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RESEARCH ARTICLE

NEUROENDOCRINE NEOPLASMOF THE SPHENOID: CASE REPORT AND LITERATUREREVIEW

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Abstract

Neuroendocrine tumors (NETs) of the sphenoidregion are extremely rare(1)and pose significant diagnostic and therapeutic challenges due to their aggressive potential and complexanatomical location. Wepresent a case of a 34-year-old male whoinitiallypresentedwith persistent headaches and subsequentlydevelopedcranial nerve involvement, leading to the diagnosis of a sphenoid neuroendocrine tumor with caver nous sinus invasion. Given the surgicalrisks, a conservative approachwithsomatostatinanalogtherapywaschosen, resulting in partial tumorresponse. However, disease progression occurredaftertreatment discontinuation. This case underscores the importance of long term foll ow up and the need for individualized treamentstrategies in managing b ase neuroendocrine tumors(2).

Introduction:-

Neuroendocrine tumors (NETs) are a heterogeneous group of neoplasmsthatoriginatefrom neuroendocrine cells and are capable of secretingvarious hormones. Whilethey are commonlyfound in the gastrointestinal tract and lungs, primaryskull base NETs, particularlythoseinvolving the sphenoidregion, are exceedingly rare. Due to their location, theyoftenpresentwith non-specificsymptomssuch as headaches, visualdisturbances, and cranial neuropathies, leading to diagnostic delays. The management of thesetumorsrequires a multidisciplinaryapproach, balancing the risks and benefits of surgical intervention, medicaltherapy, and radiotherapy. In this case report, wedescribe a sphenoid NET with cavernous sinus invasion, highlighting the diagnostic process, therapeutic consider ations, and long-term management challenges.

Case Report.

A 34-year-old male, an ex-smokerwith no significantpastmedicalhistory, presented in May 2020 with persistent headachesthatwereunresponsive to conventionalanalgesic treatment. Over time, hissymptoms progressively worsene d, and in January 2023, hedevelopedleft sided ptosis. This new symptompromptedfurtherneurologicalevaluation. Magneticresonanceimaging (MRI) of the brainrevealed a tumorlocated at the sphenoid, extendinganteriorlytoward the sellarregion and invading the leftcavernous sinus. The imagingcharacteristicsraised suspicion of a locallyaggressiveneoplasm. To furthercharacterize the lesion, a biopsywasperformed, which histologicallyconfirmed

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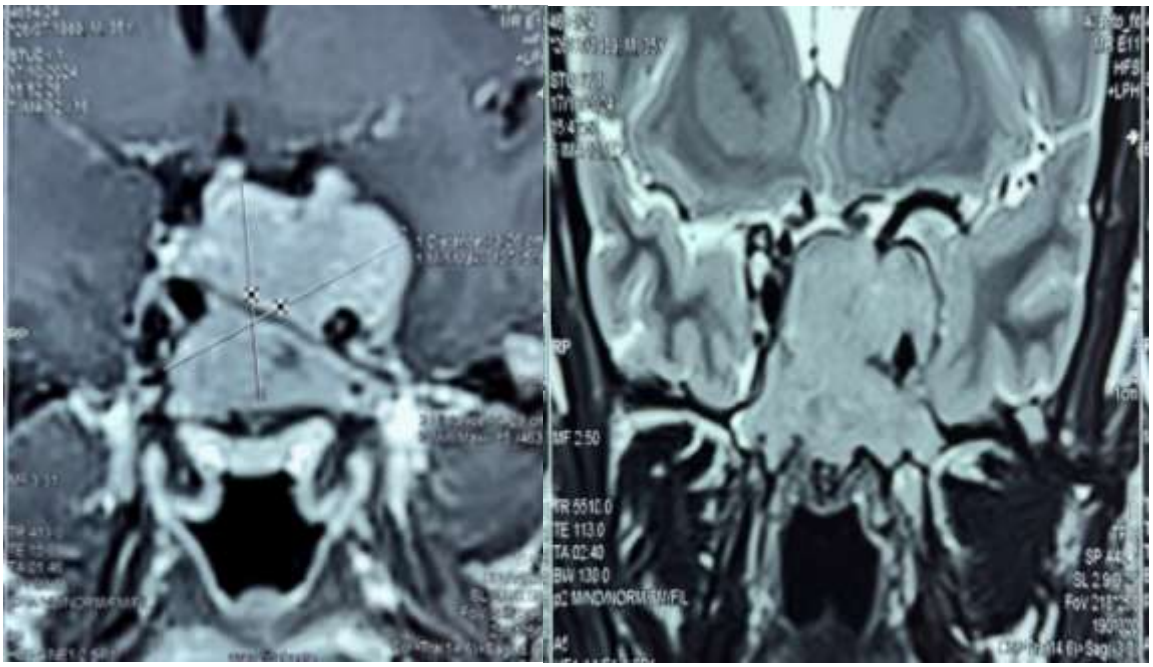
the diagnosis of a neuroendocrine tumor. Functionalimagingwith an octreoscansupported the presence of somatostatinreceptor expression, confirmingits neuroendocrine nature.



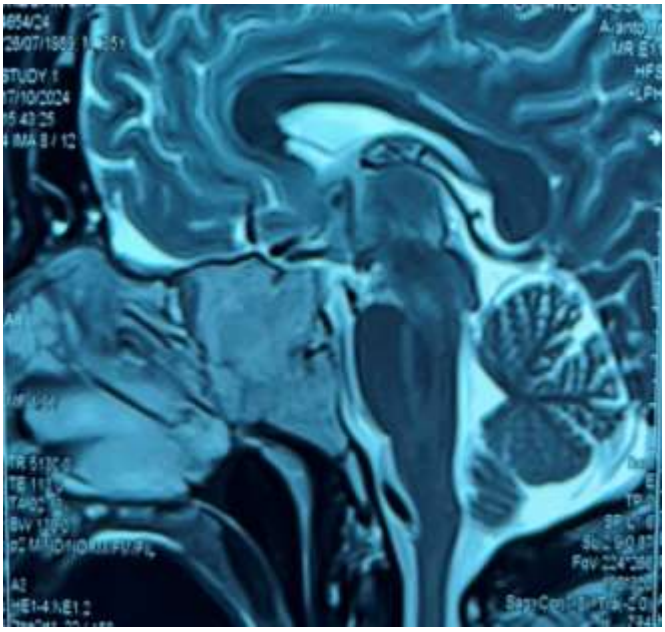


Figure 1,2,3 :Magneticresonanceimaging (MRI) of the brainshowing a tumorlocated at the sphenoidwith local extension.

Given the tumor's location and the potential risksassociatedwithsurgical intervention, the case wasdiscussed in a multidisciplinarytumorboard, includingneurosurgeons, oncologists, and endocrinologists. The consensus decisionwas to initiatetreatmentwithsomatostatinanalogtherapy (Somatuline), aiming to control tumor progression and alleviatesymptoms.

Follow-up MRI performedafterseveralmonths of therapydemonstrated a modestreduction in tumor size, decreasingfrom 33.4×40×55 mm to 33×30×50 mm, suggesting a partial response to treatment. The patient'ssymptomsimprovedslightly, althoughcompleteresolutionwas not achieved.
Unfortunately, the patient waslost to follow-up after March 2024, missingscheduledimaging and consultations. He returned in October 2024, reporting a recurrence of headaches and worsening ptosis, suggestingdisease progression. Uponreassessment, MRI wasrepeated, and the tumorexhibitedsigns of re-expansion. Given the previous partial response to somatostatinanalogtherapy, the decisionwas made to reinstateSomatulinetreatment, with plans for close monitoring and furtherconsideration of radiotherapy if necessary.

Discussion:

NETs of the skull base, particularlythosearisingfrom the sphenoidregion, remain a clinicalrarity, withlimited cases reported in the literature(3). Theirinsidiousonset and non-specificsymptomsoftenresult in delayeddiagnosis, by which time they have frequentlyinvadedcriticalneurovascular structures such as the cavernous sinus(4). The differentialdiagnosisincludespituitaryadenomas, meningiomas, chordomas, and metastaticlesions, necessitatingthoroughimaging and histopathological confirmation(5).
NETs of the skull base originatefromcells capable of neuroendocrine differentiation, thoughtheir exact histogenesisremainsunclear. Sometheoriessuggestthatthey arise fromectopic neural crestcells or remnants of neuroectodermal tissue in the sphenoidregion(6). Thesetumorscanvary in theirdifferentiation, withwell-differentiated cases exhibiting an indolent course, whilepoorlydifferentiatedNETsbehaveaggressivelywithrapid invasion of adjacent structures(7). The expression of somatostatinreceptors, as demonstrated in this case, provides an opportunity for targetedtherapiessuch as somatostatinanalogs and peptide receptorradionuclidetherapy (PRRT)(8).
Management strategies for skull base NETsdepend on multiple factors, includingtumor location, extent of invasion, functionalstatus, and patient preferences. Whilesurgicalresectionisoften the primarytreatmentmodality for accessible lesions, cases involving the cavernous sinus pose significantrisks of cranial nerve damage and vascular complications(9). In such scenarios, medicaltherapywithsomatostatinanalogs has demonstratedefficacy in controllingtumor progression, particularly in tumorsexpressingsomatostatinreceptors(10). In this case, the initial response to somatostatinanalogtherapywaspromising, but the patient'sloss to follow-up led to disease progression, reinforcing the necessity of continuous monitoring.
Additionally, radiotherapyremains a viable option for refractory cases, particularlystereotacticradiosurgery, whichcanprovidetargetedtreatmentwhileminimizing damage to surrounding structures(11). Fractionatedradiotherapycanalsobeconsidered in cases wheretumorgrowththreatens adjacent neurovascular structures(12). However, data on the long-termefficacy of radiotherapy in skull base NETsremainlimited, necessitatingfurtherresearch.
Recentadvances in targetedmoleculartherapies have opened new avenues for the management of advanced or recurrentNETs. Peptide receptorradionuclidetherapy (PRRT) usingradiolabeledsomatostatinanalogs has shownpromisingresults in metastatic and inoperableNETs by selectivelydelivering radiation to tumorcellsexpressingsomatostatinreceptors(13). However, itsefficacy in skull base NETsremains an area of ongoing investigation. Additionally, immune checkpoint inhibitors and novel tyrosine kinase inhibitorsmayplay a future role in cases resistant to conventionaltherapies, thoughfurtherclinicalstudies are needed to establishtheireffectiveness(14). Given the chronic nature of NETs, long-termfollow-up with serial imaging and biochemical markers is essential to monitor disease progression and guide treatmentdecisions(15). Patient compliance is a crucial factor in achieving optimal outcomes, as illustrated in this case where the interruption of follow-up led to tumorregrowth and symptomrecurrence.

Conclusion :

This case underscores the diagnostic and therapeuticcomplexitiesassociatedwithprimary neuroendocrine tumors of the sphenoid bone with cavernous sinus invasion. No surgical management withsomatostatinanalogscanachieve disease control in selected patients, particularlywhensurgeryiscontraindicated. However, the success of medicaltherapy relies heavily on strict adherence to treatment and follow-up protocols. A multidisciplinaryapproachremains crucial in optimizing patient outcomes in such rare and challengingpresentations.

Declaration of competinginterest:

None declaredunderfinancial, general, and institutionalcompetinginterests.

Consent:
Writteninformed Consent isobtained from the Patient for publication and anyaccompanying Images.

Ethical Approval:
This study is exempt from ethical approval in our institution since itdoesn'tinvolve experimental treatment.

References:

1. KAMBE A, NAKAJIMA S, FUKUSHIMA K, MIZUSHIMA M, SAKAMOTO M, HORIE Y, et al. PrimaryIntracranial Neuroendocrine Tumor of the Skull Base Complicatedwith Tension PneumocephalusafterRadiotherapy. NMC Case Rep J [Internet]. 2021 Sep 16 [cited 2025 Apr 26];8(1):609–15. Availablefrom: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC8769468/
2. Cassell A, Manobah B, Willie S. Diagnostic and Therapeutic Challenges of Rare UrogenitalCancers:UrothelialCarcinoma of the Renal Pelvis, Ureters and Urethra. World J Oncol. 2021 Feb;12(1):20–7.
3. PrimaryCerebellar Neuroendocrine Tumors:Chimeras or Real Entities A Case Report with a 6-Year Follow-Up | Case Reports in Oncology | KargerPublishers [Internet]. [cited 2025 Apr 26]. Availablefrom: https://karger.com/cro/article/9/2/432/89274/Primary-Cerebellar-Neuroendocrine-Tumors-Chimeras
4. Joshi VV, Thombare P, Verma M, Kamat N, Patkar D. Primaryskull base neuroendocrine carcinoma—a case report and review of literature. Radiology Case Reports [Internet]. 2020 Jul 1 [cited 2025 Apr 26];15(7):1071–7. Availablefrom: https://www.sciencedirect.com/science/article/pii/S1930043320301709
5. Klimstra DS, Modlin IR, Coppola D, Lloyd RV, Suster S. The Pathologic Classification of Neuroendocrine Tumors: A Review of Nomenclature, Grading, and StagingSystems. Pancreas [Internet]. 2010 Aug [cited 2025 Apr 26];39(6):707. Availablefrom: https://journals.lww.com/pancreasjournal/fulltext/2010/08000/the_pathologic_classification_of_neuroendocrine.2.aspx
6. Rosai J. The origin of neuroendocrine tumors and the neural crest saga. ModPathol. 2011 Apr;24 Suppl 2:S53-57.
7. Asa SL, Mete O, Cusimano MD, McCutcheon IE, Perry A, Yamada S, et al. Pituitary neuroendocrine tumors: a model for neuroendocrine tumor classification. Modern Pathology [Internet]. 2021 Sep 1 [cited 2025 Apr 26];34(9):1634–50. Availablefrom: https://www.sciencedirect.com/science/article/pii/S0893395222005439
8. Stueven AK, Kayser A, Wetz C, Amthauer H, Wree A, Tacke F, et al. Somatostatin Analogues in the Treatment of Neuroendocrine Tumors:Past, Present and Future. International Journal of Molecular Sciences [Internet]. 2019 Jan [cited 2025 Apr 26];20(12):3049. Availablefrom: https://www.mdpi.com/1422-0067/20/12/3049
9. Rangel-Castilla L, Russin JJ, Spetzler RF. Surgical management of skull base tumors. RepPractOncolRadiother [Internet]. 2016 [cited 2025 Apr 26];21(4):325–35. Availablefrom: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4899518/
10. Liu H, Wang H, Qi X, Yu C. Primaryintracranial neuroendocrine tumor:two case reports. World Journal of SurgicalOncology [Internet]. 2016 Apr 30 [cited 2025 Apr 26];14(1):138. Availablefrom: https://doi.org/10.1186/s12957-016-0887-4
11. Primary Neuroendocrine Tumor in Brain - Tamura - 2014 - Case Reports in NeurologicalMedicine - Wiley Online Library [Internet]. [cited 2025 Apr 26]. Availablefrom: https://onlinelibrary.wiley.com/doi/10.1155/2014/295253
12. Hood B, Bray E, Bregy A, Norenberg M, Weed D, Morcos JJ. PrimaryCarcinoidTumor of the Cavernous Sinus. World Neurosurgery [Internet]. 2014 Jan 1 [cited 2025 Apr 26];81(1):202.e9-202.e13. Availablefrom: https://www.sciencedirect.com/science/article/pii/S1878875013007559
13. Bison SM, Konijnenberg MW, Melis M, Pool SE, Bernsen MR, Teunissen JJM, et al. Peptide receptorradionuclidetherapyusingradiolabeledsomatostatinanalogs: focus on future developments. Clin Transl Imaging [Internet]. 2014 [cited 2025 Apr 26];2(1):55–66. Availablefrom: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3991004/
14. Albertelli M, Dotto A, Nista F, Veresani A, Patti L, Gay S, et al. Present and future of immunotherapy in Neuroendocrine Tumors. RevEndocrMetabDisord [Internet]. 2021 [cited 2025 Apr 26];22(3):615–36. Availablefrom: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC8346388/
15. Franchina M, Cavalcoli F, Falco O, La Milia M, Elvevi A, Massironi S. Biochemical Markers for Neuroendocrine Tumors:TraditionalCirculating Markers and RecentDevelopment—A ComprehensiveReview.

Diagnostics (Basel) [Internet]. 2024 Jun 18 [cited 2025 Apr 26];14(12):1289. Availablefrom: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC11203125/