Nasopharyngeal Angiofibroma

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Key Words
angiofibroma; embolization;
nasopharyngealneoplasm

Background. Nasopharyngeal angiofibromas are uncommon neoplasms, which are histologically benign but locally invasive and destructive. They mostly affect the nose and nasopharynx, but may extend to the adjacent structures. The first choice of treatment is surgery. This paper presents our experience in the management of nasopharyngeal angiofibroma.

Methods. We retrospectively reviewed 13 patients of nasopharyngeal angiofibroma attending the Department of Otorhinolaryngology at Taipei Veterans General Hospital (VGH-TPE) from 1979 to 1999. Two patients re-curred after surgery at other hospitals and were excluded from this study. Eleven patients under went the primary surgery and were included for statistical analysis. The relation ship between preoperative embolization and intraoperative effective blood loss (EBL) was also discussed.

Results. The incidence was about 1 per 5,000 otorhinolaryngology admissions. The ratio of female to male was 1 to 10. The mean age of symptom onset was 18 years. There were 7, 3 and 1 patients in IA, IB and IIA stages of Sessions’ classification. Seven patients underwent preoperative embolization. One patient was treated with stilbestrol preoperatively. Of the surgical procedures, transpalatal approach was the most often performed (8/11). Postoperative radiotherapy (1,000 cGy) was administered to 1 patient due to the suspected recurrent tumor. The average intraoperative EBL was 731.8 ml. Patients with preoperative embolization had an average EBL of 814.3 ml and those with out embolization had an average EBL of 587.5 ml. The difference of EBL between the patients with and without preoperative embolization was not statistically significant. The average follow-up duration was 118.4 months and no tumor recurred.

Conclusions. Surgery is the main therapeutic option for nasopharyngeal angiofibroma. We suggest preoperative embolization. Radiotherapy is an adju vant treatment and reserved for the patients with advanced stage or recurrent tumors. [Chin Med J (Taipei) 2001; 64:39-46]
ri orly into the sphenoid sinus, and laterally through spheno-palatine foramina into the pterygomaxillary fissure, where it may spread into the infratemporal fossa, and into the orbit through either the foramen lacerum or superior orbital fissure. Intracranial extension may also be a result of erosion through the sphenoid sinus.

The presenting symptoms are often epistaxis and nasal obstruction.

Biopsy is seldom required prior to definitive treatment due to compelling presentations, physical examinations and radiographic findings.

Surgery has been the most effective method of treatments for angiofibroma over past years. However, radiotherapy has been advocated as the initial treatment by a few investigators in spite of high risk of development of secondary malignancy in young patients. Some authors suggest that adjuvant hormone therapy may be effective.

There is no accurate incidence reported in literature to date. Harrison reported an incidence of 1 per 15,000 admissions at the Royal National Ear, Nose and Throat Hospital, London, while Jamal reported a higher incidence of 1 per 4,000 in Amman, Jordan.

In this paper, the clinical data of 11 cases with nasopharyngeal angiofibroma were presented, including age of onset, diagnostic methodology, duration of disease, staging and treatment modality, complications and recurrence. Especially, we focused on the relation between the preoperative embolization and intraoperative blood loss. The surgical procedures for nasopharyngeal angiofibroma were also discussed.

Methods

The charts from the patients with nasopharyngeal angiofibroma were reviewed. Eleven cases met inclusion criteria. There were 10 men and 1 woman. The inclusion criteria were definite diagnostic evaluation, pathological proof, institutional treatment with the primary surgerist and other adjuvant therapy, and clinical follow-up at our section.

The patients' general data, including clinical presentations, surgical procedures, intraoperative blood loss, adjuvant treatment, complications and recurrence were listed in Table 1. Ten of 11 patients (90.9%) re-
ceived CT scans (for example as Fig. 1) for localization and staging. CT scan was not performed for 1 case because it was not available in 1979. Nine patients (81.8%) underwent the preoperative angiography with selective catheterizations of the carotid arteries (Fig. 2 as for example) to determine the feeding artery. The feeding vessels were embolized with gelfoam strips 24 to 48 hours before surgery.

The staging system used in the present series was Sessions’ classification (Table 2). Seven patients were classified as stage IA, 3 as stage IB, and 1 as stage IIA. Preoperative hormonal therapy with stilbestrol (5 mg, three times a day) was administered to 1 patient for 30 days. Surgical approaches performed are shown in Table 3. Of the surgical procedures, transpalatal approach (Fig. 3) was the most often performed (8/11). All patients had the histologic confirmations of angiofibroma. Postoperative radiotherapy (1,000 cGy) was administered to 1 patient due to the suspected residual tumor.

To assess the difference of the intraoperative ef-

Fig. 1. Axial CT scan (A) and (B) showing contrast enhancement in a nasal angiofibroma (arrow) extending into the right-sided posterior nasal cavity in an 18-year-old male patient.

Fig. 2. Angiogram of the right external carotid artery before (A) and after (B) selective embolization of the right internal maxillary artery (arrowhead) using particles of Ivalon in an 18-year-old male patient.
Effective blood loss (EBL) between patients with and without preoperative embolization, the Mann-Whitney test was used for statistical analysis. The statistical data were generated by the software Statistical Package for the Social Science (SPSS).

In addition, 2 cases receiving surgery elsewhere were referred to our department due to recurrent nasopharyngeal angiofibroma. One of them had a recurrent tumor with invasion to the middle cranial fossa and was treated with hormonal therapy (stilbestrol 5mg three times a day for 30 days) and radiotherapy (1,500 cGy). After Denker’s operation and ligation of internal maxillary artery, the other patient was managed with radiotherapy 3,000 cGy due to the residual tumor of the maxillary and sphenoid sinuses. Their tumors disappeared after adjuvant treatments. They were followed up for 213 and 242 months respectively.

**Results**

The incidence of nasopharyngeal angiofibroma was 1 per 5,000 of total admissions between 1979 and 1999. The median age of symptom onset was 18 years, with a range of 12 to 74 years. The average duration of symptoms prior to presentation was 21.9 months, with a range of 15 days to 7 years. Symptoms and signs at the time of presentation are depicted in Table 4.

Of 11 patients, 2 refused angiography due to old age in 1 case and economic consideration in the other. Preoperative embolization was attempted on 9 patients but only 7 cases were successful because of anaphylactic reaction in 2 cases. The feeding vessels of all cases were internal maxillary arteries.

The average intraoperative effective blood loss (EBL) was 731.8 ml, with a range of 250 to 1750 ml. Seven patients with preoperative embolization had an average EBL of 814.3 ml and 4 patients without preoperative embolization had an average EBL of 587.5 ml. Using the Mann-Whitney test, we found no significant difference between the patients with and without preoperative embolization ($p = 0.85$). If 7 patients of stage IA tumor were considered, the difference of EBL between the 4 patients with the preoperative embolization and 3 patients without the preoperative embolization was also not significant ($p = 0.37$).

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**Table 2. Staging system of Sessions** for nasopharyngeal angiofibroma

<table>
<thead>
<tr>
<th>Stages</th>
<th>Tumor extent</th>
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<tbody>
<tr>
<td>IA</td>
<td>Limited to nose and/or nasopharyngeal vault</td>
</tr>
<tr>
<td>IB</td>
<td>Extension into ≥ 1 sinus</td>
</tr>
<tr>
<td>IIA</td>
<td>Minimal extension into PMF</td>
</tr>
<tr>
<td>IIB</td>
<td>Full occupation of PMF with or without erosion of orbital bones</td>
</tr>
<tr>
<td>IIC</td>
<td>Infratemporal fossa with or without cheek</td>
</tr>
<tr>
<td>III</td>
<td>Intracranial extension</td>
</tr>
</tbody>
</table>

PMF = pterygomaxillary fossa.

**Table 3. Surgical approaches for 11 patients with nasopharyngeal angiofibromas at Taipei Veterans General Hospital**

<table>
<thead>
<tr>
<th>Approach</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Transpalatal</td>
<td>8</td>
<td>72.7</td>
</tr>
<tr>
<td>Lateral rhinotomy</td>
<td>2</td>
<td>18.2</td>
</tr>
<tr>
<td>CWL and transantral ethmoidectomy</td>
<td>1</td>
<td>9.1</td>
</tr>
</tbody>
</table>

CWL = Caldwell-Luc operation.

**Table 4. Clinical presentations of 11 patients with nasopharyngeal angiofibromas at Taipei Veterans General Hospital**

<table>
<thead>
<tr>
<th>Symptoms and signs</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Epistaxis</td>
<td>10</td>
<td>90.9</td>
</tr>
<tr>
<td>Nasal obstruction</td>
<td>6</td>
<td>54.5</td>
</tr>
<tr>
<td>Deafness</td>
<td>1</td>
<td>9.1</td>
</tr>
<tr>
<td>Facial deformity</td>
<td>1</td>
<td>9.1</td>
</tr>
</tbody>
</table>

**Fig. 3.** Transpalatal approach. The nasopharyngeal angiofibroma (arrow) is shown after elevation of mucoperiosteal flap and removal of bone from the hard palate.
All the specimens were pathologically proved to be angiofibroma. There was no surgical complication or mortality. The average duration of the postoperative follow-up was 118.4 months, with a range of 5 to 202 months. There was no tumor recurrence during the follow-up. The overall results were satisfactory; only 1 patient complained of nasal obstruction.

**Discussion**

The clinical presentations of the nasal pharyngeal angiofibroma in our series were similar to those reported in the literature.\(^1\,^2\,^5\,^9\) However, there still were some differences from the other series in age and sex distributions. Nasal pharyngeal angiofibromas are found predominantly in adolescent males who are commonly diagnosed between 14 and 25 years of age.\(^2\) The median age of symptom onset was 18 years in our study, with 2 exceptions, 1 female patient aged 57 years and 1 male patient aged 74 years, rarely reported in the literature. The incidence was 1 per 5,000 admissions of our department, which might be relevant in comparison with other series (1 per 15,000 to 1 per 4,000).\(^15\,^16\)

Diagnosis was made mainly on a histological basis, together with clinical data such as age, sex, location of lesion, suggestive symptoms and appropriate radiological studies. Because of a high incidence of nasopharyngeal carcinoma (NPC) in Taiwan, it is important to make a differential diagnosis. The angiofibroma appears as a firm, red dish-purple, mucosa-covered, and friable or granular mass (Fig. 4) mostly over the nasal roof, while the NPC has an irregular surface mostly over the Rosenmüller’s fossa.

In our management, the contrast-enhanced computed tomography (CT) is arranged before biopsy for its high diagnostic value. If angiofibroma is highly suspected, we should keep in mind that biopsy may accompany the risk of significant bleeding.

Nasopharyngeal angiofibroma without intracranial extension is a surgically curable disease.\(^1\,^2\,^4\,^6\) Various surgical approaches have been advocated by different series.\(^1\,^2\,^4\,^6\) The main procedures in our series were transpalatal approaches and lateral rhinotomies, which can be used for most early stage tumors. Advanced dis ease affects the pterygomaxillary fossa re quired midfacial degloving ap proaches.\(^17\) Lloyd et al.\(^18\) advocated midfacial degloving approach as surgical procedure for patients who do not show sphenoid involvement on pre-operative CT scans. More radical surgery is needed to eliminate the tumor within the sphenoid because of its high risk of recurrence. The cure rate in our series was about 100%, which was better than that of the other series (75-90%).\(^5\,^7\,^10\) Predictably, lower cure rates are seen in studies with a large number of tumors with intracranial involvement.\(^16\)

The role of radiotherapy has been debated and controversial. Control rates exceeding 75% have been reported with primary radiation therapy,\(^12\,^19\) and many authors recommend external beam radiotherapy for intracranial involvement, unresectable disease and multiple recurrences.\(^2\,^13\) The potential of radiotherapy exists for the later osteoradionecrosis, de laid or abnormal bone growth, or malignancy in young patients. Hence, radiotherapy as a primary treatment should be avoided.\(^13\,^18\) In our series, 1 patient with primary tumor recurrence received the post-operative radiotherapy due to the suspected residual tumor. This residual tumor disappeared after a course of 1,000 cGy.

The tumor is characterized by high recurrence rate, re-ported as high as 22%.\(^4\) It is prone to malignant transformation.

**Fig. 4.** Endoscopic view of the right nasal cavity. The nasopharyngeal angiofibroma (T) is located posterior to the middle turbinate (arrow head).
mined by tu mor growth rate at the time of sur gery, combined with in complete sur gi cal ex ci sion, which is greatly dependent on the type of tu mor extension. Most re cur rences were found in tu mors that had in vaded and ex panded the sphenoid. It has been re ported that le sions can un dergo dero spon ta ne ous re gres sion with age, but it has not been def i nitely proven.17,18

Some au thors ad vo cate that the pre op er a tive em bol iz ation as an im portant part of man age ment to re duce in traop er a tive blood loss.20,21 In our se ries, the feed ing ves sels were em bol ized with gelfoam strips 24 to 48 hours be fore sur gery be cause the gelfoam could per sist for 72 hours. The av er age in traop er a tive EBL was 731.8 ml, with a range of 250 to 1750 ml. Seven pa tients with pre op er a tive em bol iz ations had an av er age EBL of 814.3 ml, while 4 pa tients with out em bol iz ation had an av er age EBL of 587.5 ml. This dif fer ence was not statis ti cally sig nif i cant. Those who re ceived com plete in vest i gation pro ce dures or pre op er a tive em bol iz ation might have ap parent clin i cal pre sen ta tions, such as large tu mors or deep in va sion. This might ex plain why there was no less blood loss in this group. In Lloyd et al’s opin ion,19 pre op er a tive em bol iz ation may shrink the tu mor and make to tal re moval more dif fi cult, es pe cially if there is deep in va sion of the sphenoid. Al though our re sult is sim i lar to those of some stud ies,7,10,17 we still sug gest that pre op er a tive em bol iz ation should be per formed to re duce in traop er a tive blood loss. Only 2 pa tients failed in pre op er a tive em bol iz ation be cause of the anaphylactic re ac tion. Now a days, nonionic wa ter-sol uble con trast med ia are widely used and avail able, which lower the risk of such pro ce dure.

Hor mone ther apy with stil be strol could be ad min is tered as an ad ju vant treat ment to re duce vas cu lar ity and tu mor size.13,14 Thus, it may fa cil i tate tu mor ex ti ran sion and te ler age and pre vention of re cur rence. For lo ca li zation of tu mor, the con trast-en hanced CT scan or MRI be fore sur gery is man datory. Angi ography with pre op er a tive em bol iz ation per mits a less ag gres sive sur gi cal ap proach and may re duce in traop er a tive blood loss. Most stage IA, IB and IIA tu mors can be re se cted with transpalatal or lat eral rhino tomy ap proaches, while the ad vanced stage tu mors with the midfacial de gloeving ap proach. Ra dio ther apy should be re served for cases with in tra cra nial ex ten sion or mul ti ple re cur rences. Hor mone ther apy with stil be strol could be ad min is tered as ad ju vant ther apy to re duce vascu lar ity and tu mor size.

Conclusions

In our ex pe ri ence and in lit er a ture, the treat ment for na so pha ryn geal angiofibroma with out in tra cra nial ex ten sion is sur gery. The best ap proach must be care fully planned, as it is im por tant for the ex po sure of the tu mor, rad i cal re moval, con trol of in traop er a tive hem orr hage and pre vention of re cur rence. For lo ca li zation of tu mor, the con trast-en hanced CT scan or MRI be fore sur gery is man datory. Angi ography with pre op er a tive em bol iz ation per mits a less ag gres sive sur gi cal ap proach and may re duce in traop er a tive blood loss. Most stage IA, IB and IIA tu mors can be re se cted with transpalatal or lat eral rhino tomy ap proaches, while the ad vanced stage tu mors with the midfacial de gloeving ap proach. Ra dio ther apy should be re served for cases with in tra cra nial ex ten sion or mul ti ple re cur rences. Hor mone ther apy with stil be strol could be ad min is tered as ad ju vant ther apy to re duce vas cu lar ity and tu mor size.

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