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Case Report

Sacral Dysgenesis As A Part Of Caudal Regression Syndrome - A Case Report

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ABSTRACT

Caudal Regression Syndrome (CRS) is a rare syndrome occurs when sacrum or lowest part of spine does not form fully in utero. The actual cause is known although there is a relation with maternal diabetes. The incidence is one in 25,000 live births. The diagnosis can be made by ultrasound in first trimester of pregnancy and postnatal MRI (Magnetic Resonance Imaging). We present a case of 6 year old male with sacral dysgenesis associated with other spinal abnormalities.

***Key Words**: CRS, Syndrom*

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INTRODUCTION

Caudal Regression Syndrome (CRS) is a rare syndrome characterized by incomplete development of distal spinal segment. [1] The incidence of CRS is estimated to be one in 25,000 live births. [2] It occurs due to the insult to the caudal region of the embryo in utero by as early as 4th week of gestation. In about 16% of cases it is believed to be caused by maternal diabetes and cause for 84% of cases is unknown.[3] Some use the sacral dysgenesis as a synonym for CRS. But sacral agenesis is the spinal part of the syndrome in which a part of sacrum with vertebrae caudal to it is missing or malformed, whereas CRS is the syndrome or group of symptoms.

Sacral agenesis or sacral regression result in varying degree of development failure including a partially formed or absent lower half of the spine, orthopaedic malformations, spinal cord defects and related motor and sensory deficits. Other congenital anomalies, such as genitourinary defects(i.e. unilateral or bilateral renal absence, renal displacement and fused urinary tubes), gastrointestinal defects (inability to control bowel movements and closed anus) and cardiac disease may be present. [4,5]

A preliminary diagnosis can be made in prenatal period by ultrasound, during the first trimester of pregnancy, but must be confirmed after birth. The severity of the disease is determined by examination of the newborn by postnatal ultrasound and MRI (Magnetic resonance imaging). [6]

CASE REPORT

This case of 6 year old male child with sacral dysgenesis was observed during the observation of MRI films for the measurements on sacrum with regards to the study on 'Regional & Sexual Differences on Sacrum- a study on MRI and Dry Bones'. Patient was retrospectively followed. The entire of the patient was taken in detail. MRI of the patient was carried out in the

lumbosacral spine using T1, T2W sequences in multiple planes and cervicodorsal spine screening was performed on T2W sequences in sagittal plane. The MRI images showed hypoplasia of sacrum with absence of caudal one segment of sacrum and entire coccyx vertebrae as shown in (Fig. 1). There was an abrupt, club shaped termination of spinal cord at level of T12 (Fig. 2).

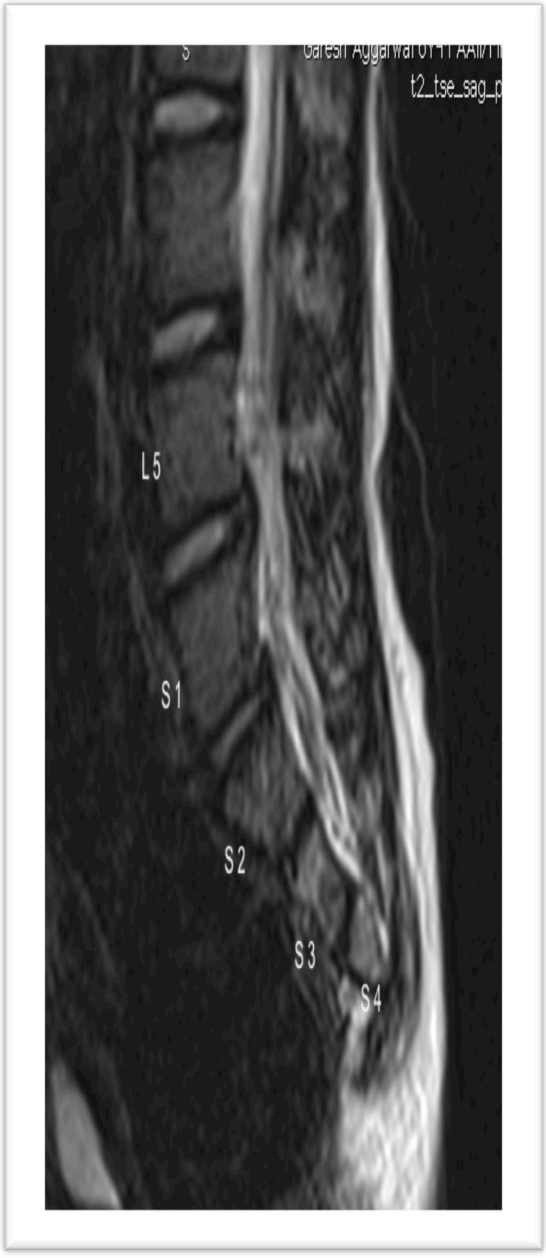

Fig 1: MRI (non contrast image) sagittal section showing absence of caudal segment of sacrum with entire coccyx vertebrae.

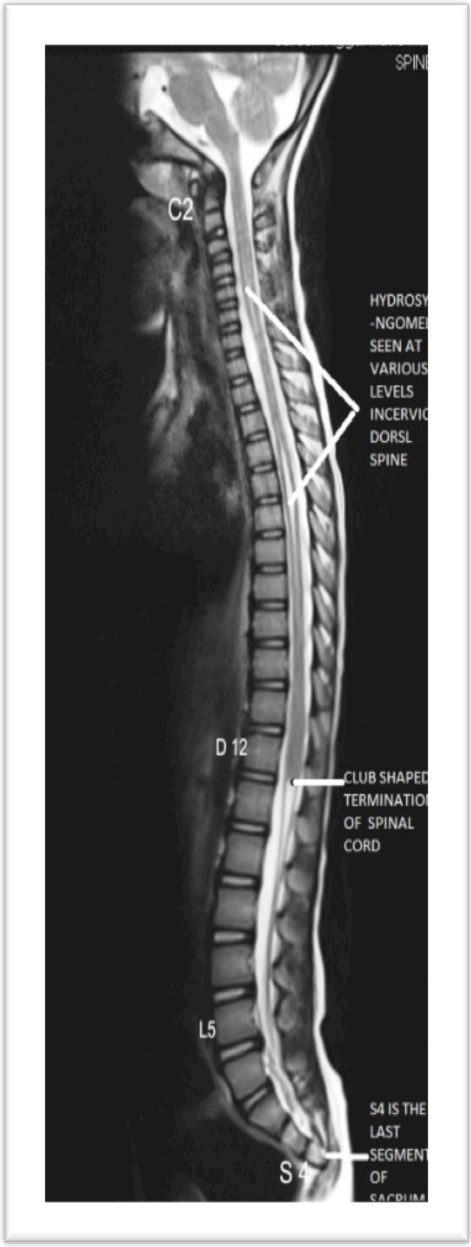

Fig 2: MRI (non contrast image) sagittal section showing club shaped termination of spinal cord with hypersensitivity at various level

Longitudinal hypersensitivity in spinal cord at multiple levels from C4-7 in cervical spine and T3-8 in dorsal spine was found which is indicative of hydrosyreniomeila. Annular disc bulge was seen at L5/S1 level. The tests and reports were discussed with clinicians till the final conclusion was made, for the case to be of sacral dysgenesis.

DISCUSSION

Caudal regression syndrome is a infrequent disorder with prevalence of 1: 25,000 live births. it consist of various degrees of development failure of lower half of spine associated with other congenital anomalies such as genitourinary defects, gastrointestinal defects and may be with cardiac defects. It occurs frequently in infants of diabetic mother. [5] Caudal regression is result of abnormal development of structures derived from the caudal mesoderm .CRS in not hereditary and recurrent risk also minute. Gene mutation in HLBX9 homeobox gene also suspected for a cause of CRS. In this case patient had complete absence of last sacral segment and coccyx vertebrae and club shaped termination of spinal cord. [2]

As the pathology is irreversible, supportive treatment can be given. It needs a multidisciplinary treatment with involvement of paediatrician, orthopaedic surgeon, urologist and physical therapist according to the severity of the disease. If the vital systems are unaffected or minimal affected, survival is a rule. These patient have normal intelligence, thus they live normaly except for neuromuscular defects in lower limb and sphincters.[3,5]

CONCLUSIONS

CRS is a congenital syndrome which results from in utero insult to the embryo. Prenatal diagnosis of condition and maternal care, especially in diabetic mothers during pregnancy can prevent such abnormalities in the new born. Severity of lesion and presence of associated anomalies affect the quality of life as otherwise the surviving infants usually have normal mental functions.

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