

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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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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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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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

mutations of *PLA2G6*, in addition, suggests that the fetal NAD is a new entity, distinct from INAD, with different molecular basis. Associated malformations suggest a wide phenotypic spectrum and probable genetic heterogeneity. Finally, fetal NAD is an additional etiology of fetal akinesia.

**ABSTRACT A8****Motor neuron disease presenting with fetal akinesia**

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By contrast to infantile spinal muscular atrophy, which usually links to deletions in the SMN genes, fetal onset motor neuron disease is poorly reported. We collected a series of twelve cases of fetal arthrogryposis (16-31 weeks gestational age) with fetal motor neuron disease and excluded infectious diseases, lysosomal storage disease and neuroaxonal dystrophy. Of these twelve, 3 were thought to be ischemic in nature with microvascular alterations and systemic or central nervous system ischemic injury. The remaining 9 all displayed marked reduction in anterior horn motor neurons. Of these 9, four demonstrated mineralised neurons, four demonstrated either neuronal loss or cavitation in the globus pallidus, and in two, degenerating neurons were detectable in the brainstem or globus pallidus. Specific sequencing of SMN1 was performed in 6 of 9 and was reported as normal. Whole exome sequencing was performed in 4 without definitive diagnosis. We conclude that fetal motor neuron disease can be distinguished from ischemic injury, is morphologically heterogeneous, may affect the globus pallidus and is rarely linked to SMN1 mutations.

**ABSTRACT A9****The central nervous system lesion in amniotic rupture sequence**

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We review the central nervous system anatomy in nine cases of amniotic rupture sequence, all of which had neuropathological examinations. Of these, four had normal brains, and in none of these was the cranial vault involved, and one had cleft lip and palate. Of the remaining five, all had portions of the scalp, calvarium and dura replaced by amniotic membrane directly overlying arachnoid. In one, the membrane covered a narrow necked large encephalocele, and the contained brain demonstrated extensive disruption and degeneration. In the remaining four, one demonstrated cranioplacental adhesion, and in three there was a broad based encephalocele covered in large part by amnion. Two of these four cases demonstrated holoprosencephaly. One case with holoprosencephaly and one without demonstrated marked aqueductal stenosis, and two of the four demonstrated aqueductal occlusion or near occlusion by neuroglial excrescences. None demonstrated ventriculomegaly. Three of these four cases demonstrate varying degrees of mechanical distortion and

secondary pathology. We conclude that brains with amniotic rupture sequence demonstrate both malformation and deformation, which likely points to the embryonic stage origin of the lesion.

**ABSTRACT A10****Chronic traumatic encephalopathy in contact sports: The Canadian experience**

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Chronic traumatic encephalopathy (CTE) is suggested to be a progressive neurodegenerative disease, characterized by tau deposits in the depth of cortical sulci in neurons and in glioneuronal complexes around blood vessels. Few studies have suggested that it is caused by multiple concussions or subconcussive brain injuries. A recent publication showed that most American football players whose brain were donated to the Boston University concussion center had CTE (Mez et al. 2017). Over the last 6 years, with the help of neuropathologist colleagues across Canada, we have collected the brains of 33 high level professional and amateur athletes. These include 5 National hockey league (NHL) players, 15 Canadian football league (CFL) players, 3 College football players, 3 College hockey players, 2 professional boxers, 1 professional bull rider, 1 BMX champion, 1 rugby player and 2 skiers. All were male and the ages ranging from 15 to 87 years. Our results indicate that only a small portion of cases have CTE. Moreover, most cases are low stage (stage 1 or 2) and this pathology is mainly seen in some of the younger players. Older players either have no pathological findings or have other neurodegenerative diseases such as Alzheimer's disease. The disparity of results between the 2 groups will be discussed.

**ABSTRACT A11****Executive dysfunction and altered cerebrovascular activity in a rodent model of vascular cognitive impairment**

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Most basic science research has focused on overt stroke caused by blockage of major blood vessels. Less attention has been paid to small vessel disease giving rise to covert stroke that often leads to vascular cognitive impairment (VCI). One reason for this may be the relative lack of relevant animal models. This talk will describe a model of VCI induced in middle-aged Sprague-Dawley rats exposed to a diet high in saturated fats, salt and refined sugar (HFSS). In Experiment 1, rats fed HFSS and subjected to a small mediodorsal (MD) thalamic stroke with or without concomitant cerebral hypoperfusion experienced significant executive dysfunction. In Experiment 2, dietary