Twentieth Clinicopathological Conference on Pituitary Disease

Thursday 1st February 2018

Royal College of Obstetricians and Gynaecologists, London, NW1 4RG

PLATINUM

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A number of companies have contributed towards the cost of this meeting, and will have an exhibition stand present.
They have had no input into the meeting organisation, the agenda or the selection of speakers, unless otherwise stated.

AGENDA

08:30 **Registration**

09:30 **Welcome and Introduction**
Dr Stephanie Baldeweg, Consultant Endocrinologist, University College London Hospitals

09:35 **Medical therapy of pituitary disease over the last 20 years**
Prof Ashley Grossman, Professor at the Universities of Oxford and London

10:00 **Forum 1 – Case Presentations – Prolactinomas**
Chairs Dr SE Baldeweg and Mr M Powell

Case 1: Late development of secondary dopamine agonist resistance and skull base invasion in a patient with macroprolactinoma
A de Bray, ZK Hassan-Smith, J Dirie, E Littleton, P Sanghera, J Ayuk, N Karavitaki

Case 2: An atypical prolactin secreting macroadenoma presenting with apoplexy in the 3rd trimester of pregnancy; management challenges
M Bakhit, E Maratos, A Ansaripour, S Al-Sarraj, T Hampton, K Barkas, S Barazi, N Thomas, J Gilbert

Case 3: Aripiprazole: monotherapy for a macroprolactinoma?
S Hussain, M Shiafkou, G Gaoatswe, JV Anderson, SA Akker

Case 4: Impulse control disorder in a patient with prolactinoma treated with cabergoline
N Samani, MW O'Reilly, A Toogood, N Karavitaki

11:00 **Tea, Coffee and Posters**

11:30 **Radiotherapy of pituitary disease over the last 20 years**
Dr Gillian Whitfield, Consultant Oncologist, Christie Hospital, Manchester

11:50 **Forum 2 – Case Presentations – Pituitary all sorts**
Chairs Dr J Ahlquist and Dr M Vanderpump

Case 5: Treatment options in a boy with gigantism and inconclusive MRI
C May, G Anderson, W Bashari, B Jafar-Mohammadi, M Gurnell, S Cudlip, A Pal, T Makaya

Case 6: Androgen deprivation in prostate cancer by way of macroprolactinoma – balancing the two pathologies
JFH Pittaway, J Shamash, WM Drake

Case 7: Treatment dilemmas in Cushing's with multiple non-curative surgeries – a patient's choice?
S Miles, S Cudlip, R Mihai, BJ Mohammadi, A Grossman, A Pal

Case 8: Hypophysitis- a marginally difficult case for discussion
U Srirangalingam, R Ronneberger, R Menon, M Rodriguez–Justo, G Webster, N Dorward

12:50 **Lunch and Posters**

13:50 **The Pituitary Foundation**
Menai Owen-Jones, CEO, Pituitary Foundation

14:00 **Forum 3 – Case Presentations – Pregnancy and fertility**

Chairs Miss JP Grieve and Prof GS Conway

Case 9: Gonadotrophic adenoma presenting with ovarian hyperstimulation syndrome
M Sorour, C Broughton, A Williams

Case 10: Diabetes insipidus and pregnancy
I Kurera, T Rehman, A Hameed, G Bano

15:00 **Tea, Coffee and Posters**

15:30 **Pituitary surgery over the last 20 years**

Mr Michael Powell, Emeritus Consultant Neurosurgeon, National Hospital for Neurology and Neurosurgery

15.50 **Forum 4 – Case Presentations – Pituitary surgery**

Chairs Prof PM Bouloux and Mr N Dorward

Case 11: A case of persistent acromegaly after surgery; management options
J Halliday, A Pal, A Brooke, SA Cudlip

Case 12: A challenging case of a recurrent growth-hormone secreting pituitary adenoma
JS Bal, D Paraskevopoulos, JC Benjamin, J Ahlquist

Case 13: Adenoma, aneurysm or apoplexy: an interesting pituitary mass
SA Tee, S Pearce, D Birchall, P Bhatnagar, RA James

Case 14: Surgical debulking of pituitary adenoma under local anaesthesia – world first!
SK Ahmed, V Hox, S Russell, T Matthews, R Chugh, S Ramalingam, H Buch, J Ayuk, N Karavitaki

16.50 **Poster and presentation prizes**

Dr Stephanie Baldeweg, Consultant Endocrinologist, University College London Hospitals

17.00 **Close**

FACULTY

PANEL & ORGANISERS

FACULTY

Dr James Ahlquist, Consultant in Endocrinology and Diabetes, Southend Hospital

Dr Stephanie E Baldeweg, Consultant Endocrinologist, University College London Hospitals

Prof Pierre-Marc Bouloux, Consultant Endocrinologist, Royal Free London

Prof Gerard S Conway, Lead in Reproductive Endocrinology, University College London Hospitals

Mr Neil Dorward, Consultant Neurosurgeon, National Hospital for Neurology and Neurosurgery

Miss Joan P Grieve, Consultant Neurosurgeon and Clinical Lead for Neurosurgery, National Hospital for Neurology and Neurosurgery

Prof A Grossman, Professor at the Universities of Oxford and London

Mr Michael Powell, Emeritus Consultant Neurosurgeon, National Hospital for Neurology and Neurosurgery

Dr Mark Vanderpump, Consultant Endocrinologist, London

Prof Gillian Whitfield, Consultant Oncologist, Christie Hospital, Manchester

ORGANISING COMMITTEE

Chair:

Dr Stephanie Baldeweg
Consultant Endocrinologist,
University College London Hospitals

Dr Stephanie E Baldeweg, Consultant Endocrinologist, University College London Hospitals

Prof Pierre-Marc Bouloux, Consultant Endocrinologist, Royal Free London

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Mr Michael Powell, Emeritus Consultant Neurosurgeon, National Hospital for Neurology and Neurosurgery

Dr Mark Vanderpump, Consultant Endocrinologist, London

PANEL MEMBERS

ENDOCRINOLOGY:

Prof Ashley Grossman
Professor at the Universities of Oxford and London

Dr Paul Carroll
Consultant Endocrinologist, Guys and St Thomas Hospital

Prof Will Drake
Consultant Endocrinologist,
Barts and the London School of Medicine

Prof Karim Meeran
Consultant Endocrinologist, Imperial College London

Dr Frankie Swords
Consultant Endocrinologist, Norfolk and Norwich NHS trust

OTHER SPECIALITIES:

Dr Katherine Miszkiel
Consultant Neuroradiologist, University College London Hospitals

Dr Gordon Plant
Consultant Neurologist, University College London Hospitals

Dr Naomi Fersht
Consultant Neuro-oncologist, University College London Hospitals

Dr Federico Roncaroli
Consultant Neuropathologist, Imperial College London

NEUROSURGERY:

Prof Ed Laws
Consultant Neurosurgeon, Brigham and Womens Hospital, Boston, USA

Prof Paolo Cappabianca
Frederick II University, Naples, Processor and Chairman, Division of Neurosurgery, University of Naples Federico II, Italy

Mr Simon Cudlip
Consultant Neurosurgeon, Oxford Radcliffe NHS Trust

Mr Alisher Abutaev
Consultant Neurosurgeon, Republic Endocrine Centre, Tashkent

Mr Rick Nelson
Consultant Neurosurgeon, North Bristol NHS Trust

1.) Late development of secondary dopamine agonist resistance and skull base invasion in a patient with macroprolactinoma

AUTHOR(S):

A de Bray, ZK Hassan-Smith, J Dirie, E Littleton, P Sanghera, J Ayuk, N Karavitaki

Abstract

We report on an 81-year old man, diagnosed with a macroprolactinoma in the early 1980s, treated with transcranial surgery and radiotherapy. He was maintained on bromocriptine, along with replacement for hypopituitarism. His prolactin was normal until 2008, when a gradual increase was noted with the patient considered to be compliant on medical treatment (2008: 739 mU/L, 2009: 2421 mU/L, 2012: 3498 mU/L). Pituitary MRI in 2009 reported largely empty fossa and in 2012 stable appearances with an expanded pituitary fossa and a small amount of residual pituitary tissue. Follow up scans in 2014 and 2015 were reported as stable with the prolactin reaching 113,566 mU/L. A switch from bromocriptine to cabergoline had no effect, with his prolactin continuing to rise, reaching 171,670 mIU/L by August 2016, and so was reverted to bromocriptine. Prolactin increased further to over 500,000 mIU/L by March 2017. The patient developed weak voice, dysarthria and weakness of the tongue and was diagnosed with left hypoglossal and recurrent laryngeal nerve palsies. Repeat imaging was reviewed in the pituitary MDT and whilst appearances in the fossa were stable, an invasive clival lesion was identified. He was not a candidate for surgery or radiotherapy; he was referred to Oncology and was commenced on temozolomide; following 4 cycles, there is good response on imaging and reduction of PRL (23,132 mU/L).

Development of secondary resistance to DA in macroprolactinomas is very rare and can be related with aggressive tumour behaviour and malignant potential. In our patient, it was manifested more than two decades after initial diagnosis highlighting the unpredictable behaviour of these tumours.

Multidisciplinary approach is required. Temozolomide monotherapy is the first line chemotherapy for aggressive pituitary tumours with positive outcome in a number of cases. Nonetheless, the long-term prognosis remains unclear.

2.) An atypical prolactin secreting macroadenoma presenting with apoplexy in the 3rd trimester of pregnancy; management challenges

AUTHOR(S):

M Bakhit, E Maratos, A Ansaripour, S Al-Sarraj, T Hampton, K Barkas, S Barazi, N Thomas, J Gilbert

Abstract

Introduction:

In 2016, the Society for Endocrinology clinical committee developed guidelines for the emergency management of pituitary apoplexy in adults, which provide a clear diagnostic and management pathway. However, in some situations the management may be more challenging.

Case report:

A 31 year-old woman, who was 34 weeks pregnant, was transferred to our unit with pituitary apoplexy after presenting to her local hospital with sudden onset severe headache, diplopia and bi-temporal hemianopia. Brain MRI showed a 22mm haemorrhagic pituitary macroadenoma displacing the optic chiasm.

Upon transfer to our neurosurgical ward, her ocular and visual signs had improved. She had a bitemporal superior quadrantanopia, visual acuity 6/6 with no ophthalmoplegia. Initial investigations demonstrated a midday cortisol of 162 nmol/L, free thyroxine 10.4 pmol/L, TSH 0.63mIU/L and serum prolactin 4543 mIU/L. She was commenced on hydrocortisone and levothyroxine. Initially she was managed conservatively but a few days later she developed a further episode of headache associated with worsening of her visual acuity and severely constricted visual fields. She proceeded to have an endoscopic transphenoidal resection. At surgery the tumour was found to be haemorrhagic, consistent with pituitary apoplexy. Her visual acuity and fields were restored to normal post-operatively.

Histopathology revealed an aggressive tumour with cells diffusely and strongly immunoreactive to prolactin and a markedly increased Ki-67 of 20%. The p53 was 30%. These findings are consistent with atypical adenoma but also raised concerns regarding pituitary carcinoma. She commenced cabergoline and following multi-disciplinary team discussions, labour was induced at 37 weeks gestation. CT chest abdomen pelvis and MRI neuroaxis did not reveal any systemic or CNS metastases.

Discussion:

During pregnancy, serum prolactin may be challenging to interpret as a tumour marker. The significance of these histopathological findings in pregnancy is unknown. The challenges of managing atypical pituitary tumours in pregnancy are discussed and the evidence is reviewed.

3.) Aripiprazole: monotherapy for a macroprolactinoma?

AUTHOR(S):

S Hussain, M Shiafkou, G Gaoatswe, JV Anderson, SA Akker

Abstract

A 39 year old man with bipolar affective disorder presented with lethargy and erectile dysfunction. Clinically he appeared eupituitary and had full visual fields to confrontation with red pin.

Investigations revealed hyperprolactinaemia (34,999munit/L) and secondary hypogonadism (FSH 1.9unit/L, LH 0.9unit/L, testosterone 6nmol/L). Pituitary MRI showed a macroprolactinoma elevating the chiasm and left optic nerve with left-sided cavernous sinus extension. A pituitary macroadenoma had been seen on an MRI scan six years previously and had grown significantly.

He had had two previous admissions with manic-depressive episodes and described his current mental state as low but stable. He was off medication at the time of review.

Treatment options discussed included: a) dopamine agonists with careful psychiatric cover; b) non-curative transsphenoidal surgery accepting the potential surgical complications and likely need for post-operative radiotherapy and c) a trial of aripiprazole.

Aripiprazole is a second generation atypical antipsychotic with partial dopamine agonist activity. Some case reports have documented successful treatment of prolactinomas, usually when it is used in combination with other treatments.

The patient was prescribed 5mg of aripiprazole and the prolactin fell to 22,637munit/L within four weeks. Over five months aripiprazole was uptitrated to 15mg with a current prolactin of 14,962munit/L and a modest reduction in tumour volume and relief of any compromise to the optic apparatus.

The management of patients with psychiatric disease and prolactinomas can be challenging due to the recognised mood disturbance caused by first-line dopamine agonists. Aripiprazole could be a useful management option for patients who are deemed high risk for first-line medications as a potential alternative to surgery and can be used as monotherapy.

We present the data and scans for this case, discuss aripriprazole's mechanism of action and side-effects as well as its use to date in the management of hyperprolactinaemia. We discuss long-term treatment options for this patient.

4.) Impulse control disorder in a patient with prolactinoma treated with cabergoline

AUTHOR(S):

N Samani, MW O'Reilly, A Toogood, N Karavitaki

Abstract

A 55-year old man presented with Bell's palsy in March 2015. Head MRI revealed a large enhancing lesion expanding the pituitary fossa and extending to the left cavernous sinus. Pituitary hormone profile showed PRL 138,738 mU/L (85-325), hypogonadotropic hypogonadism with morning testosterone 0.7 nmol/L (7-27) and normal IGF-I, ACTH reserve and thyroid hormones. The patient reported headaches, erectile dysfunction, non-existent libido and no galactorrhoea. His medical history included nothing of significance; he reported smoking 1 cigarette/day and alcohol consumption 15 units/week.

He started cabergoline 500 mcg twice/week resulting in normal PRL and testosterone, and adenoma shrinkage. In October 2015, he mentioned "increased anger" and "high libido". During subsequent clinic visits, he did not mention further problems, until August 2017, when he attended with his wife who also confirmed bursts of anger and being less risk adverse in making business decisions; he was initially advised to reduce cabergoline to 500 mcg/week. On review 6 weeks later, he had not followed this advice, as he was very satisfied and happy with the changes in his behaviour ("more active socially and sexually"). He also revealed that during the last 18 months, he experienced compulsive shopping (eg. bought 15 bicycles and two cars £60K each), took business decisions with significant negative financial sequelae and recently, had separated from his wife. He was not keen on dose reduction, but he finally agreed to take cabergoline 500 mcg/week. In the last consultation, he reported complete remission of his impulse symptoms and full insight into his previous behaviour.

Impulse control disorders are an under-recognised complication of dopamine agonist treatment in prolactinomas. Pathophysiology remains unclear (interaction with D3-receptors in mesolimbic system?). Patients may not spontaneous report these and increased awareness is needed aiming to prevent serious or catastrophic consequences for patient's lives and medico-legal issues for the clinicians.

5.) Treatment options in a boy with gigantism and inconclusive MRI

AUTHOR(S):

C May, G Anderson, W Bashari, B Jafar-Mohammadi, M Gurnell, S Cudlip, A Pal, T Makaya

Abstract

We present the case of a boy aged 10.6 years who presented to our paediatric endocrinology colleagues with tall stature. With hindsight, when compared with peers and siblings, he was noticeably tall from the age of 5. At 10 years his height velocity was 12cm/year, prompting referral and further investigation.

His height was 178.7cm (+5.79 sds above the mean), and well outside the family target centile range. IGF-1 was elevated at 78.6 nmol/L (10.6-60.8) and an OGTT demonstrated non-suppression of GH (5.5-9.1 mcg/L). His bone age was 11.03 years. The remaining pituitary profile and visual fields were normal. Initial contrast MRI was inconclusive but C11 methionine PET/MRI at Addenbrooke's identified a pituitary microadenoma as a potential surgical target.

An MDT clinic held with the patient, his parents, paediatric and adult endocrinologists and neurosurgical teams allowed discussion about medical and surgical treatment options. The team had previously debated whether puberty should be induced to accelerate epiphyseal fusion and limit growth. In view of the patients wish to prioritise school studies and go through puberty prior to surgery, medical treatment with somatostatin analogue was chosen with a view to surgery at a later date.

Gigantism is rare and typically due to macroadenomas. Genetic causes are known but not fully understood. Genetic results available for our patient are so far negative and functional imaging demonstrates a microadenoma. This rare case required an individualised approach tailored to the patient and family.

Discussion points:

- Use of functional imaging to identify surgical targets
- Pros and cons of medical and surgical treatment in a young person and tailoring treatment choice
- Should puberty be induced early in gigantism to limit growth
- Titrating somatostatin analogue to achieve ideal growth hormone values in a pre-pubertal patient
- Should pegvisomant be used as first line medical therapy

6.) Androgen deprivation in prostate cancer by way of macroprolactinoma – balancing the two pathologies

AUTHOR(S):

JFH Pittaway, J Shamash and M Drake

Abstract

A 72 year-old gentleman from Zimbabwe presented to clinic with MRI pituitary findings of 1.5x1.4x1cm pituitary macroadenoma. This had been discovered incidentally during investigation for severe headaches at another hospital. The mass was in contact with but not compressing the optic chiasm. He described no visual loss. He had an unintentional weight loss of 10kg in the last 2 months. He described decreased libido for 3 years and no erections including the mornings. His only other past medical history was glaucoma. Examination revealed small, 6-8mls testes bilaterally. Visual fields were minimally decreased temporally to confrontation with red pin. Pituitary profile revealed a raised prolactin of 19,204mU/L(0-496) after PEG precipitate. The rest of his anterior pituitary function was within normal limits. Testosterone was suppressed at 1.6nmol/L(9-27). PSA was raised at 4,426mcg/L(0-4.4) and liver function revealed a raised ALP of 758u/L(30-130). On the basis of these blood tests he was diagnosed with a macroprolactinoma and metastatic prostate cancer and referral to oncology was made.

Whole body CT and bone scans revealed widespread bony disease including a burden of disease in the skull. He was started on docetaxel and, in view of impending chiasmal compromise, low-dose cabergoline. His headaches improved but after an initial fall in PSA, his testosterone level started to rise and oncological concern was raised about the implications for his metastatic prostate cancer. Cabergoline therapy was suspended and, thus far, he remains symptom free with no visual disturbance and repeat pituitary imaging shows no compromise of the optic chiasm. His prostate cancer disease burden is currently stable on enzalutamide 18 months on from diagnosis.

This case of a male patient with metastatic prostate cancer who is biochemically hypogonadal secondary to a macroprolactinoma, presents a management dilemma of the competing imperatives of the two diagnoses. The panel's views would be welcomed.

7.) Treatment dilemmas in Cushing's with multiple non-curative surgeries – a patient's choice?

AUTHOR(S):

S Miles, S Cudlip, R Mihai, BJ Mohammadi, A Grossman, A Pal

Abstract

A 56 year-old gentleman presented with Cushing's syndrome in September 1992: investigations confirmed cortico-hypersecretion and demonstrated a raised plasma ACTH, normal pituitary MRI and adrenal hyperplasia on abdominal CT. Bilateral inferior petrosal sinus sampling (BIPSS) demonstrated similar ACTH levels peripheral to the inferior petrosal sinus after CRH stimulation on two occasions. He did not suppress on high dose dexamethasone suppression tests. He was initially offered bilateral adrenalectomy but declined. He nevertheless underwent transsphenoidal pituitary surgery in September 1994. Histology demonstrated normal anterior gland with Crooke's hyaline change but no corticotroph adenoma or hyperplasia. He continued to be hypercortisolaemic post-operatively and underwent a bilateral adrenalectomy in February 1995. This was not curative, with CT demonstrating a 6cm x 3.4cm hyperplastic remnant of the right adrenal gland.

New MRI now demonstrated a small right-sided pituitary microadenoma while two further BIPSS supported central ACTH excess, with a central to peripheral ratio of 3.8. He is now treated with metyrapone 250mg QDS and ketoconazole 200mg QDS, with a cortisol day-curve currently showing levels of 09.00h 761nmol/L, 12.00h 319nmol/L, 15.00h 195nmol/L, 18.00h 286nmol/L, 21.00h 201nmol/L (mean 352nmol/L), but he has declined further upwards titration. He has numerous complications from his Cushing's including osteoporosis with multiple rib and vertebral fractures, muscle and skin atrophy, uncontrolled hypertension, and left ventricular hypertrophy. He has been offered a remnant adrenalectomy, or transsphenoidal adenectomy and radiotherapy by our multidisciplinary team, but has declined all being particularly concerned with possible development of hypopituitarism, hypoadrenalism and/or Nelson's syndrome.

He thus has had partially-controlled Cushing's syndrome for over 20 years with significant complications and is now requesting pasireotide therapy in preference to any other intervention. We question whether this should be sought considering he has a surgical target and has declined potentially curative options, given there may be no defined period of treatment?

8.) Hypophysitis - a marginally difficult case 4 discussion

AUTHOR(S):

U Srirangalingam, R Ronneberger, R Menon, M Rodriguez–Justo, G Webster, N Dorward

Abstract

A 57-year-old man was referred with 6-year history of gradually enlarging pituitary gland and stalk suggestive of an infiltrative disease. He had a background of ulcerative colitis and scleritis maintained on low dose steroid. He was gonadotrophin and TSH deficient. There was no evidence of diabetes insipidus. The patient declined an initial pituitary biopsy. An FDG PET scan identified an enlarged sub-mandibular gland and lymphadenopathy in the neck and paratracheal region. A sub-mandibular gland biopsy demonstrated a nodular lymphocyctic infiltrate but without B cell clonal expansion by PCR. A further lymph node biopsy suggested a nodular small B cell infiltrate with immunohistochemical features consistent with a marginal zone lymphoma. PCR for B cell clonality was awaited.

His case was discussed at the Haematology Oncology MDT. In the context of a diagnosis of marginal zone lymphoma, pituitary involvement would constitute distant extra-nodal site necessitating the need for active therapy rather than a watch-and-wait strategy. The patient agreed to a pituitary biopsy to clarify involvement. Pituitary biopsy confirmed a lymphoplasmcytic infiltrate, but PCR did not demonstrate evidence of clonal B-cell expansion. A significant number of IgG4+ plasma cells (IgG4: >20 positive plasma cells / high power field) were noted however, suggestive of IgG4 related hypophysitis. The PCR for clonal expansion in the lymph node biopsy was also negative, making a diagnosis of lymphoma less likely. There was insufficient tissue from the lymph node or sub-mandibular gland biopsy to confirm an IgG4 infiltrate.

His case was discussed in the joint UCLH/Oxford IgG4 MDT. A diagnosis of IgG4-related disease with salivary and nodal involvement was favoured. It was agreed that he would have a further lymph node biopsy to confirm an IgG4+ infiltrate in the lymph nodes and he would be considered for rituximab therapy.

Discussion points

1. Need to consider a diagnosis of IgG4 related disease with an infiltrative appearance in the pituitary gland and stalk.

2. Difficulties in differentiating between lymphoma and IgG4+ plasma cells infiltrates.

3. Importance of a tissue diagnosis and the need to consider biopsy at multiple sites where diagnosis unclear

4. Link between marginal zone lymphoma and IgG4 related disease.

9.) Gonadotropic adenoma presenting with ovarian hyperstimulation syndrome

AUTHOR(S):

M Sorour, C Broughton, A Williams

Abstract

Introduction
Gonadotroph adenomas represent 40–50% of pituitary macroadenoma, however only a small subset of these tumors secrete sufficient hormone to elevate serum gonadotropin levels to produce a characteristic clinical phenotype (functioning gonadotroph tumors), they routinely present with visual field disturbance or deficiency of other anterior pituitary hormones.

We present a rare case of a delayed diagnosis of a follicular stimulating hormone (FSH)-secreting macroadenoa adenoma presenting with ovarian hyperstimulation, polycystic ovaries and abdominal pain.

Case report
We report a case of a 36 year-old lady who presented with abdominal pain and was found to have multiple bilateral ovarian cysts which were resected.

Six months later, she presented again with acute abdomen and was found to have ovarian torsion along with recurrence of multiple ovarian cysts, requiring a salpingo-oopherectomy.

On further investigations, she was found to have a serum FSH level of 9.5 IU/L (N 1.0 - 9.0 IU/L), a serum oestradiol level of 2096 pmol/L (N 161 - 774 pmol/L) and serum LH level of 0.71 IU/L (N 2.4 - 7.2 IU/L). Her serum prolactin level was 740 mIU/L (N <700 mIU/L). The remainder of the pituitary hormonal profile was normal.

Brain MRI revealed a pituitary macroadenoma measuring about 1.5 cm in its maximum craniocaudal extent with no invasion to the cavernous sinuses, just touching the inferior surface of the optic chiasm. Her visual fields testing revealed small cuts in the bi-temporal upper quadrants. She underwent endoscopic endonasal resection of her adenoma, and the histological examination confirmed a pituitary adenoma with FSH immunopositivity in keeping with gonadotroph cell adenoma.

The postoperative serum pituitary profile was normal.

Conclusion
FSH-secreting adenomas can rarely present with ovarian hyperstimulation syndrome and this diagnosis should in females with recurrent unexplained ovarian cysts.

10.) Diabetes Insipidus and Pregnancy

AUTHOR(S):

I Kurera, T Rehman, A Hameed, G Bano

Abstract

Diabetes insipidus (DI) can complicate up to 1 in 30,000 pregnancies. DI during pregnancy has a variety of causes, some that predate the pregnancy and others that begin during gestation. The presentation can involve exacerbation of central or nephrogenic DI during pregnancy, which may have been subclinical before pregnancy. Women without pre-existing DI can also be affected by the actions of placental vasopressinase which increases in activity between the 4th and 38th weeks of gestation, leading to the accelerated metabolism of AVP and causing a transient form of DI of pregnancy. This form of diabetes insipidus may be associated with increased complications of pregnancy, including preeclampsia. Management of central diabetes insipidus and transient diabetes insipidus of pregnancy can be achieved with 1-deamino-8-D-arginine vasopressin (desmopressin acetate) (DDAVP), a vasopressin analog. We present a case of 34 years old lady who had surgery for pituitary macroadenoma at the age of 30. She developed diabetes insipidus postoperatively that was treated with desmopressin. Her symptoms gradually improved over time, and she stopped her DDAVP. She conceived and at 25 weeks of pregnancy developed polyuria, nocturia, and polydipsia. She refused to restart DDAVP. At 27 weeks she noticed reduced fetal movements. Her USS showed a reduction in amniotic fluid with fetal growth restriction and occlusion of the single umbilical artery. At 28 weeks USS revealed intrauterine fetal death.

Increased awareness of diabetes insipidus in pregnancy may lead to early diagnosis and appropriate treatment that will reduce the risks of maternal and fetal morbidity. Overall, growing experience with DDAVP has shown that it is a safe and effective treatment for diabetes insipidus caused by a variety of factors.

11.) A case of persistent acromegaly after surgery; management options

AUTHOR(S):

J Halliday, A Pal, A Brooke, SA Cudlip

Abstract

We present the case of a 42-year-old Male with persistent acromegaly despite previous surgical management in another centre. He was initially diagnosed following ankle surgery when an anaesthetist noticed that he had acromegalic features. After 3 months of medical management with monthly Lanreotide 30mg he developed visual symptoms and thus proceeded to surgery. Unfortunately, post-operatively he had persistent acromegaly with an IGF-1 of 77nmol/l (7-28nmol/l) that has been managed with ongoing monthly injections of Lanreotide 120mg. Currently his IGF-1 and GH are 52.9nmol/L and 4.6mcg/L respectively. Dedicated MRI pituitary revealed no clear target for further surgery or radiotherapy. Hoping for an alternative to lifelong injections, or the possibility of reducing his dose of Lanreotide, he was referred for a further opinion from a dedicated multidisciplinary team comprised of surgeons, endocrinologists, oncologists and specialist nurses. Further specialist imaging was performed. This revealed changes consistent with functioning tumour beneath the pituitary fossa. These findings raise the possibility of further surgery becoming an option.

Discussed is the importance of investigating patients with persistent functioning pituitary tumours after initial treatment, and the value of referral to and discussion of such complex patients by a dedicated multidisciplinary team with experience of such complex cases. The value too for the patient of being able to meet and have all available treatment options discussed in a multi-disciplinary setting. Also discussed are the risks and benefits of the available management options, as well as the cost-effectiveness of the different treatment modalities where ongoing drug costs in younger patients are considerable.

12.) A challenging case of a recurrent growth-hormone secreting pituitary adenoma

AUTHOR(S):

JS Bal, D Paraskevopoulos, JC Benjamin, J Ahlquist

Abstract

41 male, presented with a two-year history of headaches. He has sleep apnoea and hypertension. He had had surgery for bladder exotrophy as a child.

Through investigation of his headaches he had a MRI of head which showed a soft tissue sellar mass approximately 15 x 10 mm and not causing compression to the optic apparatus. He did not have visual compromise. His pituitary profile revealed IGF 1229 ng/mL and growth hormone 6.53 ug/L, and the rest unremarkable. Acromegaly was diagnosed. Surgery was recommended as first line treatment with the aim to cure or reduce the growth hormone load.

He underwent trans-sphenoidal resection of pituitary adenoma. Postoperatively he felt the headaches had ceased and 2 days after surgery the IGF-1 had reduced to 20ng/mL. Histology showed a somatotroph adenoma with strong positivity in growth hormone secreting cells (Ki 67 2.2%). A month later his headaches had returned. A repeat IGF -1 was 984 ng/mL; a 3 month postoperative MRI showed an 8mm enhancing residual tumour. He underwent a revision trans-sphenoidal surgery 9 months later. The patient felt better postoperatively but at one month the IGF-1 remained high at 858 ng/mL. The histology was unchanged.

His repeat imaging showed residual adenoma. He was given the option for radiotherapy at this point but opted for a further surgery. We obtained a 11C-methionine pituitary PET scan to guide our resection. This showed a surgical target and an endoscopic resection of recurrent pituitary adenoma was undertaken in October 2017. The histology confirmed adenoma but his postoperative IGF-1 remains high at 823 ng/mL.

We have offered the patient gamma-knife radiosurgery. This recurrent adenoma has been resilient to repeated surgery and we hope radiosurgery may control the growth hormone excess. Medical therapy is planned while awaiting radiosurgery.

13.) Adenoma, aneurysm or apoplexy: an interesting pituitary mass

AUTHOR(S):

SA Tee, S Pearce, D Birchall, P Bhatnagar, RA James

Abstract

An 88 year old lady presented with sudden onset headache, nausea and diplopia waking her from sleep. She had no limb weakness, speech or swallowing difficulties. There was no preceding history of visual disturbance, fatigue or weight changes. Past medical history included gout and previous pulmonary embolism. She was taking naproxen and omeprazole, was a non-smoker and teetotal. There was no family history of stroke, aneurysms or malignancy.

On examination, she was alert and orientated, GCS 15/15. She had left sixth cranial nerve palsy, with no other positive neurological signs. BP was 222/111, with no postural drop. Systemic examination including visual fields was unremarkable, and she was clinically euthyroid and eupituitary.

Bloods showed normal FBC, LFT and U+E. Cortisol (3pm) was 901nmol/L, GH 1.55ug/l, IGF-1 8nmol/L, FSH 0.9U/l, LH <0.5U/l, oestradiol 185pmol/l, prolactin 1270mIU/L, TSH 0.61mU/l, fT4 9.1pmol/l, fT3 1.5pmol/l.

Initial CT head showed a 46x15x17mm mass obliterating the pituitary fossa, extending into the cavernous sinuses, with no optic chiasm compression. MRI pituitary with contrast however revealed the “mass” corresponded to bilateral saccular aneurysms arising from the cavernous portions of the internal carotid arteries (ICA). CT angiogram confirmed the above. Neuroradiology MDT discussion recommended conservative management. She was commenced on levothyroxine 25mcg daily for secondary hypothyroidism, 10mg amlodipine, and a reducing course of dexamethasone.

Pituitary insufficiency secondary to ICA aneurysm is rare, with a reported prevalence of 0.17%. This case illustrates that ICA aneurysms can mimic pituitary adenomas or even apoplexy both clinically and radiologically, therefore detailed imaging is imperative in establishing the diagnosis. Management options include microsurgical procedures, endovascular interventions, and correcting any pituitary dysfunction. Patients should be discussed by a multidisciplinary team and individual co-morbidities and wishes considered. Intracranial aneurysms and pituitary adenomas can also co-exist, which may necessitate aneurysm treatment prior to adenomectomy, to minimise surgical complications.

14.) Surgical debulking of pituitary adenoma under local anaesthesia – World first!

AUTHOR(S):

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Abstract

We report a world first case of pituitary surgery under local anesthesia in an 86-year old male with multiple co-morbidities and visual deterioration.

The patient had a history of non-functioning macroadenoma presenting in 2011 with an acute history of headache and diplopia; conservative management and imaging surveillance had been advised due to his extensive co-morbidities. These included chronic kidney disease; significant left ventricular dysfunction; atrial fibrillation with mitral regurgitation and dilated atrium; and was considered to be unfit for adenoma excision under general anesthesia. In spring of 2017, he presented with new-onset deterioration of his left-sided temporal vision and progressive adenoma growth on serial imaging. MRI demonstrated large macroadenoma (volume 7.98 cm3) compressing the optic chiasm. There was a new left supero-temporal field restriction leading to inability to use his mobility scooter and subsequent loss of his independence. He was referred to us and after anesthetic assessment and discussion with him and his immediate family; we recommended surgical intervention as his only chance of regaining vision and independence. He accepted the risks of surgery given that his mobility was a major element for his quality of life, but a few days later whilst awaiting an urgent surgery slot, he presented with complete blindness. We proceeded with immediate surgical intervention. Local anesthesia was achieved by a greater palatine block, followed by a left nostril transsphenoidal pituitary approach and tumour debulking. Surgical intervention, and postoperative phase were uneventful. Post-operative MRI demonstrated 85% reduction of tumour volume (residuum 1.22 cm3). Visual loss however, only improved subjectively to perception of shades of grey (from no light perception), not objectively with visual testing.

Notwithstanding, this is the first description of successful surgery of a pituitary lesion under local anesthesia, and is a paradigm shift in the field of future pituitary surgery in appropriately selected cases.

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