Basaloid Squamous Cell Carcinoma Clinically and Radiologically Masquerading as a Head & Neck Paraganglioma – A Case Report

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1. Abstract
1.1. Background: This paper reports the first case of basaloid squamous carcinoma clinically and radiologically masquerading as a Head & Neck Paraganglioma (HNPGL).
1.2. Case Presentation: A 66-year-old male presenting with unilateral hearing impairment, 7th – 12th (excluding 11th) cranial nerve palsies was diagnosed radiologically as a HNPGL by Magnetic Resonance Imaging of the brain featuring a hypointense and hyperintense punctate mass centred at the jugular fossa with intracranial extension. The ascending pharyngeal artery recognized as the major feeder was embolized by percutaneous embolization following digital subtraction angiography. Gross total resection of the tumour was followed by an uneventful postoperative recovery. Combined immunohistochemistry and histopathological morphology revealed a basaloid squamous cell carcinoma following which patient completed radiotherapy and is at 3-month follow-up currently.
1.3. Conclusion: The case report discusses the diagnostic pitfalls and management challenges of this rare entity based on prior evidence, as well as the literature review and clinical and surgical analysis of them.

2. Introduction
Basaloid Squamous Cell Carcinoma (BSCC) is a rare malignant uncommon variant of squamous cell carcinomas (SCCs) that arises usually in the neck [1]. We report a case of a patient who was diagnosed radiologically to have a Head & Neck Paraganglioma (HNPGL)(new nomenclature of Glomus Jugulare Tumor/GJT). However, subsequent histopathological and immunohistochemistry revealed a BSCC. This case highlights the importance of histological confirmation for accurate diagnosis and management.

3. Case Report
A 66-year-old previously healthy male with no history of tobacco/alcohol use/unprotected sexual activity, presented with right sided hearing impairment associated with vertigo for 6 months. No complaint of pulsatile tinnitus, headache, dysphagia, dysarthria, visual disturbances, seizures, or difficulty in walking. He had a deviation of the mouth towards the left side, with incomplete closure of the right eye (House-Brackmann Grade 2). On examination, right sided 7th, 8th, 9th, 10th, and 12th cranial nerve palsies were observed. However, the cough and gag reflex were unimpaired and taste sensation was intact. No masses or lymphadenopathy were seen.

Contrast enhanced Computerized Tomography (CECT) of brain and petrous temporal bone revealed right middle ear and mastoid air cells filled with soft tissue material and widening, erosion, and filling of the right jugular foramen with multiple vascular channels alongside erosion of the right caraticojugular spine. The non-contrast and contrast enhanced Magnetic Resonance Imaging (MRI) brain that followed revealed a lobulated rather well demarcated isointense to hyperintense mass centred in the right jugular fossa.
with intracranial extension infiltrating into the right temporal lobe; extending to the level of the odontoid and mastoid; inferiorly and laterally respectively. The mass displayed hyperintense and hypointense punctuate signals and enhanced with contrast suggestive of a HNPGL. The mass appeared to have infiltrated the sigmoid sinus while partially encasing the narrowed petrous portion of the internal carotid artery. Head, neck, chest, and abdominal evaluation suggested no evidence of distant metastasis nor possible primaries.

Digital Subtraction Angiography (DSA) conducted on the day of the surgery revealed a dominant abnormal blush in the right ascending pharyngeal artery. Selective catheterization and embolization with 300 – 500 μm Poly Vinyl Alcohol (PVA) particles resulting in achievement of complete occlusion of the vessel. Surgery to excise the right glomus tumour was conducted on the same day of the preoperative embolization. A lateral presigmoid retrosigmoid extracranial skull base approach was employed where a right sided “?” mark incision was made extending from the anterior margin of the sternoleidomastoid muscle (SCM), behind the ear extending to the temporal region. SCM was dissected from the anterior border to find the Internal Jugular Vein (IJV) and carotid artery. The muscle was dissected from the mastoid tip followed by a mastoidectomy. The external ear was closed, and malleus, incus and the ear drum were removed. The pre- and retro-sigmoid bone was drilled exposing the sigmoid sinus. The tumour was found to be extending along the IJV and a gross total excision of the tumour was done with achievement of hemostasis and preservation of the facial nerve. Tumour sections reveal infiltrative tumour composed of neoplastic cells in solid, trabecular, and cribriform growth patterns. Adenoid cystic like pattern and rosetting like acellular central canals, small broad perivascular papillary pattern and squamous metaplasia was observed. Perineural, bony and soft tissue infiltration was seen. On immunohistochemistry, positive cytoplasmic and membrane staining with Ck 5/6 and strong nuclear staining with P 63 was noted suggestive of a carcinoma of squamous, basaloïd, and adenoid cystic morphology. P16 staining was not done. Thence, a combined morphological and immunohistochemistry diagnosis of BSCC was made [2].

Post-surgical radical radiotherapy (adjuvant radiotherapy) was planned at a dose of 65 Gy to be delivered in 30 fractions pertaining to the type/location of the tumor and the post-surgical residual tumor volume. Currently, the patient has completed all 30 sessions and is pending to undergo a 3-month post-radiotherapy MRI scan with compliance and satisfaction. The patient currently suffers from ipsilateral facial nerve palsy and a hearing deficit equivalent with compliance and satisfaction. The patient currently suffers and is pending to undergo a 3-month post-radiotherapy MRI scan tochemistry diagnosis of BSCC was made [2].

4. Discussion
Jugular foramen lesions commonly include paragangliomas followed by certain other types such as meningiomas, schwannomas or even chondrosarcomas. These head-and-neck paragangliomas (HNPGLs) are rare tumours with an estimated clinical incidence of 1/100 000 patients per year. The classical main types of HNPGLs are carotid body tumours, jugular (considered GTs in the current context) and tympanic paranganglial tumours collectively considered together as JTPGLs and vagal paranganglial tumors. [3] On the other hand, metastatic localization within the region of the jugular foramen is rare accounting for around 3.5 – 36% of all secondary malignant tumours involving the temporal bone [4]. GTIs are the most common form of paragangliomas and hence the most common of all jugular foramen tumours. They are highly vascularized histologically benign lesions known to arise from the temporal bone paranganglial system. However, they are known to be locally invasive by invading adjacent bone, blood vessels and the central nervous system [5]. In the contrary, BSCC are more aggressive rare form of SCC (accounting for 5% of all SCCs) with higher potential for distant metastasis and poor prognosis [6].

When considering the demographical variations, HNPGLs are commonly seen in females in their late 40s while BSCCs are usually common in males in their 60s with history of tobacco/alcohol use [7,8]. A retrospective analysis of 20 patients diagnosed with temporal bone metastases revealed a median age of detection at 60 years with a majority being males (13 out of 20). This points out the importance of considering metastatic lesions in cases of male patients of advanced ages despite presenting with classical features of GJ tumours [9]. The clinical presentations of JTPGLs usually involve pulsatile tinnitus, ear mass, hearing loss, pain and vertigo attributed to the mass effects dominated by involvement of cranial nerves. Hence, a preoperative cranial nerve defect is frequently observed, especially in these cases of JTPGLs over other HNPGLs [8]. On the other hand, temporal bone metastases usually associate with otological symptoms of hearing loss in addition to other neurological manifestations like facial palsy, while the initial presentation of BSCCs usually include neck lymph node metastases [1,9]. This is relatable to other known cases of temporal bone metastases presenting as glomus jugulare tumours as well [10-13]. However, it is important to note that lower cranial nerve palsies involving the IXth, Xth, XIth (and XIIth) cranial nerves (CNs) which are seen in HNPGLs because of mass effect, are also considered as common features of metastatic lesions describes in literature as JF syndrome of Vernet (involving IXth, Xth, XIth and XIIth CNs), posterior laterocondylar syndrome (Vernet’s plus XIIth CN) and posterior retropharyngeal syndrome (Vernet’s plus XIth and Horner’s) [12]. Furthermore, a 1976 review of temporal bone metastases also describes a triad of facial paralysis, otalgia and periauricular swelling to alert a clinician of metastatic temporal bone disease [14]. While classical features of JTPGLs, such as hearing
loss, tinnitus and vertigo were seen in this patient; the presence of the combination of VIIth and lower cranial nerve palsy'ns highlight the importance of cranial nerve palsy's dominating over otological symptoms in exclusion of metastatic lesions and that the mere absence of the classical triad nor the various JF syndromes are not reliable indicators of benign lesions.

Diagnostic tests both biochemical and radiological play a major role in overcoming dilemmas associated with complex presentations. This is especially relevant in cases of JTPGLs, since diagnostic biopsies are discouraged or even contraindicated due to risk of precipitating hypertensive crisis, hemorrhage or tumour seeding [3]. Radiological investigations include CTs and MRIs as first line; where CT scans though less sensitive accurately define possible bone invasion and MRIs displaying characteristic ‘salt-and-pepper’ appearance [3]. An analysis of 236 patients with benign paragangliomas identified that 26 off the 27 patients with GJ tumours who underwent CT scans revealed to be true positives while all 24 patients with GJ tumours who underwent MRIs were found to be true positives, illustrating the paramount importance of imaging in patients with clinical suspicion of HNPGs [8]. On the other hand, the biochemical studies, especially plasma or 24- hour urinary metanephrine or catecholamine concentrations can incidentally be effectively used in certain instances since these tumours can be rarely secretory. This is evident by higher sensitivities of 24-hour urine studies observed in the same analysis, which revealed a combined sensitivity of 89.9% [8]. CTs and MRIs were both used in this case to arrive at the diagnosis, where the hyperintense and hypointense punctate signals in preoperative MRI corresponding to the classical appearance proved to be wrong. Endocrine studies were not conducted due to pragmatic difficulties. Hence it is important to identify that while classical appearances are highly useful in the diagnosis, metastatic lesions should nevertheless always be considered and scans involving the neck, chest and abdomen ought to be carried out too to exclude potential primaries that might have led up to a masquerading metastasis.

Angiography (digital-subtraction angiography (DSA) or MRI angiography) is also an important modality for diagnosis and definition of the vascular anatomy preoperatively but also for embolization of the major feeders where it is recommended for the management of resectable. However, the role of angiography and embolization serves in reducing intraoperative complications and facilitation of surgical radicality over curing or reducing clinical sequelae [15]. A review of 38 PGL patients in The Mayo Clinic associates pre-surgical radicality over curing or reducing clinical sequelae [15].  In ruling out metastatic lesions despite the presence of classical audiological and radiological features suggestive of JTPGLs. Furthermore, post-operative radiotherapy should only be started once a combined morphological and immunohistochemistry diagnosis is made to ensure effective treatment. We recommend further analysis to evaluate the validity of application of these hypotheses in wider patient populations.

5. Conclusion

This first case of BSCC masquerading as a HNPGL highlights that malignant tumors can create diagnostic dilemmas and hence require careful evaluation and exclusion. Furthermore, surgical experience and careful planning is essential in ensuring favorable outcomes in these patients.

6. Patient Perspective

The patient has successfully completed the follow-up radiotherapy with good compliance and is satisfied with the level of care received and outcome. However, his concern lies with the persistence of the deficits preoperatively. Patient counselling has been
able to alleviate this by explaining the current status.

7. Declarations

7.1. Funding
None

7.2. Conflicts of Interest
None

7.3. Availability of Data and Material
All patient records, operation notes and radiographic information are available in the form of hardcopies. Scanned documents can be provided on the request from the journal.

7.4. Code Availability
Not Applicable

7.5. Authors’ Contributions
Mr. Pumudu Weerasekara was the lead author of the paper and Dr. Sunil Perera was the chief supervisor of the project while being the surgeon in charge of the patient. Dr. Wasantha Rathnayake, Dr. Nadeeka Chandrarathne and Dr. Geethika Jayaweera contributed equally to the content of the paper from their respective fields.

7.6. Ethics Approval
Not Applicable

7.7. Consent to Participate
Written informed consent was obtained from the patient and can a scanned copy of the consent form can be provided upon request from the journal.

7.8. Consent for Publication
All authors unanimously agreed to provide consent to publish this paper in the journal.

References


