5-16-2022

A rare case of autoimmune glial fibrillary acidic protein astrocytopathy

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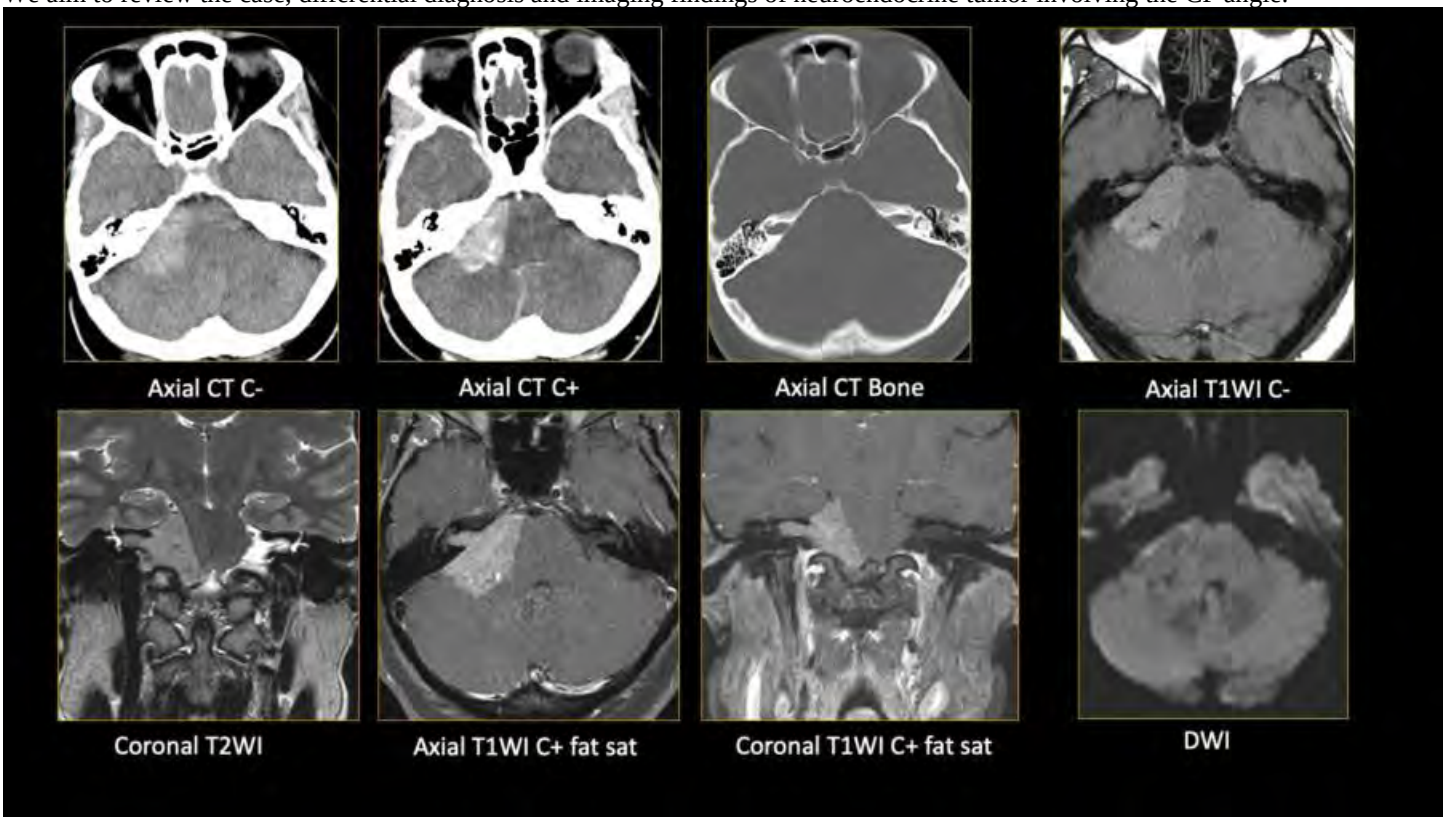
Materials and Methods
Large homogeneously enhancing soft tissue mass in the right cerebellopontine cistern and internal auditory canal isointense to gray matter on T2 and slightly hyperintense on T1.
Results
Neuroendocrine tumors infrequently metastasize to the brain with an incidence of up to 5%.(1-2) Our patient has no evidence of a primary tumor arising from another location, and it is likely that the brain is the primary source of this tumor. There are very limited reports of this phenomenon in the literature, with only case reports of possible CNS primary neuroendocrine tumors. (2) To our knowledge, this is the first described case of a neuroendocrine tumor involving the CP angle.
Conclusions
We aim to review the case, differential diagnosis and imaging findings of neuroendocrine tumor involving the CP angle.



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844

A Rare Case of Autoimmune Glial Fibrillary Acidic Protein Astrocytopathy

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Purpose
This is a 69-year-old male who presented with cognitive decline, hypophonia, dyspraxia, course tremor, myoclonic jerks, and gait dysfunction, all developing over 3-4 weeks. Initial head CT and MRI brain without contrast were negative. EEG demonstrated diffuse slowing. CSF analysis revealed elevated protein and elevated white blood cells (lymphocyte predominant). Given neurological worsening, MRI brain and cervical spine with contrast were performed and unique findings were identified, detailed below. The CSF autoimmune panel returned positive for Glial Fibrillary Acidic Protein (GFAP). The patient was started on steroids with rapid clinical improvement.
Materials and Methods
MRI brain with contrast 2 weeks after initial imaging demonstrated extensive, predominantly periventricular curvilinear enhancement in a perivascular distribution. MRI cervical spine demonstrated symmetric posterolateral pathological enhancement within the cord. Given these findings and the CSF results, a diagnosis of autoimmune GFAP astrocytopathy was established, and the patient was started on steroid therapy. Repeat MRI brain with contrast 2 weeks later demonstrated marked interval decrease in the perivascular enhancement, with mild residual periventricular enhancement. Follow-up MRI brain with contrast 5 weeks after beginning steroid therapy demonstrated complete resolution of abnormal perivascular enhancement.
Results
First described in 2016, autoimmune GFAP astryocytopathy is a rare autoimmune inflammatory central nervous system (CNS) disorder. Typical symptoms include subacute progressive encephalopathy, involuntary movements, autonomic dysfunction, and fever. Key imaging findings include curvilinear periventricular enhancement in a perivascular distribution, which can also be seen in the

infratentorium and spinal cord. Cases generally feature elevated CSF protein and CSF pleocytosis. The vast majority of cases have been in parainfectious and paraneoplastic settings. Treatment regimens are not yet well-established, but patients who are treated with early immunotherapy generally show marked resolution of symptoms over a period of weeks.
Conclusions
Autoimmune GFAP astrocytopathy is a rare, novel immunotherapy-responsive inflammatory CNS disorder with key imaging findings including curvilinear periventricular enhancement in a perivascular distribution, with similar abnormal perivascular enhancement in the infratentorium, and spinal cord involvement. The findings share similarity to CLIPPERS syndrome, though more extensive.

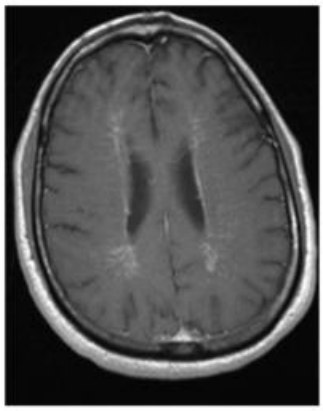

Figure 1. T1-weighted MRI brain with contrast demonstrates curvilinear periventricular enhancement in a perivascular distribution.

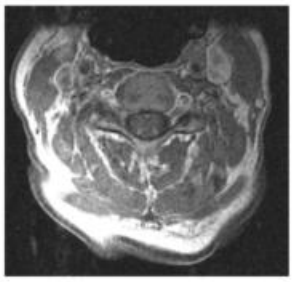

Figure 2. T1-weighted MRI cervical spine with contrast demonstrates symmetric posterolateral pathological enhancmenet within the cord.

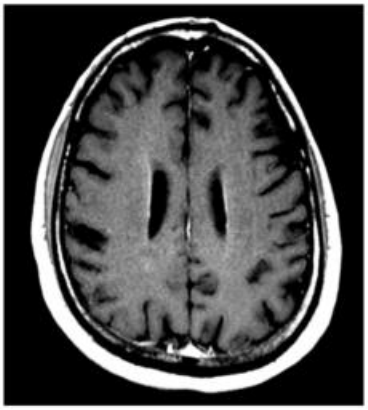

Figure 3. Two week follow up T1-weighted MRI brain with contrast demonstrates marked decrease in periventricular curvilinear enhancement with mild residual enhancement.

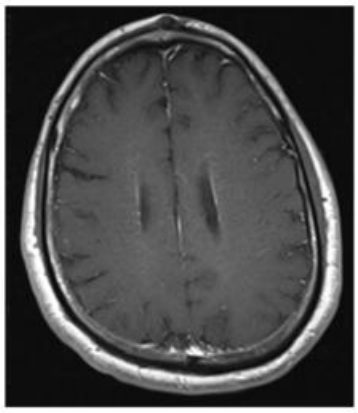

Figure 4. Five week follow up T1-weighted MRI brain with contrast demonstrates near-complete resolution of periventricular curvilinear enhancement.

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1509

A Rare Case of Diffuse Midline Glioma of the Cervicothoracic Spine

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Purpose
19-year-old previously healthy male initially presented with bilateral foot numbness and weakness that rapidly progressed up the lower extremities into the chest. Patient also complained of perfuse night sweats but no weight loss. The spinal cord lesion was thought to be inflammatory given his young age and positive ACE levels in the serum and CSF. At presentation, steroids were initiated. However, the lesion progressed on steroids and biopsy eventually confirmed a diffuse midline glioma, H3 K27-altered.
Materials and Methods
Spinal MR demonstrate a long segment expansile intramedullary cord lesion in the central aspect of cervicothoracic spinal cord that is T2/STIR hyperintense with heterogenous enhancement.
Results
Diffuse midline glioma, histone H3 K27M mutant was added to the World Health Organization Classification of Tumors in 2016 as a specific mutation of the previously classified diffuse astrocytoma. The K27M mutation appears specific for diffuse gliomas arising in midline structures. Specific gene mutation was the most important predictive factor of overall survival. No specific imaging features have been described in the literature for spinal cord lesions, but contrast enhancement and expansile increased T2/STIR signal was commonly seen. The presence of enhancement on MRI was not a predictive factor of patient outcome.
Conclusions
Diffuse midline gliomas can be found throughout midline structures of the central nervous system including the thalamus, brainstem, and spinal cord. Tumors in the thalamus and posterior fossa tend to be solid or infiltrative with infrequent necrosis. There are limited studies on spinal cord diffuse midline gliomas, however case studies report distant and early metastasis to the leptomeninges. Early detection could help facilitate timely treatment and possibly prevent metastatic disease in certain patients.