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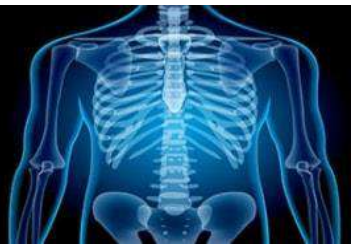

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VACTERL association with non-rotated bowel: A rare case report

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Abstract
VACTERL is an acronym for a group of congenital anomalies where V stands for vertebral defects or vascular anomalies(single umbilical artery), A stands for anal atresia, C stands for cardiac defects, TE denotes trachea-esophageal fistula, R stands for renal abnormalities and L stands for limb defects such as polydactyly or syndactyly. Atleast, three of above features are considered necessary for diagnosis of the condition [1].
It may or may not be associated with other bowel pathologies (most commonly duodenal obstruction).
If there is accompanying hydrocephalus, then the term VACTERL-H is used.
The estimated incidence is 1 in 10,000 – 40,000 births. Few familial cases have been identified and maternal diabetes and maternal diabetes is only environmental influence implicated. It is called an association, rather than a syndrome because the complications are not pathogenetically related, tend to occur more frequently than expected and are thought to be linked to embryonic mesodermal defects.
It is thought to be autosomal recessive in nature [2].
It has been statistically associated with maternal exposure to exogenous sex hormones during the first trimester of pregnancy [3].
It has also been associated with Fanconi Anemia [4].

Keywords: VACTERL, vertebra, anal, trachea-esophageal, limb defects

Introduction

Case

Here we present a case report of a 10yr old female child, a known post operative case of anal atresia, presented to OPD with chief complaints of pain around umbilicus and pain in left flank. No limb defects were noted on inspection.
A Contrast enhanced Computed Tomography (CECT) was conducted in our Radiology department.

The CECT was scanned in phases as follows

1. Non- Contrast Whole Abdomen
2. Arterial phase
3. Cortico- Medullary phase
4. Nephrogram
5. Urography phase

Following findings were noted

1. Non – rotated left kidney with duplex collecting system and bifid ureter. One of the double moiety ureters drained just medially and inferiorly to another ureter into bladder. No ectopic ureter insertion was noted. Normal enhancement and excretion of dye was noted in both kidneys.
2. Bifid spinous process of S1 vertebra with a sclerotic focus
3. Most of the jejunal and ileal loops were located on right side of abdomen with caecum and appendix located higher up just on left side of midline. Appendix was noted inferior to stomach. Ascending colon was also noted on left side of midline suggestive of Non-rotation of Bowel.
4. Spleen was also noted to be slightly lowered down in position.

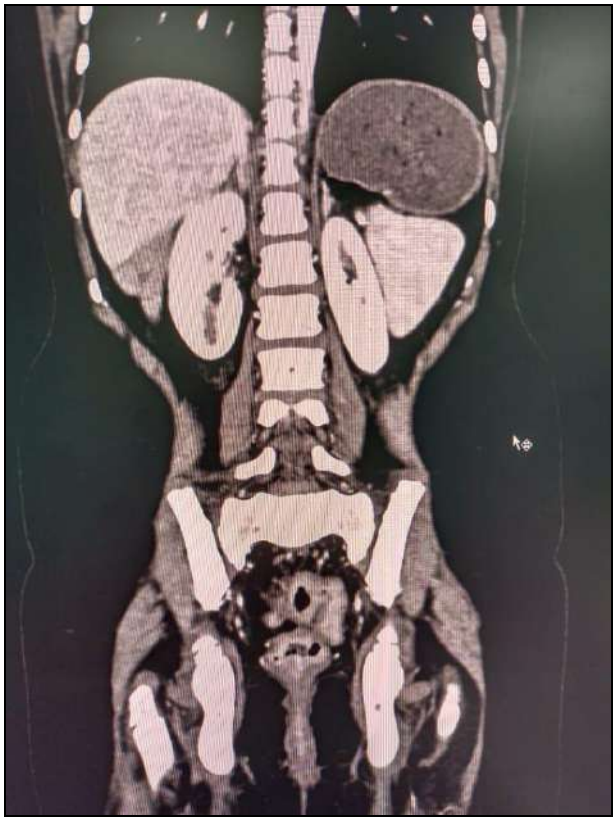

Fig 1: Represent Non – rotated left kidney with duplex collecting system and bifid ureter. One of the double moiety ureter drained just medially and inferiorly to other ureter into bladder. No ectopic ureter insertion was noted. Normal enhancement and excretion of dye was noted in both kidneys with mildly lowered down position of spleen.

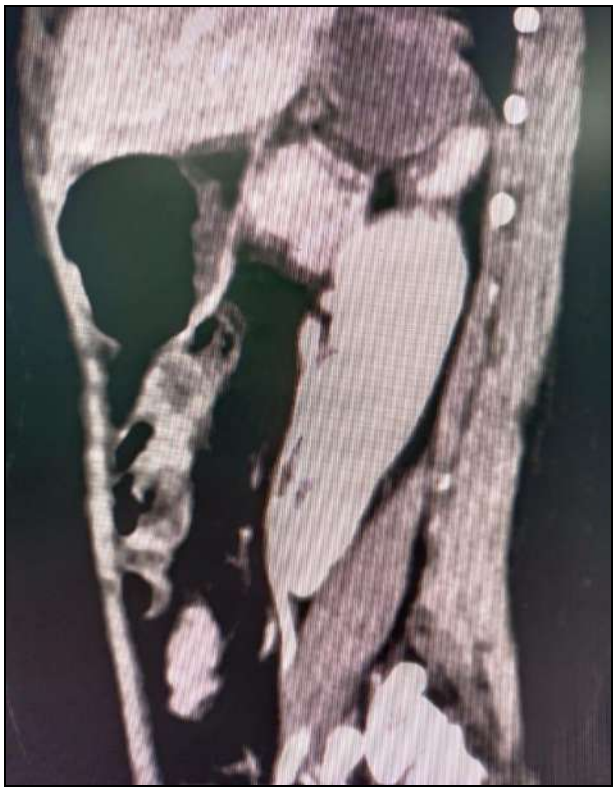

Fig 2: Non – rotated left kidney with duplex collecting system and bifid ureter. One of the double moiety ureter drained just medially and inferiorly to other ureter into bladder. No ectopic ureter insertion was noted. Normal enhancement and excretion of dye was noted in both kidneys.

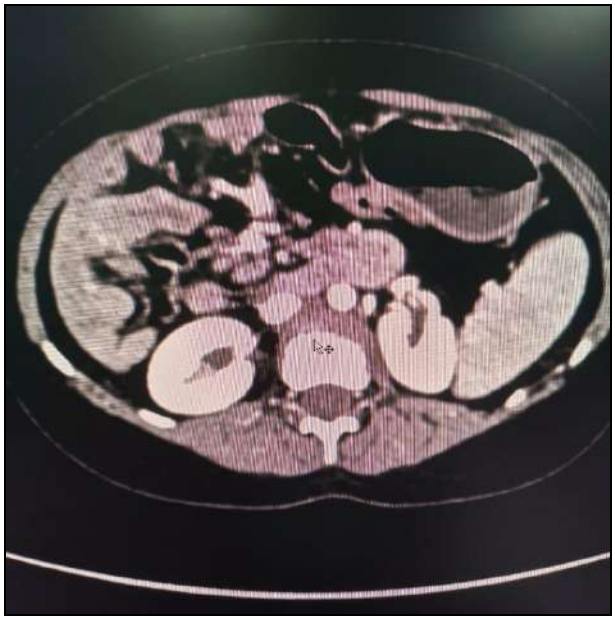

Fig 3: represents- Most of the jejunal and ileal loops were located on right side of abdomen with caecum and appendix located higher up just on left side of midline. Appendix was noted inferior to stomach. Ascending colon was also noted on left side of midline suggestive of Non- rotation of Bowel.

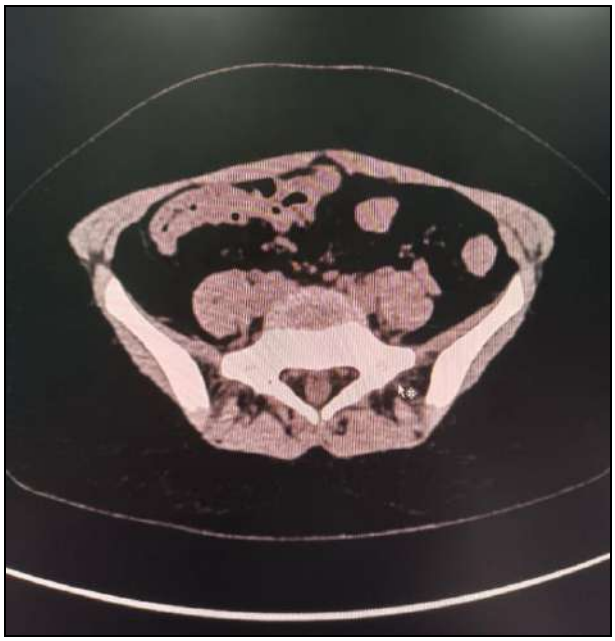

Fig 4: represents- bifid sacral spine.

Discussion

Patients with a combination of multi-system congenital anomalies affecting the vertebral column (V), anus and/or rec-tum (A), heart (C), trachea and/or esophagus (TE), kidneys (R), and limbs (L) may be diagnosed as having a VACTERL syn-drome; 3 of which are required for the diagnosis.

The Hedgehog gene (Hh) has been identified as a crucial ligand involved in the signaling pathways of organogenesis, including the development of the gastrointestinal tract [8].

Ab-normalities in the Hh signaling pathway have been implicated in the myriad of disorders associated with VACTERL syndrome and congenital duodenal obstruction

At birth, our patient presented with anal atresia which was documented and surgically corrected [5].

Ten years later, our patient presented with pain around umbilicus and in left flank.

On CECT, we detected bifid sacral spine with malrotated left kidney with duplex collecting system and bifid ureter. Thereby, along with anal atresia, three criteria required for VACTERL association have been fulfilled. Also, it was also associated with non – rotated bowel and mildly lowered spleen.

The other highlight of this case is although reports say that VACTERL babies with ipsilateral renal disorder have the same side limb defects, our case has a renal anomaly with no limb anomaly.

Conclusion

Various combinations of congenital anomalies must be in kept in mind, which may be used to assist the pediatricians in the diagnosis of such rare cases. These are rare congenital anomalies, which need team management. Thereby, a high degree of suspicion and knowledge of various combinations of congenital anomalies must be kept in mind by radiologists for betterment of these children. As children are the future of nation. Parental counseling also forms an integral part of this management.

Conflict of Interest

Not available

Financial Support

Not available

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