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RESEARCH ARTICLE

RARE CERVICAL PARAGANGLIOMAS: DIAGNOSTIC AND THERAPEUTIC CHALLENGES IN THREE CASES

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Abstract

Paragangliomas are rare neuroendocrine tumors arising from extra adre nal paraganglionic tissue derived from neural crest cells. They may dev elop anywhere from the skull base to the sacrum.Cervical paragangliom as also known as head and neck paragangliomas account for approxima tely 3% of all paragangliomas and mainly involve the carotid body ,vag al or jugulotympanicregions (1) .These tumors are typically benignbut may exhibit locally aggressive behavior. They are more frequently obse rved in females , accounting for nearly 67% of cases, and most commo nly diagnosed between the third and seventh decade of life (4).Complet e surgical resection remains the treatment of choice ;however , the rich vascular supply and proximity to critical neurovascular structures often make surgery high-risk .Radiotherapy represents a viable alternative, although its efficacy may vary. Accurate imaging plays a crucial role in diagnosis, and genetic analysis is essential for patient monitoring and long-term follow up.We report three cases of female patients presenting with painless cervical masses. Imaging confirmed the diagnosis of cervi cal paraganglioma, and biochemical assessments were performed. Due to tumor size and vascular invasion,surgical intervention was deemed u nsuitable,leading to radiotherapy as the preferred therapeutic option. Th is case series aims to provide an overview of the clinical presentation, d iagnostic approach ,and therapeutic strategies for cervical paragangliom as .

Introduction:-

Case report: -

Case 1: -

A 42-year-old female with no comorbidities or significant family history presented with a slowly progressive right lateral cervical mass evolving over eight years. CT angiography of the supra-aortic trunks revealed a highly vascularized lesion at the carotid bifurcation measuring 36 × 34 × 66 mm ,encasing both the internal and external carotid arteries over more than 180° , while maintaining vascular patency and compressing the internal jugular vein .MRI angiography showed a hyperintense lesion on both T1- and T2-weighted sequences with homogeneous enhancement measuring 41x29x64mm (Figure 1) , consistent with a Shamblin III right carotid paraganglioma. Plasma normetanephrines were elevated (6.08 times the upper normal limit) .Genetic testing revealed a pathogenic mutation in exon 1 of the VHL gene suggesting von Hippel-Lindau disease.

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^{123}I-mIBGscintigraphy demonstrated a soft-tissue mass at the carotid bifurcation measuring 39 x29 x 43 mm without tracer uptake suggesting a non-functional paraganglioma despite biochemical secretion , possibly reflecting a false-negative ^{123}I-m IBG result .

Surgery was contraindicated due to tumor size and vascular involvement as confirmed by multidisciplinary consensus. The patient receives external beam radiotherapy using intensity – modulated radiotherapy (IMRT) at total dose of 54 Gydelivered in 27 fractions. Follow-up CT angiography showed tumor regression to 34 x 23 x 44 mm (Figure 2) .Plasmametanephrines and pituitary function were subsequently monitored .

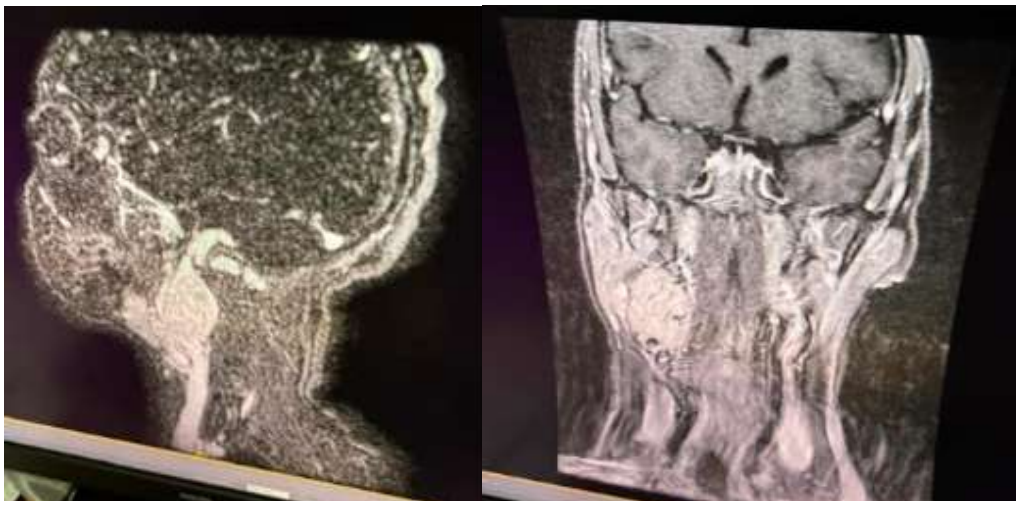

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Figure 1 : sagittal (a)and axial (b) cross-section from a CT angiography of the supra-aortic trunks showing the right carotid paraganglioma measuring 41x29x64 mm in diameter prior to radiotherapy

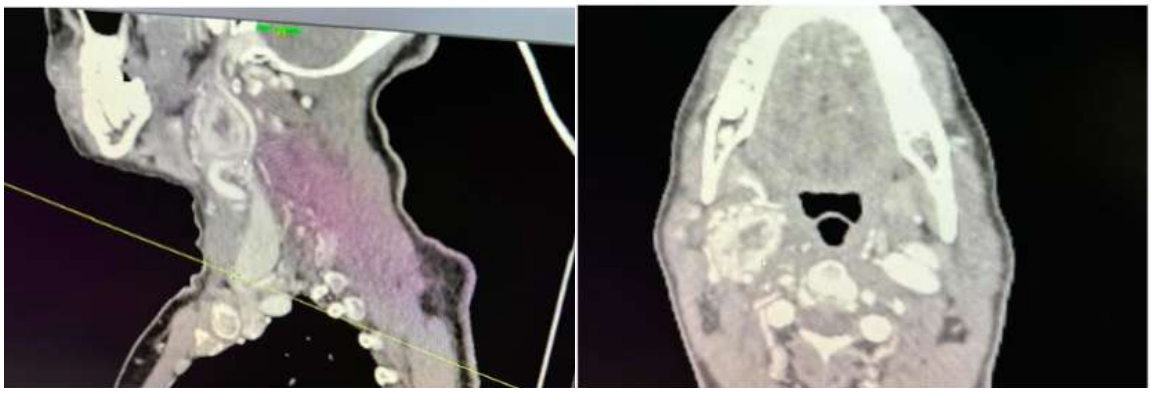

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Figure 2 : Sagittal (a) and axial (b) cross-sections from a post radiotherapy CT angiography of the supra-aortic trunks showing a reduction in the size of the right carotid paraganglioma decreasing from 39 x 29 x 43 mmto 34x23x44 mm in diameter

Case 2:-

A 38-year-old female with no comorbidities or relevant family history presented with a left lateral cervical mass progressing over two years. CT angiography revealed a heterogeneous lesion with central necrosis and intense arterial phase enhancement35 × 37 × 55 mm encasing the common carotid artery over more than 180° with preserved patency . Plasma normetanephrines were within normallimits .Genetic testing for MEN, NF1, and VHL was negative ; SDHx was not performed due to financial constraints.^{123}I-mIBGscintigraphy showed increased tracer

uptake in the cervical region, suggesting a functioning paraganglioma.The discrepancy between normal and biochemical results and imaging findings may be explained by low or intermittent catecholamine secretion.
Due to tumor extension and vascular involvement, surgery was excluded .The patient underwent IMRT with a total dose of 56 Gy in 28 fractions. Follow-upimaging demonstrated stable disease,with a residual tumor size of 37 x 30 x 57 mm.

Case 3:-
A 71-year-old female presented with a right lateral cervical mass evolving over three years. Cervical ultrasound initially identified a hypervascular, multilobulated jugulocarotidlesion measuring 44,4×25,8 mm, initially suspected to be a cystic lymphangioma .Subsequent MRI angiography confirmed a hypervascularlesion consistent with a right carotid paraganglioma, measuring 35 × 38.5 × 45 mm ,encasing the internal and external carotid arteries and compressing the internal jugular vein .

CT angiography revealed a mass beneath the mandible measuring 44 × 37 × 60 mm(Figure 3), with heterogeneous enhancement and involvement of the thyroid cartilage and internal jugular vein with loss of fat planes . Plasma metanephrines were negative, while chromogranin A levels wereelevated ,supporting the neuroendocrine nature of the tumor . Genetic testing (MEN, VHL, RET) was requested and results were pending at the time of reporting.
^{123}I-mIBGscintigraphy showed increased uptake in the right cervical region,confirming a functional paraganglioma despite negative plasma markers .Surgery was contraindicated due to tumor size and anatomical extension .IMRT was indicated , and the patient is currently awaiting treatment initiation .

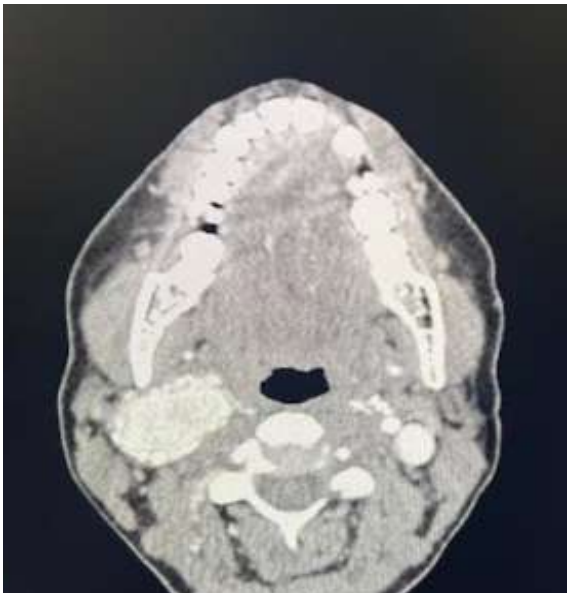

Figure 3 :Axial cross-section of a contrast enhanced CT angiography showing a right carotid paraganglioma measuring 44x37x60 mm in diameter prior to radiotherapy

Discussion:-

Head and neck paragangliomas (HNPGLs) represent approximately 70% of all extra-adrenal paragangliomas and predominantly arise from parasympathethicparaganglia .They are non-secretory and benign , with a strong female predominance and a mean age at diagnosis of around 47 years (3). Our case series is consistent with these data , as all three patients were women, although one patient was significantly older , likely reflecting a delayed diagnosis.

HNPGLs have also been associated with high-altitude residence due to chronic hypoxia, a hypothesis that may be relevant to our geographic context. Although most HNPGLs are considered non-functional, their genetic background is of major importance ,given the high rate of hereditary transmission .Up to 40% of cases are linked to germline mutations , mainly involving the SDHx gene complex.SDHB mutations are particularly associated with aggressive or metastatic disease, whereas SDHD mutations are more commonly linked to benign tumors.

In our series , genetic testing was performed in all patients, identifying a pathogenic VHL mutation in one case, consistent with group 1 mutations involving activation of the pseudohypoxic pathway . This finding led to genetic counseling for the patient and her family. However , extended genetic analysis , including SDHB immunohistochemistry or comprehensive gene panel testing , was not performed,representing a limitation of this study .

Clinically, cervical paragangliomas most often present as painless , slowly growing neck masses. All patients in our series presented with this typical feature , with additional pulsatility in two cases, suggestive of carotid body origin. Although only 4-5% of HNPGLs are reported to be catecholamine-secreting(3), one patient in our series showed elevated methoxy derivatives ,indicating secretory activity, this apparent discrepancy may be explained by the small sample size of our case series.

Radiologically, MRI angiography remains the gold standard for diagnosis ,providing characteristic imaging features including the “salt-and-pepper” appearance , and accurate vascular mapping. In our patients , MRI played a key role in confirming the diagnosis and assessing tumor extension . One case was initially misdiagnosed as a cystic lymphangioma on ultrasound , with the diagnosis subsequently corrected by MRI , highlighting the limitation of ultrasound alone.

Functional imaging using ^{123}I-MIBG scintigraphy was positive in two patients (2)(6). although^{123}I-mIBGhas limited sensitivity in non-metastatic HNPGLs , our findings illustrate its variable performance , possibly influenced by tumor size or functional status .

Surgical resection remains the only potentially curative treatment, but it is associated with a significant risk of cranial nerve injury and vascular complications ,particularly in advanced tumors .The Shamblin classification is essential for surgical risk stratification . All tumors in our series were classified as Shamblin III, reflecting advanced disease and explaining the high surgical risk . This likely reflects delayed diagnosis, with an average interval of approximately five years between symptom onset and treatment.

Due to the high surgical risk and tumor extension, all three patients were treated with intensity-modulated radiotherapy (IMRT). This approach provided good local control with minimal complications. Tumor regression was observed in one case, stable disease in the second , and treatment is ongoing in the third patient (5).

Our experience is consistent with published data demonstrating excellent local control rates with IMRT or stereotactic radiotherapy, particularly in inoperable or high-risk patients.Post-treatment surveillance requires regular clinical and imaging follow-up , with MRI recommended every 6–12 months initially and annually thereafter. Biochemical monitoring remains essential, especially in functional tumors .

The prognosis of HNPGLs is generally favorable in non-metastatic cases, with 5-year survival exceeding 90%. However, quality of life can be significantly affected by treatment-related complications including dysphonia, swallowing disorders, and psychological distress (1)(3).

This case series highlights several key points: the marked female predominance of HNPGLs, the potential for secretory behavior in tumors classically considered non-functional and the diagnostic challenges that may delay appropriate management . The identification of a VHL mutation underscores the importance of genetic screening. Nevertheless , the small number of cases, the absence of extended genetic testing and the relatively short follow-up limit the generalizability of our conclusions.(4)(6)

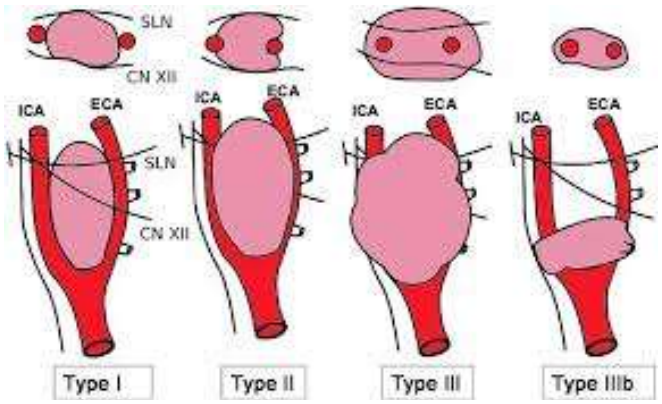

Figure 4 : Shamblin classification of cervical paraganglioma

Conclusion:-

Cervical paragangliomas are rare tumors that pose significant diagnostic and therapeutic challenges due to their vascularity , anatomical location and potential hereditary background . Our case series underscores the importance of early diagnosis , comprehensive imaging and genetic evaluation . In inoperable or high-risk cases , modern radiotherapy techniques such as IMRT represent an effective and well-tolerated alternative to surgery . Long-term multidisciplinary follow-up remains essential.

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