



Case Report

<http://wjpn.ir>**VACTERL Association in a Newborn – A Rare Case Report**Mohammad Hosein Lookzadeh^{1,2}, Fatemeh Tahghighi^{1,2*}, Abdolhamid Amooee³¹ Department of Pediatrics, Shahid Sadoughi University of Medical Sciences, Yazd, Iran² Mother and Newborn Health Research Center, Shahid Sadoughi University of Medical Sciences, Yazd, Iran³ Department of Surgery, Shahid Sadoughi University of Medical Sciences, Yazd, Iran

Received: 05 April 2018

Revised: 09 July 2018

Accepted: 11 September 2018

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Keywords:

VACTERL;

Anomaly;

Birth Defects;

Newborn

ABSTRACT

Syndrome or association VACTERL is a group of several birth defects of congenital anomalies in an individual. There must be at least 3 anomalies simultaneously for this syndrome to be referred, including spinal anomalies, anorectal anomalies, cardiac disorders, esophageal atresia with tracheoesophageal fistula, renal anomaly and limb anomalies. The organs involvement in VACTERL may present different severity and quality, from asymptomatic to life-threatening cases. Various studies have reported the other congenital associations such as cerebrovascular and pulmonary anomalies in addition to the above-mentioned called as the non-VACTERL association. The patient in this study had all 6 VACTERL syndrome criteria. The feature of this patient was the involvement of his limb and kidney anomaly, which were different on both sides. However, in previously reported cases, these two anomalies were both in one direction and on the same side. Finally, the VACTERL syndrome and Non-VACTERL Association in this patient represented in the form of esophageal atresia with trachea esophageal fistula and atrial septal defect, and the presence of a kidney with severe hydronephrosis and sacral agenesis and imperforated anus, recto vesical fistula and limb anomalies in the form of one-phalanx fingers on the left.

Introduction

VACTERL is a set of several congenital anomalies that involve different organs of the body. The prevalence of this syndrome is about one in 10000 to 40000 live births^{1,2} and is more

often seen in the infants of diabetic mothers³ and mothers who have used statin group drugs during pregnancy.^{4,5} Most cases are sporadic; however, familial cases have been rarely reported in this syndrome.⁶ Treatment usually involves supportive measures, medical treatments, and surgical repair.⁷

Case Report

The patient was a 5-hour-old female infant who was sent to a NICU Center due to severe sialorrhea and respiratory distress. The baby was born with a gestational age of 33 weeks by normal delivery from parents with a distant family relationship. The mother had no history of chronic diseases like diabetes or history of surgery and exposure to radiation or specific drug use and has been under the full supervision of an obstetrician. No abnormal condition has been reported in the embryonic ultrasound. The baby was placed under oxygen hood due to tachypnea, and then, the naso-gastric tube was inserted, which faced a barrier (Figure 1).

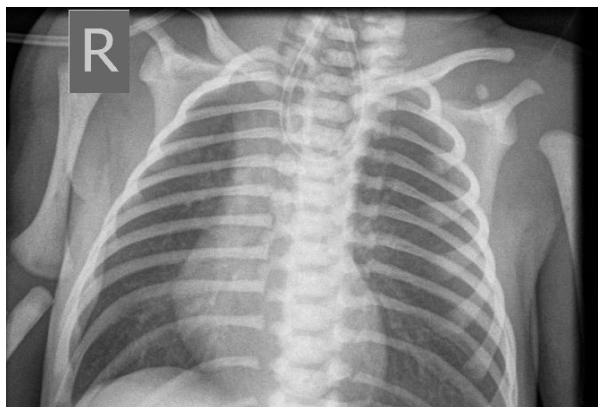


Figure 1. The picture show esophageal atresia which the NG tube recoil

It was found in other examinations that the anal is closed and meconium is excreted from the tip of Mea (through the fistula between the rectum and the urinary system) (Figure 2).

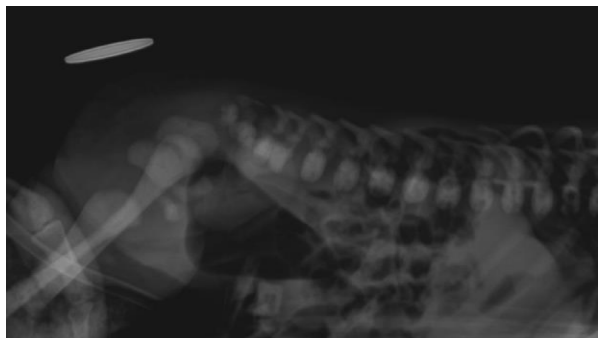


Figure 2. X-ray abdomen showed dilation of the descending and recto sigmoid colon without the gas shadow in the distal rectum and also sacral agenesis

The left hand fingers were one-phalanx (Figure 3), while other organs were normal. In search of other associated anomalies, we detected sacral agenesis in the lumbosacral radiography (Figure 2).



Figure 3. The picture show one phalanx fingers of left hand

In renal ultrasonography, the right kidney showed agenesis with severe left renal hydronephrosis (Figure 4). ASD was reported in echocardiography. None of the testicles were touched in the scrotum in the genital examination, which was reported in a two-way UDT ultrasound. The infant navel (umbilicus) contained an artery and two veins.



Figure 4. The picture show moderate to severe unilateral hydronephrosis

The newborn was undergone a surgery to repair esophageal atresia and close the fistula between the esophagus and the tracheal tube and also to place a colostomy and close the rectovesical fistula during the first week and received 4 weeks of special care, and was finally discharged with oral breastfeeding. The baby was hospitalized again after a week due to fever and lethargy and treated with antibiotics due to urinary tract infection. However, he was

subjected to vesicostomy surgery due to lack of response to antibiotic therapy, increased urea and creatinine levels, severe hydronephrosis of the left kidney and neurogenic bladder. Eventually, the patient developed seizure due to urosepsis, leading to his death.

Discussion

VACTERL was first reported in 1972 by Quan and Smith.⁸ Seventy percent of anomalies include esophagus and tracheal involvement, while the involvement of the vertebra and heart accounts for 68% and 65%, respectively.⁹ The heart involvement is more likely to occur in the forms of VSD, ASD, or Tetralogy of Fallot and less as the displacement of large vessels. Anal atresia is seen in 55% of cases. The kidneys involvement has been seen in 51% of the cases.⁹ In reported cases, 70% include other organs' involvement as defects in the development of the genitalia, anomalies of the respiratory system and vascular anomalies as umbilical cord unit artery in NON VACTERL association cases. Limb anomalies have been seen in more than 70% of cases as the small thumb, polydactyly, syndactyly (webbed fingers) and the forearm bones dysplasia.⁹ In most reported cases, limb involvement is associated with kidney involvement in the same position.⁶ The feature of the case study had all the six known anomalies in the VACTERL syndrome. However, in the previous cases, the limb and kidney anomalies were reported on one side. In case of this patient, the limb involvement was on the left side and the kidney anomalies were on the right side, which was different from all the previously reported cases.

Conclusion

Multiple congenital anomalies is rare but nowadays pediatricians and obstetricians must consult with relative parents to prevent congenital disease like this.

Conflict of Interests

Authors have no conflict of interests.

Acknowledgments

The authors would like to express their

profound thanks to all the study participants who contributed to this research.

How to Cite: Lookzadeh MH, Tahghighi F, Amooee A. VACTERL Association in a Newborn – A Rare Case Report. *World J Peri & Neonatol* 2019; 2(1): 44-6.

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