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Glomus Tumour-A Rare Presentation Case Report

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Abstract

Introduction: Glomus jugulare tumors are rare, slow-growing, hypervascular tumors that arise within the jugular foramen of the temporal bone. They are included in a group of tumors referred to as paragangliomas, which occur at various sites and include carotid body, glomus vagale, and glomus tympanicum tumors.

Case Presentation: A 4 year female child born to Non consangious parents presented with deviation of angle of mouth to the left since 2 month with drooping of eyelids and severe pain in the left premastoid region. Preliminary examination revealed purulent discharge which was foul smelling and ptosis. Child developed altered sensorium with facial weakness and was shifted in PICU and MRI brain and HRCT temporal bone s/o GLOMUS tumour. Managed with antibiotics.

Conclusion: Glomus tumors are benign, slow growing tumors originating from paraganglionic tissue, mostly located at the carotid bifurcation, jugular foramen, cervical portion vagus nerve, and middle ear cavity. Radiotherapy is treatment of choice for patients with intracranial extension, and patients with bilateral and multiple tumors, or patients who are inoperable.

Keywords: Glomus Tumour(GT)

INTRODUCTION

Glomus tumors (GT), also named as paragangliomas and chemodectomas are rare tumors, accounting for 0.03% of all neoplasms and 0.6% of all head and neck tumors ^[1]. These benign, slow-growing tumors originate from paraganglionic tissue, mostly located at the carotid bifurcation, jugular foramen, cervical portion vagus nerve, and middle ear cavity ^[2,3]. Typically, these tumors are diagnosed between the fourth and sixth decades of life. Women are affected five to six times more often than men. Familial

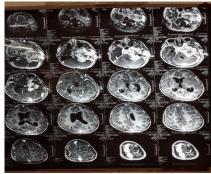
occurrence is likely to occur in 10% of patients, with an autosomal dominant inheritance. Multiple tumors are seen in 78-87% of familial paragangliomas, and the incidence of bilateral GT is 32% for familial cases and 4% for non-familial cases [4].

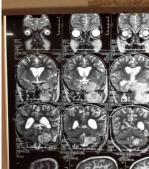
We describe a 4 year female who came with complain of deviation of angle of mouth to the left since 2 month with drooping of eyelids and severe pain in the left premastoid region with altered sensorium and facial weakness.

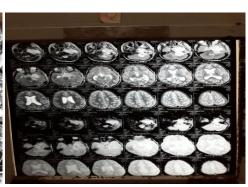
CASE PRESENTATION

A 4 year old bought with deviation of angle of mouth to the left since 2 months with drooping of left eyelid. Baby had swelling of the mastoid region with purulent foul smellin discharge and difficulty in swallowing, besides no hoarseness, dysphagia, nasal obstruction and epistaxis were reported. There are no symptoms related with the carotid artery compression. On physical examination, semi-mobile, painfull mass with a size of approximately 8×6 cm mass extending from tip of mastoid to level IV inferiorly on the

left side were palpated Baby developed altered sensorium and was managed with antibiotics.MRI brain revealed a soft tissue lesion in the skull base involving middle cranial fossa with erosion of left petrous apex and greater wing of sphenoid and extension in the middle ear, intracranial extradural extension with compression of brain stem and cerebellum .HRCT temporal bone suggestive of similar findings thus confirming Glomus tumour. Csf examination was normal.Patient was treated with antibiotics and referred to higher centre for further management.













DISCUSSION

Glomus tumors are mainly presented in two locations; cervical (carotid, vagal) and skull base (jugular and tympanic) ^[5]. These tumors are rarely seen, and have benign characteristics and a very slow growth rate of 1 mm per year ^[5]. The initial diagnosis is usually non-tender neck mass, in case of rapid growth or increase in size, because of compression to the surrounding structures, pain, hoarseness, disphagia may develop.

CT and MRI are useful diagnostic tools for assessing suspicious paraganglioma. CT can reveal the invasion of the bony structures and

intracranial extension, while MRI is better for evaluating vascular structures, extension along neural foramina, and detecting multicentricity [6]. The characteristic appearance of GT in MRI is well-defined hypointense mass with equal signal intensity to adjacent muscle on T1-weighted scans. Intense contrast enhancement is the key finding on MRI. The optimum management of GT still remains controversial. The treatment options are surgery, RT and watchful waiting. Since the metastasis rate is rare, the main aim of treatment choice is to achieve good local control without morbidity and mortality. increasing Local

treatment can be achieved with either surgically as total removal or long-term tumor control and tumor shrinkage with RT.

CONCLUSION

Although good results were achieved with complete surgical removal and total excision with microsurgical approach can often accomplished, there is a significant risk of morbidity (0-39%) and mortality (0-2.7%) with this approach [7]-[9]. Radiotherapy is treatment of choice for patients with intracranial extension, and patients with bilateral and multiple tumors, or patients who are inoperable. Glomus tumors respond to RT slowly. Residual mass persisting after RT does not indicate treatment failure. Tumor may decrease in size, but rarely disappears. Disease control is defined as the absence of progression of symptoms without any increase in size with physical examination or radiological control [10]. Glomus tumors are slow-growing lesions; therefore, it is necessary to be cautious about tumor control without increasing morbidity and mortality.

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