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Case Report

Not So Benign: A Rare Atypical Ectopic Choroid Plexus Papilloma

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Abstract

Choroid Plexus Papillomas (CPPs) are rare neoplasms (0.4-0.6 % of all brain tumors) arising from cuboidal epithelial cells of the choroid plexus. Atypical choroid plexus papillomas are even more rare and characterized by aggressive features of increased mitotic activity and frequent metastases even at diagnosis. Atypical choroid plexus papillomas accounted for 9% of choroid plexus tumors in the Surveillance Epidemiology and End Results (SEER) Database from 1978 to 2009. We describe a 56 year-old woman with a rare atypical choroid plexus papilloma ectopically located in the cerebellopontine angle and mistaken for a vestibular schwannoma or glossopharyngeal schwannoma. She demonstrated leptomeningeal seeding involving multiple cranial nerves and spinal cord. Besides papilledema she developed several neuro-ophthalmic features slowly over time from involvement of cranial nerves and subsequent intraparenchymal spread and radiation necrosis in the brainstem. Besides being rare, the cerebellopontine angle location of this tumor is also extremely uncommon making this a very unique case.

Keywords: Choroid plexus tumors; Atypical ectopic choroid plexus papilloma; Cerebellopontine angle tumors; Leptomeningeal deposits; papilledema

Introduction

Choroid Plexus Papillomas (CPPs) are rare neoplasms (0.4-0.6% of all brain tumors) arising from cuboidal epithelial cells of the choroid plexus. Atypical choroid plexus papillomas are even more rare and characterized by aggressive features of increased mitotic activity and frequent metastases even at diagnosis. Atypical choroid plexus papillomas accounted for 9% of choroid plexus tumors in the Surveillance Epidemiology and End Results (SEER) Database from 1978 to 2009. We describe an adult with a rare atypical ectopic choroid plexus papilloma in the cerebellopontine angle with several neuro-ophthalmic features that slowly manifested over time.

Case Presentation

A 56-year-old woman developed vertigo, sensorineural hearing loss, tinnitus, imbalance, and headaches. Contrast-enhanced MRI-head showed a right Cerebellopontine Angle (CPA) tumor (Figure 1A) concerning for a vestibular or glossopharyngeal schwannoma. Due to radiological and clinical worsening over time she received gamma knife radiation of total 24 gray. Despite this, the tumor enlarged and displayed a cystic component (Figure 1B). Multiple additional tiny enhancing lesions involving the bilateral cranial nerves V and VI , left cranial nerve VII and VIII (Figure 1D), and right cranial nerve IX extending into jugular foramen were seen. She continued to worsen clinically, received subtotal tumor resection that was unfortunately complicated by Cerebrospinal Fluid (CSF) leak from the right posterior auricular region. Surprisingly, histopathology disclosed an atypical ectopic choroid Plexus Papilloma (CPP) with degenerative atypia. The cranial nerve lesions and multilevel leptomeningeal deposits noted on MRI of the whole spine (Figure 1C) seemed to be metastatic deposits.

Patient received radiation of 5400 cGY in 27 fractions. The brainstem received maximum dose of 5454 cGY. Subsequently, she reported seeing black spots in the left eye, binocular transient visual loss, and vertical diplopia. Ophthalmology exam revealed visual acuity of 20/25 OU with bilateral papilledema, vitreous hemorrhage in the left eye, and small comitant left hyperphoria felt to be skew deviation. Follow-up MRI head revealed residual enhancing right CPA tumor indenting the pons and middle cerebellar peduncle and extending into the right jugular foramen with persistent enhancement of multiple cranial nerves. Lumbar puncture revealed elevated opening pressure of 31 cm H_2O with 57 nucleated cells predominantly lymphocytic, protein (35), glucose (60) and no CSF dissemination of the tumor. CSF diversion was deferred, papilledema was treated with acetazolamide.

Over time she developed right sixth and seventh nerve palsy, dysphagia, right internuclear ophthalmoplegia with upbeat nystagmus, and then downbeat nystagmus sequentially. MRI head revealed slowing enlarging right CPA tumor with leptomeningeal spread and new patchy enhancement along the right middle cerebellar peduncle and right dorsal pons (Figure 1E) concerning for intraparenchymal spread versus radiation necrosis. Spinal MRI showed new involvement of the conus in addition to the previous leptomeningeal deposits. Her various neuro-ophthalmic features seemed to emanate from cranial nerve involvement and intraparenchymal spread combined with radiation necrosis in the brainstem.

She failed a trial of bevacizumab for the tumor by developing uncontrolled hypertension. She also failed temozolomide by displaying a new area of involvement in the right temporal lobe. Currently she is on a first in-human phase I clinical trial of BXQ-350 a

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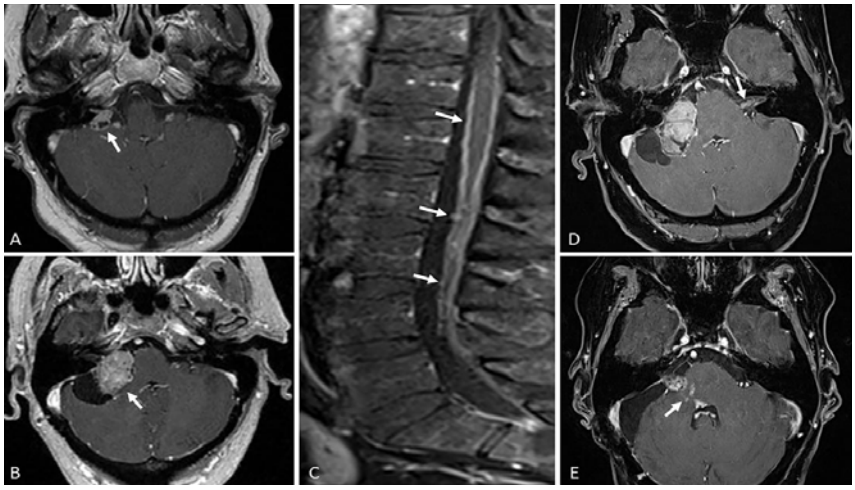

Figure 1: A) Axial T1-weighted contrast enhanced MRI of the head showing a right CP angle tumor with both solid and cystic components; B) Axial T1-weighted contrast enhanced MRI of the head 2 years later shows a significant increase in size; C) Sagittal T1-weighted fat saturated contrast enhanced MRI of the lumbar spine showing multilevel leptomeningeal deposits; D) Axial T1-weighted contrast enhanced fat saturated MRI of the head showing leptomeningeal spread along cranial nerves VII & VIII; E) Axial T1-weighted contrast enhanced fat saturated MRI of the head showing new patchy enhancement along the right middle cerebellar peduncle and right dorsal pons extending to the right facial colliculus consistent with radiation necrosis versus intraparenchymal tumor spread.

synthetic form of the human glycoprotein saposin C (NCT02859857). Her tumor remains stable on serial imaging. But for the worsening of drop metastasis in the lumbar spine she received radiation of 3000 cGy in 10 fractions using 16X photons with 3D conformal technique directed to the L-spine.

Her papilledema resolved. Her diplopia symptomatically improved with Fresnel prisms and subsequently ground in horizontal and vertical prisms in her glasses. For lagophthalmos and corneal exposure, she underwent numerous procedures including right lower lid ectropion repair, right lateral permanent tarsorrhaphy, gold weight placement, a right temporalis tendon transfer, a right static facial sling, and bilateral rhytidectomies.

Discussion

Choroid Plexus Tumors (CPTs) are rare papillary neoplasms derived from choroid plexus epithelium [1]. Within the Surveillance Epidemiology and End Results (SEER) Database from 1978 to 2009, CPT represent <1% of all CNS neoplasms in all ages and 14% of all brain tumors in the first year of life [2].

Although mostly benign, CPP can spread locally and cause a mass effect on the brain [3]. Atypical CPP (aCPP) was recognized as a distinct and rare entity (2016 WHO Classification of CNS Tumors) [4] showing increased mitotic activity and metastasis even at diagnosis. In the SEER database, CPP and aCPP accounted for 74 and 9 %, respectively, of all CPTs [2].

CPPs predominantly arises from the lateral ventricles in children and fourth ventricle in adults [5]. The CPA location is extremely rare with only approximately 9% located in this region and almost exclusively found in adults [6,7].

It is hypothesized that extraventricular CPP in the CPA may arise from ectopic choroid plexuses [9] or from the normal choroid tuft outside the foramen of Luschka [7,8], or from drop metastases or seeding along the cerebrospinal fluid pathways.

CPPs in the CPA cannot be distinguished easily from other tumors pre-operatively [1] and often mistaken for vestibular schwannomas. On CT, they may be isodense, hypodense, hyperdense or show mixed density with stippled, dense or patchy calcification occasionally. The tumor is isointense or hypointense (T1-weighted MRI) and isointense or hyperintense (T2 weighted MRI) [1] with homogenous or heterogeneous contrast enhancement. 20% display peritumoral cystic component [9], some may have an enhancing mural nodule [1].

Clinical symptoms include headache, dizziness, unsteady gait and cranial nerve deficits involving V, VII and VIII reported in about 50% cases [1]. Obstructive hydrocephalus can occur with cerebellopontine angle CPP.

5-year survival rate of 90, 77, and 58 % was seen for CPP, aCPP, and Choroid Plexus Carcinoma (CPC), respectively. Older age and male sex were prognostic for worse survival [2].

Surgical resection remains the treatment of choice. Gross Total Resection (GTR) through the suboccipital retrosigmoid approach or far lateral suboccipital approach is preferred over subtotal resection. GTR may or may not be significantly associated with improved survival [2].

Resection of enhancing mural nodules or enhancing parts of the cyst has been described but it is not necessary to remove the cyst. There is risk of tumor recurrence and malignant transformation [10]. The role of radiation for CPP or its recurrence remains to be determined.

Conclusion

We have thus described a rare case of atypical ectopic choroid plexus papilloma that displayed metastatic involvement of cranial nerves from early stages. The cerebellopontine angle location is also rare. Our patient displayed myriad neuro-ophthalmic signs sequentially overtime making this a very unique case. We postulate that it could be from the leptomeningeal and intraparenchymal

spread besides radiation necrosis.

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