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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doi:10.1017/cjn.2018.37

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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doi:10.1017/cjn.2018.38

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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doi:10.1017/cjn.2018.39

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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doi:10.1017/cjn.2018.40

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

deviation with right-sided hemi-atrophy. The patient had prior tumour debulking. Recent MRI demonstrated an enhancing posterior fossa mass extending to the skull base at the jugular foramen and another mass in the upper neck along the jugular bulb with displacement and encasement of the right common carotid artery down to C5. Resection of the neck mass reveals an anaplastic PXA. The tumour has close approximation with adjacent peripheral nerves and is positive in 2 lymph nodes. Comparison with the original tumour molecular and immunohistochemical profiles reveals a conserved BRAF V600E mutation but the transformed malignant glioma now expresses dot-like EMA positivity and ATRX is completely lost (mutated). Transformation of a PXA (WHO Grade II) into an anaplastic PXA (WHO Grade III) has been well documented, but extracranial extension is extraordinarily rare. We report herein the first documented case of a posterior fossa PXA that underwent malignant transformation and extracranial invasion to the parapharyngeal space.

Abstract A5

Contribution of chromosome 9p deletion and Bcl1, Myc and p16 immunohistochemistry to the characterization of oligodendrogliomas

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doi:10.1017/cjn.2018.41

Molecular studies suggest that anaplastic oligodendrogliomas (OIII) can be subdivided into clinically relevant subgroups. We analyzed a retrospective series of 40 consecutive OIII operated at our institution and compared them to 10 grade II oligodendrogliomas (OII). Chromosome 9p status was compared to Bcl1, Myc and p16 expression by immunohistochemistry, clinical and histological data, and to event free survival (EFS) and overall survival (OS).

Chromosome 9p deletion was observed in 55 % of OIII (22/40) but not in OII, and correlated with both OS and EFS. Bcl1 expression was significantly higher in OIII (45 % versus 14% for OII) and correlated with MIB-1 expression, vascular proliferation, tumour necrosis and a shorter EFS. Myc expression was correlated with histologic grade (27% in OII, 35% in OIII) and to a shorter EFS in chromosome 9p non-deleted OIII. p16 expression was not correlated with grade but revealed two distinct expression profiles according to chromosome 9p status. In 9p non-deleted oligodendrogliomas, p16 hyperexpression was correlated with shorter OS in both OII and OIII whereas absence of p16 expression was correlated with shorter OS and EFS in 9p deleted OIII.

Abstract A6

Eosinophil infiltrates in astrocytic tumors

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doi:10.1017/cjn.2018.42

Eosinophils may affect each stage of tumor development. The presence of eosinophils appears to correlate with longer patient survival in several non-CNS malignant tumors. Tumor-associated tissue eosinophilia (TATE) has been increasingly recognized. Two previous studies (Hayes RL, *et al.* 1995; 2001) revealed eosinophils in the intracavitary tissues of malignant gliomas following the infusion of IL-2 combined with activated autologous killer cells, but not in the primary operative specimens. Our recent study (Lu JQ, *et al.* 2014) demonstrated the infiltration of eosinophils in 19 of 44 pilocytic astrocytomas but not in 10 ependymomas. In the present study, we examined 7 cases of subependymal giant cell astrocytoma (SEGA; 4 with tuberous sclerosis; age range: 13 – 33 years; 4 males and 3 females), as SEGA is well known to contain infiltrating mast cells and lymphocytes. Five of 7 SEGA contained eosinophils that were focally scattered to rare in frequency; intratumoral and/or perivascular in location. In comparison, only one of 8 consecutive cases of glioblastoma showed infiltrating eosinophils. The incidence of TATE is significantly higher in SEGA than that in glioblastoma (71.4 % versus 12.5%; p=0.04, by Fisher's exact test) but is not significantly different from that in pilocytic astrocytoma (71.4 % versus 43.2%, previously reported; $p = 0.232$). Our findings support the concept that eosinophils may play a functional role in the development of astrocytic tumors, especially those with longer patient survival.

Abstract A7

Fetal neuroaxonal dystrophy: a new etiology of fetal akinesia

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doi:10.1017/cjn.2018.43

Neuroaxonal Dystrophies (NAD) are neurodegenerative diseases characterized by axonal "spheroids" occurring in different age groups. The identification of mutations delineated new molecular entities in these disorders. We report neuropathological data of a new form of NAD, characterized by a precocious prenatal onset, different from classical and connatal Infantile Neuroaxonal Dystrophy (INAD).

We studied 5 fetuses examined after pregnancy termination and 2 term neonates deceased just after birth, 4/7 from consanguineous parents. All subjects presented severe fetal akinesia sequence with microcephaly. In 4/7 cases, a molecular study was performed. In all cases, "spheroids" with typical immunohistochemical features were identified, with variable spreading in the central and peripheral nervous system. Basal ganglia, brainstem, cerebellum and spinal cord involvement was constant. Associated CNS malformations, unusual in INAD, were associated including hydrocephalus (2), callosal agenesis/hypoplasia (2), olfactory agenesis (1), cortical (3) and retinal (1) anomalies. None cases demonstrated mutations in *PLA2G6,* found in INAD.

The clinical and neuropathological features of these fetal cases are different from those of "classical" INAD. The absence of