



Case Report

Congenital Scoliosis Associated with Tobacco and Illicit Substance Use

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Abstract

Congenital scoliosis is a complex spinal deformity that arises during fetal development. It is often associated with other congenital anomalies, such as those found in VACTERL syndrome, which includes vertebral anomalies, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, and limb abnormalities. This case report presents a unique case of congenital scoliosis associated with VACTERL syndrome and explores the impact of substance use during pregnancy.

Keywords: Congenital scoliosis; Birth defects; Substance use; VACTERL

Introduction

The majority of congenital scoliosis cases are not inherited but instead result from an insult to the developing fetus early in gestation [1]. With other important structures developing in this same timeframe, congenital scoliosis is often associated with other malformations. VACTERL association, occurring in 1/10,000 to 1/40,000 births, is the nonrandom occurrence of at least three of the following congenital anomalies: Vertebral, Anal, Cardiac, Tracheoesophageal, Renal, and Limb [2].

Case Presentation

A 28-year-old, G1 mother was referred to a high-risk delivery center at 37-weeks gestation after a third trimester ultrasound revealed congenital scoliosis of the lower thoracic spine. Prior to the consultation appointment with the high-risk obstetrics center, the mother presented to the delivery center with premature rupture of membranes.

Maternal medical history was significant for anemia, anxiety, major depressive disorder, substance use disorder, and post-traumatic stress disorder. Pregnancy was complicated by insufficient prenatal care, trichomonas infection, fetal growth restriction, fetal spine abnormality, and maternal use of tobacco, methamphetamine, MDMA (3,4-Methylenedioxymethamphetamine), tetrahydrocannabinol (THC), and cocaine. Maternal toxicology screens during pregnancy were positive for MDMA, methamphetamines and THC. Methamphetamine use was reported to be three times per week and tobacco use was quantified at one-half pack per day.

Maternal infectious labs, including group B streptococcus, were negative. At hospital presentation, the mother's urine drug toxicology screen was positive for methamphetamines and THC. Rupture of membranes was approximately 19.5 hours prior to delivery. At delivery, the infant was vigorous with a strong cry and did not require resuscitation. The Apgar scores were 9 and 9 at 1 and 5 minutes. The birth weight was 1930 grams. The admission examination was insignificant except for visible curvature of the lumbar spine. The infant was admitted to the neonatal intensive care unit for monitoring of feeds, temperature, and vital signs due

to weighing less than 2000 grams and for further investigation of the spinal curvature.

An x-ray of the spine (Figure 1) was obtained with anteroposterior and lateral views. Imaging revealed multiple vertebral segmentation anomalies within the thoracic and lumbar spine, with evidence of dextroscoliosis of the thoracolumbar spine. There was mild kyphotic curvature of the thoracolumbar spine noted. The x-ray also showed 14 pairs of ribs. The pediatric orthopedic surgery service was consulted and recommended outpatient follow up. Due to the vertebral body anomalies in addition to congenital scoliosis, further investigation was performed.



Figure 1: Anteroposterior x-ray of the spine.

Renal ultrasound showed a solitary right pelvic kidney with normal urinary bladder. The pediatric urology service recommended outpatient follow-up. An ultrasound of the spine and contents showed a low-lying conus concerning for tethered cord. The pediatric neurosurgery team recommended an MRI of the spine in 3-6 months. An echocardiogram was performed which showed a moderate-sized secundum atrial septal defect, dilated right atrium, flattened diastolic interventricular septum position, and hyperdynamic left ventricular systolic function. The pediatric

cardiology service recommended outpatient follow-up with repeat imaging in 6 months. With this constellation of findings, the infant had three characteristic features and met criteria for VACTERL association (Vertebral - segmentation anomalies and scoliosis, Cardiac - ASD, and Renal - solitary kidney).

The infant progressed on oral feeds and took full feeds by day of life (DOL) 7. The baby was discharged on DOL 12 with recommendation for outpatient follow up with the infant's primary care provider, pediatric neurosurgeon, pediatric orthopedic surgeon, pediatric cardiologist and pediatric urologist.

Discussion

The patient was initially diagnosed with congenital scoliosis. The majority of cases involving congenital scoliosis result from an insult to the developing fetus during early stages of gestation [1]. There are other important structures developing during this timeframe; therefore, congenital scoliosis is often associated with malformations such as spinal cord, heart, and kidney anomalies. In one study, 61% of infants diagnosed with congenital scoliosis experienced aberrant development in other systems. This phenomenon was considered as early as 1972 when Quan and Smith established the VATER association (Vertebral anomalies, Anal atresia, Tracheoesophageal fistula and/or esophageal atresia, and Radial dysplasia) [2,3]. The acronym was later revised to include cardiac and renal anomalies to become what it is today—VACTERL association [2-4].

VACTERL association is the nonrandom occurrence of at least three of the following congenital anomalies: Vertebral, Anal, Cardiac, Tracheoesophageal, Renal, and Limb anomalies [5]. The infant met criteria for diagnosis of this association because she exhibited 3 of the characteristic features: Vertebral - segmentation anomalies and scoliosis, Cardiac - ASD, and Renal - solitary kidney. This case is unique because the infant had a documented exposure to tobacco and illicit substances during the embryonic period which is crucial for development of the spinal column and related structures [1]. Therefore, we have reason to believe that exposure to these drugs (tobacco, methamphetamine, MDMA, cocaine, and THC) played a significant role in her developing this rare association which is seen in 1/10,000 to 1/40,000 births [6].

The exact etiology of VACTERL association is poorly understood. Due to heterogeneity in presentation as well as the complexity of embryological development, it has not been determined if this association is due to genetics, teratogenic causes, random errors, or a combination of the three [6]. VACTERL association is caused by a combination of genetic susceptibility plus exposure to teratogens or environmental triggers in utero [6,7]. This model would support our hypothesis that the use of tobacco and illicit substances during the embryonic period played

a significant role in our patient's presentation. Research focusing on teratogens and environmental exposures has produced multiple case reports and a few retrospective trials linking specific exposures to this condition [8-12] One such study isolated tobacco as having statistically significant relationships with VACTERL association, but the results were only significant after adjusting the odds ratio for potential confounding variables [13]. Other studies have shown maternal exposure to estrogen and/or progesterone products as well as maternal diabetes to be statistically significant variables in infants who developed this association [14]. However, this patient did not have either of these exposures [6,11]. There have been no reported cases of VACTERL association caused by the use of tobacco, methamphetamine, MDMA, cocaine, or THC. Further studies are needed to determine which substances contributed to the development of VACTERL in this case.

Disclosure statement

There are no financial disclosures to be made or conflicts of interests pertaining to this case report. This case report examines the cases of a pediatric patient delivered and treated at University of Arkansas for Medical Sciences Medical Center, Little Rock, AR.

Informed Consent

Written informed consent for patient information and images to be published was provided by the patient's legally authorized representative.

Ethical Statement

This case report adheres to the relevant ethical guidelines to ensure the protection of patient rights, confidentiality, and ethical conduct. Our institution does not require IRB review and approval for case reports or series containing information from less than 4 patients.

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