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Case Report

A case report of a 33-year-old male patient having right-sided neck swelling suggesting paraganglioma

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Abstract

This case report describes a 33-year-old male patient who presented with right-sided neck swelling. The patient was diagnosed with paraganglioma, a rare neuroendocrine tumour, located in the right upper carotid space. Diagnostic methods included ultrasound (USG), magnetic resonance imaging (MRI), and fine needle aspiration cytology (FNAC). The MRI suggested a carotid body tumour, and the final diagnosis was confirmed by histopathology after surgical removal of the lesion. Surgical resection is the recommended treatment for this type of tumour.

Keywords: Paraganglioma, Susceptibility Genes, Shamblin Classification, Fisch Classification

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1. Introduction

Paragangliomas are neuroendocrine tumours arising from extra-adrenal autonomic ganglia within the paravertebral sympathetic and parasympathetic chains.[1-2] Carotid body tumours are a specific type of paraganglioma located at the carotid bifurcation and are rare chemical receptor tumours derived from embryonic neural crest cells that accounts for 0.6% of head and neck tumours. The pathogenesis of this disease remains unclear, but it is considered to be associated with chronic hypoxia of tissues and mutation of mitochondrial oxygen-sensitive genes.[2] The Incidence of the tumour is 1-2/100,000, with a slight female preponderance. The highest incidence is in the age group of 50-70 years.[3] Paragangliomas commonly present as a single, benign, unilateral tumour but 1% of sporadic and 20% to 80% of familial cases may have multiple tumours. Instances of malignancy and metastasis are rare. The difference of benign versus malignancy is based on histology. Attempts have been made to histologically distinguish benign tumors from the rare malignant paraganglial tumors, based on scoring systems that have been introduced for pheochromocytomas and paragangliomas of the retroperitoneum. However no scoring system has got majority. The most popular scoring system has been suggested by Thompson.[8] This system includes a variety of criteria, such as cellular and nuclear features, growth patterns, or occurrence of necrosis. Early identification and complete surgical resection is the key to cure and carries a favourable prognosis. The biological aggressiveness of paragangliomas can be judged by presence of distant metastases.[4] Symptoms depend on the specific locations. In contrast to paraganglial tumors of the adrenals, abdomen and thorax, head and neck paragangliomas seldom secretory relasing catecholamines and are hence rarely vasoactive. Petrous bone, jugular, and tympanic head and neck paragangliomas may cause hearing loss. The internationally accepted clinical classifications for carotid body tumours are based on the Shamblin Class I–III stages, which correspond to postoperative permanent side effects. For petrous-bone paragangliomas in the head and neck, classification proposed by Fisch is used. Head and neck paragangliomas have been associated with nine susceptibility genes: *NF1*, *RET*, *VHL*,*SDHA*, *SDHB*, *SDHC*, *SDHD*, *SDH*

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AF2 (*SDH5*), and *TMEM127*. Hereditary Head and neck Paragangliomas are mostly caused by mutations of the *SDHD* gene followed by *SDHB* and *SDHC* mutations. Multiple head and neck paragangliomas are common in patients with *SDHD* mutations and SDHB mutations are associated with malignancy.

2. Materials and Methods

A 33-year-old male patient came to the ENT department, with the chief complaint of right-sided neck swelling for two months which was insidious in onset and a gradual increase in size was noted. USG: Well-defined solid hypoechoic mass lesion seen in right upper anterior triangle neck which appears as a separate mass and superior from thyroid. MRI (neck): Well-defined enhancing lesion seen in right upper carotid space with extension up to submandibular gland with preserved fat planes, oropharynx and causing displacement of carotid and jugular vessels with maintained flow voids- the possibility of carotid body tumour (paraganglioma). FNAC: Smears show predominantly haemorrhagic material with scattered blood cells. The lesion was surgically removed and sent for HPE which suggests paraganglioma. **Figure 1**.

3. Discussion

Paragangliomas are solid, encapsulated benign tumours that are slow-growing with the potential for 10% malignant progression of patients. They are called as glomus tumours as well. Carotid body tumours and paragangliomas of the head and neck are painless, slow-growing tumours that often present as masses in the neck or lesions originating from the carotid space. The initial symptoms can also be a pulsating mass in the neck. Carotid body tumours can be moved horizontally rather than vertically, a finding known as a positive Fontaine's sign.[5] Large Carotid body tumours may affect vagal nerve and cranial nerves IX, XI, and XII sometimes. Eztremely large tumors may cause even Horner's syndrome or the facial nerve defecit.[5-6] Sometimes a carotid bruit or a pulsating mass can be detected. Differential diagnoses include aneurysm or pseudoaneurysm of the carotid artery, hematoma, glomus vagale tumour, vagal schwannoma and carotid body hyperplasia.[3] Excision biopsy is the treatment of choice for carotid body tumour paragangliomas. It was done according to SHAMBLIN' S classification[7] (based on tumour size) under GA from the right side neck swelling. According to this classifications tumour is devided into 3 groups. Group I includes small tumours that could be easily dissected away from the vessels. Group II consists paragangliomas of medium size that were intimately associated and compressed carotid vessels but could be separated with careful sub adventitial dissection. Group III consisted of tumours that were large and typically encased the carotid artery. This is tumors require partial or complete vessel resection and replacement. Radiotherapy can be tried as alternative treatment modality that can decrease the tumour size or stop its growth. It is recommended for patients who cannot undergo surgery on account of extensive involvement, multiple tumours, and high operative and anaesthetic risk.

4. Conclusion

The final diagnosis is made by MRI (Neck) suggestive of a well-defined enhancing lesion seen in the right upper carotid space with extension and relations with surrounding structures. Carotid body tumour (paraganglioma) can be benign or malignan. This can be detected histologically. Surgical resection is the gold standard treatment.

5. Source of Funding

None.

6. Conflict of Interest

None.

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