Tumours of the CNS – Orbital and Skull Base Tumours

**P-5-634** Lymphocytic hypophysitis in a 13-year-old girl

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The lymphocytic hypophysitis is an infrequent disease, usually seen in woman during the postpartum period. Up to date no cases of this disease in patients less than 20 years of age have been published. Herein we describe a 13-year-old girl with normal menses until the beginning of the symptomatology. She developed abruptly a severe headache with nausea and vomiting. Later on she developed diplopia. On admission she was acutely ill, physical exam. revealed a right convergent strabismus, reactive pupils, breast stage Tanner 5 with galactorrhea and pubic hair stage 5. A MRI revealed an intrasellar mass with moderate suprasellar extension, with contrast-enhancement with gadolinium. Lab: Sed rate: 61 mm (1st. hour), estradiol: 11 ng/mL, LH: 1.8 mIU/mL, FSH: 2.0 mIU/mL, PRL: 72.7 ng/mL, T4: 5.2 μg/dL TSH: <0.5 μIU/mL, cortisol: 14.2 μg/dL, IGFB: 3.0 U/mL, CFN was normal.

She underwent a transphenoidal removal of the mass. Histology revealed a lymphoplasmocytic infiltration of the anterior and posterior pituitary gland, with few eosinophils, within pervascular areas and surrounding nests of atrophic adenohypophysis. The clinical, endocrinological and histological patterns of the Lh described in our patient, are similar to those reported in adult patients. Lymphocytic hypophysitis must be considered as a differential diagnosis of intrasellar mass within the pediatric population.

**P-5-635** Changes induced by extrapituitary-perisellar disease on hypophyseal physiology

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**Objective:** The present work studied the changes in pituitary physiology induced by perisellar pathologies not involving the pituitary gland.

**Methods:** The clinical histories of 89 patients admitted to our neurosurgical service during the last 10 years were reviewed. Patients were affected with varied pathologies such as: Perisellar Tumors (25 cases), Cranioophyngiomas (17), Giant Aneurysm (3), Intrasellar Arachnodacode (29), Benign Intracranial Hypertension (2) Hydrocystosis (4), Sarcodeidosis (4), Primary Hypothyroidism (4) and Hydromyelia (1). Diagnoses were made by: CT, MRI, ANGIOPHAGHY, and serum hormones measurements to assess gonadal, thyroidal, adrenal, and pituitary function. Serum prolactin values were grouped as: N-Normal < 20 ng/mL, A: 20-50 ng/mL B: 50-100 ng/mL C: > 100 ng/mL. Total pituitary hypofunction was diagnosed when all axes were depressed. Partial hypofunction was diagnosed when only some of the axes were depressed. Diabetes insipidus was diagnosed by urine osmolality, volume and density. We excluded patients with hypophysal adenomas.

**Results:** 54% of all the patients had hyperprolactinemia: 23% of them had type A, 24.5% type B and 3% type C. 56.7% of all the patients had partial pituitary hypofunction, and 22.4% had total hypofunction. 36% of the patients had normal serum hormones concentrations, whereas 22.4% had diabetes insipidus. Patients with cranioophyngiomas had the highest incidence of pituitary changes, whereas the highest single hormone change among all patients was hyperprolactinemia. All patients with chronic inflammatory disease had diabetes insipidus and partial pituitary hypofunction was twice as frequent as total pituitary hypofunction.

**Conclusion:** The data suggest that changes in pituitary function are frequently the result of perisellar pathology not involving the pituitary gland. A thorough study of pituitary function appears mandatory in all patients with such pathologies.

**P-5-636** Gangliocytoma and ganglioglioma associated with hypophysal adenomas

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Neuronal tumors: Gangliocytomas or neuronal choristomas are composed of binuclear mature neurons. Gangliogliomas are mixed tumors composed of neuronal and neoplastic glial stroma. The location of these tumors in the sellar region is very rare as well as the association with hypophysal adenomas.

Three cases are reported. A 58-year-old man with acromegalic syndrome, GH basal 70 ng/mL, with a sellar tumor. The pathologic anatomy (PA) showed the coexistence of a gangliocytoma associated with an acidophilic adenoma with GH immunomarking in more than 60% of the cells.

The second case is a 29-year-old man with Cushings disease, with a sellar and suprasellar tumor and a manifest hypercortisolism. The PA confirmed a basophilic adenoma with ACTH positive immunomarking greater than 90% of the cells, coexisting with a ganglioglioma.

Third case: A 43-year-old woman with acromegalic features presented with a Cushings disease. MRI showed an intrasellar tumor. The PA showed a ganglioglioma associated with a mixed adenoma.

**Conclusion:** The rare coexistence of neuronal tumors associated with hypersecreting hypophysal adenomas and the possibility that these tumors may develop a neurosecreting activity inducing the formation of adenomas are the reason of this presentation.

**P-5-637** Cerebral perfusion pressure during transsphenoidal surgery

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Numerous maneuvers have been used to raise the intracranial pressure during transsphenoidal surgery for pituitary macroadenomas. Initially we compared the effect of intrathecal saline infusion and Valsalva maneuver both on CSF pressure (CSFP) and cerebral perfusion pressure (CPP) in twenty patients during the above procedure. The increase in CSFP produced by one Valsalva maneuver was similar to a single increment of a 2.5 ml bolus of intrathecal saline. (4 ± 2 mmHg) With Valsalva maneuvers the maximum CSFP produced was 10 ± 4 mmHg with a decrease of CPP to 50 ± 14 mmHg whereas with saline infusions the maximum CSFP was 25 ± 7 mmHg and a decrease in CPP to 59 ± 13 mmHg. The effect of intrathecal saline infusion was more sustained. In the second prospective study on a further 38 patients the effect of intrathecal air injected into the subarachnoid space during transsphenoidal surgery was compared with reference to the CSFP and the CPP. The mean rise in CSFP pressure per 2.5 ml bolus of intrathecal air was 6.2 ± 2.3 mmHg. The maximum CSFP pressure achieved was 29.6 ± 6.7 mmHg (mean ± S.D.) (range: 18-45 mmHg). The minimum CPP with the same maneuver was 55.8 ± 10.6mmHg (mean ± S.D.) (range: 44-79 mmHg).

Thus the changes in CSFP and CPP produced by intrathecal saline and air were of the same order. The injection of air was found to be useful in delineating the superior extent of the tumour during the various phases of tumour removal. The importance of monitoring the CPP regardless of the method of increasing the CSF pressure is underlined and the sustained effect of intrathecal saline and air is emphasized.

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**Thursday, 10 July 1997**

**14:00-16:15**

**P-5** Tumours of the CNS – Orbital and Skull Base Tumours

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**P-5-638** Orbital pseudotumor causing sudden exophthalmos and intraorbital hemorrhage due to acute superior ophthalmic vein obstruction: A case report

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**Introduction:** Orbital inflammatory pseudotumors cause painful exophthalmos and usually respond to corticosteroid therapy, and most of them are confined to the orbit. A case of orbital pseudotumor with sudden onset of exophthalmos is presented. The patient did not respond to steroids therapy and was improved by surgical decompression of the superior ophthalmic vein.

**Case:** This 59-year-old, otherwise healthy, female patient presented with sudden onset of severe exophthalmos and ophthalmic pain. Orbital CT scan showed increased density on upper part of the left orbit with exophthalmos. On T1-weighted image of MRI, decreased signal intensity with marginal enhancement was noted on the same site, and left superior ophthalmic vein was not seen. On angiography, the run-off of distal ophthalmic artery was not seen and