Case Report

Congenital absence of ribs: a rare association in infant of diabetic mother

Pradipprava Paria*, Sutirtha Roy, Sibnath Gayen, Probodh Ch Mondal, Prativa Biswas, Sibarjun Ghosh

Department of Pediatric Medicine, R G Kar Medical College, Kolkata, West Bengal, India

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*Correspondence:
Dr. Pradipprava Paria,
E-mail: drpradip83@gmail.com

ABSTRACT

Maternal diabetes, type 1, type 2 or gestational diabetes has an increased risk of congenital malformation in fetus. It is also the most common cause of perinatal mortality in these infants. Although various malformation reported in literature, our case of congenital absence of ribs and adjacent intercostal muscles in an infant of diabetic mother is not reported earlier. Poor glycemic control may be the cause of teratogenesis. Asymptomatic single rib absence is managed conservatively but for absence of multiple ribs, surgical repair is required.

Keywords: Infant of diabetic mother, Congenital absence of ribs, Surgical repair

INTRODUCTION

It is well known that maternal diabetes, type 1 or type 2 including gestational diabetes, increase the risk of congenital malformations. The most frequent types of malformations involve the central nervous, cardiovascular, gastrointestinal, genitourinary, and skeletal systems. The overall risk is 8-15%, with 30-50% of perinatal fatalities related to major congenital malformations. Poor glycemic control early in pregnancy directly correlates with a higher incidence of congenital malformations. While many fetal malformations occur as a result of maternal diabetes, multiple rib abnormality with absence of thoraco-abdominal muscles are not included. We here reported a neonate who was born to mother with type 2 DM, had absence of anterior and anterolateral parts of multiple lower ribs in right side, absence of adjacent intercostal muscles and associated bulging of abdominal viscera, mainly liver.

CASE REPORT

The full term female newborn was product of a non-consanguineous parent delivered by emergency cesarean section due to fetal distress to a 36 year old second gravid mother. The pregnancy was complicated by type 2 diabetes in mother and she was on insulin injection. Her HbA1C at seven month of gestation was 7.2. HIV and hepatitis B & C serology were negative. Antenatal TORCH screening was also negative. Her thyroid hormone profile was within normal limit. USG abdomen at 24 weeks of pregnancy did not reveal any obvious congenital anomaly or poly/oligo-hydroamnios. The birth weight of the child was 4000 gm, head circumference 35.5 cm, chest circumference 37 cm and length was 50cm. On physical examination, the infant had large, plethoric and moon faced with hairy pinna. Respiratory distress was there with visible central cyanosis. Temperature was 37.2°C, heart rate 134 beats/minute, and respiratory rate 84/ minute. Blood pressure was 76/56

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mmHg. Saturation was 70% without oxygen but with 6 lit/min oxygen flow it became 88-94% both in preductal and postductal sites. On systemic examination, there was obvious bulging on right lower thoracic and upper abdominal region, the protruding mass moves with respiration paradoxically. On palpation, the mass is soft, globular and with liver like consistency. Bilateral air-entry was there, but a bit diminished in right inframammary, infraaxillary and infrascapular region. On cardiovascular examination, apex was on left 4th intercostal space, just inside the midclavicular line. No other added sound was there over whole of the precordium except s1 & s2. Other systemic examination was within normal limit. Spine was normal, no evidence of any neural tube defect or caudal regression syndrome was there (Figure 1).

We resuscitated the baby with oxygen, fluid and antibiotic. We had applied adhesive strapping for flail chest. CT scan thorax showed, congenital absence of anterior and anterolateral parts of multiple lower ribs in right side, absence of adjacent intercostal muscles and associated bulging of adjacent abdominal viscera, mainly liver. Bronchial tree was normal (Figure 3 & 4).

On investigation, complete blood cell count was: Hb: 21.43 gm%, TC: 12,000/cmm (N 78, L 17, E 4, M 0, B 0) and platelet count: 1.4 lac/cmm. Serum sodium and potassium were 132 mmol/L and 5.5 mmol/L respectively. Serum urea/creatinin (26/0.78) was also within normal limit. The newborn had maintained her blood sugar throughout the first 48 hours of observation. Portable chest X-ray showed 11 pairs of ribs, deformed 5th rib with wide gap between 5th and 6th, scoliosis in mid thoracic region (Figure 2).

No significant abnormality was in liver, gall bladder, urinary bladder and pelvic organs but left kidney was enlarged without any focal lesion and right kidney was hypoplastic (Figure 5).
Our patient presented with respiratory distress since birth with deficient anterior and anterolateral parts of multiple lower ribs in right side and associated absence of adjacent intercostal muscles. After extensive search we could not find out such a type of presentation of infant of diabetic mother in literature. Ribs develop from cartilaginous costal processes of the developing thoracic vertebrae. Development begins at 9 weeks; secondary ossification centers appear at 15 years. The first seven “true” ribs connect to the sternum via the costal cartilages by day 45. The lower five “false” ribs do not articulate with the sternum. The etiology of rib absence remains unclear. It was postulated that a local blood-supply insufficiency might result in rib dysplasia during the embryonic period. The absence of parasternal 2nd-5th ribs is common and usually accompanied with dysplasias of mammary glands, subcutaneous tissue and pectoralis muscles. Absence of 6th-12th ribs is rare and usually accompanied with meningocele, diaphragmatic eventration, congenital heart disease etc. Short ribs do not extend as far anteriorly as the sternum. As a result volume of the chest is diminished which restricts respiratory motion and causes respiratory insufficiency. Short ribs constitute an integral part of several syndromes. It is more common to see 11 pairs of ribs in the absence of associated anomalies; this situation occurs in 5%-8% of normal individuals.

Usually, asymptomatic single rib absence is managed conservatively. But when several ribs are absent paradoxical breathing may occur i.e., malacic zone is depressed during inspiration and prominent during expiration. Such infants usually manifest dyspnea and cyanosis soon after birth. Emergency operation is necessary in these cases. If the range of rib absence is small, local compression bandaging is used to control paradoxical breathing. However, immediate operation should be performed for infants with repeated infection in the respiratory tract. Options are: for small-range rib absence, adjacent self-rib transplantation, and coverage by the skin flaps by muscles or polyethylene mesh. Large-range rib absence is treated with sternum-reversal operation. The symptoms of untreated patients can be improved with increasing age, but severely deformed spinal column and thorax may be inevitable.

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REFERENCES


