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Case Report

Primary Secretory Paraganglioma of Facial Nerve Canal: A Case Report

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Paragangliomas (PGLs) of Head and Neck region account for 0.6% of Head and Neck Tumours. These may originate in paraganglionic tissues in the area of carotid bifurcation, vagus nerve, tympanic plexus and very rarely along vertical Facial nerve canal (FNC). We intend to describe a rare case of primary paraganglioma of FNC associated with hypoxia of submarine environment, its characterization and multidisciplinary approach towards its management.

KEYWORDS: Facial nerve canal, paraganglioma, secretory

INTRODUCTION

Head and neck PGLs embryologically originates from extra-adrenal paraganglias of neural crest. These account for 0.03% of human tumours and 0.6% of head and neck tumours.[1-3] Jugulotympanic paragangliomas are known to involve the vertical segment of the FNC by erosion of the surrounding bone, however, primary PGL arising from the vertical segment of the FNC is an extremely rare entity.[4-7] Of all head and neck PGLs, 2-3 % are secretory.[8] The association of hypoxia related to high altitude in development of PGLs is well known.[9] To the best of our knowledge, associated hypoxia in a submarine environment have not been described in context to paragangliomas. Here, we describe characterization and management of a case of primary secretory paraganglioma of facial nerve canal in a submarine sailor.

CASE PRESENTATION

A 43 years old male patient, a submarine sailor by profession was referred to our centre from a tertiary care hospital for further management, as a case of paraganglioma of left ear canal. The patient had history of mass in his left ear canal for last 4 years. Patient was a known case of Anxiety Disorder with history of episodic sweating, nervousness, flushing and palpitations for the last 5 years and on medication for the same.

At the tertiary care hospital he underwent partial excision of the mass. As per the available documents, the mass was intraoperatively found to be highly vascular and only the External Auditory Canal (EAC) part of mass was removed. Histopathological report was suggestive of paraganglioma and immunohistochemistry (IHC) report was positive for vimentin, synaptophysin and chromogranin. Patient was then referred to us for stereotactic radiotherapy of residual lesion.

Evaluation at our centre revealed a well healed post aural scar and normal facial nerve function. Hearing threshold was within normal limits on audiometry. Previous High resolution computed tomography of Temporal bone (HRCT-TB) revealed a well- defined 7mm x 11 mm soft tissue mass in the left EAC eroding the posterior canal wall and extending about 11 mm x 13 mm x 18 mm posteriorly into the mastoid cavity (Figure 1). It was a discrete mass along and lateral to vertical segment of FNC. HRCT-TB (Figure 2) and 3 Tesla

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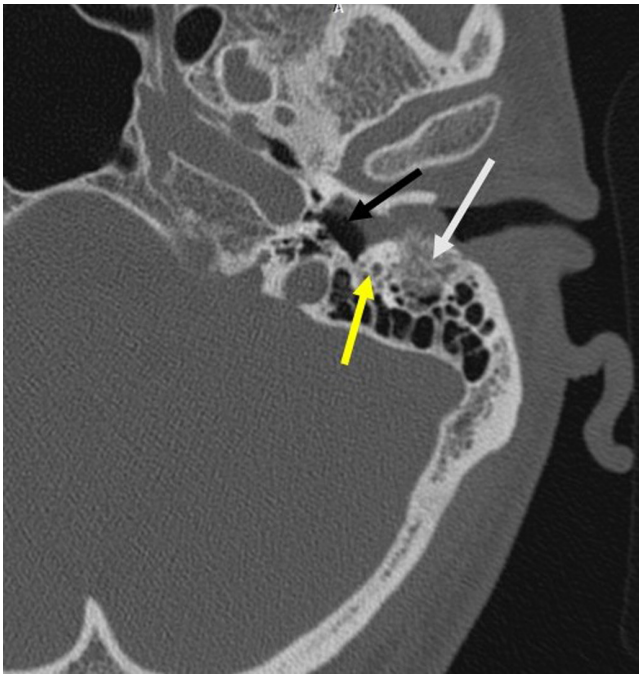

Figure 1. HRCT Temporal bone of left side showing soft tissue mass in left EAC, eroding posterior wall and continuing into left mastoid cavity (white arrow). Intact Facial nerve (yellow arrow) and middle ear (black arrow) are also depicted.

Contrast enhanced MRI (CE-MRI) of Brain and neck was repeated at our centre. They showed a 9mmx 12mm enhancing mass posterior to left EAC in the mastoid lateral to vertical segment of FNC (Figure 3). Gallium 68 Dotatate scan showed somatostatin receptor expression in left temporal bone in the region of diagnosed mass. Serum catecholamines and 24 hours urinary Vanillyl Mandelic Acid (VMA) values were raised. His investigations revealed raised hemoglobin level (19 gm %) and raised hematocrit. Further evaluation by clinical hematologist detected raised Erythropoetin (EPO) levels and was diagnosed as secondary polycythemia due to hypoxia (associated with submarine environment). However, the patient did not have any symptom associated with polycythemia.

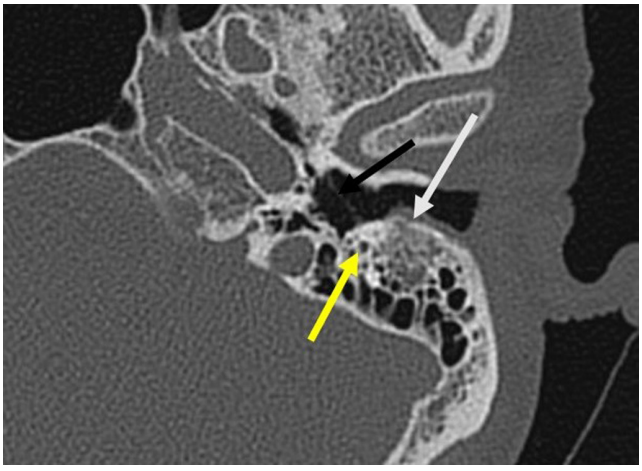

Figure 2. HRCT Temporal bone of left side showing soft tissue mass in left mastoid adjacent to facial nerve. Note: the EAC part of tumour is not there except for minimal soft tissue. Intact Facial nerve (yellow arrow) and middle ear (black arrow) are also depicted.

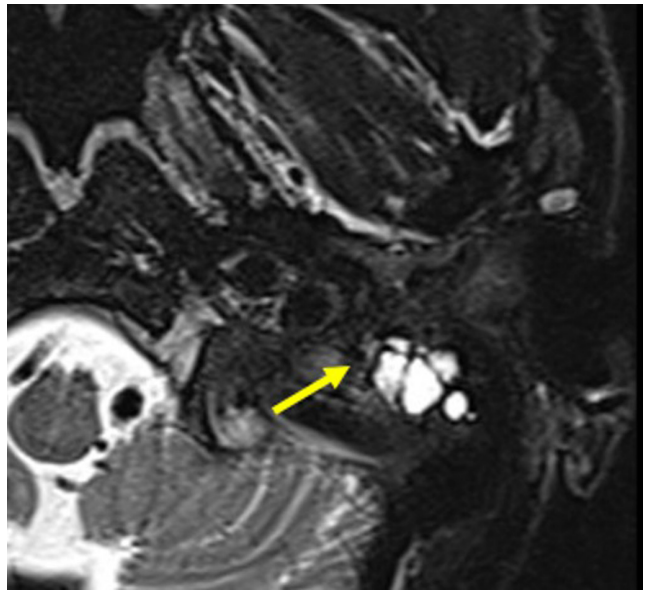

Figure 3. 3 Tesla Contrast enhanced MRI of Brain and neck showing a 9mm x 12mm enhancing mass posterior to left EAC in the mastoid adjacent to vertical segment of Facial nerve (arrow).

Based on clinical history, radiological findings, lab investigations, previous histopathological and IHC reports, a diagnosis of Secretory PGL of left FNC was made.

Since the paraganglioma was secretory in nature, the patient was started on alpha blocker followed by combination of alpha and beta blockers by endocrinologist for adequate optimization of blood pressure prior to surgery.

The patient underwent transtemporal resection of tumour under general anaesthesia. Intraoperatively, the tumour was highly vascular infiltrating the mastoid air cells lying lateral to vertical segment of FNC from second genu to stylomastoid foramen. Gross total tumour removal was achieved by a combination of drilling, curettage and bipolar coagulation. The facial nerve was delineated and was found to be intact. Final Histopathology and IHC report was suggestive of paraganglioma (Figure 4) and was positive for vimentin, synaptophysin and chromogranin (Figure 5).

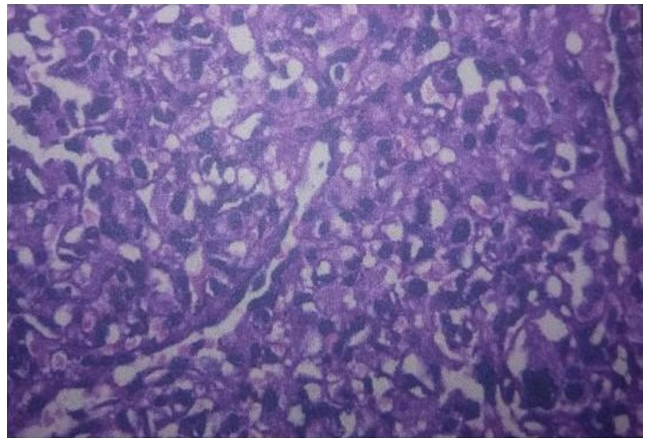

Figure 4. H&E staining showing features consistent with Paraganglioma.

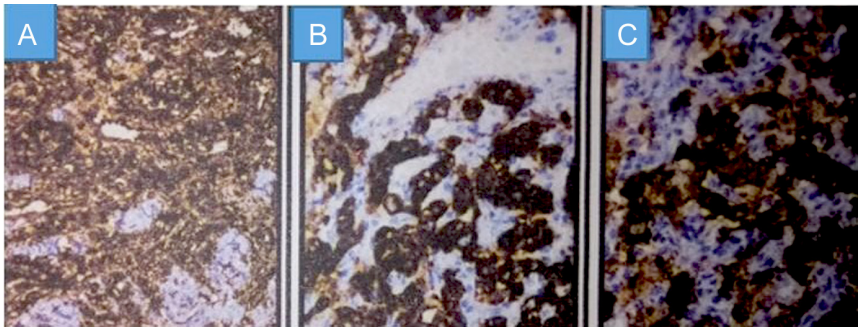

Figure 5. Tumour cells were positive for Vimentin (A), synaptophysin (B) and chromogranin (C).

The secretory symptoms disappeared within one week of surgery. The mastoid cavity is well epithelialized and he is both clinically and radiologically disease free at 18 months post-surgery. Patient has been advised for yearly clinical and radiological follow-up.

DISCUSSION

Paraganglias follow path of adventitia of Jugular bulb, carotid body, Jacobson's nerve and very rarely along Arnold's branch of Vagus nerve. Guild based on his study of human temporal bone sections in 1953 published a paper wherin he reported presence of paraganglionic tissue along course of Arnold's nerve.[10] It was also reported that, Arnold's nerve has a variable course and it may enter the facial canal either via a separate mastoid canaliculus or traverse externally and enter stylomastoid foramen to course along mastoid segment of facial nerve canal. He also stated that glomus formation in the FNC are without doubt associated with Arnold's nerve.[10,11] Based on anatomical findings of Guild, published operative and imaging findings; Frisch CD et al suggested that these tumours arise from paraganglioma surrounding this branch of Arnold's nerve in mastoid segment of FNC. It was also proposed to use the term 'Paraganglioma of Facial Nerve Canal' as it would provide a consensus in reporting.[12] We agree with the authors' suggestion and have used this term in the present case report.

Primary PGL of FNC is very rare and only 15 cases have been reported so far in the literature. Presentation may be with aural fullness, mass in ear canal, conductive hearing loss or facial tics or paresis/palsy of facial nerve, mass behind intact tympanic membrane or dysguesia.[7] In our case, presentation was as left ear canal mass without compromised facial nerve function.

The differential diagnosis of paraganglioma of FNC includes facial nerve neurinoma and hemangioma. CT reveals moth–eaten appearance due to permeative osteolysis in paraganglioma which is absent in neurinoma. Hemangioma is similar to PGL on CT occasionally shows calcification. Hemangiomas and neurinomas usually arise from geniculate ganglion while PGL of FNC arise from its vertical portion.[13] Gallium 68 Dotatate scan has been considered reliable imaging modality in view of high specificity in localizing the disease in cases of paragangliomas, especially extra adrenal.[8,14] In our case, the radiological findings were consistent with PGL of vertical part of FNC.

Secretory PGL produce symptoms like headache, palpitation, malar flushing and sweating.[8] Several cases have been reported of PGL developing in patients with chronic hypoxia associated with high altitude.[9] Primary and secondary polycythemia are differentiated on basis of raised hematocrit with low EPO in the former and raised EPO in the later.[15] In a submarine, which is a closed unit, when the carbon dioxide level goes up from normal 0.2 % to 1.0-1.5%; the respiratory quotient gets disturbed and hypoxia occurs in crew members.[16] Our patient was a submarine sailor, had secretory symptoms prior to the diagnosis of paraganglioma of FNC and was erroneously managed as anxiety disorder. Our patient's blood parameters were consistent with secondary polycythemia due to hypoxia.

Hence, in this patient consistent and recurrent exposure to hypoxic environment of a submarine may have induced symptoms of episodic flushing, palpitations, sweating due to secretory nature of already existing slow growing PGL of facial nerve canal was wrongly diagnosed as anxiety disorder.

Once facial nerve function has been compromised, surgical resection of Facial nerve canal PGL followed by nerve grafting provides freedom from disease and clinical symptoms with shorter follow-up. However, a 'wait and scan' strategy may be justifiable in patients with no evidence of facial nerve paralysis or who are unwilling for or unfit to undergo surgery (due to advanced age or comorbidities). With a normal facial nerve function, a tumor biopsy with transmastoid facial nerve decompression is acceptable prior to which Digital subtraction angiography must be advocated to evaluate tumor vascularity.[7,8] In this case, although the patient had normal facial nerve function but in view of secretory nature of the tumour with troublesome symptoms and constant exposure to hypoxic environment due to his professional need; we recommended surgical excision even if it had resulted in some form of facial nerve paresis. Biopsy prior to definitive surgery is not advisable in an already radiologically and clinically proven case of PGL as it exposes patient to undue risk of bleeding without any beneficial clinical outcome. The patient was young adult with good resectability access with no adjacent vital structures at-risk hence favourable for complete, surgical excision of tumour. This gave our patient, a tumour-free and secretory-symptom free life with decreased time-period under surveillance.

CONCLUSION

To the best of our knowledge, this is the first reported case of secretory Paraganglioma of FNC which may be associated with hypoxia of submarine environment. Awareness about the rare location of the pathology along with good radiological investigation,

histopathological examination with immunohistochemistry will help in reaching the diagnosis. Good surgical planning will help in gross total resection of tumour with preservation of facial nerve function.

Informed Consent: Written informed consent was obtained from the patients who participated in this study.

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Conflict of Interest: The authors have no conflict of interest to declare.

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