Intracranial hypertension syndrome and multiple cranial neuropathy related to atypical radiologic presentation of an intradural chordoma

N. Oliveira, L. Vasconcellos, N. canedo, J. Esperanca. Neurology, Federal University of Rio de Janeiro (UFRJ), Rio de Janeiro, Brazil

Introduction: We report the unusual case of an intradural chordoma with atypical clinical and radiologic presentation.

Case report: 36 year-old male, report in 2011 the beginning of bilateral hearing loss associated with vertigo and followed by bilateral hyposmia, decreased visual acuity in the left eye and trigeminal autonomic headache for 5 months. The physical examination showed bilateral hyposmia, Foster–Kennedy syndrome in the right eye, hypoesthesia of the left face and bilateral deafness. Laboratory exams showed CSF with one cell, hyperproteinorachia (837 mg/dL), 210 erythrocytes/mL, glucose 60 mg/dL; VDRL, cryptococcal latex, culture for common germs, tuberculosis and cysticercosis all negative. Serology for histoplasmosis and paracoccidioidomycosis were also negative. MRI scan showed thickening of the meninges and multiple round lesions less than 1 cm each, hyperintense in T2 and FLAIR, well circumscribed, under the dura and around the cerebellum, intraparenchymal in some areas, without Gadollinium enhancement. Biopsy of the lesion showed physaliphorous cells embedded in mucin matrix, suggesting an intradural chordoma. Follow-up showed no improvement of the neurological symptoms, but the patient decided not to go under any new surgeries. A few months after the surgery, the patient died of the disease.

Discussion/conclusion: Intradural chordoma is a rare entity, with still controversial treatment regimen. This case report shows this rare entity with an even rarer radiologic presentation, posing a tricky differential diagnosis and serves as an alert for the issue.

doi:10.1016/j.jns.2015.08.269

Primary intramedullary glioblastoma multiforme of the spinal cord: the uncommon disease — 55 years of experience

Dept of Spine Surgery, National Institute of Clinical Neurosciences, Budapest, Hungary; Dept of Neurosurgery, Semmelweis University, Budapest, Hungary; Dept of Pathology, Semmelweis University, Budapest, Hungary

Background: Primary glioblastoma multiforme (GBM) in the spinal cord is a very rare entity and carries a very dismal prognosis. There is a short time from onset to the time of diagnosis and is followed by progressive neurological deterioration and death.

Objective: The authors report ten cases of GBM treated between 1959 and 2015 and discuss the clinical and diagnostic features and treatment possibilities.

Patients and methods: Patients were evaluated earlier with myelography, later with computer tomography used either with or without myelography. The last 20 years the magnetic resonance imaging (MRI) became the diagnostic procedure of choice.

Results: GBM was proved histologically in all patients. A median survival time from operation to death was 7.5 months (from onset of symptoms until death was 12 months). Early results after surgery were as follows: four improved, three unchanged, three worsened. The radio-chemotherapy group survived significantly longer (15 months).

Conclusion: According to literature we suppose that the treatment is primarily debulking of the tumour, but it cannot be radical in most of the cases. When surgery is followed by irradiation and chemotherapy, the survival time can be improved. Since spinal GBM is a very rare lesion, there is still not enough experience with its treatment strategy, but postsurgical radio-chemotherapy seems to be more effective.

doi:10.1016/j.jns.2015.08.270

Intracranial hemorrhage in setting of glioblastoma with venous thromboembolism

G. Stevens, M. Khoury, M. Syneon, S. Sakrut, N. Edwin, A. Khorana, G. Barnett, D. Peereboom, M. Ahluwalia. Brain Tumor and Neuro-oncology Center, Cleveland Clinic, Cleveland, USA; Taussig Cancer Institute, Cleveland Clinic, Cleveland, USA
Background: Venous thromboembolism (VTE) is a complication of glioblastoma. Anticoagulating patients with glioblastoma carries a theoretical risk of intracranial hemorrhage (ICH).

Methods: We performed a retrospective review of consecutive glioblastoma patients (2007–2013) diagnosed with VTE.

Results: 173 (33.1%) of 523 glioblastoma patients had VTE events. Seventeen (10%) had ICH: 6 (35%) subdural hematomas and 11 (65%) intratumoral hemorrhage. In total, four patients with ICH required neurosurgical intervention. Enhancement in the area of subsequent intratumoral hemorrhage was noted in 9 of 10 with available pre-ICH scans. Multivariable regression did not show associations between ICH and tumor enhancement diameter or vascular-endothelial-growth-factor inhibitor use. Fifteen (16%) patients receiving anticoagulation had ICH compared to 2 (2.6%) not receiving anticoagulation (p = 0.005). The type of anticoagulant used was not associated with development of ICH. Median survival from non-distal VTE diagnosis to death were 8.0 and 3.5 months (p = 0.05) in patients receiving anticoagulation and those not on anticoagulation, respectively.

Conclusion: Patients with glioblastoma and VTE on anticoagulation have increased incidence of ICH. However, development of ICH was not associated with lower median survival from time of VTE. Intratumoral hemorrhage occurred within the enhancing portion of tumor; however, no relationship was identified between the development of ICH and (i) the median diameter of enhancement or (ii) type of anticoagulant used. However, patients with absence of enhancing tumor did not have intratumoral bleed suggesting a gross total resection may limit this adverse outcome.

doi:10.1016/j.jns.2015.08.271

---

A chilean case of chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids (CLIPPERS): case report

A. Galilea izquierdoa, F. Muñoz, R. Valenzuela, R. Gejman, M. Raue.

aNeurología, Hospital Clínico Universidad Católica, Santiago, Chile; bAnatomia Patológica, Hospital Clínico Universidad Católica, Santiago, Chile

Chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids (CLIPPERS) is an inflammatory disorder that involves predominantly the brainstem. The specific etiology is unknown and a wide differential diagnosis is needed. It has a characteristic MRI appearance with punctate and nodular enhancing lesions in pons, cerebellum and brachium pontis. The biopsy reveals perivascular lymphocytic infiltrate. It typically has a marked response to glucocorticoids, but with frequent relapses after tapering. A 44 year old woman was admitted on July 2014 with a history of subacute gait ataxia, left facial hypoesthesia, followed by left brachiocrural paresis, paresthesia, dysphagia and dysarthria. MRI showed multifocal infiltrative lesions involving pons and bilateral cerebellum. Cerebrospinal fluid analyses revealed increased protein levels and intrathecal IgG synthesis. Rheumatologic study and thorax–abdomen and pelvis CT were normal. PET-CT showed hypermetabolic lesions in posterior fossa. Lesion biopsy showed a polyclonal perivascular lymphoid infiltrate with CISH negative for Epstein–Barr. After 5 consecutive daily boluses of 1 g methylprednisolone she had a dramatic improvement that has continued after a slowly tapering of steroids, plus azathioprine. Control MRI after 3 month showed partial regression of lesions.

doi:10.1016/j.jns.2015.08.272

---

Down-regulation of ribosomal protein S15A inhibits proliferation of human glioblastoma cells in vivo and in vitro via AKT pathway

Y. Xu, Y. Liu, Y. Yao. Neurosurgery, First Affiliated Hospital Dalian Medical University, Dalian, China

Background: Ribosomal protein s15a (RPS15A), a highly conserved cytoplasmic protein, promotes mRNA/ribosome interaction in translation. Recent evidence showed that RPS15A would be essential for tumor cell growth.

Objective: The aim of this study was to investigate whether RPS15A has prognostic relevance in human glioblastoma.

Material and methods: Levels of RPS15A expression were measured in glioblastoma and normal brain (NB) tissues samples. Stably decreased expression of RPS15A was established by the pLVTHM-GFP lentiviral RNAi expression system. Molecular mechanisms and the effect of RPS15A on cell growth and migration were investigated using MTT assay, wound healing assay, transwell migration assay, Flow cytometry analysis, western blot assay,
immunofluorescence double labeling and tumorigenesis in nude mice.

**Results:** Here we report that RPS15A is specifically overexpressed in human glioblastoma tumor tissues, and RPS15A knockdown inhibits proliferation and migration of glioblastoma cells in vitro. Lentivirus, transduced with shRPS15A, lead to depression of phosphorylated AKT, cell cycle arrest in G0/G1 phase in U87 cells, and also reduce expression of cell cycle regulation protein CDK4 while enhancing expression of p18\textsuperscript{INK4C}. Furthermore, the growth of glioblastoma cell-transplanted tumors in nude mice is inhibited by transduction with Lv-shRPS15A.

**Conclusion:** Our findings indicate that RPS15A promotes cell proliferation and migration in glioblastoma for the first time. RPS15A might play a distinct role in glioblastoma and serve as a potential target for therapy.

doi:10.1016/j.jns.2015.08.273