Juvenile nasopharyngeal angiofibroma in a tertiary centre: ten-year experience

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ABSTRACT

Introduction: This is a retrospective study that aimed to examine the outcomes of patients presenting with juvenile nasopharyngeal angiofibroma (JNA) at a tertiary centre in Malaysia.

Methods: The demographical data, clinical presentation, investigations as well as treatment of 13 JNA patients were reviewed and collected from the medical record office at our centre from 1995 to 2005.

Results: All JNA patients were male and the average age at diagnosis was 17 (range 14–28) years. They presented with recurrent painless spontaneous epistaxis, nasal obstruction, nasal discharge, a reduced sense of smell, snoring, headache and facial swelling. One patient was at stage I, eight were at stage II, three at stage III and one patient was at stage IV, based on the Fisch classification. Angiography showed that nine tumours were supplied by both internal maxillary arteries of the external carotid system, and only four tumours received blood supply from the ipsilateral internal maxillary artery. All 13 patients underwent primary surgical resection. The overall recurrence rate was 38.5 percent for the first procedure and 60 percent for the second procedure. No major complications occurred in this group of patients as a consequence of treatment, neither for the primary tumours nor for the recurrences.

Conclusion: JNA is a rare vascular benign tumour with highly exclusive persistence and recurrence, and typically affects adolescent boys. The management of JNA presents a challenge to ENT surgeons. Preoperative angiography and embolisation minimise intraoperative blood loss and the current shift in the treatment to endoscopic excision in selected lesions reduces perioperative morbidity.

Keywords: angiofibroma, juvenile nasopharyngeal angiofibroma, nasopharyngeal tumour

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INTRODUCTION

Juvenile nasopharyngeal angiofibroma (JNA) is a rare, histologically benign, highly vascular and locally invasive tumour that occurs primarily in male adolescents, with an average age of onset at 15 years. It makes up about 0.05% of all head and neck neoplasms.[1] This tumour originates from the posterolateral wall of the nasal cavity in close proximity to the superior aspect of the sphenopalatine foramen. It may extend from the nasal cavity to the nasopharynx, the paranasal sinuses, the orbit and the pterygopalatine fossa, and may even invade the skull base or extend intracranially.[2] Histopathologically, JNA is an unencapsulated tumour that is made up of wide vascular spaces with single-layered linings, thus the propensity for haemorrhage. The blood supply of JNA comes primarily from branches of the external carotid system, although feeders from the internal carotid artery could contribute to its vascularity. The cause of this tumour remains unclear.

Most patients present with symptoms of severe, recurrent, spontaneous epistaxis with persistent nasal obstruction and discharge. As the tumour expands, facial deformities, proptosis, blindness, cranial nerve palsies and headaches may occur. Several staging systems have been proposed, although lately, the Fisch classification seems to be the most commonly used.[3] Because of its high vascularity, JNA has always presented a treatment challenge to surgeons. In the past, surgical excision of the tumour was complicated by extensive haemorrhage. With the improvements in diagnostic imaging techniques and the use of preoperative angiographic embolisation, most authorities today recommend surgery as the preferred treatment method for JNA. Irradiation has been used as an adjuvant to subtotal resection, for both unresectable lesions and those with intracranial extension.[2] The purpose of this study was to look at the outcomes with the treatment of 13 JNA patients at a tertiary centre in Malaysia.

METHODS

This is a retrospective review of 13 patients with JNA treated

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Table I. Stages of the Fisch classification system.[3]

Stage	Description
I	Tumours limited to the nasal cavity nasopharynx with no bony destruction.
II	Tumours invading the pterygomaxillary fossa, paranasal sinuses with bony destruction.
III	Tumours invading the infratemporal fossa, orbit and parasellar region remaining lateral to the cavernous sinus.
IV	Tumours with invasion to the cavernous sinus, optic chiasmal region and pituitary fossa.

Table II. Tumour stage and recurrence of angiofibroma.

Fisch's stage[3]	No. (%) of patients	No. of first recurrence	No. of second recurrence
I	1 (7.7)		
II	8 (61.5)	2	1
III	3 (23.1)	2	1
IV	1 (7.7)	1	1

Table III. Tumour stage and surgical treatment.

Surgical treatment	No. of patients	Fisch's stage (no. of patients)
Transnasal endoscopy	2	I (1); II (1)
Lateral rhinotomy	8	II (7); III (1)
Lateral rhinotomy & transpalatal	2	III (2)
Midfacial degloving	1	IV (1)

at our centre from 1995 to 2005. The demographical data, clinical presentation, investigations as well as treatment of these patients were reviewed and collected from the medical records. All tumours were classified according to the Fisch classification system[3] on the basis of computed tomography (CT) (Table I).

RESULTS

JNA was diagnosed in 13 patients at our centre from 1995 to 2005. All were male and the average age at diagnosis was 17 years with a range of 14–28 years. All the cases were referred from private or government general practitioners. Ten out of 13 patients were Malay, two were Chinese and one was Indian. All of them presented with recurrent painless spontaneous epistaxis. Other symptoms included nasal obstruction (76.9%), nasal discharge (76.9%), a reduced sense of smell (61.5%), snoring (38.5%), headache (23.0%) and facial swelling (7.7%). Nasal endoscopical examination of the nasal cavity and nasopharynx revealed a nasal mass arising from the nasopharynx in all the patients. No biopsies were taken in any of these patients as JNA was the primary provisional diagnosis.

All patients underwent contrast-enhanced CT for the initial assessment, and were classified according to the Fisch classification;[3] one patient (7.7%) was at stage I, eight (61.5%) were at stage II, three (23.1%) were at stage III and one patient (7.7%) was at stage IV (Table II). All patients underwent selective angiography. Angiography showed nine tumours (69.2%) supplied by both internal maxillary arteries of the external carotid system and only four tumours (30.8%) received blood supply from the ipsilateral internal maxillary artery. Two tumours (18.2%) also received blood supply from the internal carotid artery. Internal maxillary artery embolisation was carried out in all 13 patients, 24 hours prior to surgery. For all embolised tumours, an immediate control angiogram showed devascularisation of the parts of the tumour fed by branches of the external carotid arteries. No complications were associated with embolisation.

All 13 patients underwent primary surgical resection – eight through a lateral rhinotomy approach, two through a combined lateral rhinotomy and transpalatal approach, one through a midfacial degloving approach, and two through a transnasal endoscopic approach. One patient each at stages I and II underwent surgical resection via transnasal endoscopy. Seven patients at stage II and one at stage III underwent surgical resection via lateral rhinotomy. Two patients at stage III underwent surgical resection via combined lateral rhinotomy and the transpalatal approach. One patient at stage IV underwent the midfacial degloving approach to remove the tumour (Table III).

The average operation duration was 4 (range 3–7) hours. The transnasal endoscopic approach required the shortest operative time, averaging about two hours. The average surgical blood loss was 1,500 (range 500–3,000) ml. Both of the patients who were operated on via the transnasal endoscopic approach had the least amount of blood loss, which was 500 ml. The average surgical blood loss for the nine tumours supplied by both internal maxillary arteries of the external carotid system was 1,800 ml compared to only 1,000 ml of average blood loss of the four tumours receiving blood supply from the ipsilateral internal maxillary artery. The average operation duration for the nine tumours supplied by both internal maxillary arteries of the external carotid system was five hours, compared with 3.5 hours of average operation duration for the four tumours that received blood supply from the ipsilateral internal maxillary artery.

The patients were transfused with packed cells intraoperatively, with a minimum of two units and a maximum of seven units, and an average of four units. All patients were monitored in the intensive care unit for an average of 1.38 days postoperatively. Nasal packing remained in place for an average of 5 (range 3–7) days. All

the nasal packing was removed in the operating room. The average length of hospital stay was 10.9 (range 6–18) days. The two patients who were operated on via the transnasal endoscopic approach had the shortest hospital stay, averaging four days, without any complications.

Two out of eight patients in stage II, two out of three patients in stage III and the only patient in stage IV showed a recurrence of the tumour averaging at 9 (range 6–27) months. In three of these five patients, further recurrences occurred, two after six months and one 20 months after the second surgery. In the five patients with the first recurrence, four of them received radiotherapy treatment and one patient underwent transnasal endoscopic removal of the recurrent tumour. For the second recurrence, two patients post-radiotherapy were treated via a craniofacial approach and one repeated with transnasal endoscopic removal of the tumour (Table II). The overall recurrence rate was 38.5% for the first procedure and 60% for the second procedure. It is noteworthy that no major complication occurred in this group of patients as a consequence of treatment for the primary tumours, nor for the recurrences. The average length of follow-up was 26 (range 12–96) months. Two out of 13 patients defaulted after 12 months of follow-up, three were discharged after five years of being disease-free, and the other eight patients are currently still under follow-up.

DISCUSSION

The demographical data and clinical presentation reviewed from this retrospective study are comparable to studies done previously.[2,4-7] The typical case presentation is usually of an adolescent male with an average age of 17 years presenting with spontaneous recurrent epistaxis, nasal obstruction and discharge. Other clinical presentations depend on the extent of the tumour.[6,7] In this study, ten out of 13 (76.9%) JNA patients were of Malay ethnicity, but this data cannot be used to suggest that JNA is more common in this ethnic group, as the hospital is located in a community where the Malays form the majority of the population; and the sample size was also small.

The diagnosis of JNA is essentially based on a careful history, nasal endoscopic examination and is supplemented by imaging studies using computed tomography or magnetic resonance imaging. Biopsies to establish histological diagnosis are contraindicated and with advancement in angiography, definite diagnosis and embolisation of the tumour-feeding vessels can be performed at the same setting. It still remains open to debate whether preoperative embolisation helps to obscure tumour extensions intraoperatively, increasing the likelihood of incomplete tumour removal. However, the reduced blood loss during surgery facilitated the removal of these tumours dramatically and rendered intraoperative or postoperative blood transfusions unnecessary.[4]

In our study, preoperative angiography showed nine tumours (69.2%) supplied by both internal maxillary arteries of the external carotid system and only four tumours (30.8%) received blood supply from the ipsilateral internal maxillary artery. This was also shown in the study done by Pryor et al,[6] where 16 of the JNAs were predominantly supplied by the external carotid artery system, with equal numbers displaying unilateral or bilateral contributions. We also noticed that for the tumours that were supplied by both internal maxillary arteries, the average surgical blood loss was relatively more, and the average operation duration was comparatively longer than the tumours which were only supplied by the ipsilateral internal maxillary artery.

Various methods have been used to treat patients with JNA in the past. However, this historical information indicates that surgical resection and irradiation therapy are the most successful treatments.[2] Therefore, surgery with different approaches depends on the stage of the tumour, and this remains the primary mode of treatment. Radiotherapy is reserved for unresectable lesions.[5] In our centre, all 13 patients underwent primary surgical resection. The approach to tumour resection was dependent on the staging of the tumour based on the Fisch classification;[3] treatment of stage I and stage II tumours with minimal paranasal sinus involvement was via the transnasal endoscopic route unless intraoperative complications develop, especially uncontrolled massive bleeding endoscopically. Treatment of stage II tumours which invaded the pterygomaxillary fossa, and stages III and IV tumours was resected via the open approach. Radiotherapy was only offered for recurrent tumours.

Recurrence is a conspicuous feature of the natural history of JNA. Recurrence rates as high as 30%–50% have been reported by McCombe et al.[8,9] Harma reported a 46% recurrence in a cohort of 49 patients, 28% of which were multiple recurrences.[10] In 1992, Gullane et al reported a 36% recurrence after surgery in a series of 14 patients.[11] In our study, five out of 13 patients (38.5%) had recurrent tumours, comparable to the other reports by McCombe et al, Harma and Gullane et al.[8-11]

Advances in endoscopic endonasal surgery coupled with the success of preoperative arterial embolisation have allowed select cases of JNA to be managed endoscopically. The endoscopic approach has permitted the complete resection of suitable lesions with very little morbidity. Intra- and postoperative complications are reduced with endoscopic excision, compared to the standard surgical approaches, in selected lesions including Sessions et al's classification stage IIC lesions.[12] In addition, the endoscope has been used to improve visualisation in traditional open

approaches and the reported recurrence rates of properly-selected endoscopically resected tumours have been low. Wormald and Van Hasselt reported no local recurrence in seven patients with endoscopically-resected JNA.[13] From 1995 to 2005, most of our patients were operated on using the traditional approach at our centre because of the late presentation. With advances in the management of JNA, current trends are shifting towards the endoscopic approach for the removal of tumours in selected lesions. Further studies need to be carried out in the future to assess the outcome of the endoscopic approach at our centre.

In conclusion, JNA is a rare, vascular, benign tumour with high exclusivity, persistence and recurrence, and typically affects adolescent boys. The management of JNA presents a challenge to ENT surgeons. Preoperative angiography and embolisation minimise the intraoperative blood loss and the current shift in the treatment to endoscopic excision in selected lesions reduces perioperative morbidity. Larger demographical studies are needed to determine whether JNA is linked to a particular race only, especially in our multiracial society.

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