CASE REPORT

Acromegaly Presenting as Psychotic Disorder in a Patient with Familial Autosomal Dominant Polycystic Kidney Disease

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Abstract

Objective: To describe a rare case of Acromegaly presenting as psychotic disorder in a woman with familial autosomal dominant polycystic kidney disease (ADPKD). Method: Single case report. Result: We describe the case of a 50 year old woman, who presented with an acute psychotic episode. She had polycystic disease of kidney and liver and family history of polycystic disease. She presented with persecutory delusions, perceptual abnormalities, disorganisation and marked fluctuation in her behaviour. An urgent CT and subsequent MRI scan revealed a pituitary macroadenoma, extending into the cavernous sinus. The initial diagnosis of prolactinoma was revised to acromegaly. Her symptoms responded to combination of olanzapine and valproic acid, followed by transsphenoidal resection of the adenoma. Conclusion: This case highlights the need for investigation, especially of neuroimaging, in atypical presentations of psychosis, which may be first manifestation of rare disorders like acromegaly (German J Psychiatry 2006; 9: 136-138).

Keywords: acromegaly, psychotic disorders, polycystic kidney, autosomal dominant, pituitary neoplasms

Received: 24.7.2006
Published: 1.10.2006

Introduction

Acromegaly is a rare disorder with a reported annual incidence of 3 per million population. The clinical presentation of acromegaly consists of both physical features, which are characteristic and psychological symptoms. The psychological features of acromegaly have been described (Bleuler, 1951), consisting primarily of fluctuation in mood and change in personality. There have been few reports of acromegaly presenting as an acute psychotic episode. Further the association of acromegaly with familiar autosomal dominant polycystic kidney disease (ADPKD) has not been reported.

We report here a 50 year old woman with acromegaly presenting with psychotic illness, with history of autosomal dominant polycystic disease and family history of polycystic disease. Acromegaly due to GH hormone producing pituitary macroadenoma was confirmed on endocrinological investigations.
Case Report

BK is a 50 year old married Asian woman who was referred to the Home treatment (crisis resolution) service, with history of agitation and disruptive behaviour. Prior to the current episode, BK had been functioning well, caring for her family and working part time.

She had apparently been withdrawn and feeling depressed for 5 months, presenting with increased anxiety and perplexity for 4 to 5 days, prior to assessment in the Accident & Emergency department. She had not been sleeping well. She was labile in mood, with marked pressure of speech and paranoid delusions. BK believed that she was being subject to black magic by a family member that someone was spying on her and her family and expressed concern for the safety of herself and her family. Her cognitive functions were grossly intact, but detailed assessment was not possible. General physical examination revealed a firm, non-tender swelling in the right hypochondrium and epigastrium, with no deficits on neurological examination.

BK had history of hypertension and autosomal dominant polycystic kidney disease (ADPKD), with both renal and hepatic cysts. She was born in Nairobi and came to the UK when very young. There was no significant birth, developmental or childhood history. Her mother suffered from depression and received ECT. Her sister suffered from delusional disorder and there was family history of ADPKD.

She was commenced on Home Treatment. Home Treatment (Crisis Resolution) is 24 hour a day, 7 days a week service, which provides intensive support in the community to patients with mental health problems. She was initially prescribed zopiclone 7.5 mg at night and olanzapine 10 mg daily, which was increased eventually to 10 mg BD, in addition to lorazepam 1 mg TDS, in view of the deterioration in behaviour. She was admitted from delusional disorder and there was family history of ADPKD.

On admission, she presented with flight of ideas and disorganised behaviour, with her trying to disrobe in public in the ward. She continued to express paranoid delusions, as at presentation. She believed on occasions that the food was poisoned and that there were spirits in her room. She responded rapidly to intramuscular lorazepam 2 mg, when she was found to be mute and catatonic. She was distressed by fleeting auditory hallucinations of male voices. A score of 24/30 was obtained on Mini-Mental State Examination (MMSE) with deficits being in orientation to date and in concentration. On one occasion, she received intramuscular haloperidol and lorazepam to manage her physically aggressive behaviour. She was detained involuntarily under the Mental Health Act. BK received a course of vitamin B12 injections for low B12 level and she continued to receive treatment for hypertension. She was transferred twice to the Medical Ward, for hypokalemia and for bilateral pedal oedema.

The patient’s mental state continued to remain the same for nearly the next 2 months. As there was no improvement despite more than 2 weeks of treatment with olanzapine, this was stopped and risperidone 1 mg **_mante_** and 2 mg **_nute_** was commenced. However she developed skin eruptions, which improved with antihistamines and after stopping risperidone. BK was then started on treatment with aripiprazole. As she did not respond to 15 mg per day of aripiprazole, the medication was changed to olanzapine 10 mg per day and valproate semisodium 500 mg BD was added. There was a gradual improvement over the subsequent month with this combination of medication.

An urgent CT scan of head revealed an 11mm pituitary mass with some scalloping. MRI scan confirmed the presence of a pituitary macroadenoma, predominantly left sided and extending laterally into the cavernous sinus, and encasing the intracavernous component of the left internal carotid artery. There was an elevation of the optic chiasma. The initial impression was one of a pituitary adenoma, a prolactinoma, which was thought to be an incidental finding. Right temporal hemianopia could not be excluded on examination of visual fields. Routine blood tests (except for elevated ESR) and EEG were normal. Investigations revealed a prolactin level of 1986, growth hormone (GH) 26 and insulin-like growth factor 1 (IGF 1) 66.3 (8.9 to 32). Cushing’s disease as a cause was ruled out by performing low dose and an overnight dexamethasone suppression test and by measuring 24 hour urinary free cortisol. GH levels were not suppressed with the glucose tolerance test, which confirmed the diagnosis of acromegaly. IGF with growth hormone was between 22.7 and 17.8 mIU/L and an IGF 1 of 66.3 was strongly suggestive of acromegaly.

BK underwent transphenoidal surgery and has been recovering from it. She continues to be stable in her mental state, with euthymic mood and no psychotic symptoms. Though there were no physical signs of acromegaly, subjectively she reports that her shoe size has gone down two sizes since the surgery. She has little recollection of the time spent on Home Treatment or in hospital.

Discussion

The physical manifestations of acromegaly are well known, with many so-called giants in mythology and history now thought to have suffered from acromegaly, with Goliath’s defeat has been attributed by some to bitemporal hemianopia, secondary to a pituitary tumour. In 1886, Pierre Marie gave his name to the condition of acromegaly co-occurring with pituitary tumours (Pearce, 2002). The psychological features as well as changes in personality characterized by lack of initiative and spontaneity have been described (Bleuler, 1951; Lishman, 1997). Bleuler is said to have concluded that acromegaly was not associated with psychotic disorders, whilst others have reported marked psychomotor retardation and depression. However in a study of 51 patients with acromegaly, quoted by Lishman (1997), there was no increase in psychiatric morbidity in general or in depression in particular. Though there are reports of emotional problems in pituitary disease, a study of 93 patients with pituitary adenoma by Korali and colleagues (2003) could not find an increased risk of mental disorders.
There could be various reasons for the unusual presentation of acromegaly as a psychotic episode. The psychotic episode could be relapse of a pre-existing psychotic illness or could be related to treatment of acromegaly with dopamine agonists like bromocriptine. In patients with well established physical features of acromegaly, the associated stress may manifest as psychological symptoms. Finally, Jaquet (1997) hypothesized that there is an underlying dopaminergic abnormality in acromegaly, given that dopamine agonists have been used to reduce growth hormone (GH) levels and this abnormality could be related to the psychotic presentation. Given that acromegaly is a rare disorder, it is highly unlikely that its association with psychosis is purely by chance. Notwithstanding the lack of information regarding the pathophysiology, this case reinforces the need to rule out an organic cause for an atypical presentation of psychosis. Further, the role of routine neuroimaging needs to be re-examined due to false negative results on neurological examination. Further, MRI provides information which is not otherwise available (Marro et al., 1997).

In addition to acromegaly and pituitary adenoma, our patient had history of familial autosomal dominant polycystic kidney disease (ADPKD). We examined the possible relationship between ADPKD, acromegaly and pituitary adenoma. ADPKD is a common inherited disorder (Gabow, 1993), with cysts described mainly in ‘ductal organs’. The intracranial manifestations include arachnoid cysts and saccular (Berry) aneurysms. We were able to find only one case report of hypophysal incidentaloma in a patient with ADPKD but there have not been any reports of an association between acromegaly and familial ADPKD. As noted above, it is highly unlikely to be a chance association and hence this case might be the first in a series of case reports of associations between acromegaly and ADPKD, with as yet unknown significance.

References


