VOLUME-6 | ISSUE-4 | APRIL - 2017 • ISSN No 2277 - 8179 | IF : 4.176 | IC Value : 78.46

PARAGANGLIOMAS IN ENT & HEAD NECK; OUR EXPERIENCE

ENT

KEYWORDS:

RAUF AHMAD — Professor and Head Department of Otorhinolaryngology and Head Neck Surgery Government Medical College Srinagar, Jammu and Kashmir India

ZAFARULLAH BEIGH — Senior Resident Department of Otorhinolaryngology and Head Neck Surgery Government Medical College Srinagar, Jammu and Kashmir India

TABISH MAQBOOL — Junior Resident Department of Otorhinolaryngology and Head Neck Surgery Government Medical College Srinagar, Jammu and Kashmir India

Introduction

Head and neck paragangliomas (HNPs) are tumors of the autonomic system. They arise from chromaffin cells of the parasympathetic paraganglia of the skull base and neck, and are also called glomus tumors [1–5]. Paraganglia are chemoreceptor organs that are distributed throughout the body. They include glomus jugulare and glomus tympanicum within the temporal bone, carotid body tumors arising from the carotid body and also along nerves in the head and neck like the vagus and laryngeal nerves.

Clinically, patients with paragangliomas present with diverse signs and symp-toms. They may have a family history of paragangliomas and can present with multicentric tumors regardless of sporadic or familial origin. [6-8]

Over a ten-year period, we encountered 14 cases of craniocervical PGs including seven cases of glomus jugulares, two cases of glomus tympanicums and five cases of carotid body tumors (CBTs).In the era of safe embolization protocols and so-phisticated surgical approaches to the skull base, surgery has become the preferred treatment method for paragangliomas.[6-7]

Paragangliomas are rarely malignant, representing a small subset of extra-adrenal paragangliomas with a propensity for regional lymph-node and distant meta-static disease, primarily to bone, the lungs, and liver.6-8A higher rate of malignancy has been observed in sporadic paragangliomas compared with the familial type, with the exception of the familial syndrome associated with SDHBmutation (rate of malignancy,30%–70%).[9,10] .By comparison, the rate of malignancy for vagal paragangliomas is between 16% and 19%, between 5% and 6% for jugu-lotympanic paragangliomas, and between 3% and 4% for carotid body tumors.9 No histological criteria exist to diagnose malignancy in primary tumors. The health care professional can make the diagnosis only if malignancy is confirmed by the presence of a tumor in the lymph nodes or the disease has metastatically spread to distant sites [6-8].

Materials and Methods

This study is a retrospective study of 29 cases of PGs in head and neck treated over a period of ten years from 2007 to 2016 in the Depart-ment of Otolaryngology—Head & Neck Surgery, Goverment Medical College, Srinagar J&K. This included 10 glomus jugulare, 6 tympanicum tumors ,11 CBTs and 2 Glomus Vagale tumors. The age, gender and medical history & examination findings of these patients were recorded from case records. Typical clinical findings of craniocervical PGs included hearing loss, pulsatile tinnitus, giddiness, ear pain, bleeding from the ear, nerve palsies and neck mass.Signs included perforated tympanic membranes (TMs),aural polyp with or without bleeding, pulsatile neck swelling & cranial nerve deficits.

A thorough physical examination was done to rule out associated lesions else-where and to look for neurological deficits. Once clinically diagnosed as PGs, the patients were subjected to a contrast enhanced CT scan (CECT). A bilateral carotid angiography was done in all cases of glomus jugulare.

Apart from routine blood investigations, urinary catecholamines levels were determined in all patients. Ultrasound abdomen was done in all cases. Balloon occlusion test was done in few cases of CBTs (left sided and shambling III CBTS), Surgical procedure depended on the lesion and its extent.

Results; results are given in table 1-6

Table 1 ; Distribution of paraganglioma

TYPE OF PARAGANGLIOMA	NO.OF PATIENTS	PERCENTAGE(%)
Carotid Body Tumour	11	37.93
Glomus Jugulare	10	34.48
Glomus Tympanicum	6	20.68
Glomus Vagale	2	6.89
Total	**29**	**100**

Table 2 ; Glomus juglare tumour

Fisch Type	No. of patients	Percentage
A	0	0
B	0	0
C1	8	80
C2	1	10
D	1	10
TOTAL	**10**	**100**

Table 3 ; Glomus tympanicum tumour

Fisch Type	No. of patients	Percentage
A	5	83.33
B	1	16.66
C	0	33.33
D	0	33.33
TOTAL	**6**	**100**

Table 4; carotid body tumour

Schamblin Tumour Type	No. of patients
I	2
II	7
III	2
TOTAL	**11**

Table 5 Signs & symptoms

	Pulsatile Tinnitus	Hearing loss	Blood stained Discharge	Vertigo	Pulsatile neck swelling	CN palsies Vii x xii	Aural polyp	Neck mass
Jugulare	10	8	7	2		2 4 4	4	2
Tympanicum	6	6	6	2			2	
CBTs					11			
Vagale								2

Table 6 Surgical techniques and result

Tumor	Surgical technique	Result
Glomus tympanicum Type A Type B	Transcanal Transmastoid,facialnr esses	Cured, on follow up Cured, on follow up
Glomus jugulare Type C1 Type C2 Type D	Fisch Type A approach Fisch Type A approach	Cured, on follow up Cured, vii nerve palsy Recurrence, advised RT, lost for Follow-up
Carotid body tumors Type I Type II Type III	Transcervical Transcervical Transcervical	Cured, on follow up Cured, on follow up Cured, on follow up
Glomus vagale	Transcervical	Cured, on follow up

Discussion;

all head and neck PGs arise from the parasympathetic nervous system. These tumorshave been referred to by various terms, but generally the site of origin defines the name given to these tumors (e.g.,carotid body tumor (CBT) or carotid paraganglioma, jugular paraganglioma, vagal paraganglioma or glomus intravagale etc.). Glomus tumors occur at a rate of approximately one case per 30,000 individuals and represent 0.012% of all tumors diagnosed in humans [6].

Paragangliomas can be familial or non-familial. Baysalet al. [11], suggest that germ line mutations in the mito-chondrial complex II genes, SDHB, SDHC, and SDHD,cause hereditary PGs. Their studies conclude that muta-tions in SDHD are the leading cause of head and neck PGs.SDHD and SDHB mutations accounted for 70% of familialcases and approximately 8% of non-familial cases.

The most common presenting symptom of glomus tumors is pulsatile tinnitus (80%) followed by hearing loss (60%)[12]. Browne's sign is an uncommon physical finding (10%-30%). Cranial nerve deficits may be seen with larger tumors. Deficits of cranial nerves IX and X are most commonly seen but CNs VII, VIII, XI, and XII can also be affected. In our study all the patients of glomus jugulare and glomus tympanicum had tinnitus as main symptom. All patients of glomus tympanicum had hearing loss as complaint while as 8 out of 10 glomus jugulare patients had hearing loss as complaint. Two patients from both glomus juglare and tympanicum group had vertigo as chief complaint. all 11 patients of CBT presented with pulsatile neck mass. 2 Glomus vagale patients presented with isolated neck mass.

In the head and neck region,vagal paragangliomas are historically considered the paragangliomas with the greatest propensity for malignant behavior [13]. Less than 10% of paragangliomas in the head & neck are malignant. For paragangliomas without clinical evidence of lymphadenopathy, removal of adjacent regional lymph nodes is recommended to identify possible occult metastasis that would reclassify the tumor as malignant [14]. None of patients in our study had malignant paraganglioma.

Because of slow expansile growth of these lesions, the lower cranial nerves dysfunction is of gradual onset and in most patients, they are well tolerated as a result of gradual compensation. The anterior extension encasing cavernous sinus and internal carotid artery may produce Horner's syndrome and III, IV, V and VI nerves palsy. Intracranial extension can produce posterior fossa symptoms as nystagmus, ataxia, hemiparesis and increased intracranial.
tension. The extracranial extension along the internal jug-ular vein can produce a visible mass in the oropharynx or a palpable mass in the neck. The intraluminal growth can block venous drainage and occlude the sigmoid sinus and if present bilaterally, increased intracranial tension can occur. The intracranial extension superiorly can produce the cerebellopontine angle syndrome (deafness, tinnitus, VII nerve palsy) and those extending still laterally can produce bloody otorrhoea, a visible mass through tympanic membrane and a bruit over the mastoid. In our study two patients in glomus juglare group had cranial nerve vii, x and xii palsy. Other 2 patients had isolated cranial nerve x and xii palsy. Tumor mass presenting as Aural polyp was seen in 4 patients of glomus juglare and 2 patients of glomus tympanicum.

CBT usually presents between the fifth and seventhdecades of life as a lateral neck mass discovered by the patient or by an examiner incidental to other complaints.Rarely are symptoms secondary to the endocrine products of the tumor. Some patients report a thrill or pulse-like sensation, headache, and occasionally voice changes and vertigo. Classic physical findings are those of a pulsatile lateral neck mass that exhibits medial and lateral mobility but not superior or inferior mobility. Specifically, the eighth through twelfth cranial nerves may be affected. In a series of 27 CBTs reported by Dardik et al. [15], 78% had neck symptoms, 19% had ear pain and 11% were asymptomatic.

In our series, all cases had neck symptoms and signs
In cases of skull base PGs, CT & MRI can be complementary imaging techniques; especially before surgical resection [16]. CT scanning is the most useful modality for the evaluation of PGs of the temporal bone (glomus tympanicum and glomus jugulare) and exquisitely demonstrates the bony detail of the middle ear and jugular foramen. CT scanning can differentiate PGs from other causes of pulsatile tinnitus, including an aberrant carotid artery or high riding jugular bulb and can obviate the need for an unnecessary and dangerous biopsy [17]. For sus-pected carotid body and vagal PGs or for patients with a palpable neck mass, thin section CT scanning is performed with the use of contrast material from skull base to the thoracic inlet at 2.5- or 3-mm intervals in the axial plane.

we do not embolize tumors pre-operatively,our intra-operative blood losses are higher than that quoted by some series where embolization was performed.Balloon occlusion test to detect efficacy of contralateral blood flow through circle of Willis was done in all left sided and shambling II and III CBTs.

During tumor resection, care should be taken toprevent kinking or spasm of the major vessels. Low dose heparin (approximately 0.5 mg heparin) with temporary carotid clamping for dissection of the posterior carotid bifurcation is advisable

Radiotherapy (RT)—Patients with large tumors wherethe morbidity is high and for recurrent tumors are advised RT. In case of malignancies, post-operative radiotherapy can be given as in cases of squamous cell carcinomas. Stereotactic radiosurgery and stereotactic radiotherapy are new additions in the treatment of PGs.in our study we advised radiotherapy to two patients of glomus jugulare, both of them were operated previously by some other surgeon and they had recurrence of tumour with intra cranial extention.

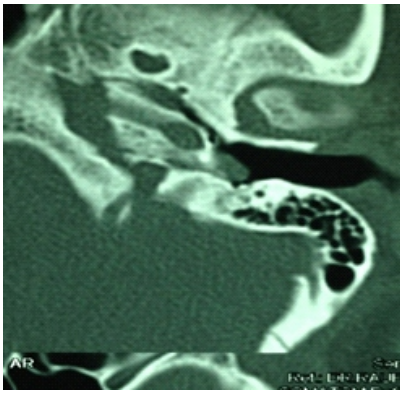


Figure 1; glomus tympanicum type A

VOLUME-6 | ISSUE-4 | APRIL - 2017 • ISSN No 2277 - 8179 | IF : 4.176 | IC Value : 78.46

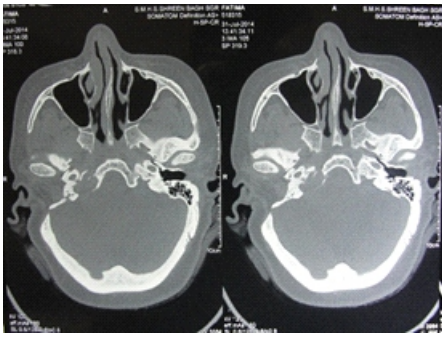


Figure 2; glomus juglare type C



Figure 3; intra operative pic of CBT excision.

References

1. Boedeker CC, Ridder GJ, Schipper J. Paragangliomas of the head and neck: diagnosis and treatment. Fam Cancer. 2005;4(1):55-9, http:// dx.doi.org/10.1007/s10689-004-2154-z.
2. Jackson CG, Glasscock ME, 3rd, Harris PF. Glomus Tumors. Diagnosis, classification, and management of large lesions. Arch Otolaryngol. 1982;108(7):401-10.
3. Kollert M, Minovi A, Mangold R, Hendus J, Draf W, Bockmuhl U. [Paraganglioma of the head and neck—tumor control, functional results and quality of life]. Laryngorhinootologie. 2006;85(9):649-56. Parag-angliome des Kopf-Hals-Bereiches—Tumorkontrolle, funktionelle Ergebnisse und Lebensqualitat nach Operation.
4. Sykes JM, Ossoff RH. Paragangliomas of the head and neck. Otolaryngol Clin North Am. 1986;19(4):755-67.
5. Wasserman PG, Savargaonkar P. Paragangliomas: classification, pathology,and differential diagnosis. Otolaryngol Clin North Am. 2001;34(5):845-62, v-vi, http://dx.doi.org/10.1016/S0030-6665(05)70351-0.
6. Hu K, Persky MS. The multidisciplinary management of para-gangliomas of the head and neck, part 2. Oncology (Williston Park). 2003;17(8):1143-1161.
7. Hu K, Persky MS. Multidisciplinary management of paragangliomas of the head and neck, Part 1. Oncology (Williston Park). 2003;17(7):983-993.
8. Persky MS, Hu KS. Paragangliomas of the head and neck. In: Harri-son LB, Hong WK, Sessions RB, ed. Head and Neck Cancer: A Multi-disciplinary Approach.3rd ed. Philadephia: Lippincott Williams & Wilkins; 2009: 655-687.
9. Suarez C, Rodrigo JP, Mendenhall WM, et al. Carotid body para-gangliomas: a systematic study on management with surgery and radio-therapy. Eur Arch Otorhinolaryngol. 2014;271(1):23-34.
10. Timmers HJ, Gimenez-Roqueplo AP, Mannelli M, et al. Clinical aspects of SDHx-related pheochromocytoma and paraganglioma. Endocr Relat Cancer. 2009;16(2):391-400.
11. Baysal BE, Willett-Brozick JE, Lawrence EC, Drovdlic CM,Savul SA, McLeod DR, Yee HA, Brackmann DE, Slattery WH 3rd, Myers EN, Ferrell RE, Rubinstein WS (2002) Prevalence of SDHB, SDHC, and SDHD germ line mutations in clinic patientswith head and neck paragangliomas. J Med Genet 39(3):178–183
12. ackson CG (2001) Glomus tympanicum and glomus jugulare tumors. Otolaryngol Clin North Am 34(5):941–970
13. Rinaldo A, Ferlito A, Myssiorek D, Devaney KO (2004) Which paragangliomas of the head and neck have a higher rate of malignancy? Oral Oncol 40:458–460
14. Lee JH, Barich F, Karnell LH, Robinson RA, Zhen WK, Gantz BJ, Hoffman HT, American College of Surgeons Commission on Cancer; American Cancer Society (2002) National Cancer Data Base report on malignant paragangliomas of the head and neck.Cancer 94(3):730–737
15. Dardik A, Eisele DW, Williams GM, Perler BA (2002) A con-temporary assessment of carotid body tumor surgery. Vasc En-dovascular Surg 36(4):277–283
16. van den Berg R, Verbist BM, Mertens BJ, van der Mey AG, van Buchem MA (2004) Head and neck paragangliomas: improved tumor detection using contrast-enhanced 3D time-of-flight MR angiography as compared with fat-suppressed MR imagingtechniques. AJNR Am J Neuroradiol 25(5):863–870
17. Lustrin ES, Palestro C, Vaheesan K (2001) Radiographic evalu-ation and assessment of paragangliomas. Otolaryngol Clin North Am 34(5):881–906