Caudal Regression Syndrome with bilateral renal agencies in an infant of diabetic mother

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Summary

We report a 36 year old Omani lady with Type 2 diabetes who gave birth to pre-term child with 'mermaid deformity'or Sirenomelia.

Case Report

A 36 year old diabetic lady was referred to us in the fourth month of her pregnancy. She had been pregnant 11 times including the present one. She had 8 full term normal deliveries out of which 7 were alive and healthy. The 8th was a full term stillborn 6 years ago. Diabetes was discovered when a raised blood sugar was found after the stillbirth. She was educated about diabetes in detail and was told to report to the hospital as soon as she wanted to conceive so that she could be shifted to insulin therapy and controlled meticulously. All the treatment options were discussed with her. When the diet alone could not bring diabetes under control, she opted for oral hypoglycemic drugs. Her 9th and 10th pregnancies ended in abortions of 2 months each. Her blood sugar status was not definitely known as she disliked coming to the hospital, however from whatever hospital records were available, the control was not exactly ideal. Though several attempts were made to improve her control of blood sugar the success was limited as she could not restrict her traditional diet which consisted of large amounts of dates and sweet fruits. In the present pregnancy she was more receptive to the idea of better control because of trauma of two successive abortions. She came for follow up more often than before but not as frequently as she could have for higher control, and certainly not early enough for preconceptional control of her diabetes. The first time we saw her in her 4th month of pregnancy. Her random blood sugar was 12 mmol/l at that time. She was immediately put on insulin therapy and blood sugar was brought down to a fasting value of 5 mmol/l and post prandial of 8 mmol/l. The blood sugar was done 5 or 6 times during pregnancy which was the maximum possible given the poor compliance of the patient but was reasonable, the values being in the range mentioned above. An ultrasound done in the 6th month of pregnancy revealed bilateral renal agencies and oligohydramnios. She went into labour in the 31st week of pregnancy and reported to the labour ward in full dilatation with absent membranes. Within fifteen minutes of admission she delivered vaginally a child with gross deformities.

The child weighed 1.480 Kg and on examination, the head, neck, chest and heart were normal. The abdomen was soft. The axial skeleton was poorly developed with exaggerated spinal curvature and narrow pelvis. The sacrum and perineum were absent and the external genitalia and anal openings were absent. Instead, the pelvis gave origin to a single curved structure in the place of two legs ending into a foot like structure with 8 toes. The child was having gasping respiratory movements and the heart rate was 60 per minute. Prematurity of 26 weeks was noted. Resuscitation was done with sugar and oxygen. Incubator care was given but the child survived only for 7 hours and expired. No radiological examination was consented to by the mother.

Discussion

Sacral agencies syndrome in its extreme form presents as sirenomelia or mermaid deformity. As a group it represents a host of abnormalities ranging from lower lumbar to sacral spinal agencies or hypoplasia. [1,2,3]. Lower limb underdevelopment is a common feature in most of these disorders. Fused ilia or an iliac amphiarthosis may also occur.[4]. In those with sacral agencies as compared to sacral hypoplasia, neurologic deficit is more pronounced and is associated with clubfeet deformity, wasting of the muscles of the lower limbs and incompetence of urinary and anal sphincter. The reason for these neurologic deficits in these cases is because of underdevelopment of the corresponding nerve roots and distal part of spinal chord. Multiple somatic abnormalities like intraspinal lipomas, tuft of hair, dimple or subcutaneous lipoma may be found in these children. [5].

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In our case the lower limbs were fused to form a mermaid like structure and these were no genitalia or anus. In addition our patient also had severe intrauterine growth retardation as evidenced by discrepancy in gestational age and morphological age of the baby and renal agencies.

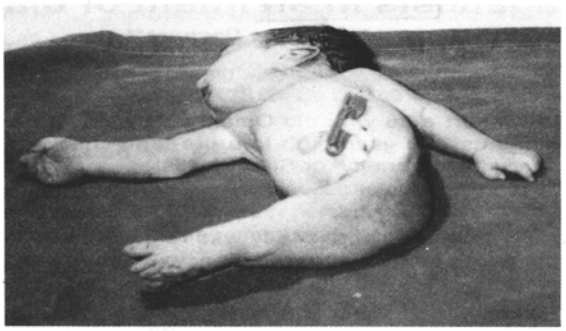
In less complete varieties there may be only sacral hypoplasia and urinary tract infection and therefore it has been suggested that the sacrum should be examined in all cases of urinary tract infections in the child born to diabetic mothers.[3,4,6].

Nineteen percent of the children with sacral agencies are born to diabetic mother.

On the other had congenital malformations are more common along the infants of diabetic mother with poor control of diabetes, the proportion being 22% in one study, but the proportion decreased to 3% in the group of infants of diabetic mothers with good control of diabetes at 14 weeks in the same study.

Although the blood sugar was within the acceptable range whenever measured we can not be very sure about the glycaemic control as the glucose profile and HbA1 could not be done due to the poor compliance of the patient.

Fig. 1



Caudal Regression Syndrome
with Renal Agenesis

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