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Case Report

Paraganglioma of the Cerebellopontine Angle Masquerading as Vestibular Schwannoma: A Rare Diagnostic Pitfall

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ABSTRACT

Paragangliomas are rare neuroendocrine tumors arising from sympathetic or parasympathetic paraganglia. Cerebellopontine angle (CPA) paragangliomas are exceptionally uncommon and may mimic more frequent CPA lesions such as vestibular schwannoma or meningioma on imaging. We report a 27-year-old male presenting with progressive right-sided hearing loss and ipsilateral facial weakness. Initial radiological evaluation suggested vestibular schwannoma. The patient underwent right retrosigmoid craniotomy and tumor excision. Histopathology and immunohistochemistry confirmed a neuroendocrine tumor consistent with paraganglioma (chromogranin and synaptophysin positive) with a Ki-67 labeling index of 10%. The patient subsequently received adjuvant radiotherapy and remains neurologically stable at follow-up. This case highlights the diagnostic difficulty of CPA paragangliomas and emphasizes histopathological confirmation and appropriate management.

Keywords*: Paraganglioma, Cerebellopontine angle, Vestibular schwannoma, Neuroendocrine tumor, Retrosigmoid craniotomy, Case report.*

INTRODUCTION

Paragangliomas are rare neuroendocrine tumors arising from sympathetic or parasympathetic ganglia and are known to occur in diverse sites such as the abdomen, pelvis, carotid body, and head and neck region.[1] These tumors account for a small proportion of head and neck neoplasms, estimated at approximately 0.6%.[2] While head and neck paragangliomas commonly arise from the carotid body, jugular bulb, or tympanic plexus, paragangliomas arising in the cerebellopontine angle (CPA) are exceedingly rare.[3]

The clinical presentation of paragangliomas is variable and may reflect either mass effect or catecholamine secretion. Although many head and neck paragangliomas are typically non-secretory, catecholamine-producing tumors may present with hypertension, tachycardia, and sweating.[6,7] A significant diagnostic challenge arises due to overlap of radiological features with more common CPA pathologies such as vestibular schwannoma and meningioma.[3] The rarity of this lesion and its potential for significant intraoperative vascularity make accurate diagnosis and management crucial.[3,10]

We report a rare case of CPA paraganglioma in a young male patient, initially suspected to be vestibular schwannoma on radiology, and confirmed as paraganglioma on histopathology and immunohistochemistry.

CASE PRESENTATION

A 27-year-old male from Lakhimpur, Goalpara, Assam presented with complaints of decreased hearing in the right ear for one year and ipsilateral facial weakness for nine months. The hearing loss was insidious in onset with no aggravating or relieving factors. The facial weakness progressed gradually. There was no history of dysphagia, breathing difficulty,

fever, or vomiting. There was no known history of hypertension, diabetes mellitus, bronchial asthma, tuberculosis, or prior surgery.

General and systemic examinations were within normal limits. Neurological examination revealed right-sided facial weakness with Glassgow coma scale was 15.

Routine laboratory investigations including hemoglobin, total leukocyte count, renal function tests, and urine analysis were within normal limits.He underwent NCCT BRAIN which showed

Right CPA mass lesion (~5.0 × 4.1 cm) with surrounding vasogenic edema and mass effect causing compression of the fourth ventricle leading to supratentorial ventriculomegaly and nearby brainstem structures.He underwent emergency left MPVP shunt placement.

Later on MRI BRAIN done which shows Ill-defined hetero-intense lesion at the right CPA junction, with blooming foci on GRE suggesting hemorrhagic elements. Widening of the right internal auditory canal was noted with mass effect on cerebellar hemisphere and effacement of fourth ventricle. Mild enhancement seen post contrast most likely s/o right vestibular schwannoma.

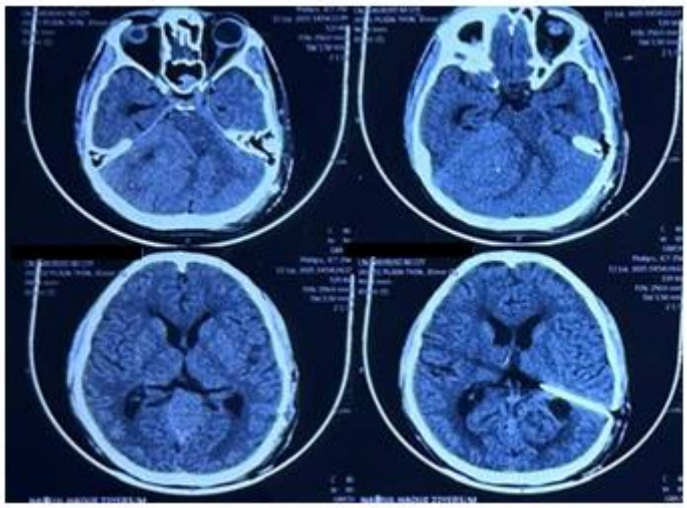

Fig 1: CT scan sections showing hypointense lesion in the right cerebello-pontine region with left medium pressure ventriculo-peritoneal shunt in situ.

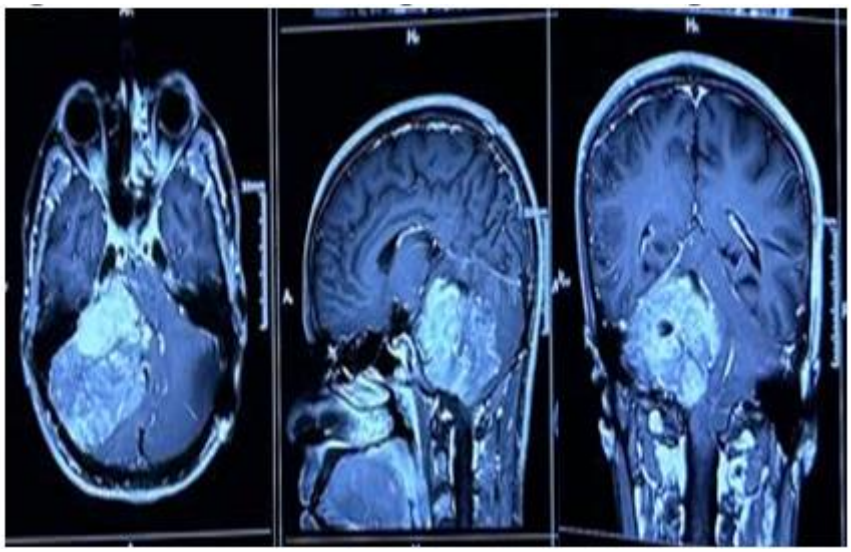

Fig 2: T1-contrast MRI showing a contrast enhancing lesion in right cerebello-pontine region with widening of right Internal Acoustic Canal showing mass effect over the right cerebellar hemisphere with effacement of fourth ventricle.

The patient underwent definite surgery for the tumor-**right retrosigmoid (RMSO) craniectomy with tumor excision** under general anesthesia. Intraoperatively, a solid-cystic mass (~4 cm) was identified in the right CPA, extending and displacing the brainstem. Excised tumor specimen was sent for histopathological examination.

Histopathology was suggestive of a **neuroendocrine tumor Grade 2** with **aberrant desmin expression**. Immunohistochemistry shows

- **Chromogranin: Positive**
- **Synaptophysin: Positive**
- **Desmin: Positive**
- **Ki-67 index: ~10**

The patient was then discharged on postoperative day 5. Postoperatively, he received **adjuvant radiotherapy**. The patient is doing well at 2 months follow-up with no additional neurological deficits.

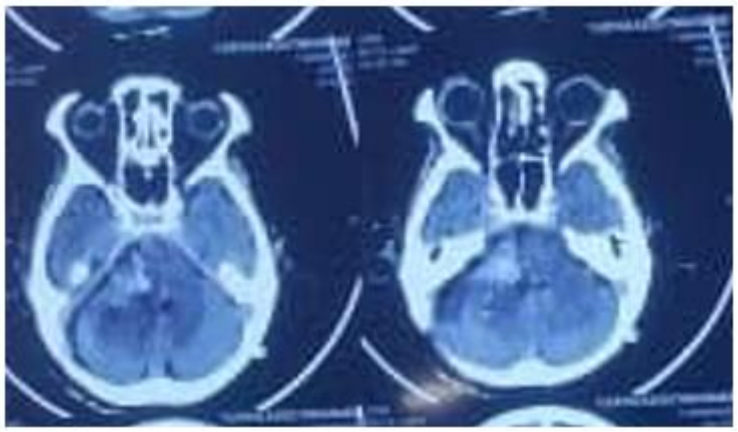

Fig 3: Postoperative non-contrast CT scan of the brain demonstrating postsurgical changes following excision of the tumor, with evidence of resection cavity in the CP angle region and no residual mass effect or acute hemorrhage.

DISCUSSION

Paragangliomas are rare neuroendocrine tumors with high heritability, and several cases are associated with genetic mutations involving succinate dehydrogenase (SDH) complex genes.[5,6] These tumors may arise sporadically or as part of inherited syndromes.[6] Their clinical behavior is unpredictable: although many are benign, some demonstrate aggressive local invasion or metastasis.[1,3]

CPA paragangliomas are extremely uncommon and can masquerade as vestibular schwannoma due to overlapping radiological characteristics.[3] In our case, the preoperative radiological diagnosis was vestibular schwannoma, largely due to CPA location and widening of internal auditory canal. However, the presence of GRE blooming foci (suggestive of hemorrhage) and heterogeneous enhancement could represent clues toward a vascular tumor rather than typical schwannoma.

One of the classic MRI features of paragangliomas is the "salt-and-pepper" appearance caused by flow voids from hypervascularity and hemorrhagic foci.[3] This feature was well demonstrated in previously published CPA paraganglioma case reports, where lesions show intense enhancement and marked vascularity on imaging.[3]

Biochemical testing, including plasma free metanephrines or 24-hour urinary catecholamines/metanephrines, is recommended when a paraganglioma is suspected, especially in patients with unexplained hypertension or adrenergic symptoms.[6,7,8] In our patient, no catecholamine-related symptoms were present, and therefore biochemical evaluation was not performed.

Surgery remains the mainstay of treatment for accessible head and neck paragangliomas.[4] In CPA lesions, the retrosigmoid approach provides excellent exposure of the CPA with adequate tumor resection potential and acceptable morbidity.[3] However, intraoperative bleeding can be significant due to tumor vascularity.[3,10] Preoperative embolization is often recommended in selected cases to reduce intraoperative blood loss and improve surgical visualization.[9,10] Our case did not undergo embolization, and tumor excision was successfully performed.

Histopathology remains definitive for diagnosis. Paragangliomas typically show a zellballen (nested) pattern and immunopositivity for neuroendocrine markers including chromogranin and synaptophysin.[3,6] The Ki-67 index in our patient was approximately 10%, indicating moderate proliferative activity, supporting adjuvant radiotherapy consideration. Radiotherapy has been shown to improve local control, particularly in tumors where complete surgical clearance is difficult or when histological features suggest higher risk.[4,9]

This case adds to the limited literature on CPA paragangliomas and emphasizes the importance of keeping paraganglioma as a rare differential diagnosis for atypical CPA tumors, especially when imaging shows internal hemorrhage, marked vascularity, or enhancement patterns inconsistent with schwannoma.

CONCLUSION

CPA paragangliomas are exceptionally rare and can mimic vestibular schwannoma on imaging. Accurate diagnosis requires high suspicion and confirmation by histopathology and immunohistochemistry. Surgical excision remains the mainstay of treatment, with radiotherapy as an important adjunct in selected cases. Early recognition of this rare entity can help optimize perioperative planning and improve outcomes.

Patient Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images

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