Clinical History:

This 13-year-old girl with diabetes has presented in the last month with 4 episodes of macroscopic haematuria. The blood pressure had been normal, and platelets and coagulation values were within normal limits, but urine analysis showed red blood cells without white blood cells. Physical examination was unremarkable.

Imaging Findings:

US showed normal-sized kidneys and Doppler evaluation revealed normal arterial flow and normal resistive indexes. The renal veins were asymmetrical and the left renal vein (LRV) had a larger calibre near the kidney (up to 8mm), with only 2 mm of anteroposterior diameter after the cross of the superior mesenteric artery (SMA). Thus the anteroposterior diameter ratio (APDR) was 4.0. Doppler assessment of the LRV illustrated turbulent flow and peak velocities of 77cm/s distal to the SMA and 18cm/s proximally, with a peak velocity ratio (PVR) of 4.3. The right renal vein had a homogenous calibre and normal flow characteristics and velocities.

Discussion:

The nutcracker syndrome (NS) is caused by the compression of the left renal vein (LRV), due to a reduced angle between the superior mesenteric artery (SMA) and the aorta. This anatomical finding has been known to anatomists for over 70 years [1] and is called the Nutcracker phenomenon [2]. In the nutcracker syndrome, compression of the LRV causes an increase in pressure which can result in the development of collateral vessels. LRV hypertension can cause local damage so this syndrome can present with haematuria and orthostatic proteinuria [3]. LRV hypertension also causes the development of varices in the renal hilum and eventually varicocele which can lead to infertility, or ovarian vein syndrome and abdominal pain [4].

Venography of the LRV with assessment of the pressures of the LRV proximal to the compressed portion and in the inferior vena cava is the direct diagnostic method. The normal renocaval gradient is 1 mmHg, but in NS it may be greater than 3 mmHg [5, 6]. The LRV can be seen to be dilated with slow washout of contrast from the vein, and there may be collateral vessels. However, this is an invasive and time-consuming method not indicated when severe symptoms are absent [7].

Contrast-enhanced Computed Tomography (CT) and Magnetic Resonance (MR) can depict the compressed portion of the LRV with proximal dilation and the collateral vessels. However, CT entails ionising radiation and MR may require sedation in younger children.

US can also show the distension of the LRV and be used to measure the diameter to calculate the diameter ratio.
Because dilatation of the proximal LRV has been documented in healthy individuals, and nutcracker syndrome can occur in the absence of LRV dilatation, the diameter ratio may not be as informative as the peak velocity ratio (PVR) [3, 8]. Doppler US is the first-level imaging technique. It allows the study of flow direction and speed [7].

Kim's [3] study on adults presents cut-off points of 5.0 for both the diameter and PV ratios, and suggests the average between these as a means to increase sensitivity and sensibility. In children the US study can be limited as the very sampling area is smaller than in adults [8].

Park's study on children with orthostatic proteinuria [8] suggests 3.98 for PVR and 4.16 for anteroposterior diameter ratio (APDR).


Takebayashi et al. [9] add the finding of collateral vessels as a diagnostic criterion (however, they may be absent early in the disease history).

A variety of surgical procedures can be performed when symptoms are severe, but NS in childhood may resolve spontaneously [10] so renal biopsy can be delayed if NS is diagnosed, and employed only if haematuria persists.

**Differential Diagnosis List**: Nutcracker syndrome

**Final Diagnosis**: Nutcracker syndrome

**References**:


Description: The anteroposterior diameter of the left renal vein measured medial to the superior mesentric artery is 2mm. Origin:
Description: The anteroposterior diameter of the left renal vein measured lateral to the superior mesenteric artery is 8mm. Origin:

Description: Flow velocity on the left renal vein medial to the superior mesenteric artery is 18cm/s. Origin:
Description: Flow velocity on the left renal vein lateral to the superior mesenteric artery is 77 cm/s.
Origin: