Craniopharyngioma with intratumoral bleeding coexisting with cerebral venous sinuses thrombosis in a young female; a case report

Abstract

Introduction: Craniopharyngioma is a slow growing intracranial neoplasm, mostly benign in nature and arise from remnants of Rathke’s pouch. Incidence is equal among both sexes and can present in all age groups. Acute presentations with headaches are less common especially among adults. Herein we report an adult female with a history of long standing secondary amenorrhea first detected to have a craniopharyngioma when presented acutely with headache and double vision secondary to intratumoral bleeding.

Case presentation: A 26 year old female with history of secondary amenorrhea and primary hypothyroidism for many years presented with acute onset headache and double vision. On examination she was found to have bilateral papilloedema, left sided lateral rectus palsy and bitemporal hemianopia. She was averagely built and had normal secondary sexual characteristics. A non contrast computerized tomography of the brain revealed a hyperdensity in the pituitary area and magnetic resonance imaging of the brain was suggestive of a craniopharyngioma with bleeding in to tumour. Hormonal assays were in keeping with primary hypothyroidism and hypogonadotropic hypogonadism. Since she refused surgery we managed her conservatively with pharmacotherapy including hydrocortisone, thyroxin replacement therapy and gonadotropin treatment for secondary amenorrhea. Review at one month revealed she was free of headache and lateral rectus palsy has resolved but she persisted to have bitemporal hemianopia.

Conclusion: In adults craniopharyngiomas could present acutely with headache and double vision due to the rare complication of bleeding in to the tumour.
sided lateral rectus palsy. Her visual acuity was normal and visual field analysis showed bitemporal hemianopia. Examination of the respiratory system and abdomen revealed no abnormalities.

With the available information on admission we considered the diagnosis of pituitary apoplexy and commenced immediate management including intravenous hydrocortisone and intravenous fluid resuscitation. Her random blood glucose was 114mg/dL and serum sodium and potassium levels were 140mmol/L and 4.5mmol/L respectively. On admission, she was imaged with a non contrast computerized tomography (CT) of brain, which showed a hyper dense lesion in the region of pituitary fossa (Figure 1), without cerebral oedema. It was compatible with our clinical diagnosis and we proceeded investigations which revealed following hormone profile. Random Cortisol level was 1120mmol/l, Serum free thyroxin - 15.6pmol/L (10 – 28), TSH – 9mUL (0.4 – 4), Serum adrenocorticotropic hormone (ACTH) – 26.5pg/ml in the morning (10 – 60), Serum Prolactin - 1308mIU/L, follicular stimulating hormone (FSH) - <0.66mIU/mL (21 - 131), luteinizing hormone (LH) - <0.21mIU/mL (13 - 88), Serum Oestradiol – 20.9pg/ml (85 -498).

Clinical findings, hormonal and biochemical profile were indicating that her pituitary- gonadal axis was affected. She was imaged with magnetic resonance imaging (MRI) brain, which showed supracellular solid tumour with evidences of internal haemorrhages (Figure 2)– likely to be a craniopharyngioma (15mmx17mmx12mm), with optic chiasmal compression and no extension to cavernous sinus or brain stem. Pituitary gland is visualized separately in the pituitary fossa. MRR showed chronic venous thrombosis in superior sagittal, right transverse, and right sigmoid sinuses with multiple draining collateral cerebral veins without evidences of venous infarction. Other laboratory exams were; White cell count – 15400/mm 3 - Neutrophils-86%, Lymphocytes – 9%, Haemoglobin – 12.5g/dl, Platelet – 200 x 10 9/mm 3, Serum creatinine - 86 µmol/l, Serum albumin – 38g/dl, Total protein – 55g/dl, aspartate transaminase (AST) - 40 IU/L, alanine transaminase (ALT) – 38 IU/L, erythrocyte sedimentation rate (ESR) – 45mm in 1 hour, We did not commence her on anticoagulants since there was intratumoral bleeding and the venous sinus thrombosis was chronic without evidence of any infarctions.

Discussion

With the most likely diagnosis of craniopharyngioma with intratumoral bleeding causing bitemporal hemianopia, ophthalmoplegia and hypogonadotrophic hypogonadism we referred the patient for neurosurgical team. While waiting in the neurosurgical ward for surgery over a period of one week she was treated with intravenous hydrocortisone, and she was continued on oral thyroxin 100 µg daily. Her headache has gradually improved over a period of one week. Following explanation regarding the need for surgery patient did not consent for operation and wanted a conservative management. She was commenced on gonadotropin therapy in addition to hydrocortisone and thyroxin replacement therapy. Her oral thyroxin dose was increased to 100 µg daily. Review after one month of this treatment revealed that she had resolution of headache and lateral rectus palsy. But she had persistent bitemporal hemianopia. It was decided to follow her up monthly in the clinic and a repeat MRI brain was planned to perform in six month to assess the size of the tumour with re-explanation regarding the need for surgery.

Conclusion

Although craniopharyngiomas are usually slow growing tumors with indolent course, acute presentations with headache, double vision and ophthalmoplegia are possible even in an adult patient in case of intratumoral bleeding.
Acknowledgment
None.

Conflict of interest
None.

References