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Case Report

Rare Primary Neuroendocrine Neoplasm (NENs) Brain Tumor, Case Report and Literature Review

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Abstract

Rare Neuroendocrine neoplasms (NENs) are malignancies rarely involving the central nervous system. A 42-year-old man was referred to our clinic with chronic intermittent frontal headache and left hemiparesis. After an initial assessment with CTs and MRIs, proper basal ganglia involvement with a heterogeneous brain tumor was found. The patient underwent brain tumor resection, and pathology showed Neuroendocrine neoplasms. Surprisingly, tumor manifestation was similar to metastatic NENs. It is essential that the prognosis between primary NENs and metastatic NENs to the brain is utterly different, and treatment options are not identical. The patient was discharged concession with left hemiparesis and referred to a neuro-oncologist for further treatment.

Key words: neuroendocrine neoplasm; brain tumor; central nervous system

Background

The Central Nervous System (CNS) contains multiplex cells competing to bring out neuroendocrine neoplasms (NENs). [1] neuroendocrine cells are distributed all around organs and are well represented throughout the respiratory system, gastrointestinal tract, and CNS. [2] NENs are developing in all previous systems, and rare reports discussed NENs derived primarily in the brain. [3] These tumors are still sporadic, and they count for only 4%–6% of all extracranial malignancies. While there has been an increasing incidence of NENs over these years, they are diagnosed as secondary due to extra-CNS origin. [4] When NENs are diagnosed as the primary tumor, the extra-cranial source should be assessed on further diagnostic imaging. Brain metastases are correlated with a poor prognosis, with overall survival of 8 months from CNS involvement to diagnosis. [5,6] The incidence of patients with NENs having brain metastases is <5%, and many originate from the lungs. [5] Moreover, 1.4% of metastatic brain tumors are NENs. Notably, when patients present with metastatic NENs to the brain, assessment for other NENs origin is advised.

Case Presentation

Here, we describe a case of a 42-year-old previously healthy man who presented with symptoms of intracranial hypertension such as early morning headache and continuous blurred vision. Initial imaging showed an intracranial heterogenous brain tumor. An MRI of the brain demonstrated a heterogenous 2 cm lesion in the right thalamus, which entered lateral ventricle near the foramen of Monro. (Figure-1) He has undergone surgery, and diagnostic pathologic tests reported neuroblastoma. (Figure-2) But referral pathologic center had the positive test of synaptophysin and chromogranin, that neurofilament staining was positive, either. Pathology reported high-grade neuroendocrine carcinoma, with a Ki-67 of 45%-50% positive. Epithelial membrane antigen (EMA), glial fibrillary acidic protein (GFAP), neuronal nuclear antigen (NeuN), S-100, thyroid transcription factor-1 (TTF1), CDX2, and a pancreatic battery including insulin, gastrin, somatostatin, glucagon, vasoactive intestinal peptide (VIP) and pancreatic polypeptide (PP) were all negative. While the patient was assessed by PET/CT scan and whole-body MRI, which all were harmful for any evidence of extracranial sources of NENs brain tumor.

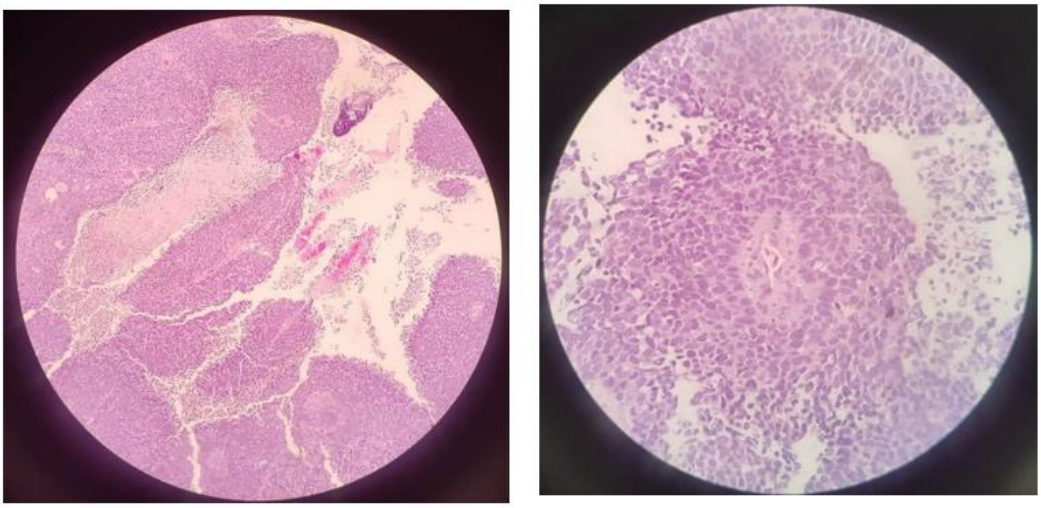

Figure 1: MRI Imaging of primary NENs brain tumor. A: MRI T1 sequences with right thalamic involvement enhanced with

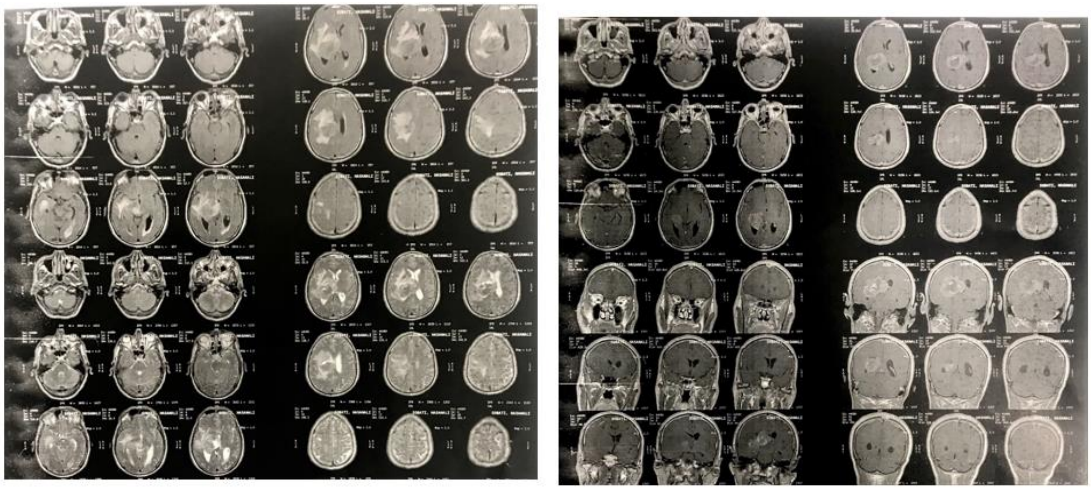

Figure 2: Pathologic pattern of NENs tumor

Treatment

Subsequently, after surgery, the patient developed mild left hemiparesis and worsening mental status with a Glasgow Coma Scale of 14. After initial brain tumor surgery, GCS was about 12-13. The left-sided hemiparesis improved with physical therapy. The decision was made to proceed with further treatment with referral to radio-oncologist.

Discussion

Based on our knowledge, rare reports discussed NENS in literature. Neuroendocrine cells are available in different organ systems, and initial NENs can develop in all organs. [7] The setting of the primary brain tumor inside the thalamic area also present notable importance. The thalamic and pre-ventricular nucleus is a group of neuroendocrine cells isolated next to the ventricles in our patient's primary NEN area. NENs can be classified clinically as functional or nonfunctional if increased hormone production is needed. [8] In addition, articles categorized NENs as their anatomical location and Ki-67 presenting. [8] Ki-67 marker uses as a functional degree of proliferation because it is secreted by active cells. [9] Research suggested a possible primary CNS NEN in a 77-year-old woman with overall survival of 1.5 years with confirmed histology. Regarding our case, other cases demonstrated non-CNS primary NENs. Secondary NENs manifested in the pineal gland, pituitary, and along the meninges, which surprisingly thalamic, are was rare. [10]

In our case, several indications reinforce the speculation that a brain tumor was indeed a primary NEN. First, tumor location is in an area with known neuroendocrine cells. [11] secondary, most metastatic brain mass most commonly demonstrated in the location of the gray-white junction. [11] thirdly, extensive workup with CT and PET/CT imaging showed no other extra-cranial sources. This strongly suggests that the brain was the primary and only disease site.

The gold standard protocol for assessing NENs, both CTs and FDG PET/CTs. Which has a sensitivity of 95% for assessment of a primary NENs brain tumor. [12] similar to our case, if NEN is diagnosed as high grade, FDG PET/CTs have magnificent sensitivity in finding primary sources. Our patient had multiple PET-CTs around his brain tumor course that was negative to find another primary location.

More research are required on the exact treatment of primary brain NENs. As a common idea, in metastatic brain NENs, reports has demonstrated radiation therapy plus surgery are more effective. In addition, platinum-based chemotherapy such as cisplatin is suggested in high-grade primary NENs. Consequently, in primary NENs brain tumors, brain tumor resection and chemo-radiotherapy should be considered for treatment.

Conclusion:

Finally, although many diagnosed NENs tumors originate from other organs such as respiratory tracts, gastro-interstitial and supra-renal, in rare conditions, NENs are manifested as primary brain tumors. In This situation, precise pathologic evaluation with a whole organ assessment is needed to rule out metastasis. Furthermore, based on the patient's condition, open brain tumor resection with other chemo-radiotherapy is effective for most excellent outcomes.

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