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General Paediatrics Section

Diastematomyelia with Spina Bifida Occulta and Bilateral Intrathoracic Kidneys

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ABSTRACT

A 7-year-old girl had presented with nocturnal enuresis. On investigation, she was found to have spina bifida, diastematomyelia and focal syrinx. on MRI the X ray of her spine showed scoliosis. Her intra venous pyelogram revealed bilateral intra thoracic kidneys.

Key Words: Ectopic kidneys, Urinary incontinence, Spinal dysraphism

INTRODUCTION

Nocturnal enuresis is grouped under vegetative disorders as an elimination disorder. Five years of mental age is compatible with the continence of urine. Though genetic factors have been mentioned to play a role, the aetiology is complex due to the involvement of physiologic and psychological factors [1]. It is essential to rule out urinary infections, as it is the most remediable cause of the symptoms. The further diagnostic evaluation includes ultrasonography of the bladder and the renal system [2]. Children with nocturnal enuresis are more likely to have abnormalities of the urinary tract.

CASE REPORT

A 7-year-old female child had presented with a history of urinary incontinence since birth in the form of nocturnal enuresis and pain on walking since 2 months. There was no complaint of dysuria, haematuria or pain in the abdomen. The child did not have an altered sensorium or seizures.

On examination, the child was found to be afebrile, with a pulse of 78 per minute and a respiratory rate of 28 per minute. The weight of the child was 13.8 kg and her height was 94 cm, which was less than 3 percentile. Spine examination revealed scoliosis in the thoracic region with deviation towards the left. Her respiratory system cadiovascular and neurological examinations were within normal limits.

The blood investigations revealed a haemoglobin of 10.7 gms%, a total count of 16700 / cubic milliliter, polymorphs 75 percent (%), lymphocytes 20%, monoocytes 01% and basophils and eosinophils 04%.

Her urine examination was unremarkable and her urine culture was sterile. Her blood urea was 20 mg% and her serum creatinine was 0.47 mg%. Her electrolytes were Na (Sodium)- 140 meq/L (miiliequivalents per liter), K (Potassium) –4.6 meq/L, Cl (Chlorides)- 107 meq/L and BSL –102 mg%.

The USG of her abdomen and pelvis revealed an ectopic right kidney most probable intra thoracic. Intravenous urography [Table/Fig-1] was done, which had the following impressions:

It was suggestive of bilateral simple renal ectopia with an intra thoracic location. Her right kidney was located at a higher level than her left kidney. Both her kidneys had normal functions. There was spina bifida of the thoracic spine with widening of the inter pedicular distance. Scoliosis of the lower thoracic spine was also present.

A renal scan revealed ectopically located, bilateral, normally functioning and excreting kidneys with no evidence of scars.

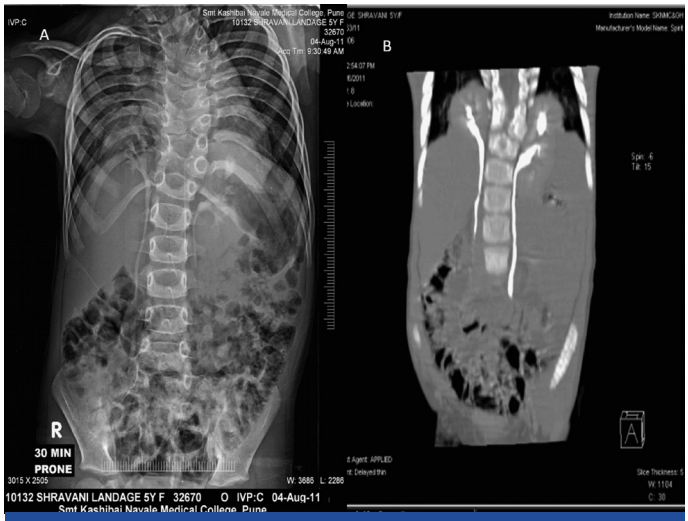
Intravenous urography showed bilateral thoracic kidneys [Table/Fig-1]: A (IVU) and B (CT- IVU)

MRI of the thoracic spine and the whole spine [Table/Fig-2] screening showed scoliosis of the dorsal spine with a convexity towards the left side and a Syrinx. Hemivertebrae were noted at the D4 and the D7 levels with multiple vertebral anomalies. The cord was low lying and it ended at the L3 level. A focal syrinx was noted within the cord and it extended from the D5 to the D8 level. Diastematomyelia was noted at the lower end [Table/Fig-3]. The kidneys were thoracic in location.

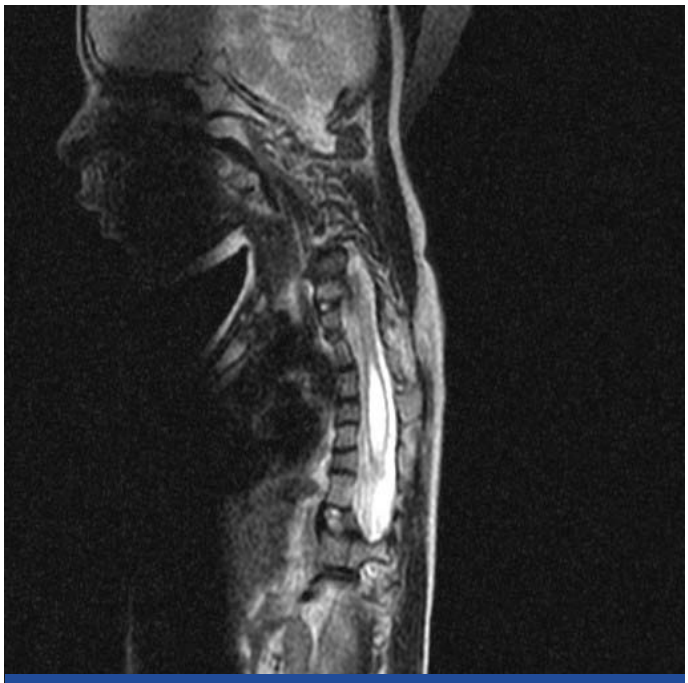
DISCUSSION

Bed wetting or nocturnal enuresis is categorized into the primary and secondary types. The primary type is also called as the persistent type and the secondary type is also known as the regressive type. children with Primary noctural enuresis are likely to have urinary tract abnormalities. In cases with a strong suspicion, it is mandatory to get ultrasonography and uroflowmetry done [1]. Our case was also associated with congenital scoliosis. Around the sixth week of gestation, the neural elements along with the spine are formed. The children with these findings are more likely to have visceral and intra-spinal abnormalities. Once a spinal abnormality is diagnosed, it is important to rule out malformations in other systems as well. Though our patient did not have any cardiac problems, cardiac anomalies may be found in some children.

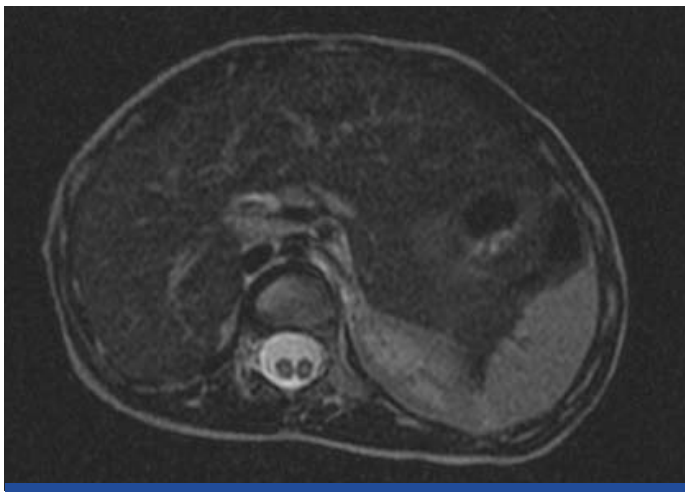
20 to 40 percent patients have intra spinal anomalies, the most common one being occult spinal dysraphism [3]. Disturbances in the embryologic formation of the spinal canal and the cord in the process of canalization and de-differentiation, which starts in the 28th week of gestation, results in lesions at the caudal end of the neural tube. They are less obvious and are covered with skin; hence, they are referred to as occult lesions. The clinical problems appear later; they may not appear at the beginning. As the child grows, there may be urinary or faecal incontinence, difficulty in



[Table/Fig-1]: A (IVU) and B (CT- IVU) shows bilateral thoracic kidneys



[Table/Fig-2]: Syrinx



[Table/Fig-3]: Diastomatomylia

locomotion and pain in the back or legs. Careful assessment of the urinary tract and the treatment of the urinary tract infections is very important to prevent progressive renal disease. Spina Bifida Occulta (SBO) is occasionally associated with syringomyelia, distematomyelia and a tethered cord.SBO may be associated with a pathology and significant sequelae, although a majority of the lesions pose no clinical threat. Sharma et al., in their case report, have described a patient with a Wilms tumour in the lumbosacral region. It was associated with Diastematomyelis and SBO [4]. The reversal of this deficit is unusual and a halting of the neurologic deterioration is a more realistic goal. An early diagnosis of this lesion, before the age of 3-years, may be associated with better surgical outcomes [3]. Holman CB et al., opine that diastematomyelia is an uncommon malformation wherein the neuraxis is separated. They have described a case of increasing curvature of the spinal cord [5]. Diastematomyelia and diplomyelia consist of a division of the spinal cord and the meninges into two halves over several segments. They may be associated with a bony spur or a fibrous septum. This bony spur may originate from the posterior part of the vertebral body and extend posteriorly. In 50% of the cases, this defect involves the lower lumbar vertebrae. It may be in the form of abnormalities of the vertebral bodies like fusion defects, hemivertebrae, hypoplasia, kyphoscoliosis, spina bifida and myelomeningocele. The symptoms include scoliosis, low back pain and incontinence. In their study, MRI clenched the diagnosis, it being the study of choice [6]. Prenatal ultrasound can also detect whether the diastematomyelia is isolated or whether it is an association with any serious neural tube defects. Syringomyelia is characterized by a fluid-filled cavity within the spinal cord. While its pathogenesis is currently debated, the relationship of syringomyelia with other conditions such as the Chiari I malformation and cord/column trauma, is well accepted [7]. It may or may not communicate with the cerebro spinal fluid. It rarely produces symptoms in childhood, but a rapidly progressive scoliosis may be the first indication.

The patient who was reported also had renal ectopia though it was asymptomatic. The ectopia results from an incomplete or abnormal process of ascent and rotation as the kidneys normally ascend from the pelvis into their normal position behind the ribs. They can be related to other problems such as VUR / blockage or obstruction. Thus, ectopic kidneys have to be evaluated [8]. Yusuf Ersham reported two cases of ectopic renal tissue and ectopic pelvic kidney, both of which were associated with split cord malformation [9]. Congenital thoracic ectopic kidney is a rare developmental anomaly, with the thorax being the rarest location. This anomaly is usually asymptomatic and it can be discovered incidentally on routine chest radiography. At least 200 cases of thoracic kidney have been described, with a vast majority of cases being documented in adults [10]. One case report presented seven persons from three generations who had combinations of acral, renal and ocular defects. The renal anomalies varied from a mild rotation to renal ectopia [11]. Here, we have presented the interesting clinical images of a child with spina bifida occulta, diastematomyelia and bilateral intra thoracic kidneys, which were discovered while we were investigating for nocturnal enuresis.

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