

*Case Report***Paraganglioma: Carotid Body Tumor Presented As Painless Neck Swelling**

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ABSTRACT

A 28 year old male patient presented with swelling at left submandibular area of 5 months duration, gradually enlarging in size. There was no history of any predisposing factors. Ultrasonography of neck showed a well defined, round to oval homogenous mass lesion at the bifurcation of left common carotid artery. Increased vascularity on Colour doppler was noted. MR angiography showed splaying of left internal and external carotid artery at the bifurcation of common carotid artery. Excision of mass was done. The histopathological examination showed paraganglioma: carotid body tumor left side. We are presenting this case for its clinical, radiological and histopathological findings.

Keywords: Carotid artery, Paraganglioma, Head and Neck Mass.

INTRODUCTION

The carotid bodies are reddish brown, ellipsoid structures lying embedded in the adventitia of carotid artery bifurcation. ^[1] Their function is involved in the reflex control of heart rate, blood pressure and respiration via chemoreceptors. The tumors of this tissue are very rare constituting less than 0.5% of all body tumors. ^[2] Carotid body tumors can be a diagnostic challenge for the clinician.

CASE HISTORY

A 28 year old male patient presented with swelling in left mandibular region of 5 months duration. It was painless gradually increased without any compression effects. No other significant contributory history was present. Clinically swelling was

suggestive of? Cervical lymphadenopathy? Vascular tumor. Radiological evaluation using Computed tomography angiography revealed a hypervascular mass with enlarged feeding arteries. Plain MR angiography of neck revealed splaying of left internal and external artery at bifurcation of common carotid artery. The left subclavian, brachiocephalic trunk, right common carotid and bilateral vertebral arteries appeared normal. Chest X ray and USG of abdomen and pelvis were normal. Tumor was surgically excised and sent for histopathological examination.

Gross- A single globoid, well circumscribed, firm, grey brown mass (m) 2.2x2x2 cm.(Figure 1)) was received. Cut section revealed solid, smooth rubbery grey

brown mass with focal areas of hemorrhages (Figure 2).



Figure 1: Photograph of gross specimen showing globoid mass.



Figure 2: Cut section showing solid grey brown mass with focal areas of haemorrhage.

Microscopy- Multiple section studied showed tumor cells arranged in nests, clusters, trabecular and characteristic “Zellballen” growth pattern with intervening delicate fibrovascular tissue (Figure 3). Individual tumor cells were round with hyperchromatic nuclei, dispersed chromatin and abundant granular cytoplasm (Figure 4). Areas of focal hemorrhages were noted. Capsular or neuronal invasion was not seen. On the basis of microscopic features, histopathological diagnosis of carotid body paraganglioma of left side was made..

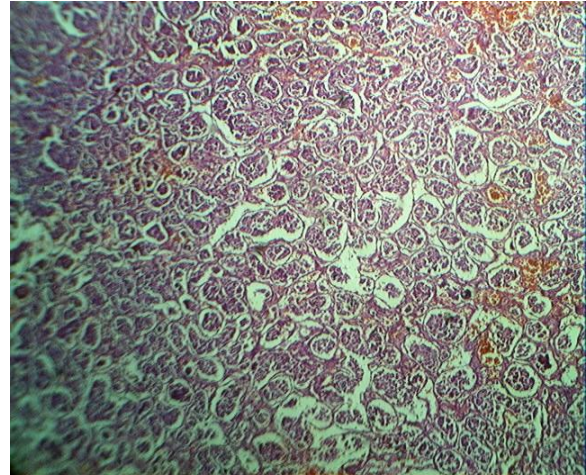


Figure 3: Photomicrograph showing tumour cells arranged in nested pattern separated by fibrous septa – Zellballen pattern. (H & E stain, 100x)

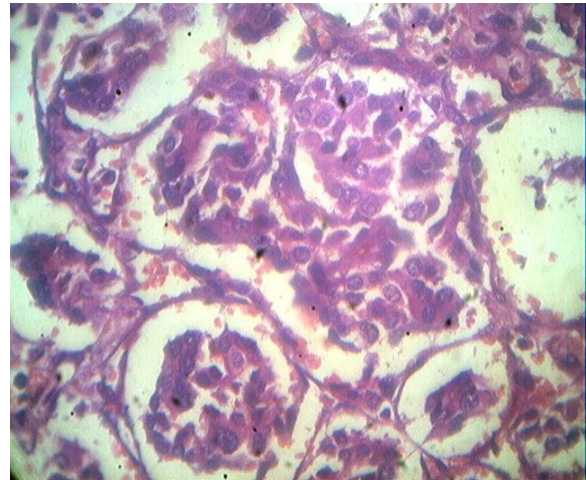


Figure 4: Photomicrograph showing high power view.(H & E stain, 400x)

DISCUSSION

Carotid body tumor was first reported by Marchand [3] in 1891. Tumors of carotid body were originally described as chemodectomas, now considered as a part of widely described group of tumors known as paragangliomas. [1] Paragangliomas are derived from neural crest cells and can be found in nerve perineurium or vascular adventitia from the skull bone to the pelvic floor. [4] The paragangliomas of head and neck region are rare and represent 0.6% of all head and neck tumors. [5] They are divided according to location-carotid body tumor at carotid bifurcation, the glomus

jugulare tumor at jugular foramen, vagal paraganglioma and glomus tympanicum tumor at the middle ear. These tumor may synthesize and secrete catecholamines, although less than adrenal pheochromocytoma.

Carotid paraganglioma occurs at any age but they are typically diagnosed between the third and sixth decade of life. [6] These tumors are sporadic in 90% of cases and familial in 10% of cases. In our case it was of sporadic type. It has been estimated in sporadic setting that 4-8% of patients with carotid body paragangliomas have bilateral tumor and occasional there may be multiple paragangliomas. [7] Carotid body is more freely movable horizontally than vertically (Fontaines sign). Pulsation over tumor can be felt. Symptoms related to compression of adjacent structures may be seen clinically. Differential diagnosis of cervical lymphadenopathy, carotid aneurysm, metastasis, tracheal cleft cyst should be considered.

Imaging procedures like USG, Color Doppler, CT, MRI, Angiography of neck are helpful tool for diagnosis and determination of surgical management. Preoperative embolisation can decrease blood loss.

Our case underwent surgical excision of lesion. Post-operative period was uneventful and patient on regular follow up is doing well.

Our histopathology showed two types of cell proliferation-

Type 1 (chief) cell with copious cytoplasm and large round/ oval nuclei with cytoplasm containing dense coarse granules.

Type 2 (sustentacular) cells - elongated cells closely resembling Schwann cells.

Multiple sections from tumor showed tumor cells arranged in clusters, nesting and trabecular pattern set in a fibrous stroma with prominent vasculature and characteristic "Zellballen" pattern was noted. In our case

there was no capsular or neuronal invasion. Abnormal mitotic activity, multinucleate giant cell were not seen.

Our case was benign carotid paraganglioma, occasional cases show neuromelanin pigmentation and amyloid deposition. 10% cases may show malignant changes.

On histopathology carotid body tumor differential diagnosis includes neuroendocrine tumor, medullary carcinoma of thyroid, hyalinizing trabecular adenomas of thyroid.

The treatment of choice for carotid body tumor is surgical resection with overall good prognosis. Regular follow up is necessary to look for recurrence or metastasis.

CONCLUSION

Carotid body paragangliomas are extremely rare tumors. Thorough clinical assessment of the patient of neck masses must therefore be performed pre-operatively to achieve better patient management.

REFERENCES

1. McPherson GAD, Halliday, Mansfield AO. Carotid body tumors and other cervical gliomas: Diagnosis and management in 25 patients. *Br J Surgery* 1989;76:33-6.
2. Cottrell ED, Smith LL. Management of uncommon lesion affecting the extracranial vessels. IN: Rutherford RB(ed). *Vascular Surgery*. Philadelphia: W.B. Saunder. 1995, PP.1623-27.
3. Marchand F- Contributions to knowledge of normal and pathological anatomy of the carotid and the glandula. *Int. BeiterWissn Med*1891;1: 535-81.
4. Mendenhall WM, Amdur RJ, Vaysberg M, Mendenhall CM, Werning JW: Head and neck paragangliomas. *Head Neck* 2011; 33:1530-1534.
5. Boedeker CC, Ridder GJ, Schipper J: Paragangliomas of the head and neck:

- diagnosis and treatment. *Fam Cancer* 2005; 4:55-59.
6. Myssiorek D. Head and neck paragangliomas: An overview. *Otolaryngol Clin North Am* 2001;34: 829-36.
 7. MR Naniwadekar, SV Jagtap, AY Kshirsagar et al: Fine needle aspiration diagnosis of carotid body tumor in a case of multiple paraganglioma presenting with facial palsy. *Actacytologica* 2010, 54(4),635-639.

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