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Megacalycosis with Ipsilateral Segmental Mega-Ureter - A Rare Entity

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1. Abstract

Congenital megacalycosis of Pugivert is a rare clinical entity which denotes presence of dilated calyces most often without any functional abnormality and Segmental mega-ureter implies large, focal segmental ureteral dilatation producing a distorted ureter. Megacalycosis with an ipsilateral segmental mega-ureter is a rarer association of which only a few cases have been reported after its original description. It is diagnosed incidentally or the patient might present with urinary tract infection or calculus formation and usually respond to conservative management. We report a case of this rare condition in a 14-year-old boy who presented occasional colicky abdominal pain was managed conservatively and kept on follow-up. This case report is to emphasize this rare, relatively harmless co-existence of these two conditions.

2. Introduction

Congenital megacalycosis of Pugivert is a rare clinical entity which denotes presence of dilated calyces most often without any functional abnormality [1-6]. This term was first coined by Pugivert in 1964. The following criteria of dilated calyces, polycalycosis and renogram showing no pelvicalyceal system obstruction should be met for a diagnosis of megacalycosis. It is most often seen with polycalycosis which implies increased number calyces usually greater than 22-25 minor calyces [7]. Megacalycosis with an ipsilateral segmental mega-ureter is a rarer association. Only a few cases have been reported of this association after its original description [8]. Patients typically present with urinary tract infection or calculus formation and usually respond to conservative management. This case report is to emphasize this rare, relatively harmless co-existence of these two conditions.

3. Case report

We report a case of 14-year-old male patient who had presented with occasional right flank pain. He did not have any urinary tract infection and calculus in the past. The physical examination was not contributary. Biochemical tests of the renal function were within normal limits. Urine routine and microscopy were unremarkable. The ultrasonogram of the abdomen showed enlarged right kidney with normal corticomedullary differentiation. The calyces dilated with a normal calibre pelvis and upper ureter. Intravenous pyelography showed dilated, increased number of calyces, 35 in number, with a normal appearing pelvis and a dilated ureter in its lower third with a gradual tapering and cut-off near the vesico-ureteric junction. DPTA renogram showed differential renal function of 45% on the right side with no evidence of obstruction. The above investigative findings were suggestive of megacalycosis with ipsilateral segmental megaureter. He is being managed conservatively with follow-up plan of renal biochemical testing and ultrasonogram on yearly basis (Figure 1).

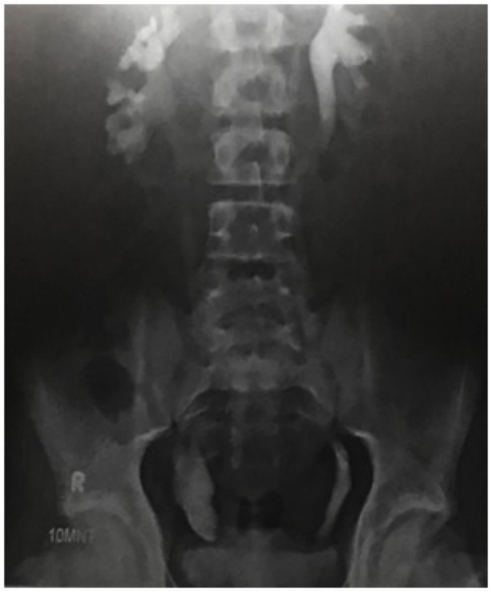

Figure 1: Intravenous pyelogram of the patient taken at 10 mins showing right Megacalycosis(inordinate number of dilated calyces) and segmental dilated distal ureter with maximum diameterwith tapering towards vesico-ureteric junction

4. Discussion

Congenital megacalycosis (CM), first described by Pugivert in 1963 is a congenital urinary anomaly consisting of oversized calyces and a renal pelvis of normal calibre [1-6]. In addition to dilatation, the calyces maybe increased in number (polycalycosis) usually more than 20-25 [7]. The renal pyramids are under developed which causes lack of projection of the papillae into the calyces eventually producing the dilatation of the calyces without neither fornix nor papillae impressions [9]. The defect is mostly unilateral, shows male predominance and usually does not disrupt the renal functions. There is no cortex abnormality such as scarring or signs of chronic infection.

Segmental megaureter is segmental dilatation of ureter and is a rare entity, and only 12 case reports are found in the literature [10]. This condition is marked by large, focal segmental ureteral dilatation producing a distorted ureter. Distal most part of the ureter may be normal, stenotic, or atretic. They usually have a dysplastic or disorganized muscle coat. It may be associated with megacalycosis and hypoplastic, dysplastic or non-functioning kidney [10]. Stasis of urine in enlarged, redundant lower ureter may promote the formation of urinary calculi and infection. Patients with usually present with symptoms of pain, hematuria, fever and dysuria.

Diagnosis is by a combination of imaging modalities with the ultrasonogram showing dilated pelvicalyceal system without cortex abnormality. In Intravenous pyelogram, the presence of normal calibre renal pelvis interposed between the dilated collecting system and the distal dilated ureter without evidence of vesicoureteral reflux implies the coexistence of ipsilateral idiopathic megaureter [8]. Voiding cystometrogramis done to rule out VUR and diuretic renogram to look for any obstruction and assess the functional status of the kidney.

It is rare for megacalycosis and primary segmental megaureter to be co-exist in the same patient. Blanca Vargas et al reported five cases of congenital megacalycosis with primary megaureter where the patients were operated initially with incorrect diagnosis of obstructive uropathy and only later this rare association was recognised [9]. Four cases of this association were presented in 1987 by G A Mandell et alemphasising recognition of their coexistence and the importance of careful interpretation of urogramswhich would avoid needless operations [8].

Ranawaka et al had showed that patients with ureteral diameter less than 10 mm could be managed conservatively with prophylactic antibiotic without renal function deterioration ofwhile ureteral diameter of > 10 mm were more prone to febrile UTI and stone formation.[11] Recurrent UTI and calculus warrants surgical intervention in the form of excision of the aperistaltic segment and reimplantation of the ureter [11-12]. Conservatively managed patients should ideally be kept on close follow-up with periodic ultrasonogram with addition of functional studies when required until the stability and innocuity of the dilatation is established [11].

Our patient now into adolescence, with minimal symptom of occasional colicwas diagnosed with the megacalycosis with ipsilateral segmental megaureter and a preserved renal function. Hence, he was put on prophylactic antibiotic and kept on follow-up.

5. Conclusion

As happened with our patient, the rarity of this condition leads to diagnostic doubts and incorrect diagnosis, whereincareful interpretation of intravenous urogram can clarify the diagnosis and rule out the possibility of infundibular stenosis and other causes of hydroureteronephrosis. The recommendation is to follow-up the patient into adulthood due to the reported increased incidence of urinary infections and urolithiasis. This condition must be kept in mind in the diagnostic workup of congenital hydronephrosis and infundibular stenosis.

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