CLINICAL VIGNETTE

Infratentorial Presentation of Astrocytoma as a Primary Brain Tumor

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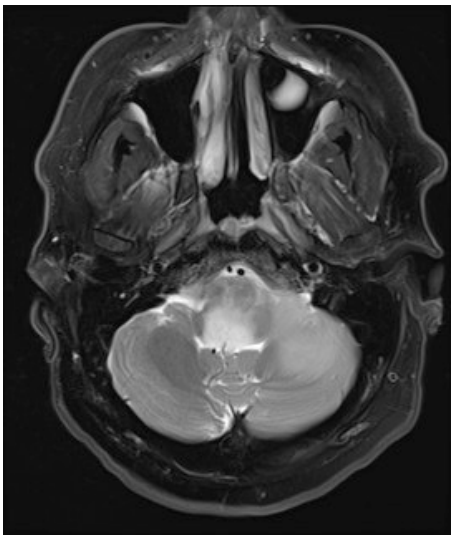
Case Report

A 43-year-old male with Obstructive Sleep Apnea treated by Uvuloplasty 11 years prior, presented with blurry vision for the past 3 months. The patient also felt that his "balance was off" having to "catch himself from falling". He developed a "nasal" voice after the uvuloplasty but over the past 3 months he began slurring words, which is completely different from how his voice sounded over the past 10 years since the surgery. He denies weakness, dizziness, fevers, chills, nausea or vomiting. His friends noted that he walks and speaks as if he is drunk although he does not drink alcohol or use any illicit drugs. He smokes cigarettes. The only medication that the patient takes on a regular basis is omeprazole 20 mg daily for GERD.

Physical exam was essentially normal except for the neurologic exam. His speech was somewhat slurred but coherent. He was alert and oriented to person, time, place and situation. He had both horizontal, oscillatory and vertical nystagmus. He could not perform tandem gait and had a positive Romberg. His gait was wide based. Motor was 5/5 throughout and sensory and proprioception were intact throughout.

MRI of the brain revealed diffuse brainstem infiltration bilaterally, more prominent dorsally than ventrally, and infiltration of the left cerebellar hemisphere. Swelling of the brainstem resulted in crowding at the foramen magnum with partial obstruction of the fourth ventricular outlet foramina causing dilatation of the ventricular system greatest along the fourth ventricle (i.e., mild obstructive hydrocephalus).

The patient was admitted to the hospital and Neurosurgery performed a left suboccipital craniectomy and biopsy of the left cerebellar lesion. Pathology yielded a low grade astrocytoma (WHO Grade 2).



MRI of Brain showing cerebellar and brainstem involvement of astrocytoma

Epidemiology

Glioma is not rare, accounting for approximately one-third of all primary brain tumors (PBTs)[1]. Of these, approximately 9.1% are classified as low grade astrocytoma (LGA)[2]. This case of LGA is rare, both in its anatomic location and age of onset. Infratentorial PBTs are rare in adults[1,2,3].

The Central Brain Tumor Registry of the United States (CBTRUS) (n=326,711) found 95.7% of all PBTs were supratentorial with cerebellar and brain stem tumors accounting for only 2.7% and 1.6% of all PBTs, respectively. Infratentorial PBTs are much more common in children than adults, accounting for greater than 26% of all PBT's in children aged 0-19 years old. In contrast, infratentorial tumors account for only 2.7% of all PBTs in adults aged 20 and older. This number drops to 2.2% when examining adults 35 and older[2].

In a review of 1,283 adult (>18 years) patients diagnosed with glioma, Strauss et. al. reported that only 4.4% (n=57) were posterior fossa tumors with 18 (31.5%) of these classified as primary cerebellar tumors and 21 classified as primary brainstem tumors (37%) . The median age at diagnosis was 40 years (range 19-81 years). Of 57 patients ,12 had grade II astrocytoma. 11 of these were classified as brainstem, and 1 as cerebellar[1].

Both astrocytomas and infratentorial PBTs are more common in men than women[2].

Discussion

The rotatory nystagmus and blurry vision seen in our patient can be explained by tumor involvement of the brainstem into the medulla and medial longitudinal fasciculus. The wide base gait and speech abnormalities are explained by cerebellar involvement giving cerebellar ataxia and cerebellar dysarthria.

Neurosurgery felt surgical cure was not possible given the location of the tumor. Radiation oncology recommended fractionated external beam radiation therapy. Hematology/Oncology concurred, given the low grade astrocytoma. The patient continued on radiation therapy and was doing well at the time of this article.

Surgical resection of low-grade gliomas (LGGs) has been shown to improve overall survival (OS) with greater benefit seen in those patients with full versus partial resection [3,4]. Some studies suggest more complete resection may prevent disease progression to a high grade glioma but this evidence is inconclusive[3,4].

Use of early radiation therapy for LGGs, as opposed to waiting for progression to initiate therapy, was established by the EORTC randomized controlled trial that showed an increase in progression free survival (PFS) (5.3 years vs. 3.4 years) despite failing to show a significant improvement in OS (7.4 years vs. 7.2 years)[5]. This increase in PFS has been confirmed in multiple subsequent trials and is radiation therapy is now a standard treatment for LGGs[4].

The role of chemotherapy in the management of LGGs remains unclear and until recently these tumors have been considered resistant to medical therapy[3,6]. A Radiation Therapy Oncology Group (RTOG) study is currently examining radiation therapy alone vs. radiation therapy plus chemotherapy (procabazine, lomustine, and vincristine) in 251 adults who were either aged 40 years or older or with subtotal resection of LGG[7]. Improvements in both PFS and OS (13.3 years vs. 7.8) years have been observed with adjuvant chemotherapy[6,7]. A similar trial by RTOG on the benefit of adjuvant temozolamide therapy on LGGs is ongoing and preliminary results show a 3 year OS of 73.1%[6].

Although treatment of LGGs in adults is well established in the literature, treatment of infratententorial LGGs, and specifically LGAs, is poorly understood. Recently, however, Bagley et al[8] found that patients with cerebellar grade II gliomas had a more favorable OS than those with supratentorial grade II gliomas. This interesting finding suggests that the disease course of infratentorial LGGs in adults differs from that of supratentorial tumors, with need for additional studies.

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