Abstract A1

An autopsy case of small vessel, childhood, primary angiitis of the central nervous system (SVcPACNS): suggestions for establishing specific histologic criteria for diagnosis

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SVcPACNS is a rare inflammatory/immune disorder that typically affects the *small blood vessels* of the brain. SVcPACNS differs from most adult forms of PACNS by being predominantly lymphocytic, non-granulomatous and non-necrotizing. Previously healthy children are typically affected by range of signs and symptoms, including: seizures, headache, cognitive decline, behavior/personality change, focal neurological deficits and potentially a decreased level of consciousness. Treatment protocols featuring *induction* (steroids and cyclophosphamide) and subsequent *maintenance* phases (e.g., mycophenolate mofetil) have been demonstrated to yield favorable outcomes. Since SVcPACNS is characteristically angiography negative, the diagnostic gold standard is brain biopsy. Interpretation of these biopsies is often challenging given the histologic overlap between SVcPACNS and encephalitis. Distinguishing the foregoing is critical since the treatment of these entities is significantly different.

Herein, a rare autopsy case of SVcPACNS in a 4 year old male is presented. This case provides a unique opportunity to review the Alrawi criteria for the histologic diagnosis of PACNS and establish/refine criteria specific to SVcPACNS. Generally, such criteria should feature: 1) an *intramural and lymphocyte predominant* infiltrate devoid of multinucleated giant cells; 2) *structural vessel alterations* lacking fibrinoid necrosis; 3) *perivascular pathology* supportive of an angiocentric process; 4) the *absence of encephalitis*; and, 5) the *absence of a concurrent systemic or rheumatic illness* that could account for the CNS findings.

Abstract A2

Implementation of the 2016 WHO CNS Classification for infiltrating glioma: the VGH experience

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The classification system for gliomas has undergone significant revisions in the last several years, with the incorporation of molecular criteria and removal of mixed histologic diagnoses. Large-scale molecular studies have elucidated the biological characteristics of low grade gliomas, and enabled grouping based on IDH mutational and 1p19q codeletion status that outperforms histology in predicting patient outcomes. Mutations in *ATRX* and *TP53* are largely mutually exclusive of 1p19q- codeletion in the context of mutant IDH, and are useful in screening patients for further molecular studies.

At our institution, new testing methods for 1p19q codeletion and ATRX were implemented in 2014, and in this presentation we review all cases submitted for 1p19q testing since this time. In comparing histologic to molecular diagnoses, the majority of histologic oligodendrogliomas indeed have 1p19q codeletion, while oligoastrocytomas and GBMOs largely are re-classified as astrocytomas and glioblastomas, respectively. We have also found that loss of ATRX nuclear expression associated with *ATRX* mutation is highly indicative of 1p19q retention, however the immunohistochemical test can be challenging to interpret and there have been a small number of discordant results.

Abstract A3

Anaplastic ependymomas with ganglionic differentiation

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Anaplastic ependymomas are relatively uncommon, WHO grade III tumours that can occur in any location of the central nervous system. Low, as well as high grade tumours may show an additional component, most frequently cartilage or bone. Ganglionic differentiation has been demonstrated in only very few cases, usually young individuals. The purpose of this communication is to describe six examples of anaplastic ependymomas with a definite ganglionic component. All tumours were supratentorial and occurred in adults, 36-81 years of age. With the exception of one with a cystic component, all showed a diffuse MRI pattern and variable enhancement. All tumours displayed necrosis, vascular proliferation and marked pleomorphism, due to a mixture of epithelioid, giant, rhabdoid and gemistocytic, as well as clear and undifferentiated cells. Immunohistochemistry revealed reactivity for EMA (often atypical), GFAP and markers of neuronal differentiation, usually synaptophysin and chromogranin. All displayed high MIB-1 and occasional P53-positive nuclei. Two cases showed sarcomatous differentiation with desmin, smooth muscle actin and very rich reticulin staining. Electron microscopy revealed "zippering" intercellular junctions, basal bodies and cilia. Neuronal differentiation was expressed by neurosecretory granules and/or rich endoplasmic reticulin, large nuclei with nucleoli, and rare neuritic processes with microtubules. Differentiation of these rare tumours from glioblastomas might be important for the future development of tumour-specific molecular therapies. Electron microscopy is highly recommended for correct diagnosis of atypical variants of Anaplastic Ependymoma.

Abstract A4

Extracranial invasion of a recurrent, transformed anaplastic pleomorphic xanthoastrocytoma: a case report

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Pleomorphic xanthoastrocytoma (PXA) is a rare tumour comprising <1% of all primary central nervous system tumours and the majority (~98%) occur supratentorially. We report on a 40-year-old female with a past medical history of a rare posterior fossa/cerebellar PXA who presented with a right-sided neck mass, decreased shoulder power and longstanding right tongue

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