A case of ketotic hypoglycaemia with cataract

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Introduction

Ketotic hypoglycaemia (KH) is the most common cause of hypoglycaemia in non-diabetic children[1]. Ross SG, *et al*[2] first described KH in 1924. Reduced oral intake due to prolonged fasting, diarrhoeal illness and vomiting are identified triggers of KH. The condition is commonly seen among children aged 18 months to 5 years, and they grow out of it by 8-9 years[3]. A cataract is a common cause of visual impairment in children, accounting for 5-20% of blindness worldwide[4]. Cataract is seen in a number of metabolic disorders in children, and KH has been identified as a cause of cataract in childhood[5]. Here we describe a 4-year-old child with congenital cataract presenting with recurrent episodes of KH.

Case report

A 4-year-old boy who was diagnosed to have infantile cataract, was brought to the accident and emergency department with a history of difficulty in waking up in the morning. On admission, the child was drowsy, but the rest of the neurological examination was normal. His capillary blood sugar level on arrival was 24mg/dL. He was given a 10% dextrose bolus followed by an infusion and he recovered well after the initial management. There were ketone bodies in his urine. Blood ketone bodies were not measured due to unavailability. The rest of the basic biochemical investigations were normal, including venous blood gas. After emergency management, a detailed history revealed that he had missed his usual dinner the previous night. However, there was no associated febrile illness or vomiting. There was no history suggestive of ingestion of oral hypoglycaemic drugs at home. His weight, height and head circumference were between median to -1SD. Abdominal examination revealed no hepatomegaly or splenomegaly. He had a similar presentation nine months back, and at that time, the diagnosis was made as KH.

He was the second twin born to non-consanguineous healthy parents. The mother did not have gestational diabetes or pregnancy-induced hypertension. He was born via an elective caesarean section at 38 weeks of gestation. His birth weight was 2.15kg, and the birth weight of his twin sister was 2.5kg. Both had average Apgar scores at birth. Breastfeeding was established within the first few hours of birth. He had developed a convulsion on day 2, and his blood sugar at the time of seizure was 18mg/dL. He needed one intravenous 10% dextrose bolus followed by an infusion for 12 hours. There was no history of refractory neonatal hypoglycaemia. He was commenced on empirical antibiotics after a full septic screen. He had a negative septic screen and antibiotics were stopped after 48 hours. Neonatal discharge records revealed that he had ketone bodies in the urine at the time of hypoglycaemia. He was discharged on day 7, and the discharged weight was 2.2kg. He was thriving well while on exclusive breast milk. At the age of 5 months, he was diagnosed to have bilateral lamellar cataracts. He underwent corrective surgery at the age of 5 months and he was regularly followed up by an ophthalmologist. Subsequently, he had developed right-sided glaucoma as a complication of surgery. There is a history of developmental delay in the early years but currently he has age appropriate development.

He was extensively investigated to find a link between hypoglycaemia and cataract. His renal functions, liver functions and bone profile were normal. Urine for reducing substances (Clinitest and Clinistix) was negative, and serum galactose levels were normal. The short synacthen test did not show any adrenal insufficiency. Toxoplasma, rubella, cytomegalovirus and herpes simplex virus IgM antibodies (TORCH screening) were negative. His serum triglycerides, cholesterol, lactate, uric acids, acyl carnitine profile and amino acid profile were normal. In the absence of other aetiology, the diagnosis was made as cataract associated with KH and parents were educated about the management of KH.

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Discussion

Though KH is associated with childhood cataract, the exact mechanism is not known[3]. This association was first described by Scheie HG, *et al*[5] in 1964. Subsequently, Merin S, *et al*[6] in 1971 described a case series of 13 children with hypoglycaemia and infantile cataract. The study revealed that 7 out of 13 had KH, and this association was more common among males who were born with low birth weight. The most common type of cataract was lamellar cataract. Similarly, in this case, the child was born with low birth weight and had bilateral lamellar cataracts[6]. Furthermore, Merin S, *et al*[6] stated that the cataract was not "congenital" but "infantile" because the abnormality was not evident at birth. Though this association was described a few decades back; an extensive search in PubMed and Google Scholar did not reveal recent studies related to this association.

In 1975, Chylack LT[7] carried out a study among rats and showed that hypoglycaemia results in the reduction of hexokinase activity in the lenses, which would lead to the development of cataract. Further, the study revealed that 20 hours of hypoglycaemia leads to lamellar cataract and prolonged hypoglycaemia more than 48 hours results in nuclear cataract[7]. Though cataract can occur irrespective of the aetiology of hypoglycaemia, there is a high tendency to develop cataract among children with KH compared to idiopathic hypoglycaemia[8]. Wets BJ *et al*[8] in 1982 evaluated 40 children with KH for the development of cataract. Of them, 15 had cataracts, and on three occasions, the development of the cataract preceded KH. Moreover, the study revealed that the mean time interval between the development of cataract and hypoglycaemia was three and a quarter years. However, the study has not discussed whether these children had neonatal hypoglycaemia or not, especially among children who developed cataract before the first episode of KH. In addition to cataract, infantile glaucoma, nystagmus, esotropia, and optic atrophy have been associated with hypoglycaemia in children[6]. This child had developed glaucoma in one eye as a complication of cataract surgery. Other ocular abnormalities were not evident in this child.

Conclusion

It is essential to conduct further studies to assess the exact prevalence and risk factors of cataract in children with hypoglycaemia. Moreover, clinicians should be aware of the possibility of developing cataract in children with neonatal hypoglycaemia and KH.

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