated group of 12 patients, 2 of stage I and 7 of stage II remained stable.\textsuperscript{5,7} If the AVM is symptomatic, complete excision (preceded by embolization) is the treatment of choice.\textsuperscript{6} Ligation of the arterial vessels alone or partial excision should be avoided, because a new collateral circulation will form, triggering further enlargement.\textsuperscript{15,16} In our scoping review, 17 of 20 patients who underwent initial embolization exhibited aggravated or persistent AVMs. Wu et al. reported that the mean lag time between the last embolization and ultimate resection was 5.6 years (range: 2–8 years).\textsuperscript{1} Based on these results, embolization is best employed only to reduce blood loss and facilitate surgical extirpation.

Conclusion

Our case was initially misdiagnosed as an otophleboloma because of the cauliflower-like auricle, and the patient presented 3 years later with massive bleeding. To accurately diagnose an AVM, meticulous physical examination including palpation and auscultation and imaging are essential. In most cases of auricular AVM, optimal treatment is a combination of super-selective embolization and complete surgical excision.

Funding

This work was supported by a grant from the Korea Health Technology R&D Project through the Korea Health Industry Development Institute (KHIDI), funded by the Ministry of Health & Welfare, Republic of Korea (grant number HI14C2264).

Conflicts of interest

The authors declare no conflicts of interest.

References