

Research Article

The Role of Intravenous Urography with Erect Film and Retrograde Pyelography in Revealing Patho-Etiology of the Loin Pain and Haematuria Syndrome by Discovering its Overlooked Link with Symptomatic Nephroptosis

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- Intravenous urography
- Retrograde pyelography
- Loin pain haematuria syndrome
- Nephroptosis

Abstract

Introduction and objectives: To report the role of intravenous urography with erect film (IVU-E) and retrograde pyelography (RGP) in resolving the puzzle of Loin Pain and Haematuria Syndrome (LPHS) by revealing its patho-etiological overlooked link with Symptomatic Nephroptosis (SN). We demonstrate that renal pedicle stretch causes neuro-ischaemia as evidenced by the new IVU 7 sign and the damaged renal medullary papilla shown on RGP.

Materials and methods: Images are reported from a series of 190 SN patients. Repeated standard imaging was invariably normal, when supine. However, 190 patients demonstrated SN of > 1.5 vertebrae on repeating IVU-E. Of whom 36 (18.9%) patients developed recurrent episodes of painful hematuria for which no organic pathology was detected on all standard imaging, when supine- thus fitting the definition of LPHS. The IVU 7 sign, with its horizontal and vertical segments, represents the renal pedicle at supine and erect IVU films, respectively was used for measuring renal pedicle stretch causing renal vessels stenosis and ischaemia. The RGP demonstrated the internal renal papillary damage causing LPHS.

Results: Of 190 with SN on IVU-E, 182 were females and 8 males. The mean age was 28.8, duration of symptoms 15.7 and hospital follow up 6.6 years.

Patients showed no abnormality on IVU or ancillary imaging when supine. All patients showed renal drop of >1.5 vertebrae (>5 cm) on erect IVU film. Stretch/rotation of renal pedicle causing neuro-ischaemic pain of LPHS was demonstrable on the right side in 72 (37.9%) and bilaterally in 7 patients.

Complications of SN on IVU-E included both obstructive and neuro-ischaemic: obstructive complication included ballooned renal pelvis, hydronephrosis and upper pole diverticulum. Neuro-schaemic complications induced by pedicle stretch and rotation/twist were haematuria of the LPHS affecting 36 (18.9%), auto nephropexy affecting 12 right kidneys, upper pole calyctiasis with extra-vasation affecting 28 (15.8%) right kidney and 2 bilateral that are best shown on RGP. Renal atrophy affected 4 right kidneys. Upper pole infarction affected 2 kidneys. Retrograde pyelography (RGP) also demonstrated upper pole calyctiasis with extra-vasation. Surgical treatment was used in 28 patients; 10 had simple nephropexy and 18 had RSD&N for severe LPHS. Four of the patients treated with simple nephropexy had recurrence of LPHS while those who had RSD&N were all cured.

Conclusion: Upright IVU film and RGP are essential for the diagnosis of SN complicating into LPHS. The new IVU 7 sign affirms that pedicle stretch causes ischaemic nephropathy. Renal sympathetic denervation and nephropexy is curable for LPHS but simple nephropexy is not.

INTRODUCTION

Symptomatic Nephroptosis (SN) has been known for centuries but was disparaged >80 years ago [1,2]. The Loin Pain and Haematuria Syndrome (LPHS) was reported in 1976 [3]. The use of IVU with erect film (IVU-E) and retrograde pyelography (RGP) in revealing patho-etiological overlooked link between SN and LPHS and the introduction of a new curative surgery are new discoveries [4-6].

This article is based on a prospective 10 years observational study on loin pain haematuria syndrome (LPHS) complicating symptomatic nephroptosis (SN) [1,2]. Episodic loin pain (LP) or LPHS that has no detectable pathology is frustrating to both patient and urologist. I inherited 30 cases from my predecessor at King Khaled Hospital, Najran, Saudi Arabia. Repeated investigations and imaging including CT and MRI and MRA were all normal-being feasible only on supine posture. A patient showed me a maneuver to bring her own right kidney down to right iliac fossa and keeping it there for examination (Loin grip sign). Ever since I diagnosed 1.7 new cases/m and found the underlying cause of SN and LPHS.

The main management problem of loin pain was the lack of demonstrable pathology on repeated imaging, when supine. The underlying SN though well known [3] was disparaged [4] and LPHS though well documented, its existence may be doubted [5] and both are extremely problematic to manage [5-8]. Demonstrable renal pathology of loin pain and haematuria was invariably lacking on all supine imaging of the received protocol [5-8]. Urinary tract infections (UTI) may affect a few patients during the occasional episodes but UTI, stones and organic causes play no role in the pathogenesis of LPHS. Many complex ramifying management problems of SN and LPHS are well known, but have no solutions. Some of the problems were communicated [9,10] and the illusive overlooked link of SN with LPHS was pointed out recently [11,20].

The objective of this study was to answer the following questions:

- Is chronic episodic LP/LPHS genuine?
- Is there an organic pathology? How to reveal it?
- Why is the Standard Imaging Protocol (SIP) negative?
 - Defining:
 - Nephroptosis means mobile kidney.
 - Standard Imaging Protocol (SIP) as in current use.
 - Upright Imaging Protocol (UIP) in which an IVU with erect film (IVU-E) is done.
 - Demonstrating:
 - Features and complications of SN seen on UIP not on SIP
 - Patho-etiology link of SN with LPHS.
 - Discussing:
 - Reasons for disparaging SN, ignoring it by Textbooks and overlooking it in routine urological practice

- USA Authorities ignores both SN & LPHS, UK Authorities ignores SN and what the leaders choose to ignore, others do not see.

Nephroptosis is differentially diagnosed from ectopic Kidney by blood supply and mobility. In SN the kidney drops >1.5v (>5 cm) on erect IVU film causing pain with/out haematuria. In LPHS no detectable anomalies is seen on SIP. Every pair of IVU films, shown here, shows left Supine (S) and right Erect (E) for comparison.

Measuring renal drop, laterality, pelvi-ureteric junction (PUJ) kink and pelvicalyceal dilatation (PC) as evidence of obstruction on IVU E, have established renal pedicle stretch and rotation as evidence of renal neuro-ischaemia. Although SN is known, it was disparaged >60 years ago and omitted from all textbooks. SIP is constantly normal- at supine posture. UIP (**IVU-E**) is rarely requested, hence chance diagnosis of SN is unlikely and diagnosis is easily missed. Many of SN features and some complications were documented >70 years ago. The link of SN with LPHS is a new discovery explaining its real patho-etiology [1,2].

Here we report the evidence that IVU-E and RGP are vital diagnostic tests for revealing the patho-etiological link of LPHS with SN.

MATERIAL AND METHODS

Photographs taken from a prospective study on LPHS and SN are used to demonstrate the value of IVU-E and RGP in revealing the patho-etiology of LPHS. The IVU-E demonstrates the IVU7 sign which shows the degree of stretch in renal pedicle on erect posture which in turn causes vascular stenosis and ischaemia. This is affirmed by the tube stretch hypothesis as well as confirmation of renal vessels stenosis on surgical exploration.

RESULTS

The results of this study demonstrate that IVU-E reveals the IVU7 sign that measures vessel stenosis on stretch at erect posture (Figure 1). This is further confirmed by the tube stretch hypothesis (Figure 2). Also on surgical exploration both renal artery and veins are severely stenosis and stenosis causes renal ischaemia (Figure 3). A normal RGP in a case of SN that does not suffer the LPHS is shown in (Figure 4).

The upper pole of the right kidney is the first and most severely affected in LPHS (Figure 5,6). Later on the medullary damage spread to other calyces most commonly of the right kidney. This creates calycio-venous fistula (Figure 7) that causes bleeding in one direction and contrast leakage in the other. Figure (8) shows advanced renal mudullary papillary destruction in LPHS more severe on the right side. Figure (9) compares IVU to RGP of the same patient with LPHS. The IVU looks normal while the RGP shows severe multiple bilateral papillary necrosis. Also CAT and MRI are normal. Again (Figure 10) compares the IVU and RGP of the same LPHS case. The IVU looks deceitfully normal while RGP reveals the massive medullary destruction.

DISCUSSION

The presented photographic evidence demonstrate that IVU-E and RGP are mandatory for the proper assessment of



Figure 1 Shows renal pedicle mapped on a supine IVU film (Horizontal. Left) and erect film (Vertical, right) making a figure of 7 where the renal pedicle is stretched to 3 times its normal length, causing stenosis and ischemia.

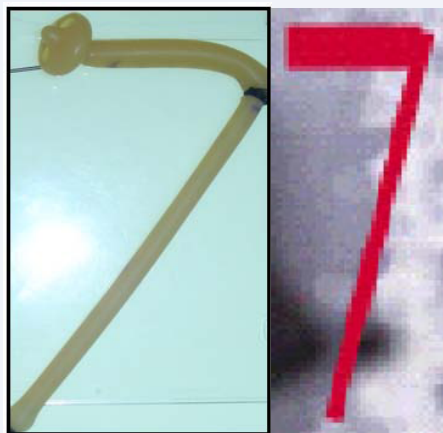


Figure 2 shows the rubber tube stretch hypothesis where the lower half of the tube is stretched to double the length of horizontal half as compared with IVU 7 sign which confirms that stretch of a tube or artery causes stenosis of the lumen.

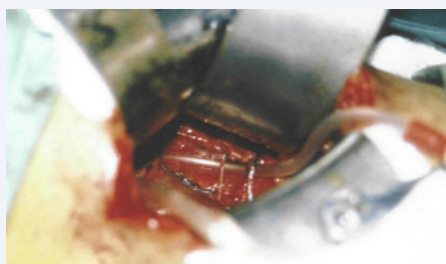


Figure 3 shows severe stenosis of the renal artery and veins in a case of LPHS. The Nelaton tube represents the diameter of normal renal artery.

LPHS as it reveals the underlying the patho-etiology. The IVU-E demonstrates the IVU7 sign that is a measure of pedicle stretch and in turn the degree of vascular stenosis that causes ischaemic damage to the kidney affecting mostly the renal medulla. The upper pole is the first and most severely affected. The right kidney is also the most commonly affected in females suffering LPHS. The RGP accurately assesses the renal damage that is proportional with the chronicity of the case.

It is worth noting that this is the first report to document the role of RGP in revealing the patho-etiology of LPHS. RGP was not previously used in the investigation of LPHS or SN. The IVU-E was not previously used in investigating LPHS. It is observed that the whole renal pedicle is stretched which causes renal ischaemic neuropathy. Hence the operation of renal sympathetic denervation and nephropexy (RSD&N) was designed and proved 100% curable in all 18 cases who consented for this surgery. This makes all other useless surgeries in LPHS obsolete.



Figure 4 shows a normal RGP in a case of SN that does not suffer the LPHS.



Figure 5 shows RGP at supine and erect posture with destruction of the upper pole papilla leaking contrast into vein in a case of LPHS. The upper pole is the first and most to be affected.



Figure 6 shows RGP with destruction of the upper pole papilla leaking contrast into a diverticulum in a case of LPHS.



Figure 7 shows RGP with multiple pyelocalyctais with erosion of medullary papillae and contrast leakage into veins (papillary venous fistula)- bleeding in LPHS occurs in the opposite direction.

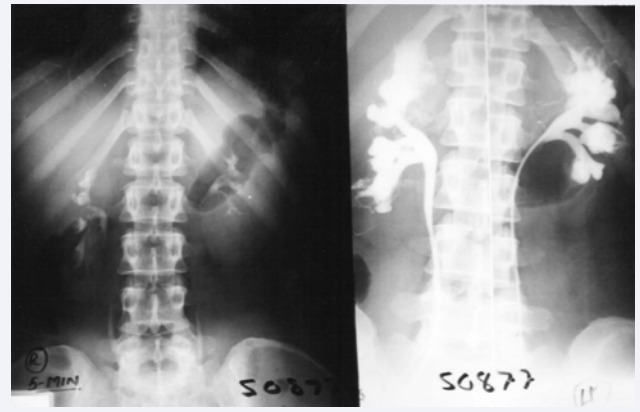


Figure 10 shows IVU (left) and RGP (right) of the same patient with chronic long standing LPHS. The IVU looks deceptively normal while RGP shows massive bilateral destruction of renal papilla.



Figure 8 shows advanced renal medullary papillary destruction in LPHS more severe on the right side.

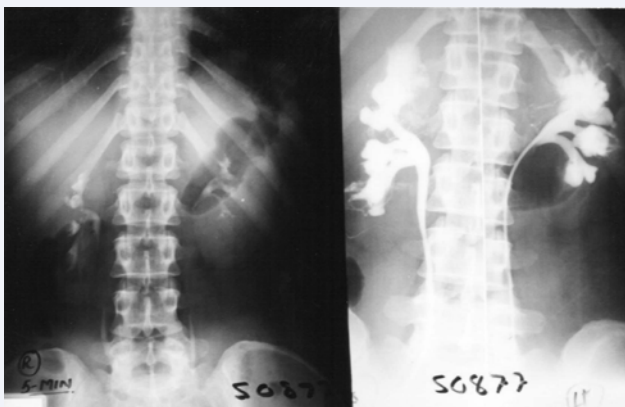


Figure 9 shows IVU (left) and RGP (right) of the same patient with LPHS. The IVU looks normal while the RGP shows multiple bilateral papillary necrosis.

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