A Case of Secretory Glomus Jugulare

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Introduction

Glomus jugulare tumours are highly vascular tumours arising from the chief cells of the paraganglia in the adventitia of the dome of the jugular bulb. Estimated incidence is 1 per 1.3 million population, of which 1- 5% are malignant [1,2]. The benign tumours are locally aggressive. Growth rate is about 0.8mm per year [3]. Glomus secreting catecholamines occur symptomatically in about 1- 3% of cases [4]. One such case which was not diagnosed for six years is being reported.

Case Report

A 68 year old lady, reported with complaints of weakness right side of the face since 15 days. She gave history of recurrent pain, progressive deafness and a buzzing sound right ear for the last six years. There was no history of otorrhoea or vertigo. On the basis of a computerised tomography (CT) scan report suggesting cholesteatoma, she was advised a mastoid exploration. Past history revealed hypertension and atrial fibrillation of five years duration.

On examination, pulse was irregularly irregular and blood pressure 150/70 mm of Hg. Carotid pulsation on the right was very prominent and a bruit was present. There were no other findings on systemic examination. Otoscopy revealed a red smooth, faintly pulsatile bulge in the region of the eardrum (Fig. 1). There was no discharge or clinical evidence of cholesteatoma. Hearing was nil and facial nerve palsy on right was of lower motor neuron type. Blood sugar was marginally raised. Electrocardiogram showed atrial fibrillation. T_3 , T_4 and thyroid stimulating hormone (TSH) levels were normal. Radiography of chest showed cardiomegaly. 2D echocardiography gave ejection fraction values of 71%. Ultrasonography of the abdomen was normal. Contrast enhanced computed tomography (CECT) head and neck showed an enhancing mass lesion in the right jugular fossa extending up to the right carotid canal (Fig. 2). The lesion showed erosion and destruction of the petrous part of temporal bone, middle ear region and aberrant hypervascularity of internal and external carotid artery. These

features suggested a Glomus Jugulare. Colour doppler neck reported fistulous communication between the internal jugular vein (IJV) and branches of the internal carotid artery (ICA) in the intracranial portion. Magnetic resonance angiography (MRA) and magnetic resonance venography (MRV) showed evidence of expansion of the right jugular fossa with an ill defined lobulated 2.5 x 2.1 cm speckled mass lesion with mixed signal intensity below the middle ear cavity (Fig. 3). Vanillylmandelic acid (VMA) in 24 hour urine by high performance liquid chromatography (HPLC) was 8.73 mg/g creatinine (normal range 1.60 – 4.20). On this basis a 'Secretory Glomus Jugulare' was diagnosed. The cardiac status of the patient was managed with antiarrhythmic drugs, anticoagulant and diuretic. As per classification devised by Oldring and Fisch, the tumour was Type D with intracranial extension and would require a posterolateral approach if surgery was contemplated. Anaesthetic management could be challenging and control of hypertension requires alpha and beta blockers initially. After embolization and surgical resection, loss of vasoconstrictor tone could result in circulatory collapse, requiring massive intravenous infusions and the use of antigravity suits to the lower body to increase venous return. Modalities of treatment were explained to the patient and surgery was not considered in the setting of an uncontrolled cardiac status and associated co-morbidities at her age. Radiotherapy was offered but she declined and the patient preferred not to go for any further treatment.

Discussion

The commonest mode of presentation of a glomus jugulare is a pulsatile tinnitus with a conductive deafness in the initial stages and later a pulsatile red mass behind the drum (the rising sun sign) [5]. Alfred and Guilford in 1962 found that the average delay in diagnosis was six years. Among the various modalities of management 'no active therapy' is considered one. Radiotherapy causes perivascular fibrosis and this slows the progress of the tumour [5]. However it could not have benefited our patient since catecholamine secretion is not affected

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Fig. 1 : Glomus as seen on otoscopy

by the application of radiation [6]. Complete surgical resection is ideal but not always possible due to proximity of important neurovascular bundle and difficult approach. Several studies using gamma knife surgery show that there has been no change in tumour size or volume [7,8]. A variety of functional imaging studies for neurosecretory tumours are also available [9]. Radioactive labelled meta-iodobenzyl guanidine (MIBG) is taken up by the neurosecretory cells because of structural similarity to noradrenaline. Therefore ^{123}I -MIBG scan can help in detecting secretory paragangliomas. The sites picked up by the scan can be targeted with ^{131}I -MIBG loaded with substantial radioactivity in malignant cases. CECT and MR studies help in the diagnosis, while MRA, MRV and colour doppler helps in planning preoperative embolization. Direct biopsy is required occasionally to exclude a squamous cell carcinoma.

This case highlights the relevance of 'no active therapy' in select elderly patients with glomus jugulare. Surgery in such a patient under the mistaken diagnosis of cholesteatoma could have been disastrous.

Conflicts of Interest

None identified

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Fig. 2 : Mass lesion (arrow) expanding and eroding the right jugular fossa and extending into the petrous temporal bone

Fig. 3 : MRA shows enhancing mass lesion in right jugular fossa with aberrant vascularity

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