An Unusual Presentation of Type 1 Diabetes

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Introduction

Hyperglycemic hyperosmolar state (HHS) and diabetic ketoacidosis (DKA) are life-threatening emergencies in diabetic patients. While DKA at presentation of Type 1 Diabetes (T1D) in children represents about 25% of cases, HHS is very unusual as a first presentation of T1D in children and adolescents. The exact incidence in the pediatric population is not known.

Case Presentation

History:

- 10 year old Haitian-Sudanese boy presented to ER for obtundation
- 10 day history of polydipsia, polyuria, later vomiting
- On day of presentation, he had difficulty rousing

Presentation in the ER:

- VS: HR 140 bpm BP 87/61 mmHg
- GCS 8
- Initial labs (see laboratory results): BG 130.4 mmol/L, serum Na 125 mmol/L, high anion gap metabolic acidosis, 4+ glucosuria and 1+ ketonuria

Initial Management:

- NS bolus 20cc/kg x3 for hypovolemic shock
- DKA protocol: insulin 0.1U/kg/hr, double maintenance IV fluid with KCl
- Urgent CT and MRI head (normal)

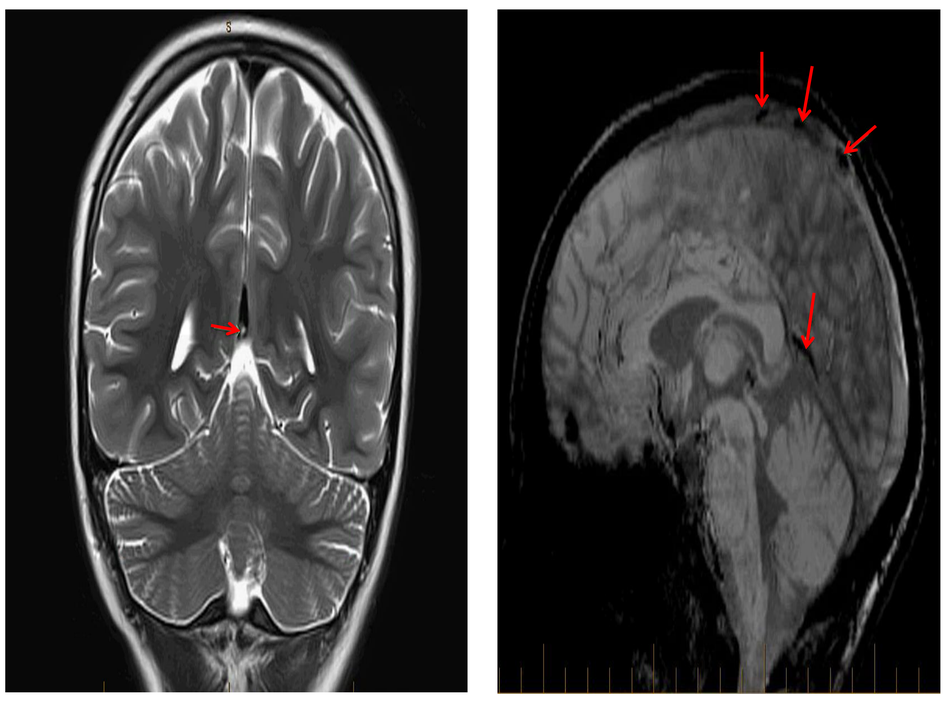
Hospital Course & Complications:

- Admission to ICU for profound dehydration and diminished LOC requiring intubation and inotrope s
- Double maintenance of IV fluid continued
- Agitation on day 3, found to have superior sagittal and straight sinus thrombosis, no hemorrhage (see image)
- Unilateral vocal cord paralysis without clear etiology
- Non-pressure ulcers over the ischia region and in the left gluteal fold, requiring surgical debridement x2

Condition at Discharge:

- Residual unilateral vocal cord paralysis
- Grossly normal neurological exam
- BG were well controlled on 1.2 U/kg/day of insulin

Figure 1: Brain MRI



MRI brain shows non-occlusive thrombus in the straight sinus (left) and superior sagittal sinus (right) with no hemorrhage.

Table 1: Summary of laboratory findings

	References	Unit	Initial	6 h	12 h	24 h	48 h
Venous pH	7.34 - 7.44	pH	7.10	7.15	7.19	7.22	7.28
pCO2	41 - 51	mmHg	30.7	58.3	48.1	45.4	41.3
bicarbonate	21 - 26	mmol/L	9.2	18.8	17.6	17.4	18.5
Base excess	-3.0 to - 3.0	mmol/L	-18.7	-8.2	-9.2	-8.8	-6.9
Glucose		mmol/L	130.4	51.2	40.0	35.0	22.7
Sodium	137 - 144	mmol/L	125	162	170	176	169
Urea	2.0 – 6.8	mmol/L	25.5	24.1	21.2	19.6	13.3
Creatinine	31 -63	umol/L	352	306	277	212	165
S-Osmolality	282 -296	mOsm/kg	451	440	417	390	372
Urine ketones	negative		trace	negative	negative	negative	negative

Discussion

- The classic presentation of HHS in children is usually in obese adolescents with T2D.
- Diagnostic criteria for HHS: BG >33 mmol/L, S-osmolality >330 mOsm/kg, absence of significant ketosis and acidosis.
- The symptoms tend to occur more gradually over a relatively long period.
- Excessive drinking of soft drinks and juice was part of the cause of the presenting severe hyperglycemia .
- Fluid deficit is estimated to be double that associated with DKA alone, and as such tends to be grossly underestimated.
- Our patient presented with both HHS and mild DKA and fortunately survived with few sequelae.
- At follow-up he had progressed to have honeymoon period lasting five months and continues to have well-controlled diabetes with recent HbA1c of 7.1% on 0.75 U/kg/day of insulin

Conclusion

- HHS usually presents with extreme hyperglycemia, hyperosmolality and profound dehydration and absence of significant ketosis and acidosis.
- Although HHS and DKA have been described as distinct entities, one third of patients exhibit findings of both conditions.
- DKA usually develops faster than HHS, hours to days in comparison to days to weeks in HHS.
- HHS requires more aggressive treatment in terms of volume expansion and electrolyte deficit replacement than those with isolated DKA to avoid the risk of thrombosis. The degree of volume deficit is frequently underestimated.

References

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