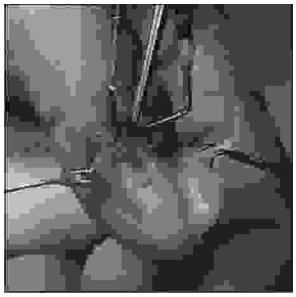
ORIGINAL PROF-1087

GLOMUS TUMOURS



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ABSTRACT... Glomus tumours of the temporal bone occur in the region of the jugular bulb and middle ear. They are rare, highly vascular, slow growing tumours and most are benign. Tumours that originate from the jugular bulb and extend to involve the middle ear are referred to as glomus jugulare tumours. Those that are found in the middle ear around the otic ganglia in the tympanic plexus are known as Glomus tympanicum. These tumours occur predominantly in women in the fifth and sixth decades of life. Because of the insidious onset of symptoms, these tumours often go unnoticed and there is often a significant delay in diagnosis. Morbidity in these cases is determined by their size and position. **Objectives:** 1. To study the age/sex incidence of patients suffering from glomus tumours of the temporal bone. 2. To see various clinical presentations with which these tumours present and their variation according to the age and sex. **Design:** Our study design was non-inter-ventional descriptive. **Settings:** This study was performed at CMH Rawalpindi and Multan from January 2000 to June 2002. **Subjects:** We included ten patients of glomus tumours of the temporal bone in our study. 8 out of these were females and two were males. Though most of the patients belonged to middle age group yet few were also from the younger and older groups. **Interventions:** All the patients underwent CT scan with and without contrast, MRI of the requisite site and carotid angiography. **Results:** We found that these tumours were predominantly present in females, mostly in the middle age group. Individual symptoms were studied in detail and their presence was found to be directly proportional to the increasing age of patients. **Conclusion:** It is concluded that due to the slow growth of this tumour the diagnosis is often delayed until it is extensive. Therefore clinicians should be more vigilant about this rare disease and must keep it in their differentials.

Key words: Glomus tumours, Paragangliomas.

INTRODUCTION

Glomus tumour of the temporal bone occurs in the region of jugular bulb and the middle ear[1, 2]. They are rare, highly vascular, slow growing tumours and are mostly benign. Tumours that originate from the jugular bulb and extend to involve the middle ear are referred to as

glomus jugulare tumours. Those found in the middle ear around the otic ganglia in tympanic plexus are known as glomus tympanicum. These tumours occur predominantly in women in the fifth and sixth decades of life. Because of the insidious onset of symptoms, these tumours often go unnoticed and there is often a significant delay in diagnosis. Morbidity in these tumors is determined by their size and position.

The study was carried out by us to see age and sex incidence of patients suffering from glomus tumours of temporal bone and also to see various clinical presentations with which these tumours present and their variation according to age and sex. Being a very rare disease, the clinicians are liable to miss the diagnosis because of little clinical experience.

The aim of the study was to promote awareness amongst the clinicians that they should have a suspicion of this disease if the patient presents with pulsatile tinnitus deafness, cranial nerve palsies, non-specific headache and pulsatile red mass behind the tympanic membrane. This purpose has been selected with the aim of early diagnosis, timely management thus better prognosis.

Glomus tumours arise form paraganglionic chemoreceptor cells and are part of the APUD system of the cells of neural crest origin. Paragangliomas of the jugulo-tympanic area are the most common tumours involving the middle ear. In 1941, Guild described the distribution of normal glomus bodies within the temporal bone. Then in 1945, Rosenwasser correctly recognized and reported the first case of a tumour arising from these cells. After this report, many new cases were reported and many older cases were reclassified as true glomus tumours .

These neoplasms can occur at any age but the peak incidence is in the fifth decade. Glomus tumours occur four to five times as often in women than as in men and there is predilection for Caucasians[3,4,5,6]. The majority of these tumors arise from along the jugular bulb or Jacobson's nerve (85%), while 12% arise from glomus bodies overlying the promontory and 3% have their origin attributed to Arnold's nerve. The most common presenting symptoms include conductive hearing loss and pulsatile tinnitus[7,8]. Other symptoms may include aural hemorrhage or otorrhoea, otalgia and facial palsy. When the tumours enlarge within the jugular foramen, neuropathies of IX, X, XI and XIIth cranial nerves occur. Anterior expansion towards the clivus may produce Abducent and Trigeminal palsies. Aquino's sign is blanching of the tympanic mass with gentle pressure on the carotid artery. The sign of Brown is the pulsation sign elicited by pneumatic compression and abolished with further compression. These tumours may be multicentric[9,10]. Association of these tumors with tumors of cells of neural crest origin (MEN 1 and pheochromocytoma) is not uncommon. This must be considered to rule out the devastating pheochromocytoma. Malignancy occurs in one to three percent of these tumours[11].

Evaluation includes a careful history and physical examination. High resolution computed tomography provides accurate information about bony destruction and the extent of the disease[12,13]. Arteriography remains the most useful radiological modality available for the assessment of a glomus tumour. Prior to angiography, serum catecholamines and urinary vanillylmandelic acid (VMA) levels should be obtained to screen for tumours which secrete these vasoactive compounds[12]. Therapeutic options for glomus tumours of the temporal bone include surgical excision, radiotherapy[14], combined therapy and in selected cases observation. Surgical excision offers the only chance for total tumour eradication[13,15].

PATIENTS AND METHODS

MATERIAL AND METHODS

This was a hospital based non international, descriptive study of 10 patients who presented to combined military hospital Rawalpindi and Multan with a suspicious of glomus tumour of temporal bone and diagnosed as such after thorough investigations. Because this disease is very rare therefore convenience sampling was done and

first ten patients who reported to us were included in study. Data collection was based on observational facts. This study was carried out from January 2000 to June 2002. Approval was taken from hospital ethics committee.

CMH Rawalpindi and Multan are tertiary care hospitals in Military set up. Military personal and their families are being referred from all over the country. In addition large numbers of civilian patients also consult. These are well-established centers for investigations/ treatment.

INCLUSION CRITERIA

1. All age groups
2. Both sexes.
3. Already operated cases of glomus tumours that presented in ENT OPD for follow up.

EXCLUSION CRITERIA

1. Patients failing to follow up for initial six months post operatively.
2. Patients unwilling to participate in the study.

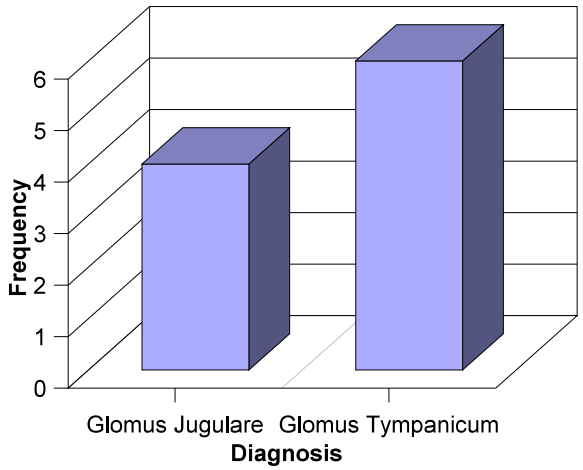
DATA COLLECTION AND ANALYSIS

Our patients (10 in no) presented with deafness, pulsatile tinnitus, non-specific headache, hoarseness and dysphagia. We collected information regarding the age of patients, sex and their presenting complaints. Diagnosis of glomus tumours was based on history, clinical examination as well as investigations including pure tone audiometery, blood complete picture, urine RE, X-Ray chest (PA view), plain X-Ray mastoids, CT scan with and with out contrast or MRI of the requisite site, carotid angiography and 24 hrs urinary VMA estimation. In addition; ECG, LFT's, Serum urea and electrolytes were performed in order to assess heart, liver, kidneys in patients above 40 years of age. Operated cases were followed up monthly for first year post operatively and twice monthly for second year post operatively. Patients were examined for cranial nerves; recurrence and CT scan/MRI was done in suspicious cases only. Statistical analysis of data was performed using SPSS package.

RESULTS

As glomus tumours are a rare occurrence so we could only collect 10 such cases from January 2000 to June 2002. Six cases out of them (60%) were of glomus tympanicum tumour i.e.; type A whereas remaining four cases (40%) were diagnosed as glomus jugulare, out of which one case was of type B (25%), 1 case of type C (50 %) and one type D (25%) - (Fig-1).

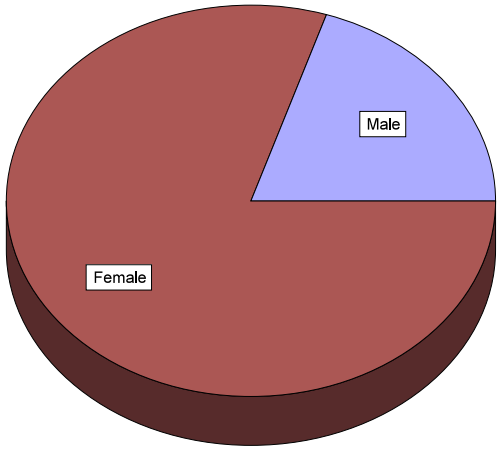
Fig-1. Diagnosis of glomus tumours of temporal bone



Eight out of our 10 patients were females (80%) whereas only two patients were males (25%) making female to male ratio 4:1 - (Fig - 2).Most of our patients belonged to middle age group i.e., between 45 to 65 years of age but younger and older age groups were also involved. Mean age of females turned out to be 44.63 years whereas it was fifty years in case of male patients making the total mean age of 45.70 yrs - (Fig -3).

Nine out of 10 cases presented with deafness, whereas only 1 patient had no such complaint. All the patients 6 in number later diagnosed to have glomus tympanicum tumour had deafness (100%) whereas 3 out of 4 patients of glomus jugulare had it (75%). All such patients were found to have conductive deafness. Over all 100% male and 87.5% female patients were affected whereas only 1 female patient (12.5%) was spared.

Fig-2. Sex incidence of glomus tumours of temporal bone.



Sex of the patient

Fig-3. Age incidence of glomus tumours of temporal bone

Pulsatile tinnitus showed the same percentage as of deafness being absent in the same case that had no complaint of deafness. Non specific headache overall, was found present in only 5 cases (50%). Two out of 3 cases of glomus jugulare had this complaint (75%) whereas only 2 patients of glomus tympanicum had it (33.3%) and 4 were spared (66.66%). It also showed the tendency in the upper limits of middle age group i.e; around 52.80 yrs. Four out of 8 females patients had headache (50%) and rest 4 had not (50%). 1 out of 2 male patients had headache (50%). Pulsatile red mass behind the tympanic membranes was found in 8 out of 10 cases (80%). Out of the 2 patients (20%) that had no such mass, 1 case was of glomus jugulare and the other was glomus tympanicum tumour. Both these patients were females. Hoarseness was present in 4 out 10 cases (40%), whereas it was absent in rest 6 patients (60%). It also showed involvement of upper limits of middle age (mean =56.75 years). 3 out of 7 females (37.5%) had hoarseness whereas remaining 5 (62.6%) had not. All 3 cases of glomus jugulare had hoarseness whereas 6 cases of glomus tympanicum tumour had no such problem.

Dysphagia was present in only 3 out of 10 cases (30%) and was absent in 70% cases. 3 patients out of 4 that had glomus jugulare (75%) had this complaint. 2 were females (60%) and 1 was male (33.33%). It was absent in 1 out of 4 cases of glomus jugulare (25%) and in all 6 cases of glomus tympanicum (100%). Mean age of patients affected turned out of to be 57 years. We found cranial nerve palsies (either IX or X CN or both) in all 4 cases of glomus jugulare (nerve paralysis or Horner's syndrome). No cranial nerve palsy was found in any case of glomus tympanicum. Overall percent was 40 % - (Table -I).

Table-I. Mean age of different clinical presentations

Clinical presentation	No. Of patients	Mean age of presentation	%age
Deafness	9	46.33	90%
Pulsatile tinnitis	9	46.33	90%
Headache	5	52.80	50%
Pulsatile red mass behind tympanic membrane	8	47.88	80%
Hoarseness	4	56.75	40%
Cranial nerve palsies	4	56.78	40%
Dysphagia	4	56.75	40%

DISCUSSION

It is seen that glomus tumours of temporal bone mostly involve females of middle age group and the clinical features are in direct proportion to the increasing age. These results are comparable to contemporary research.

No local author has yet undertaken any study about this disease and no material is available in the texts. Results of our study are thus compared with foreign based studies.

In a study of glomus tumours of the temporal bone carried out by Pemberton LS et al in 2005, the median age at presentation was 55 yrs (Range 23-82 yrs).

In 1975, Jacobs IN et al carried out a study which revealed Glomus tumours of temporal bone in patients as young as 6 months. Same results were shown in another study conducted by Bartles LJ et al in 1988.

Taylor DM et al in 1965, found that though these glomus tumours were most common in patients aged 40-60 years, yet they encountered two cases presenting in the eight decade of life.

In our study we were able to collect the data from 10 patients suffering from glomus tumours of temporal bone, including both glomus tympanicum and glomus jugulare tumours. Our patients that included both male and female ranged in age from 30-71 years with the mean age of about 46 about years. This is in line with the studies carried out in the west.

In a study made by Spector GJ in 1973, 46 patients of glomus tumours of temporal bone were analyzed and female to male ratio was shown to be 6 : 1. This ratio according to our study turned out to be 4:1.

Rohit et al in 2003, showed 17 cases of glomus tympanicum that were treated by surgery and all were females (100%).

Our society is basically a male dominated society. Health of females is not only poor but is given least priority. They are mostly dependant on the male members, especially in rural areas and have no access to medical resources that are otherwise also quite meager. That is why though our study showed female predominance in this regard; our ratio of female involvement was less than the studies carried out in other parts of the world.

In 1994, Watkin et al analyzed 61 cases of glomus tumours of temporal bone and found that deafness was the presenting feature in 69% cases, pulsatile tinnitus in 55% cases and hoarseness and dysphagia due to cranial nerve deficit in 16% cases each. In addition to above 15% cases also had a non-specific headache, 8% had imbalance, 6% had otorrhea and only 3% cases had symptoms related to endocrine syndrome. Their study also revealed that 75 % cases had a pulsatile red mass behind the eardrum on examination. Facial palsy on affected side was seen in only 8% cases.

In 2004, Liu JF et al studied ten patients of glomus tumours and found that hearing loss and pulsatile tinnitus were the most common presenting symptoms. Same results were published previously by Tatla T et al in 2003.

Our study revealed no patient with imbalance, otorrhea or endocrine disturbance. Moreover none had facial nerve palsy. We found in our study that deafness and pulsatile tinnitus were the most common presenting features being present in 90% cases. Headache was the presenting complaint in only 50% cases, hoarseness in 40% cases and dysphagia in only 30% cases. Examination of patients in our study revealed pulsatile red mass behind the tympanic membrane in about 80% cases and cranial nerve palsies (IX, XCN) in 40% cases. The symptoms and signs seen in our patients gave more or less mixed picture. Though there was no variation found in their character with age and sex yet our study did indicate their increased frequency of occurrence with increase in age i.e. older the patient the more variety of symptoms and signs he had as compared with the younger group.

CONCLUSION

It is concluded that the glomus tumours of the temporal bone are slow growing, highly vascular, benign but locally invasive and destructive tumours that occur predominately in females. In rare circumstances they become malignant. These tumours can affect any age group, however; it has a propensity for people in middle ages which is the most productive part of life. The severity of symptomatology is directly proportional to increasing age of patients.

Thorough clinical examination and otoscopy findings supplemented by CT scan and MRI give early diagnosis of the disease. This is a key stone in the decision making for the future management of these patients thus preventing them from further misery of the disease process which although is rare but devastating. Clinicians should therefore, be more vigilant about this tumour so that it is detected at early stages before extensive damage is done.

It is further concluded that more locally based studies are required on the subject to lay down principles to help such patients in our setup.

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