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RESEARCH ARTICLE

A RETROSPECTIVE STUDY ON THE OCCURRENCE AND PRESENTATION OF JUVENILE NASOPHARYNGEAL ANGIOFIBROMA

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ABSTRACT

Juvenile Nasopharyngeal Angiofibroma (JNA) is a rare breed of tumour found in adolescent males and there is a paucity of Indian studies on this subject. This is a retrospective observational study of JNA patients who presented at the V.S.S. Medical College and Hospital, Burla, Odisha between October 2009 and September 2012. Total 3953 cases of mass in head and neck region were presented at our department during that period. Out of those there were 17 cases of JNA. The incidences, age and sex distribution, symptoms and management approaches were recorded. The mean age at presentation was 16.94 years. The surgical approaches used were endoscopic endonasal approach and lateral rhinotomy approach. The recurrence rate was 35.3%.

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INTRODUCTION

Juvenile Nasopharyngeal Angiofibroma (JNA) is a relatively rare breed of tumor occurring mainly in adolescent boys. It accounts for 0.5% of all head and neck tumors (Chandler *et al.*, 1984). The tumor originates from the postero-lateral wall of the nasal cavity near the superior margin of the sphenopalatine foramen (Zito *et al.*, 2001). Typically, patients present with unilateral nasal obstruction and discharge or epistaxis. As the disease aggravates, facial deformities, proptosis, blindness and cranial nerve palsies may occur (Yang *et al.*, 1998). The diagnosis of JNA is essentially based on a careful analysis of ailing history and nasal endoscopic examination, supplemented by imaging studies using Computed Tomogram (CT) and Magnetic Resonance Imaging (MRI). Biopsies to establish histological diagnosis are contraindicated. The blood supply of these lesions is primarily from the External Carotid Artery (ECA) and in some cases from the Internal Carotid Artery (ICA) (Li *et al.*, 1998).

MATERIALS AND METHODS

The study was carried out in the department of ENT and Head and Neck surgery, V.S.S. Medical College, Burla between October 2009 and September 2012. The work includes all cases of masses in head and neck region presented to the outdoor. Such cases, when referred from surgical specialities, are also included in this work. Total 17 cases of JNAs were reported and their case records were evaluated.

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Observations

Incidence

Total number of patients presenting with mass in head and neck region from October 2009 to September 2012:- 3953
Out of these 3953, number of cases of JNAs:- 17
Hence incidence rate is:- 0.43%

Most common presentations are nasal obstruction and recurrent painless spontaneous epistaxis.

Majority of patients presented in a late stage due to stoic attitude of people in this part of country.

Recurrence Rate

6 cases of recurrence have been observed within 2 years. This implies recurrence rate of 35.3 %.

DISCUSSION

Head and neck tumours are commonly encountered in otolaryngological practice. From October 2009 till September 2012, total 3953 cases of head and neck masses were encountered in our department, out of which 17 cases were juvenile nasopharyngeal angiofibroma. This reflects the incidence of around 0.43% which is comparable to the study that reports the incidence to be around 0.5% (Chandler *et al.*, 1984). JNA, as the name suggests, is the disease affecting young persons (Zito *et al.*, 2001; Yang *et al.*, 1998). In our study, the average age of presentation is 16.94 years which is

Age and Sex Distribution

NO. OF CASES	SEX	AGE OF PRESENTATION
1	MALE	21 yrs
2	MALE	14 yrs
3	MALE	16 yrs
4	MALE	16 yrs
5	MALE	24 yrs
6	MALE	14 yrs
7	MALE	15 yrs
8	MALE	17 yrs
9	MALE	13 yrs
10	MALE	15 yrs
11	MALE	16 yrs
12	MALE	23 yrs
13	MALE	15 yrs
14	MALE	17 yrs
15	MALE	19 yrs
16	MALE	15 yrs
17	MALE	18 yrs

Average age at diagnosis is 16.94 years.

CLINICAL PRESENTATIONS

SYMPTOMS	NO OF CASES
Nasal obstruction	15
Recurrent painless spontaneous epistaxis	11
Headache	6
Unilateral nasal discharge	6
Nasal intonation of voice	6
Facial swelling	5
Conductive hearing loss	4
Snoring	1
Reduced sense of smell	1

STAGING

(Based on FISCH Classification)

STAGE	NO. OF CASES
Stage I	1
Stage II	4
Stage III	12
Stage IV	0

comparable to the observations made in the literature. In all the cases, the disease was diagnosed with the male group, indicating its predilection for male sex (Schick *et al.*, 2003). As per our study, the most common symptoms are nasal obstruction and recurrent painless spontaneous epistaxis. This has been corroborated in a study by Windfuhr JP, Remmert S. *Acta otolaryngol. Oct. 2004. medscape articles*. Surgery is the recommended treatment for JNA (Zito *et al.*, 2001; Yang *et al.*, 1998). Our standard care of management has been surgery. Out of 17 cases, primary surgical resection by endoscopic endonasal approach was done in 1 case and lateral rhinotomy approach was done in rest 16 cases. Pre-operative embolisation has been noted to reduce intra-operative blood loss (Li *et al.*, 1998; Schick *et al.*, 2003; Moulin *et al.*, 1995). The reported

recurrence rate following treatment of JNA varies from 0 to 55% (Mc Combe *et al.*, 1990; Herman *et al.*, 1999). In our study the recurrence rate is 35.3%. The high rate of recurrences in JNA following surgery is attributed to the advanced stage of disease at presentation, lack of encapsulation of the tumour and its propensity for submucosal spread.

Conclusion

Surgery remains the mainstay of therapy for JNA. Accurate staging, preoperative embolization, state-of-the-art surgical approaches like endoscopic excision for limited stage disease and maxillary swing approach for advanced stage disease, account for better surgical outcome. The high recurrence rate is a result of advanced stage tumors at presentation. Aggressive re-excision should always be considered for resectable recurrent disease and RT should be reserved for unresectable residual or recurrent disease.

Abbreviation

JNA- Juvenile Nasopharyngeal Angiofibroma

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