Case Report

GLOMUS JUGULARE TUMOUR MISDIAGNOSED AS A PAROTID NEOPLASM: A Case Report and Literature Review

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ABSTRACT

A variety of tumours can present as swellings in the parotid region. Patients with such tumours usually present to the dental surgeon or at the Ear, Nose and Throat (ENT) Clinic. In order to improve on early recognition and appropriate referral, the clinicians need to improve on their depth of knowledge regarding patterns of presentations of rare pathologies such as glomus jugulare tumour.
This case report identifies common presenting signs and symptoms of a glomus jugulare tumour which would help the clinician to arrive at an appropriate diagnosis and thus enable the patient get a timely treatment. The report identifies the relevant clinical, radiologic and pathologic parameters of this rare lesion.
There are currently different management strategies for the management of glomus jugulare tumours ranging from conservative management to stereotactic radiosurgery.
This report offers guidelines for clinicians to decide on the best treatment modality based on the patient's presentation.

KEY WORDS: Glomus jugulare, diagnosis, treatment

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INTRODUCTION

Glomus jugulare tumour (GJT) also known as jugulotympanic paraganglioma, is a rare, slow growing, locally aggressive neoplasm arising from the paraganglia within the adventitia of the jugular bulb at the skull base. A variant arises from the paraganglia situated near the middle ear surface of the promontory and the bony wall of the tympnanic canal, and here referred to as glomus tympanicum.[1,2,3]

GJT expresses in spontaneous or familial variety. They are commoner in females than in males with the familial variant having more male predilection.[4] The age range is reported to be between 13 - 85 years with a peak age incidence of between 4^{th} – 5^{th} decades of life.[5]
Glomus tumour grows in multiple directions, compressing adjacent cranial nerves and invading vascular structures with some spreading intracranially. Implicitly, these tumours are associated with pulsatile tinnitus, hearing loss and (or) a middle ear mass. However, large GJTs with an intracranial extension are relatively rare. Due to the inaccessibility of the location of the disease, most patients present with an advanced disease at diagnosis.[2,5,6]

Several classification systems for GJTs have been proposed. However, the more common are

the classifications by Jenkins - Fisch[7] and Jackson-Glasscock and co-workers.[8] The Jenkins-Fisch system classifies GJT into 4 types designated A - D, depending on the tumour size and relationship to the anatomy of the petrous bone. Type A tumours are limited to the middle ear cleft. Type B tumours are limited to the tympano-mastoid area. Type C tumours involve the infralabyrithine compartment and extend to the petrous apex while Type D tumours extend intracranially and are further subdivided by diameters into types D1 (< 2cm) and D2 (> 2cm).

Glomus tumour is a relatively rare tumour in Nigeria. A pubmed search revealed only 4 Nigerian case reports.[9,10,11,12] Further pubmed search of the English literature for GJT with a parotid region presentation, revealed only one report.[13]

A case of GJT is presented with the aim to document and draw attention to this rare disease's ability to be misdiagnosed as a parotid neoplasm, a neoplasm often referred to the Oral and Maxillofacial surgeon / Ear Nose and Throat surgeon's clinics.

CASE REPORT

A 29-year old female was referred to the Oral and Maxillofacial Surgery (OMFS) outpatient's clinic of the study institution on account of a left parotid swelling of 8 years duration. The swelling was painless and has been slowly increasing in size. The patient later developed dysphagia, hoarseness, reduced sense of hearing on the left ear, tinnitus and also a history of associated weight loss. Pulsatile tinnitus was elicited. There was no facial nerve involvement. There was no familial history of the disease or history of alcohol ingestion or tobacco use.

Further examination revealed a cachectic patient with a swelling on the left parotid region extending from the upper preauricular region to the upper cervical third superio-inferiorly. The posterio-anterior extension was from pre to post-auricular regions. It measured about 14cm X 10cm in its widest dimension (Fig 1). The swelling appeared diffuse, firm, non-tender and fixed. The mass was non pulsatile, there was no bruit on auscultation. There was also no associated cervical lymphadenopathy. The tympanic membrane appeared bluish on otoscopy (done by the ENT surgeons).
Intraoral examination revealed presence of hemiatrophy of the left side of the tongue with restriction of tongue movement and deviation to the ipsilateral side (Fig 2 and Fig 3). There was however no gait disturbance.

An Impression of a parotid neoplasm (malignant) was made to rule out GJT. A fine needle aspiration cytology (FNAC), radiological investigations (conventional CT scan and Carotid CT angiography) were requested. The CT scan revealed a large moth eaten appearing mass around the region of the jugular fossa of the left skull base with intracranial extension into the middle and posterior cranial fossa (Fig 4). The CT Angiography showed a highly vascular tumour with patency of the carotid vessels (Fig 5). A diagnosis of glomus jugulare Type D2 (Jenkins-Fisch classification) was made based on clinical and radiological findings. FNAC revealed a benign neoplasm with a request for an incisional biopsy. The histologic sections showed nests of neoplastic cells, which were arranged in an organoid (Zellballen) fashion, with intervening fibroma secular stroma between the nests. The cells had abundant eosinophilic cytoplasm, with round to oval fairly uniform nuclei with evenly dispersed chromatin. No area of glandular differentiation was seen. The overall histologic picture was consistent with glomus jugulare.

After an evaluation of the clinicopathologic findings at the Multidisciplinary Tumour Board meeting, it was concluded that the tumour was not amenable to surgery because of the involvement of vital structures and intracranial involvement. Radiotherapy was advised and patient was referred to the oncology unit. After a review of the patient by the oncologist, it was decided that the patient should be treated with conventional radiotherapy. Patient had 6 cycles of radiotherapy over a period of 6 weeks. Patient was reviewed again at the OMFS clinic 6 months post radiotherapy and it was found that the effect of radiotherapy on the tumour mass was moderate (Fig 6 & 7). Patient said she was advised to undergo a second course of

radiotherapy but she declined on account of financial constraints. Upon approval from the institutional ethics committee, the patient's data was retrieved for publication.

DISCUSSION

Glomus tumours are rare, benign tumours that can develop in several locations in the body including areas in and around the middle ear. These tumours are often extremely vascular due to their intimate involvement with vascular tissues.[5]

Our patient presented with a tumour, which appeared on CT scan as a mass around the jugular fossa of the middle and posterior skull base. Pathognomonic CT scans are said to provide adequate information on the likely pathology and thus there is usually no need for an incisional biopsy.[14] Genetically, there has been reports of germ line mutation in one of the genes SDHB, SDHC and SDHD, as being related to the solitary GJT while the gene (PGL1) that was recently identified at the 11q23 locus, has been related to the familial types.[15,16]
Glomus tumor is reported to be rare in blacks with limited reports in Nigeria.[10] Godwins et al.[11] did present three cases of chemodectomas (paragangliomas, glomus tumours, carotid body tumours), at the Jos University Teaching Hospital in 2009. Two of them were female while the third was male. Another case series of a ten-year (1992 - 2001) retrospective study carried out in North Eastern Nigeria by Ngadda and co-workers (12) found two cases (1.4%) of glomus tumour out of 139 cases of vascular tumours. The said study reported hemangioma as being the commonest vascular tumour accounting for 50.3%.

GJT has a high female preponderance with a highly variable age incidence that is sometimes reported to be in the 5th or 6th decade bracket.[4,5] Our case was a female who presented in the 3rd decade. No known ethnic or racial predilection is reported in the literature. The incidence of malignancy in glomus tumours is less than 5%. The commonest presentation is hearing loss and pulsatile tinnitus with 60-80% of patients presenting with these complaints. Others are cranial nerve (vagal, hypoglossal and accessory nerves) neuropathy leading to lingual hemiatrophy, dysphonia and dysphagia.[5,6,17] Our patient presented with all these symptoms. It is reported to occur more commonly on the left side, an occurrence our case expressed.[18,19]
There has been no reported case of glomus tumour presenting as a parotid neoplasm in Lagos Nigeria with only one case reported in the English literature.[13] Our diagnosis was based on the history, clinical observation and advanced radiological imaging as well as histopathological report.

A rare occurrence of palpitations and headaches is reported to be suggestive of excess catecholamine secretion. A urine test for elevated epinephrine, dopamine and vaniylmandellic acid can confirm this suspicion and an MRI of the chest and abdomen can be used to detect additional extracranial lesions.[20]
The treatment options are governed by many factors comprising; patient factors (age, performance status, co-morbidity, informed patient choice), tumour related factors (volume, evidence of growth) and treatment factors (possibility of satisfactory surgical excision, possibility of achieving disease control with different treatment modalities, operative risks and risk of adverse radiation reactions).[19,21] The four options of management for patients with glomus tumors are: Microsurgical removal, stereotactic radiosurgery (SRS), fractionated radiotherapy and conservative management by radiological surveillance.[5,6,19,22]

Conservative management was not an option in this case as the patient presented late with a type D2 (Jenkins - Fisch Classification). The intimate relationship of glomus jugulare tumours to neural-vascular structures makes complete resection without unacceptable side effects often impossible. Conversely, their high vascularity makes them especially sensitive to radiation treatment.

Treatment options will take into consideration the precise anatomical position of the lesion, its size, (dictating the perceived risk of alternative intervention with microsurgery) and clinical presentation (clinical symptoms and signs of brain stem dysfunction may necessitate surgical removal, which could be complete or partial).

All these perceived risk would influence the choice of treatment option.[18,22]

Our patient had a large tumour, 14cm by 10cm in its greatest dimension. The option of combined surgery by the Head and Neck and the Neurosurgical unit of the study institution were considered at the Multidisciplinary Tumour Board meeting. The size of the tumour, its associated neural symptoms (tinnitus, dysphonia, dysphagia) and presence of intracranial extension observed in the patient precluded such an option. A two-fold increase in morbidity or mortality often result from total or subtotal resection of an intracranial glomus tumour.[23] Fractionated radiotherapy is believed to be effective but due to its propensity to encompass a wide field, it has been reported to induce bone necrosis of the skull base.[11,21] Other potential complications include troublesome reduction of saliva production, brainstem radionecrosis, radiation-induced stenosis of the internal carotid artery leading to hemiplegia and secondary malignancy.[12,19,21] Stereotactic Radiosurgery (SRS/SRT) is another viable option. SRS is a highly conformal radiotherapy with treatment to a precisely delineated target volume, delivered using stereotactic localization techniques. The procedure allows radiation to be limited to the target area and thus helps spare the surrounding tissues as much as possible.[21,24]

Comparative studies of all treatment options have reported radiotherapy and radiosurgery as having better outcome than microsurgery. Microsurgery has been reported to show higher tumor control failure. They equally reported more complication rates. The incidence of cranial nerve palsies after treatment were significantly higher in surgical than in radiotherapy or radiosurgery series. The researchers tended to suggest that surgery should be considered only for selected cases.[24,25] Some researchers have suggested that clinical signs and symptoms of brain stem dysfunction may necessitate surgical removal either complete or partial.[18,22]

Because we do not have facilities for SRS and patient could not afford to seek care elsewhere, the option of SRS was dropped and patient was referred to the oncology unit for conventional radiotherapy. The patient had 6 cycles of radiotherapy (over a period of 6 weeks). However, the effect of radiotherapy on tumour size was moderate. The dimension of the mass was 12.5cm x 9cm, 6 months postradiation (Fig 6 & 7). Patient had however not developed any additional signs and symptoms.

Glomus jugulare tumour is a rare neoplasm in Nigeria, which can present as a parotid swelling. Some surgeons may not be familiar with its presentation and tumours presenting late like they do, may likely be misdiagnosed. Late presentation makes surgery an unattractive option. Every stakeholder in head and neck surgery (oral and maxillofacial surgeons, dental surgeons, ENT, and General surgeons) must be able to recognize this tumour on presentation so as to improve the chances for surgical treatment. A high index of suspicion and detailed evaluation of masses presenting in the parrotid region will result in better outcome.

The likelihood of late presentation in a less endowed environment like Nigeria is high and treatment options for such patients would be limited to conventional radiation at best. Surgical removal by radiosurgery for asymptomatic small tumours would not be an option in our environment due to lack of facilities. However, surveillance of small asymptomatic GJT is possible in Nigeria.

Conflict of Interest: None declared

Fig 1. A 28 years old female with a left parotid swelling of 8 year duration.

Fig 2. Left Lingual Hemiatrophy

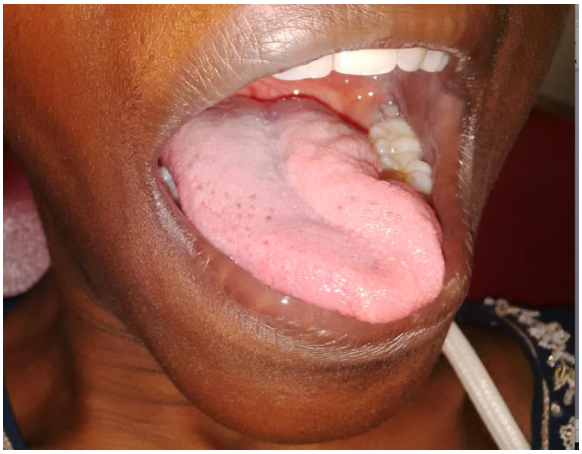

Fig 3. Tongue Restriction on ipsilateral deviation.

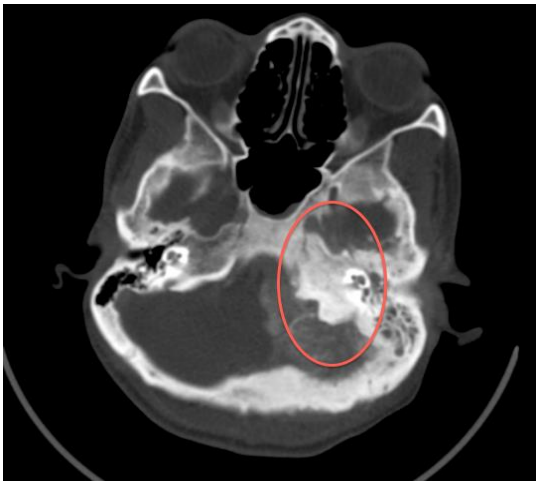

Fig.4. CT Scan, axial section showing mass around the jugular fossa.

Fig.5. CT angiogram showing mass around the jugular fossa patency and flaying of the carotid vessels.

Fig. 6. Post Radiotherapy (6 months)

Fig. 7. Post Radiotheraphy (6 months)

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