Management of Head and Neck Arteriovenous Malformations - Team work Counts

Naresh K. Panda1*, Prabhat Thakur4, Ramesh K. Sharma2, Gautam Biswas1, Roshan Verma1 and Niranjan Khandelwal2
1Department of Otolaryngology Head and Neck Surgery, PGIMER, Chandigarh, India
2Department of Plastic Surgery, PGIMER, Chandigarh, India
3Department of Radiodiagnosis and Imaging, PGIMER, Chandigarh, India

Abstract

Purpose: To demonstrate the management protocol in head and neck vascular malformation

Methods: This is a retrospective review of 12 patients of arteriovenous malformation managed at a teaching hospital. Medical records were examined for age at first diagnosis, disease course, prior treatments, and age at presentation, management, therapeutic outcomes, impact on quality of life and photograph at time of presentation.

Results: Twelve patients with head and neck arteriovenous malformation presented to our centre. There was equal distribution of males and females with an average age of presentation being 24 years (range 13-40 years). Ten patients out of 12 underwent embolization which was followed by surgery. Complete excision was achieved in eight cases while partial resection was achieved in two cases. Three patients had complications while getting treated.

Conclusions: Head and neck AVM can be presented as expansile, invasive and locally aggressive lesions which require detailed evaluation and multidisciplinary approach for treatment.

Keywords: Vascular Malformation; Surgical Excision; Multidisciplinary Approach

Introduction

Arteriovenous malformations (AVM) are congenital malformation sharing communication between arteries and veins (arteriovenous shunting) which lack normal capillary networks. A nidus is an area with abnormal vasculature and shunting [1]. The initially quite lesion may progress to expansile mass during puberty or adolescence. Once it progresses, it causes bleeding, ulceration, pain, and cardiac volume overload [2-5].

Complete removal of the lesion is the treatment of choice [2-5]. Complete removal may be difficult due to diffuse involvement in the head and neck region. They are also known to increase in size during adolescence and after an attempted treatment [5-7]. The various modes of treatment that are available commonly include either transarterial vessel occlusion or ablative surgery. Single modality treatment may not be able to result in significant long-term reduction in clinical features. The combination therapy is now considered to be the treatment of choice [2].

We have managed in our centre during the last ten years. Incomplete removal and resultant residual lesions have led the patients to attend our hospital for treatment. The aim of the present series is to present the course of the AVM presenting in our Institute and their surgical management.

Methods

This was a retrospective analysis of 12 cases of arteriovenous malformation of head and neck region that were managed in our department between August, 2004 and December, 2014. The records were evaluated for the age of presentation, previous surgery and treatment options. The AVMs were subsequently staged using the Schobinger staging system (Table 1).

The records were reviewed for various variables like age, sex, past treatment and progression. The treatment and outcome of the lesions were also looked at. A prior consent was taken to take pictures of the patients. The diagnosis of arteriovenous malformation was established by getting clinical history, performing physical examination, obtaining radiographic images, and pathological results. MRI and arteriography revealed feeding arteries with dilated draining veins around a centralized nidus in each of these patients. Flow voids suggestive of fast flow lesions, were present on MRI/MR angiography. Multiple feeding arteries were present in each arteriovenous malformation. Angiography was followed by embolization in the treatment phase of some patients. All the patients underwent diagnostic angiography to verify the associated shunt and know the feeding arteries. The lesions were classified as AVMs if there was a clearly identifiable nidal component; otherwise they were classified as fistulas. PVA (polivinyl alcohol) and Gel foam were the most common materials used. Surgical resections were performed with the aim to remove the session completely. Reconstruction included both local and free flaps. The final diagnosis was confirmed by histopathological examination.

Results

Twelve patients with head and neck arteriovenous malformations presented to our centre. Among them six were male and six were female. The average age at presentation was 24 years (range 13-40 years). The summary of clinical features is presented in table 2. A diagnosis of haemangiomia was most commonly made before they presented to our centre. The continuous growth and local invasion prompted all patients to search for a second medical opinion. The photographs of the patients illustrating the clinical presentation of AVMs are shown in Figure 1-8.

The data regarding the extent of disease at the time of presentation at our centre is also detailed in table 2. Each patient

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 (quiescence)</td>
<td>Warm, pink-blue, shunting on Doppler examination</td>
</tr>
<tr>
<td>2 (expansion)</td>
<td>Enlargement, pulsation, thrill, bruit, tortuous veins</td>
</tr>
<tr>
<td>3 (destruction)</td>
<td>Dystrophic skin changes, ulceration, bleeding, pain</td>
</tr>
<tr>
<td>4 (decompensation)</td>
<td>Cardiac failure</td>
</tr>
</tbody>
</table>

Table1: Schobinger staging of arteriovenous malformation [14].
was diagnosed with head and neck AVM consistent with stage II or stage III of disease. The diagnosis was based upon vascular expansion, soft tissue invasion, pulsation, skin infiltration, mottling, and ulceration. There was a history of enlargement of the lesion after puberty. Surgical removal with preoperative embolization was performed in ten patients with an aim to completely remove the lesion. Due to the extensive nature of the disease, the complications included blood loss and incomplete removal. The embolization procedures were well tolerated by all the patients.

Table 2: Showing the patient details about management.

<table>
<thead>
<tr>
<th>S. No</th>
<th>Age</th>
<th>sex</th>
<th>Location</th>
<th>Grade</th>
<th>Preoperative embolization</th>
<th>Surgical resection</th>
<th>Previous surgery</th>
<th>Follow up complication</th>
<th>Revision surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>30</td>
<td>F</td>
<td>Pinna</td>
<td>3</td>
<td>yes</td>
<td>Total resection and reconstruction</td>
<td>0</td>
<td>Flap failure, three months</td>
<td>1</td>
</tr>
<tr>
<td>2</td>
<td>24</td>
<td>F</td>
<td>Pinna, neck</td>
<td>3</td>
<td>yes</td>
<td>Ligation of EJV, IJV, ECA and reconstruction of pinna</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>3</td>
<td>23</td>
<td>F</td>
<td>Pinna</td>
<td>3</td>
<td>yes</td>
<td>Total resection and reconstruction</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>4</td>
<td>14</td>
<td>M</td>
<td>Cheek, Face</td>
<td>3</td>
<td>no</td>
<td>Excision and reconstruction</td>
<td>0</td>
<td>Flap failure</td>
<td>1</td>
</tr>
<tr>
<td>5</td>
<td>13</td>
<td>M</td>
<td>Pinna</td>
<td>2</td>
<td>yes</td>
<td>Excision and reconstruction</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>6</td>
<td>20</td>
<td>F</td>
<td>Forehead</td>
<td>3</td>
<td>no</td>
<td>Total excision</td>
<td>2</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>7</td>
<td>38</td>
<td>M</td>
<td>Submandibular region, parapharyngeal space, vallecula</td>
<td>3</td>
<td>yes</td>
<td>Partial resection</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>8</td>
<td>22</td>
<td>F</td>
<td>Face</td>
<td>3</td>
<td>yes</td>
<td>Excision</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>9</td>
<td>27</td>
<td>M</td>
<td>Face, oral cavity and neck</td>
<td>3</td>
<td>yes</td>
<td>Partial glossectomy</td>
<td>2</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>10</td>
<td>17</td>
<td>M</td>
<td>Pinna</td>
<td>2</td>
<td>yes</td>
<td>Excision and reconstruction</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>11</td>
<td>4</td>
<td>F</td>
<td>Face</td>
<td>3</td>
<td>yes</td>
<td>Excision</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>12</td>
<td>20</td>
<td>M</td>
<td>Submandibular region</td>
<td>2</td>
<td>yes</td>
<td>Excision</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

Figure 1: Thirty years female with right pinna AVM.

Figure 2: Showing dilated and tortuous arteries supplying pinna.

Figure 3: Showing bare cartilage after excision of malformation.

Figure 4: Post operative picture after three months.
Out of 12 cases, five cases of pinna AVM underwent embolization with surgical removal. This was followed by reconstruction of pinna with temporoparietal flap, among which two cases required revision surgery due to flap failure. One patient who underwent embolization and partial removal of necrotic portion of tongue had an extensive malformation of the face, oral cavity and tongue. Three patients out of twelve had previous surgery before presenting to our institute. Three patients required re-surgery, two of them were with AVM of pinna, in which failure of flap occurred and one was with AVM of cheek who presented with recurrence. In two patients, complete removal of the lesion could be achieved without the need for embolization. Most common complications were pain, bleeding and failure of reconstruction flap requiring revision surgery.

The normal daily functions of these patients improved following intervention. In each case, these improvements were attributed to a decrease in bleeding episodes, improvement in appearance, elimination of pain, and return to normal daily activities.

**Discussion**

Vascular malformations are quiescent lesions, as evidenced by endothelial turnover [3]. Arteriovenous malformations usually progress over time and have likelihood of recurrence after surgery. It may be due to failure of the primitive arteriovenous shunts.

Although the presence of a quiescent arteriovenous malformation may be troublesome, it is the increase in size of the lesion that is the primary cause of morbidity. Arteriovenous malformation may enlarge because of increased blood flow causing collateral formation, dilatation, and thickening of adjacent artery and veins [1,8]. Other arteriovenous shunts may open, stimulating hypertrophy of surrounding vessels from increased pressure [1,5,8].

Angiogenesis (growth of new blood vessels from pre-existing vasculature) or vasculogenesis (de novo formation of new vasculature) may explain expansion of arteriovenous malformations. Although neovascularization may be a primary stimulus for expansion, it also could be a secondary event. For example, ischemia, which is a potent stimulator of angiogenesis, is a well-known cause of enlargement of arteriovenous malformation after proximal arterial ligation or trauma [1,5,7].

Seccia, et al [9], have reported that the chances of cure are better if the lesions are treated early. Early diagnosis is best established with MRI, arteriogram, and CT angiography illustrating high flow lesions with flow voids. Surgical excision along with embolization, or the combination of these modalities, has been attempted in the management of AVM [10]. Some evidence suggests that surgery with preoperative embolization and complete removal of the lesion offers the best chance for cure [2,3,5,7,11]. Unfortunately, recurrent, recidivistic, or dormant disease cannot be avoided in spite of best therapy. It is a frequent finding despite the type of therapy. Role of embolization in high flow arteriovenous malformation initially thought to have limited value, now has better outcome with new embolization material like onyx(ethylene-vinyl alcohol copolymer) [12]. Close observation of arteriovenous malformation, similar to the management of malignancies, is therefore required. It is difficult to assess the outcome of patients with vascular malformations. The algorithm provided by Wu, et al. [5], reflects the unusual behaviour of these vascular anomalies. The pathogenesis of AVMs remains unclear. Hemodynamic contributions and vascular remodelling have been attributed to the development of these rare vascular anomalies [2,3,5-8,13]. Hormones, trauma, and previous surgery have also been implicated in rapid growth. Medical options are limited for vascular malformations. Primary embolization should be regarded only potentially curative as an often recanalization and reexpansion will occur over a course of time. Hence surgical excision is the mainstay of treatment in these lesions. However
Complications like bleeding and recurrence are common and may have to be dealt. Palliative embolization may be indicated for relief of symptoms i.e. haemorrhage, pain or ischaemic ulcerations when the lesion cannot be surgically removed.

This article is designed to provide a description of the disease progression, treatment and its outcome. This case series demonstrates that combined modality therapy is critical in managing patients with arteriovenous malformations. The 12 selected cases simply illustrate the disease course and subsequent management of arteriovenous malformations of the head and neck with a multidisciplinary approach.

References


