Original Research Paper

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Otorhinolaryngology

SYNCHRONOUS CAROTID BODY AND GLOMUS JUGULARE TUMORS: A CASE REPORT

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KEYWORDS :

INTRODUCTION

Carotid body tumor and glomus jugulare tumor are highly vascular neuroendocrine tumors which are classified as paraganglioma. These are usually slow growing benign tumors.

Although benign in nature they sometimes show malignant potential. Paraganglioma is a very rare tumour and B/L concurrent carotid body with glomus jugulare tumor is extremely rare. We are reporting a case of 32-year-old female who was suffering from B/L concurrent carotid body tumor and glomus jugulare tumor.

Case Report

A 32 year old normotensive and non diabetic female was admitted with complaints of difficulty in hearing for 5 year in right ear. She also complaint about mass in right EAC which was gradually in progressive for last 6 months in addition to this she also complaint about occasionally headache-vertigo difficulty in closing the eye, difficulty in making facial expression for last 2 year.

On examination facial palsy present. The mass was painless non-tender and well defined.

Although no family history was reported, genetic evaluation for SDH mutation was advised due to young age and multicentric presentation She did not give any positive family history.

- CT (Computer Tomography) head and neck angiography there is well defined irregular shaped strongly enhancing mass lesion splaying the internal and external carotid arteries. Measuring 19x12.6mm on right side and 10.6x10.5mm on left side.
- A large similar morphology ill defined strongly enhancing mass region measuring 34x23x35mm (AP x TR x CC) size created at right jugular foramen and superiorly eroding the bones of floor of middle ear causing destruction of bone of middle ear ossicles and inner ear and extending into external auditory cannal.
- Another similar morphology strongly enhancing lesion 10x15mm seen at left jugular foramen.

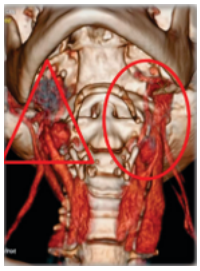

Image A

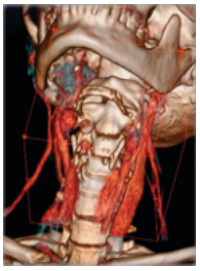

Image B

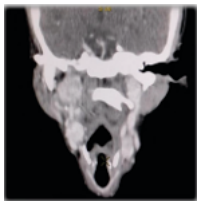

Image C

- **CT (Computer Tomography) Head and Neck Angiogram:** Multiple (4) stronglly enhancing mass lesions seen at B/L carotid bifurcation and B/L juglar foramen (Image C)

DISCUSSION

Definition :

Carotid body tumor and glomus jugulare tumor are highly vascular neuroendocrine tumor which were collectively previously called chemodectoma (now obsolete). Now they are called paraganglioma). There usually are located in the jugular fossa, carotid bifurcation, vagus nerve and other parts of the body. They arise from specialized neural crest cells associated with autonomic ganglia).

Demography

Head and neck paragangliomas are rare tumors, with an estimated overall incidence of approximately 1 per 100,000–300,000 population annually. Carotid body tumors represent the most common head and neck paraganglioma, accounting for nearly 60–70% of cases. The estimated incidence of carotid body tumors is approximately 0.5–1 per 100,000 population, whereas glomus jugulare tumors are considerably rarer.

A slight female predominance has been reported, with a male-to-female ratio ranging between 1:2 and 1:4 in most series. The typical age of presentation is between 40 and 60 years, although familial or SDH-associated cases may present at a younger age.

Multicentric disease occurs in approximately 10% of sporadic cases and up to 30–40% of hereditary cases, particularly in association with SDHD mutations.

Pathophysiology

They are usually benign, hyper vascular, slow growing tumor (<2 cm/5 years) but rapidly growing tumor may also occurs. Only 1% to 5% of them are malignant). Paragangliomas can secrete catecholamines and serotonin regardless of their location, which can result in episodic arrhythmias, hypertension, diaphoresis, headaches, and potential hypertensive crises. Any manipulation of the tumor, such as with surgery, angiography, embolization, or even palpation of the mass, could re- lease these vasoactive substances and precipitate a hypertensive crisis. For this reason, patients suspected of having an actively secreting tumor should be screened preoperatively by testing their urine for vanillylmandelic acid and 5-hydroxyindoleacetic acid).

Paraganglioma may occur in two patterns :

1) Familial : non multicentric up-to 50%.
2) Nonfamilial : multicentric 5%. These tumors usually possess secretory granules (even the functionally inactive tumors) and may actively secrete catecholamines (similar to pheochromocytomas, occur in only 1–4% of gastro jejunal tumor).
3) Up to 30–40% of head and neck paragangliomas are associated with germline mutations, most commonly involving the succinate dehydrogenase (SDH) gene complex, particularly SDHB, SDHC, and SDHD. SDHD mutations are strongly associated with multifocal and bilateral head and neck paragangliomas. Given the young age of the patient and the presence of synchronous bilateral carotid body and jugular tumors, genetic counselling and SDHx mutation testing should be strongly considered. Identification of SDH mutations has important implications for surveillance, family screening, and long-term follow-up.

Carotid Body Tumors

Possibly most common is paraganglioma (some study mentioned Pheochromocytoma is more common). Five percent cases may be bilateral but incidence increases up to 26% in familial cases (autosomal dominant). Carotid body tumors consist of epithelioid cells grouped into cords or clusters, also known as Zellballen. Typically, the cells are polyhedral with granular cytoplasm. VEGF (Vascular endothelial growth factor) and platelet- derived endothelial cell growth factor are expressed in carotid body tumors and may contribute

to the extreme vascularity of these tumors.

Glomus Jugulare Tumor
They are very vascular. Main feeders are from the ECA (especially inferior tympanic branch of ascending pharyngeal artery, and branches of posterior auricular, occipital, and internal maxillary), with additional feeders from petrous portion of the ICA.

Clinical Features
Clinical features of paraganglioma can be divided in two categories :
1) due to release of chemical mediators : these features developed due to release of catecholamine, serotonin and kallikrein, which can result in episodic arrhythmias, hypertension, diaphoresis, headaches, and potential hypertensive crises. Any manipulation of the tumor, such as with surgery, angiography, embolization, or even palpation of the mass, could re- lease these vasoactive substances and precipitate a hypertensive crisis.
2) due to specific location of the paraganglioma.

Clinical Features of Glomus Jugulare
Patients commonly present with hearing loss and possible tinnitus, dizziness and ear pain may also occur. hearing loss may be conductive due to obstruction of the ear canal or sensory neural due to infection of the labyrinth. Various combination of lower cranial nerve palsy may occur. Ataxia and or hydrocephalus can occur.

Clinical Features of Carotid Body Tumor
Usually it is painless slow growing mass present in the upper part of the neck. Large tumors may cause cranial nerve involvement especially vagus and hypoglossal nerve.

Investigation
Investigations for paraganglioma is divided in to two broad groups.

For Diagnosis
For diagnosis of glomus jugulare tumor both CT, MRI and angiogram are usually done. MRI is done for location and extent of tumor, CT scan for better assessment of the bony involvement of the skull base and angiogram for conformation of diagnosis (rule out vestibular schwannoma), ascertain patency of both jugular vein (JV). For diagnosis of carotid body tumor MRI (or CT) is used to evaluates the extent of tumor and intracranial extensions. Carotid angiogram is highly crucial to demonstrates patency of the ECA, ICA and predominant blood supply of the lesion (usually ECA with sometimes contribution from thyrocervical trunk and vertebral artery). Characteristic findingsis splaying of bifurcation.

Endocrine Study
Fractionated plasma metanephrine, 24-hour urinary catecholamines and clonidine suppression test. For glomus jugulare audiometric and vestibular testing also should be performed for diagnosis of hearing loss, prognosis and medicolegal purposes.

Management
Surgery is the treatment of choice for paraganglioma. Both carotid body and glomus jugulare tumor are attached with vital neurovascular structure and there is higher chance of neurovascular injury during the surgery. Endovascular approaches are also used as solely or part of adjuvant treatment to surgery.

Medical Management
Alpha and beta blockers given before surgery-blocks possibly lethal blood pressure lability and arrhythmias. Adequate blockade takes approximately 2–3 weeks of alpha blocker and at least 24 hours of beta blocker therapy; in emergency, 3 days of treatment may suffice.

Radiotherapy
The role of radiotherapy in managing carotid body tumors remains unclear.

Role Of Endovascular Surgery
One of the most significant advances in management has been preoperative, transvascular embolisation of these tumours.

Fractionated Radiotherapy and Radiosurgery
Role of radiation is still controversial and are still reserved for inoperable, surgically unsuitable, residual or in recurrent cases.

Management
Management of head and neck paragangliomas depends on tumor size, location, functional status, genetic profile, patient age, and cranial nerve involvement. A multidisciplinary approach involving otolaryngology, neurosurgery, radiology, endocrinology, and interventional radiology is essential.

Preoperative endocrine evaluation with plasma-free metanephrines and 24-hour urinary catecholamines should be performed to exclude functional tumors. In catecholamine-secreting tumors, adequate alpha-adrenergic blockade followed by beta-blockade is mandatory prior to surgical intervention.

Surgical excision remains the primary treatment modality for carotid body tumors, particularly in young patients with operable disease. Shamblin classification aids in predicting surgical morbidity.

For glomus jugulare tumors, management options include micro-surgical resection, stereotactic radiosurgery (SRS), or fractionated radiotherapy. SRS has emerged as an effective alternative for skull base tumors with high local control rates and lower cranial nerve morbidity.

Preoperative transarterial embolization is recommended to reduce intraoperative blood loss in selected large, hypervascular tumors.

Genetic counselling and SDHx mutation analysis should be considered in young patients and those with multicentric or bilateral disease.

Long-term radiological surveillance is mandatory due to risk of recurrence or delayed progression.

Prognosis and Complication
Glomus Jugulare Tumors
The most common complications are cerebrospinal fluid fistula, facial nerve palsy, and varying degrees of dysphagia. Dysfunction of any of the cranial nerves VII and XII can occur.

Other possible complication include aspiration, delayed gastric emptying, ileus and excessive blood loss. Even after gross total tumor removal, recurrence rate may be as high as one third.

Carotid Body Tumors
Possible complication includes baroreceptor failure (33%), cranial nerve deficit (17%), vascular injury and reconstruction (28%), arterial thrombosis (7%), and death (3%).

CONCLUSION
Synchronous bilateral carotid body tumors and bilateral glomus jugulare tumors represent an exceptionally rare presentation of multicentric head and neck paraganglioma. Early diagnosis through advanced imaging and endocrine evaluation is critical. Contemporary management requires individualized, multidisciplinary planning incorporating surgical resection, stereotactic radiosurgery, genetic evaluation, and long-term surveillance. Given the patient's young age and multicentric disease, genetic testing and lifelong follow-up are strongly recommended to optimize outcomes and detect recurrence early. Genetic evaluation for SDHx mutations should be considered in young patients with multicentric or bilateral paragangliomas.