and DI. The most common (MC) presentation is of DI. Among anterior pituitary hormone deficits, central hypogonadism is the MC. Systemic high dose glucocorticoids (GC) are the mainstay of initial treatment. Optic atrophy is the MC neuro-ophthalmologic manifestation of NS. Only 10 - 20 % of patients have been found to have biochemical improvement of HP axis in response to high-dose GC treatment. Since pituitary involvement commonly leads to permanent endocrine deficits, it is essential to recognize it early and to treat it aggressively to prevent further permanent hormonal and neurologic deficits.

Neuroendocrinology and Pituitary

NEUROENDOCRINOLOGY AND PITUITARY CASE REPORTS

New Treatment Options for Pituitary Granulomatosis With Polyangiitis

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Background: Granulomatosis with polyangiitis (GPA) rarely involves the pituitary gland and has been reported in 1% of all cases of GPA. Most frequently, it presents with pituitary mass effect and diabetes insipidus (DI). To date, there are no treatment guidelines for this rare condition. **Case Presentation:** Case 1: A 55 year old female with a history of ANCA-positive pulmonary GPA, previously treated with glucocorticoids and immunosuppressants, presented two years later with cranial DI and bitemporal hemianopia. MRI showed a large sellar mass with suprasellar extension. High dose glucocorticoids resulted in good clinical and radiological response. Further treatment consisted in a combination of Cyclophosphamide and Rituximab (RTX). Further doses of RTX are planned aiming for a period of B-cell depletion. Case 2: A 38 year old female, presented with polyuria, recurrent nosebleeds, headaches and a left visual field defect. Pituitary profile revealed ACTH deficiency and MRI showed a heterogenous cystic lesion with peripheral enhancement and stalk thickening. Steroid replacement led to immediate improvement in her symptoms. Cranial DI was confirmed and raised Proteinase 3 (PR3) antibody suggested GPA. A combination of prednisolone and Methotrexate led to significant improvement of MRI appearances and declining PR3 antibody levels. For remission maintenance, two cycles of RTX were given with further radiological and biochemical improvement, and, following dynamic assessment of her HPA axis, she could be fully weaned off steroids. Case 3 is a 47 year old female with a history of childhood asthma. She was found to have cavitating lung lesions. ANCA positivity confirmed GPA and she was commenced on high-dose steroids. During follow-up, she developed headache, polyuria and polydipsia. MRI pituitary showed a suprasellar lesion and pituitary biopsy revealed inflammatory hypophysitis. Cranial DI was confirmed by water deprivation testing. Previous allergic reactions to both RTX and Ofatumumab precluded anti-CD40 monoclonal antibodies and she was commenced on Azathioprine. A further recurrence of pituitary GPA necessitated escalation of the steroid dose and switch of azathioprine to mycophenolate mofetil. She remains in remission and her steroids reduced to a maintenance dose. **Conclusion:** GPA pituitary has been observed to occur at variable time after diagnosis often in the absence of any other systemic features. A combination of glucocorticoids and RTX has been approved for severe relapsing pulmonary GPA, however, limited data is available for pituitary GPA. In this case series, the response to high dose steroids and RTX for remission maintenance has been encouraging. Experience with 'conventional' immunosuppresants remains limited and therapeutic responses remain variable. Further clinical studies are required to establish effective treatment for pituitary GPA.

Neuroendocrinology and Pituitary

NEUROENDOCRINOLOGY AND PITUITARY CASE REPORTS

Occult Carcinoid Causes Cushing's Syndrome

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Introduction: Lung neuroendocrine tumors (NETs) are a common cause of ectopic ACTH-secretion. Only 3% of NETs cause Cushing's syndrome[1]; the majority are small and may be radiographically occult. Additionally, the responses to dexamethasone, metyrapone, and oCRF can be indistinguishable from that observed in pituitary Cushing's disease[2]. These challenges lead to misdiagnosis and unnecessary procedures. **Case Description:** 45-year-old male presented for resection of right-sided pulmonary carcinoid tumor. He was diagnosed with Cushing's syndrome in 2011 when he experienced uncontrolled hypertension and excessive weight gain with elevations in cortisol and ACTH levels. He underwent transsphenoidal pituitary resection in May 2011 for a possible pituitary microadenoma. Postoperative worsening hypertension, weight gain, and striae led to bilateral adrenalectomy in November 2011, complicated by retroperitoneal hemorrhage and resuscitation-induced pulmonary edema. On chest CT in early 2012, an 8 mm incidental right pulmonary nodule was documented. 8 years later, chest imaging for mild COVID-19 infection again revealed a right-sided pulmonary nodule. He underwent CT-guided biopsy of the nodule in July 2020 with pathology demonstrating carcinoid histology. Right lower lobectomy was performed. Pre-operative ACTH was 1673 pg/mL (normal: 10-60 pg/mL) and post-operative ACTH was 16 pg/mL. The resected tumor stained positive for ACTH, confirming that carcinoid tumor was the source of Cushing's for the past decade.
Conclusion: The diagnosis of ectopic Cushing's syndrome can be elusive, leading to surgeries with significant morbidity, as seen in our patient. In one retrospective review, 14% of patients with Cushing's had transsphenoidal resection before they were diagnosed with an ectopic source[3]. Ectopic Cushing's syndrome can be a challenging diagnosis to make and a multidisciplinary approach with close collaboration between endocrinologist, radiologist, surgeon and pathologist may increase the diagnosis accuracy.