Rare disease

Kleine Levin syndrome in Malaysia: first typical case

Rafidah Bahari,¹ Syed Hassan Ahmad²¹Psychiatry Department, Cyberjaya University College of Medical Sciences, Cyberjaya, Selangor, Malaysia;²Universiti Teknologi MARA, Selayang, Selangor, Malaysia**Correspondence to** Dr Rafidah Bahari, rafidahbahari@cybermed.edu.my**Summary**

A 13-year-old boy presented with a 2 weeks history of tearfulness, childish behaviour, separation anxiety, hypersomnia, hyperphagia and sexual disinhibition following a brief episode of fever. He had been experiencing the episodes since he was seven. The episodes lasted from a few days to 3 weeks and would normally occur once in a year. Most of the time it started with fever and resolved spontaneously. In the past he described auditory hallucination but not this time. Examination revealed a slightly overweight adolescent male appearing appropriate to his age. During the session he was restless, sleepy and burst into tears frequently. He constantly asked his mother when he could go home to sleep. He was very childish and clung to his mother. He was given a trial of risperidone 1 mg to be taken once a day for 3 days. On follow-up he had completely recovered.

BACKGROUND

Kleine Levin syndrome is a rare illness typically presenting with periods of hypersomnia, megaphagia, hypersexuality and other cognitive and behavioural disturbances.¹ Usually affecting adolescent males, episodes normally last up to a few weeks and terminates with total and spontaneous recovery. Possibly first reported by Brierre de Bosmont in 1862, the condition however received its name from Willi Kleine who, in 1925, reported a series of cases of periodic hypersomnia and also Max Levin who described a case of periodic hypersomnia and excessive appetite in 1930.² Since then, only 186 cases were reported in America, Europe, Africa, Middle East and Asia in particular India, Japan, Taiwan, China and Pakistan.³ More recently, a case was reported in Thailand,⁴ but so far none from Malaysia. It is believed that this is the first report of such a case from this country.

CASE PRESENTATION

The patient was a 13-year-old Malay boy who came to the clinic with a 2 weeks history of hypersomnia, fatigability and increased appetite. He also displayed childish behaviour during the episode such as clinging to his mother, ringing her at work excessively and repeatedly asking the same questions. During this time he also lost his inhibition in which he sometimes walked around naked at home. He needed to be prompted to look after his cleanliness. According to his mother, the first episode began when he was 7-years-old. Since then it generally recurred once every year. In the beginning, the episodes only lasted a few days but later it became longer. This time it was the longest so far thus necessitating the consultation. The episodes would often be preceded by fever. There used to be associated hallucinatory experiences but not recently.

Apart from mild asthma, the patient was otherwise fit and healthy. He was developmentally normal and had normal IQ. However, his results in the recent major

examinations were poor due to the fact that he had a recurrence of illness at the time the examinations was being held. He had many friends and was a normally serious boy who was slightly shy.

His parents were divorced when he was 6-years-old but his mother had since remarried. At the time of the presentation, he lived in a boarding school during term time and was home during school holidays. He had good relationship with his biological and step father, but was not close to either of them. He was however close to his paternal grandparents and his mother. There were no other difficulties of note.

INVESTIGATIONS

Clinical examination revealed a slightly overweight boy with dark circles under his eyes. Examination of cognitive function was difficult to perform due to his poor cooperation. He was disorientated, and his attention as well as memory was severely affected. Systemic examination was unremarkable. No other investigations were indicated.

DIFFERENTIAL DIAGNOSIS

The main differential diagnosis in this patient was obstructive sleep apnoea. However, although he was slightly overweight, there were no other clinical signs or symptoms to support this diagnosis. Other differential diagnoses include excessive sleep with mood and behavioural changes due to brief psychotic disorder and major depressive disorder but again the clinical picture would not be consistent both illnesses.

TREATMENT

Unfortunately there was no evidence that any medication is beneficial for this condition. He was tried on a small dose of risperidone, but the mother reported no change in his condition. A few days after his presentation at the clinic, his symptoms improved.

OUTCOME AND FOLLOW-UP

A follow-up 1 week after the initial presentation showed that the patient had recovered fully. He was a completely different boy, polite, quiet and slightly serious but all his behaviours were appropriate. When asked, he had no recollections of what had happened to him in the past few weeks. He and his mother was given information about the condition and advised to be aware of any signs of recurrence. He was to be followed up in due course.

DISCUSSION

Klein Levin syndrome is an extremely rare disorder. Only about 200 cases had been reported⁵ globally over the span of 50 years. Only a handful of these were from the Asian region and so far, no case had been reported from Malaysia. When he was seen at the clinic, the patient was 13 but it had first presented when he was 7 years old. In a systematic review cited earlier, Arnulf *et al* stated that the median age of onset was 15 years but it ranged from 4 to 82 years.³ So, the age of presentation is appropriate for this patient. Since the first presentation, the patient's illness recurred yearly. He had been attending different doctors throughout the years but investigations had not revealed anything abnormal. Ramdurg in his review wrote that the median duration of the illness was 4 years and on average it terminated at the age of 23.⁶ He also found that patients on average experienced 12 episodes of illness in their lifetime. He also stated that on average, patients were their normal self for 6 months. The patient in this report had brief episodes of recurrences, ranging from 3 days to 3 weeks, with complete remissions for about 11 months. His precipitant was usually a mild flu-like illness which was supported by the literature to be the most common precipitant.⁷

Apart from hypersomnia, during the episodes the patient demonstrated disruptions in his cognitive function, in particular attention, orientation and memory. He had no recollections about what had happened during the episodes. He also presented with hypersexuality and hyperphagia. This was in keeping with a more recent review by Arnulf *et al* in which found that in episodes of illness, all the 108 patients he studied had symptoms of hypersomnia and cognitive disruptions while 66% had hyperphagia and 53% of which they were mainly male, presented with hypersexuality.⁸

In the literatures, there was no recommendation on specific investigations to diagnose the condition and it was done on the basis of the clinical presentation alone.⁹ However, a sleep study may be beneficial to rule out other sleep disturbances and evaluate the extent of the illness. However, since the episodes occur in an unpredictable

manner and are self-limiting, it is impossible to catch them during the episodes and perform the sleep study.

As for the management, different medications have been tried such as antipsychotics, antiepileptics, antidepressants and stimulants with some benefit, but good quality research is lacking in this area.¹⁰

In conclusion, the patient presented with typical symptoms of Kleine-Levin syndrome. It is a rare disorder, and so far there were no reports of patients from Malaysia, making this the first in the country. So far there good evidence-based management recommendations are lacking but fortunately the episodes are self-limiting. The patient and his mother were given education on the disorder and he was to be followed-up in due course.

Learning points

- Kleine Levin syndrome although rare presents all over the world in a typically characteristic manner.
- Although episodes appear harmless and self-limiting, it can be extremely disruptive due to its unpredictable and distressing symptoms.
- Currently no treatment or preventive measures have been consistently effective, and more research is needed in this matter.

Competing interests None.

Patient consent Obtained.

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