RARE-11. PRIMARY INTRACRANIAL LEIOMYOSARCOMA IN A PATIENT WITH NEUROBROMATOSIS TYPE 1

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Primary intracranial leiomyosarcoma (LMS) is very rare, with only a few reported cases. Only one prior case report of intracranial LMS in a patient with neurofibromatosis type 1 (NF1) was identified. We report a case of primary intracranial LMS with NF1. Our patient is a 17-year-old female without history of immunocompromise presenting with severe headaches representative of right frontal hemorrhagic tumor found to be primary intracranial LMS. In prior reported cases, most primary intracranial LMS were treated with sarcoma chemotherapy and radiation therapy. Our patient underwent multiple resections, as well as focal radiation. Her chemotherapy initially included ifosfamide, carboplatin, and etoposide, but when she failed etoposide twice due to severe allergic reaction, she completed treatment successfully with the combination of ifosfamide and doxorubicin. She continues to be doing well with no evidence of disease at 41 months post-treatment.

RARE-12. PITUITARY ADENOMA SURGERIES IN COV-19 ERA: EARLY LOCAL EXPERIENCE FROM EGYPT

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Background: The pandemic of COVID-19 has a great impact on all health-care services worldwide. Neurosurgical recommendations are to postpone the endoscopic endonasal pituitary surgeries during the pandemic. We would like to express our experience with urgent pituitary adenomas during the current COVID-19 pandemic. Methods: In our country, COVID-19 has started to become a paramount problem by March 2020. Nine cases of pituitary adenomas have presented with urgent manifestations. The endoscopic endonasal approach was performed in eight patients, while a cranialotomy was selected for a recurrent pituitary adenoma. Pre- and postoperative thorough clinical evaluations with chest CT scans were performed. Other strict infection control measures have been applied. Results: In 8 weeks duration starting from the past days of February 2020, we have operated on four females and five males of pituitary adenomas. Visual deterioration was the main presenting symptom. The driving factor for surgery was saving vision in eight patients. Fortunately, the postoperative course was uneventful for all patients. No suspected COVID-19 infection has been reported in any patient or health-care team except one patient. In our city, PCR test was routinely not available. Conclusion: In the era of COVID-19, strict infection control precautions should be employed to limit the possibility of transmission of any possible infection to patient or any of the surgical team. We believe that the risk of getting such infection is not increased by the endonasal approach. Long-term follow-up and large numbers of prospective studies are recommended to delineate the impact of COVID-19 infection on pituitary surgeries.

RARE-13. PRIMARY EMPTY SELLA SYNDROME AND PSEUDO-TUMOR CEREBRI: CORRELATION OF ASSOCIATION AND SURGICAL PLAN

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Introduction: Management of primary empty sella syndrome (ESS) is generally remaining a neurosurgical challenge due to lack of a well-standardized approach. Pseudo-tumor cerebri (Benign or idiopathic intracranial hypertension) is commonly associated condition. In this study, we have demonstrated the relationship and surgical plan and outcome of such cases. Methods: We retrospectively study Case Data of primary empty sella syndrome (ESS) for two years who were diagnosed radiologically as ESS. Fundus and other ophthalmological examinations were done. Lumbar puncture and cerebrospinal fluid (CSF) manometry was done in all patients. All patients’ data were collected and analyzed. Results: Basically, 24 patients (18 females and 6 males) were radiologically diagnosed as EES. 13 females and only one male were having symptoms of BH. 17 patients (70.83%) had headache as the first presentation. Second most common presentation in our study was visual in 14 patients (58.3%). Two patients (8.3%) had pituitary hypersecretion namely growth and prolactin hormones. In those (58.3%) confirmed to have BH Theco-peritoneal shunts were inserted. Incidental cases (29.17%) without symptoms were followed up. Conclusion: Although (ESS) is a well-known radiological hallmark for BH, not much is understood about the pituitary hypersecretion. Interestingly, pituitary hyperfunction may be the first presentation in some rare cases. Generally, natural history of that entity was benign. Frequent follow-up by neurosurgeons and increased awareness of associations are advised. We believe a more prospective large number cohort is important to outline the natural history.

RARE-14. DISRUPTION OF GEMC1-MCIDAS MULTICILIATION PROGRAM PROMOTES CHOROID PLEXUS CARCINOMA

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Tumors of the choroid plexus (CP) are rare primary brain neoplasms mostly found in children. CP tumors exist in three forms: CP papilloma (CPP), atypical CP, and CP carcinoma (CPC). Though CPP is more benign, only a few cases of CPC have been reported, with a high mortality rate and a tendency for recurrence and metastasis. CPC tumors are thought to arise from CP epithelial cells that secretes cerebral spinal fluid and generate multiple cilia on their apical surface. Here we show that aberrant NOTCH and Sonic Hedgehog signaling in mice drive CPC tumors that resemble CPC in humans. In contrast to CP epithelial cells with clusters of multiple cilia, NOTCH-driven CP tumors were monociliated, and disruption of the NOTCH complex restored multiciliation and decreased tumor growth. NOTCH suppressed multiciliation in tumor cells by inhibiting the expression of Geminin Coiled-Coil Domain Containing 1 (GEMC1), and multiciliate differentiation and DNA synthesis associated cell cycle protein (MCIDAS), early transcriptional regulators of multiciliated cell (MCC) differentiation. Consistently, Gemi1/Mcidas deficiency led to a lack of MCCs in the CP, and impaired the correction of the multiciliation defect in tumor cells by a NOTCH inhibitor. Disturbances to the GEMC1 program are commonly observed in human MCCs characterized by solitary cilia and frequent somatic TP53 mutations. Accordingly, CPC driven by deletion of tumor suppressors Tip33 and RB1 exhibits a cilia-focussed consequence to loss of Gem1/Mcidas expression. Taken together, these findings reveal that the GEMC1-MCIDAS multiciliation program in the CP is critical for inhibiting tumorigenesis, whereas a defective multiciliation program promotes CPC and may represent a therapeutic avenue for this cancer.

RARE-15. THE MOLECULAR PROFILE OF SECONDARY MENINGIOMAS IN SURVIVORS OF CHILDHOOD NON-CENTRAL NERVOUS SYSTEM CANCERS

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Introduction: Cranial irradiation remains part of childhood cancer therapy and secondary meningiomas are a late effect. Secondary meningiomas are reported in patients who received low and high dose cranial irradiation and arise >20 years post exposure. The molecular and genetic profile of primary meningiomas as has been well studied; however, only a few studies have been unique in radiation-induced meningiomas (RIM). Methods: We identified patients followed up at the Childhood Cancer Survivor Clinic, Stollery Children’s Hospital who had a history of non-central nervous system malignancies and received cranial irradiation who developed brain tumors between clinic inception in 1971 and June 2013. Whole exome sequencing (WES) as well as DNA methylation profiling were performed for patients where tumor and germline DNA were available. Results: Of 96 patients who received cranial irradiation, 16 (16.7%) developed meningiomas. The 16 patients included 15 with primary tumors in the brain and one with a primary tumor in the spine. All 16 patients received 2000–2400 cGy, suggesting a threshold dose. 9/16 (56%) had WHO Grade 2 meningiomas or greater and 7/16 (44%) were infiltrative. Post-surgical recurrences occurred in 43%. Patients experienced considerable morbidities directly attributable to the meningiomas or their treatment. 14 patients had samples suitable for further analysis. Preliminary