

## Neuro-oncology

## PP3220

**Disseminated cystic lesions: a case of disseminated oligodendroglial-like leptomeningeal tumor**

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**Introduction:** Disseminated oligodendroglial-like leptomeningeal neoplasms are an unusual presentation of oligodendroglial tumors and mostly seen in pediatric patients. Disseminated cystic lesions are mostly seen along the surface of the posterior fossa especially cerebellum. Brain stem, medial and inferior of temporal and frontal lobes are also affected. Spinal cord involvement is common in most of the cases. We present an atypical case of disseminated oligodendroglial-like leptomeningeal tumor with an older onset and preserved spinal cord parenchyma.

**Methods:** EEG, brain and spinal MRI, brain biopsy were performed for diagnosis.

**Results:** A 22-year-old female referred to our hospital after having two generalized tonic clonic seizures and auras of smell and nausea. Seizures that started two months before admission, lasted 2 minutes with tonic posture on her right hand and deviation of head and eyes. Brain MRI showed multiple hyperintense cystic and nodular lesions on T2 weighted images. Some of them had contrast enhancement on T1 weighted images. Spinal MRI was normal. CSF evaluation was normal, except for the elevated opening pressure. Pathological evaluation showed a nodular lesion consisting of oligodendroglial-like cells which were negative with NeuN, synaptophysin, pNF, GFAP, CD34, Ki-67, IDH-1, p53 staining and mostly resembled disseminated oligodendroglial-like leptomeningeal tumor morphologically. The patient is stable with antiepileptic treatment.

**Conclusions:** Disseminated oligodendroglioma-like tumors are a rare form of low-grade glial tumors. Differential diagnosis includes parasitic infections and other glial and sarcomatous neoplasms with leptomeningeal involvement. Radiological and pathological evaluations are essential. Diagnosis is difficult because of the imprecise pathological characteristics of the neoplasm.

**Disclosure:** Nothing to disclose

## PP3221

**Paraparesis and sciatic pain caused by bilateral sacral solitary fibrous tumors: when the least plausible cause is the diagnosis**

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**Introduction:** Solitary Fibrous Tumors (SFT) are rare soft-tissue tumors of pleural or, less often, extrapleural location. Although sciatic pain is very common, extraspinal compression of the nerve is extremely rare.

**Results:** A 44-year-old man, healthy apart from a totally resected left pelvic SFT in 2010, presents at the emergency department with bilateral sciatic pain and gait disorder. He had progressive sciatic pain at the right for eleven months, and left for one month, that aggravated walking and sitting. Numbness and tingling of the right foot for 6 months. One month before, he developed constipation and effortful micturition, after a week, paresis of the right foot and leg, and later also his left foot. Neurological examination disclosed an asymmetric paraparesis (Right leg grade 2, left leg grade 4 MRC), normal deep tendon reflexes apart from abolished right aquilian reflex and indifferent right plantar response. He also showed reduced tactile and algic sensation distally in both legs. At this point cauda equina syndrome due to compressive lesion was suspected and spinal MRI was performed. With a normal MRI, an extraspinal cause was pursued, pelvic CT showed bilateral sacroiliac lytic mass lesions. Histology was compatible with bilateral SFT recurrence and the patient was oriented to chemotherapy and radiotherapy.

**Conclusions:** To our knowledge this is the first report of sciatic pain as manifestation of SFT. Progressive compression of the sacral plexus and sciatic nerve in the thigh was responsible for the symptoms and its appropriate investigation allowed the diagnosis.

**Disclosure:** Nothing to disclose

## PP3222

**Primary central nervous system lymphoma with diffuse lesions in an immunocompetent patient**A. Dulamea, A. Buture, E.A. Solomon*Fundeni Clinical Institute, Bucharest, Romania*

**Introduction:** In immunocompetent patients, lesions of primary central nervous system lymphoma (PCNSL) are usually solitary, located in a cerebral hemisphere, thalamus/basal ganglia, corpus callosum, periventricular region and cerebellum. Authors report the case of an immunocompetent patient with an unusual aspect of lesions.

**Methods:** A 61-year-old woman presented a vestibular syndrome. Two weeks later behavioral changes appeared, apathy, Millard Gubler syndrome, then tetraparesis, right ataxia, aphasia, urinary incontinence. Brain MRI showed: multiple lesions with hyperintensity on T2/FLAIR, isointensity on T1, with moderate water restriction in diffusion sequences, some of them with moderate contrast enhancement, imprecisely delimited, located in bilateral fronto-insular periventricular white matter, corpus callosum, bilateral capsulo-lenticular, right caudat nucleus, with nodular appearance in the anterior pole of frontal lobe, left midbrain, right pons, left superior cerebellar peduncle. The differential diagnosis was made between: acute disseminated encephalomyelitis, lymphoma, gliomatosis cerebri, viral encephalitis and prion disease.

**Results:** CSF examination showed: 7 monocytes/mm<sup>3</sup>, normal glucose and protein, protein 14-3-3 and oligoclonal bands were negative. Blood biochemistry and CBC were normal. HIV, cytomegalovirus, Epstein Barr virus and herpes simplex virus serology were normal. Bone marrow aspirate was normal. Corticosteroids produced temporary clinical remission. The brain stereotactic biopsy established the diagnosis: non Hodgkin lymphoma with large B cells, CD20 positive. Spinal cord MRI, contrast enhanced CT of chest, abdomen and pelvis, and dilated eye examination were normal. Methotrexate, idarubicin and cytarabine produced partial remission.

**Conclusions:** This case showed that diffuse lesions may be also found in immunocompetent patients with PCNSL.

**Disclosure:** Nothing to disclose

## PP3223

**Multiple myeloma relapse presenting as meningeal myelomatosis**A. Caetano<sup>1</sup>, D. Zhang<sup>2</sup>, C. Sepúlveda<sup>3</sup>, A. Reichert<sup>4</sup>, M.D.L. Guerra<sup>5</sup>, L. Alves<sup>1</sup>

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**Introduction:** Leptomeningeal infiltration by monoclonal plasma cells is a rare clinical presentation in patients with multiple myeloma (MM), occurring in around 1% of such patients.

**Case report:** We report the case of a 70-year-old male patient, diagnosed in early 2012 with Durie-Salmon Stage III A non secretory MM that presented as a sacral plasmacytoma. He underwent radiotherapy, four chemotherapy cycles with cyclophosphamide, bortezomib and dexametasone, followed by autologous bone marrow transplant, with very good response. He relapsed two months later, needing another 6 chemotherapy cycles, with adriamycin, carmustine, cyclophosphamide and melphalan, having good response with disease control (marrow samples with 1.2% plasmacyte count). One week after the last cycle, the patient presented with a progressive encephalopathy, gait abnormalities and a right third cranial nerve palsy. The head CT scan was normal, excluding space occupying or lytic lesions. A lumbar puncture revealed mild pleocytosis (227 leucocytes/microL) with predominantly aberrant plasmacytes (around 96%, CD38+). He was diagnosed with meningeal myelomatosis and started intrathecal chemotherapy.

**Conclusions:** MM presenting as meningeal myelomatosis is a rare neurological entity. It is usually associated with multifocal (and variable) neurological deficits and should be suspected in patients with MM even without evidence of intracranial masses or cranial intraparenchymal infiltration. The treatment options include intrathecal chemotherapy but the prognosis is generally very poor.

**Disclosure:** Nothing to disclose

## PP3224

**Two cases of chronic polyradiculoneuritis revealing asymptomatic multiple myeloma**

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**Introduction:** The spectrum of multiple myeloma's (MM) neurological manifestations is diverse including mainly the spinal cord compression. The involvement of the peripheral nervous system (PNS) is rare. We report two cases with MM revealed by isolated chronic polyradiculoneuritis (CPRN).

**Case reports:** A 34 year-old woman (1) and a diabetic 66 year-old man (2), were admitted for progressive weakness and numbness of four limbs. Neurological examination showed quadriplegia with areflexia predominantly in the lower limbs. Electromyography and neuromuscular biopsy findings were consistent with demyelinating CPRN. Laboratory tests revealed a monoclonal IgG-lambda antibody. Cerebral spinal fluid examination showed increased protein level with normal cell count. Spinal MRI disclosed an osteolytic lesion the right iliac crest for both patients, associated with multiple locations on the vertebrae, and hepato and splenomegaly in one case 1. The diagnosis of MM was confirmed by the presence of a bone marrow plasma cell infiltration. Search for Bence Jones proteinuria and amyloidosis was negative in case 1. The treatment started with systemic chemotherapy.

**Discussion:** Involvement of the PNS is rare in MM. Axonal sensorimotor polyneuropathies are the most common, and usually associated to the osteosclerotic form. CPRN are exceedingly rare, but frequent with solitary plasmacytoma. The pathophysiology of these neuropathies remains obscure with numerous theories including amyloidosis, ischemia, toxic metabolic factors (circulating abnormal protein) and myelin antibodies. The management of the neuropathy consists in treatment of the tumor.

**Conclusions:** These observations emphasize the need for systematic testing for plasma cell proliferation in patients with unexplained lasting CPRN.

**Disclosure:** Nothing to disclose

## PP3225

**CNS disorders induced by radiation therapy of the brain**

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**Introduction:** CNS radiation therapy is commonly used to treat a variety of CNS tumors including primary and metastatic tumors of the brain or in an attempt to prevent the development of metastases like in small cell lung cancer. The treatment dose and volume depend on the histology, location and size of the tumor. Radiation administered to treat CNS neoplasms can result in acute, subacute, and delayed neurologic syndromes. Most important in clinical practice is radionecrosis which occurs 1 to 3 years after radiation. Symptoms depend on the location and size of necrotic lesion. Their presentation can mimic tumor recurrence. Cerebral radionecrosis often occurs after focal EBRT or brachytherapy.

**Methods:** This is a case study about 60-years-old man with initially squamous cell carcinoma of the lung in IIIA stage with complete pathologic response on the treatment of primary tumor and disease free survival of 2 years when he developed headaches accompanied by ataxia, right homonymous hemianopia and hemiparesis. Brain MRI revealed brain metastases in cerebellum, left occipital and temporal lobe. The whole brain RT (30Gy: 3Gy/10fr) was delivered with sequentially chemotherapy.

**Results:** The patient had 12 months disease free interval, then the same symptoms in more aggressive form occurred and brain MRI showed focal radionecrosis.

**Conclusions:** RT of the brain can lead to necrosis and the dose which gives a 5% probability of a given late effect 5 year after treatment for whole brain RT is 45Gy. Sometimes necrosis can occur with even lower doses which depends of endogenic factor.

**Disclosure:** Nothing to disclose

## PP3226

**Bing Neel syndrome mimicking Lyme neuroborreliosis**

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**Introduction:** Central nervous system (CNS) affection in Waldenström Macroglobulinemia (WM) is called Bing Neel Syndrome (BNS).

**Methods:** We report on a patient with peripheral paresis of the VII<sup>th</sup> cranial nerve.

**Results:** An 82-year-old man with ongoing WM was referred to our hospital due to intractable nocturnal pain in his legs. Clinical manifestation was a mild hemi paresis on the right and a painful sensory and motor polyneuropathy of both legs. CCT detected a small old left middle cerebral artery infarction. Cerebrospinal fluid (CSF) showed 400cells/ $\mu$ L. Penicillin G 20 MegaIE per day was given due to the assumption of CNS affection in Lyme disease (LD). Nocturnal pain improved but motor function of both legs worsened. Peripheral paresis of the VII<sup>th</sup> cranial nerve on the right, impairment of cognition and urinary retention developed within days. EMG displayed the active axonal demyelination mostly of motor fibers and MRI confirmed affection of the equine cauda. CSF FACS analysis showed 98% clonal CD 19<sup>+</sup>, CD20<sup>+</sup> and CD79<sup>+</sup> B-cells. CSF was negative for LD IgM, confirming BNS. The patient refused chemo or radiotherapy and died within two months.

**Conclusions:** Among the 35 BNS cases in English literature, impairment of cognition or vigilance, headache, body weakness and aphasia are frequent symptoms but mimicking of Neuroborreliosis was not reported before. Only FACS analysis of CSF confirmed the diagnosis of BNS in this case. Due to the pure prognosis of BNS, hematologists need to be alert to neurological symptoms in their patients.

**Disclosure:** Nothing to disclose

## PP3227

**Progressive bilateral ptosis in a patient with midbrain metastasis and chronic inflammatory demyelinating polyneuropathy**

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**Introduction:** A midbrain lesion is a rare but recognised cause of bilateral ptosis. This presentation in isolation, without vertical gaze involvement, is exceptionally rare, but may herald a sinister aetiology that should not be missed.

**Methods:** Case report of a 68 year old woman.

**Results:** A 68-year-old lady with a history of chronic inflammatory demyelinating polyneuropathy presented with progressive asymmetrical bilateral ptosis, but no change in her limb function. Brain MRI showed a midline midbrain lesion displacing the cerebral aqueduct. Chest CT scan revealed a left upper lobe mass, confirmed histopathologically as a poorly differentiated adenocarcinoma.

**Conclusions:** The clinical presentation of isolated bilateral ptosis resulting from a midline midbrain metastasis is rare but important. In this patient's case, it was essential to seek a central cause, even in the context of a plausible peripheral explanation.

**Disclosure:** Nothing to disclose

PP3228

Abstract withdrawn

PP3229

**Breast carcinoma presenting as paraneoplastic brainstem syndrome associated with anti-Ri antibodies**

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**Introduction:** Anti-Ri antibodies are onconeural antibodies mainly associated with breast and gynecological cancers and small cell lung carcinoma, being mostly associated with cerebellar degeneration and opsoclonus-myoclonus. Symptom onset often begins prior to the diagnosis of systemic cancer, such that identification of antineuronal antibodies may facilitate diagnosis.

**Clinical case:** Female, 49 years old, with a two-month history of diplopia and gait instability. On examination there was a right convergent squint, horizonto-rotatory nystagmus in lateral gaze and an unstable normal-based gait. Brain MRI was normal except for two small arterial aneurisms. Demyelinating, infective causes, vitamin deficiency, toxic exposure and celiac disease were excluded. CSF flow-cytometry was negative for malignant cells. Autoimmune screening showed positive anti-Ri antibodies, leading to the assumption of paraneoplastic syndrome. Steroid treatment was unsuccessful, and monthly intravenous immunoglobulin was then commenced, with slight improvement. All investigations in search for malignancy were unremarkable, except for breast echography which showed a small nodule in the superior-external quadrant of left breast. Breast MRI was subsequently performed, showing two small nodules in the same location. Core biopsy showed invasive ductal carcinoma grade II. The patient was submitted to conservative surgery followed by radiotherapy and chemotherapy, with progressive improvement since surgery and only slight residual gait imbalance.

**Conclusions:** This case illustrates that the diagnosis of paraneoplastic syndromes requires a high level of suspicion. Given the onset of symptoms can pre-date the diagnosis of systemic cancer, a thorough investigation is warranted, as treatment of underlying malignancy is the most effective step in controlling the neurological disorder.

**Disclosure:** Nothing to disclose



## PP3230

**Choroid plexus papilloma (4<sup>th</sup> ventricle) with spread leptomeningeal**

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**Introduction:** Choroid plexus papilloma (CPP) is a rare, histologically benign intracranial tumor, but may present local recurrence or leptomeningeal dissemination. In adults it's usually located in the fourth ventricle. Clinical manifestations result from increased intracranial pressure or hydrocephalus caused by obstruction of cerebrospinal fluid flow or increased production. We report a case of choroid plexus papilloma of the fourth ventricle with diffuse leptomeningeal seeding.

**Methods:** A 35-year-old woman, with a history of chronic migraine, presented with oppressive headache beginning two months before, and characteristics significantly different from their usual migraine crisis. Physical examination revealed a papilledema. Neuroimaging findings are described, treatment performed and histopathology.

**Results:** Neuroimaging studies showed a space-occupying lesion in the fourth ventricle and cystic lesions scattered throughout the subarachnoid space, affecting supratentorial, infratentorial and spinal regions. Completed resection and histopathology of the tumor, confirmed the origin: CPP, with disseminated metastases. After surgery, the patient is asymptomatic. Radiotherapy will be considered in the future.

**Conclusions:** The CPP dissemination is a rare event. An appropriate differential diagnosis and histological study are very important, to get a definitive diagnosis. Treatment with complete resection of the tumor is the best procedure and derivation of hydrocephalus, when present, must be considered. Adjuvant chemotherapy and radiotherapy have been demonstrated to increase survival in the treatment of choroid plexus carcinoma and may be indicated for aggressive disease.

**Disclosure:** Nothing to disclose

## PP3231

**Immunological findings in a patient with Surgicel granuloma (case report)**

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**Objectives:** The immune status of the patient with postoperative intra-cerebral aseptic granuloma 7 months after the removal of fibrillar-protoplasmic astrocytoma of the right frontal lobe was observed.

**Methods:** We have studied immune status of the patient with the determination of the content of CD4+/8+, CD16+, CD25+, CD38+, CD54+, CD95+, CD150+, antibodies to neuroantigen. Cell counts were measured using immunoenzymometric PAP method. Antibodies to neuroantigen studied by Dechtyarenko T.V. described method.

**Results:** We demonstrated a reduction in the immunoregulatory index CD4+/CD8+ to 1.72, significant increase in the absolute content of CD25+ to 719 cell/mcl, a reduction of the content of phagocytizing neutrophils to 1500 cell/mcl, a substantial increase in the absolute content of CD38+ to 754 cell/mcl, a significant increase in the level of antibodies to neuroantigen to 22. The amount of lymphocytes expressing molecule of the adhesion ICAM-1 CD54+ (565 cell/mcl) exceeded the normative indices significantly. We found an increase in the content of oncomarker level such as CD150+ to 712 cell/mcl. Furthermore, a quantity of CD95+ (502 cell/mcl) exceeded the normative indices two-fold.

**Conclusions:** For the first time we carried out a study to investigate the immune status of the patient with postoperative intra-cerebral aseptic granuloma formed 7 months after the removal of low grade astrocytoma of the right frontal lobe. The revealed changes describe the presence of the significant immunosuppressive effects, deep disturbances of the processes of intercellular interaction of immunocompetent cells, increased proapoptotic readiness of cells together with the presence of the expressed autoimmune aggression.

**Disclosure:** Nothing to disclose

## PP3232

**Malignant optic pathway glioma in adults with clinicopathologic and molecular features**

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**Introduction:** Malignant gliomas of the optic pathway are rare, and their genetic alterations are poorly known.

**Methods:** We report a 64-year-old woman with an anaplastic astrocytoma arising from optic pathway together with the molecular features.

**Results:** She presented with a progressive visual field loss and underwent a biopsy of mass lesion in optic chiasm. After receiving radiosurgery plus concomitant chemotherapy with Temozolomide, she remained stable without aggravation of her visual field defects for 10 months after her initial presentation. Histopathology revealed a hypercellular tumor composed of predominantly pleomorphic astrocytes displaying indecisive biphasic pattern. Molecular analysis by immunohistochemistry showed focal expression of MGMT and p53, whereas no expression was detected in EGFR or mutant IDH1. Sequence analysis revealed wild type IDH1, IDH2, and BRAF. BRAF-KIAA1549 fusion was not detected.

**Conclusions:** The presented molecular analysis did not show conclusive molecular changes that specifies glioma type, and which indicated that malignant optic gliomas in adults may share common molecular genetic features with conventional primary glioblastoma.

**Disclosure:** Nothing to disclose

## PP3233

**Ganglioglioma centered in the superior medullary velum**

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## PP3234

**A case of cerebral metastasis mimicking acute inflammatory demyelinating polyradiculoneuritis (Guillain-Barré syndrome)**

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## PP3235

**A hard to diagnose gliomatosis cerebri case**

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## PP3236

**Consecutive occurrence of two primary central nervous system tumors in the same localization**

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## PP3237

**Metastatic brain neuroendocrine tumor originated from liver misdiagnosed glioblastoma multiform. A first case report**

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## PP3238

**Cystic falx meningioma: a case report**

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## PP3239

**First described case of brain metastases heralding a squamous cell carcinoma of the supraglottic larynx**

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## PP3240

**A case of a patient with radiation anaplastic meningioma after X-ray therapy of anaplastic oligodendroglioma**

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## PP3241

**Brain tumor – is it so obvious to diagnose?**

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## PP3242

**Paraneoplastic autoimmune brainstem encephalitis: a case report**

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