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FRALEY SYNDROME - A CLINICAL CASE

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Abstract. *Fraley syndrome (FS) is a renal vascular malformation in which the branches of the renal artery cross and compress the upper renal calyx or the upper part of the renal pelvis, causing their obstruction, which can be visualized by modern imaging methods. In the diagnostic medical center "Euromed" in Chisinau, Republic of Moldova, 35 angiographies of abdominal aorta were performed on patients between 18-75 years old, among whom a 20-year-old man was diagnosed with Fraley syndrome. According to clinical data taken from medical records, the patient had recurrent hematuria. According to angiographic findings, two renal arteries were identified in the right kidney. The first artery, originating from the aorta, above the renal artery, defined as arteria segmenti inferioris renis, due to its descending course, crossed arteria segmenti superioris renis at the level of the renal gate and continued in the lower renal segment, compressing the upper renal calyces. The accessory renal artery, originating from the renal artery itself, was also identified, going directly into the parenchyma of the lower pole of the kidney. FS is a rare cause of intrarenal collecting system obstruction. This case emphasizes the importance of angiography for the detection of this syndrome.*

Keywords*: Fraley syndrome, renal artery, angiography.*

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СИНДРОМ ФРЕЙЛИ - КЛИНИЧЕСКИЙ СЛУЧАЙ

Аннотация. *Синдром Фрейли (СФ) – это порок развития почечных сосудов, при котором ветви почечной артерии пересекают и сдавливают верхние почечные чашечки или верхнюю часть почечной лоханки, вызывая их обструкцию. Данный синдром можно выявить с помощью современных методов визуализации. В диагностическом медицинском центре «Евромед» г. Кишинева, Республики Молдова были проведены 35 ангиографий брюшной аорты пациентам в возрасте от 18 до 75 лет, среди которых у 20-летнего мужчины был обнаружен Синдром Фрейли. Согласно клиническим данным медицинской карты, у пациента наблюдалась периодическая гематурия. Ангиографическое исследование выявило в правой почке две почечные артерии. Первая артерия, берущая начало от аорты, выше почечной артерии, определяемая как arteria segmenti inferioris renis, из-за её нисходящего хода, на уровне ворот почки пересекала arteria segmenti superioris renis и продолжалась в нижнем почечном сегменте сдавливая верхние почечные чашечки. Также была выявлена и добавочная почечная артерия, берущая начало от самой почечной артерии, идущая непосредственно в паренхиму нижнего полюса почки. СФ является редкой причиной обструкции внутрипочечной собирательной системы. Данный случай подчеркивает важность ангиографии для выявления данного синдрома.*

Ключевые слова*: синдром Фрейли, почечная артерия, ангиография.*

Introduction. The kidney, as an organ of vital importance, maintains the body's hydrosaline and acid-base balance, contributes to the regulation of arterial pressure. The good function of the organ is due to its vascularization by the renal arteries (RAs), which usually arise from the abdominal aorta (AA) at the level of the first or second lumbar vertebrae (L1-L2).

Congenital renal malformations as well as anatomic variants of renal arteries are more common compared to other organs supplied by branches of the abdominal

aorta, constituting approximately 18.0%, and in approximately 30% of cases a supernumerary or accessory renal artery may be present [5].

Accidental injury or ligation of atypical arteries during surgery can cause extensive hemorrhage, ischemia of a portion of the kidney, or necrosis of the kidney.

Also, one of the disorders of the renal accessory arteries is their stenosis, which can cause secondary or renovascular hypertension and non-traumatic bleeding. Sometimes, renal artery branches, due to their atypical course, can affect the proximal collecting system and cause filling defects either in the calyces, pelvis or proximal segment of the ureter.

The aim of the study. Description of a relevant case of Fraley syndrome identified by abdominal aortography.

Material and Methods. The study was performed on aortograms of 35 patients, examined between 2023-2024, in the Euromed Diagnostic Medical Center and MSPI Institute of Emergency Medicine, Chisinau, Republic of Moldova. Only those patients who signed an informed consent for participation were included in the study.

Results and Discussion. Out of the total number of studied angiographs, Fraley syndrome (FS) was established in a single case only, in a 20-year-old patient, who according to his medical records had intermittent hematuria.

Angiographic data: both kidneys of normal configuration and size, with homogeneous parenchyma and unaltered cortico-medullary distinction.

Arteria renalis dextra (ARD), diameter (d) 0.6 cm; *arteria renalis sinistra* (ARS) - d=0.58 cm. Both arteries with normal course (Figure 1).

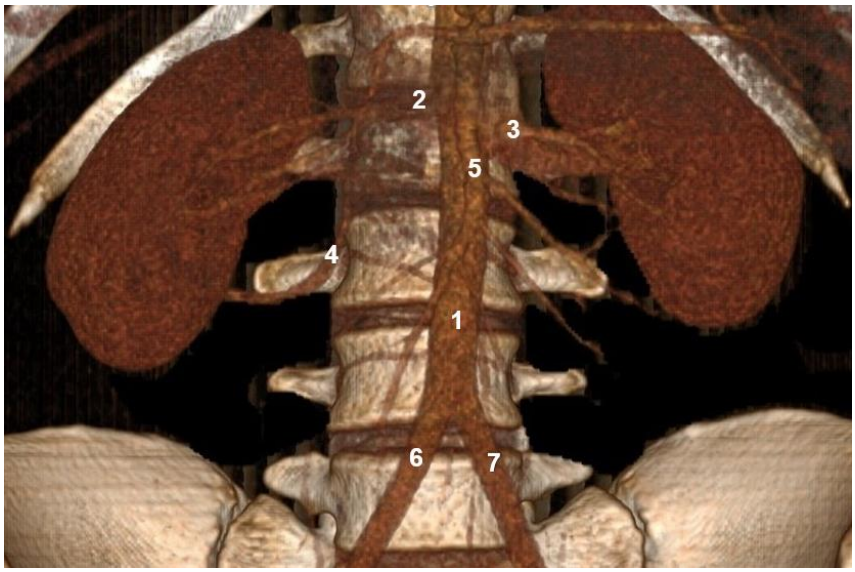

Fig.1. Angioarchitectonics of the abdominal aorta: 1 - aorta abdominalis; 2 - a. renalis dextra; 3 - a. renalis sinistra; 4 - a. polaris inferioris renis; 5 - a. mesenterica superior; 6, 7 - aa. iliaca communis (angio-CT).

On the right side, two supernumerary renal arteries were identified: the first artery with origin from the abdominal aorta, above the origin of the right renal artery, determined as arteria renalis segmentaris inferioris (d=0.22 cm), due to its

descending course, at the level of the renal hilum it intersected the arteria segmenti superioris renis, after which it entered the renal sinus and continued to the parenchyma of the lower renal segment. The second supernumerary artery started from the right renal artery itself, considered as arteria polaris inferioris (d=0.57 cm), because it did not penetrate the renal hilum, but descended to the lower pole of the organ, where it penetrated the renal parenchyma (Figure 2).

Abdomino-pelvic CT with contrast showed dilation of the superior group of renal calyces, compressed by the arteria segmenti inferioris renis, much higher to the point of intersection of the arteria segmenti inferioris renis with the arteria segmenti superioris renis. Thus, due to the uretero-vascular conflict characteristic of Fraley's syndrome, the patient was diagnosed with degree II-III hydronephrosis of the right kidney (Figure 2).

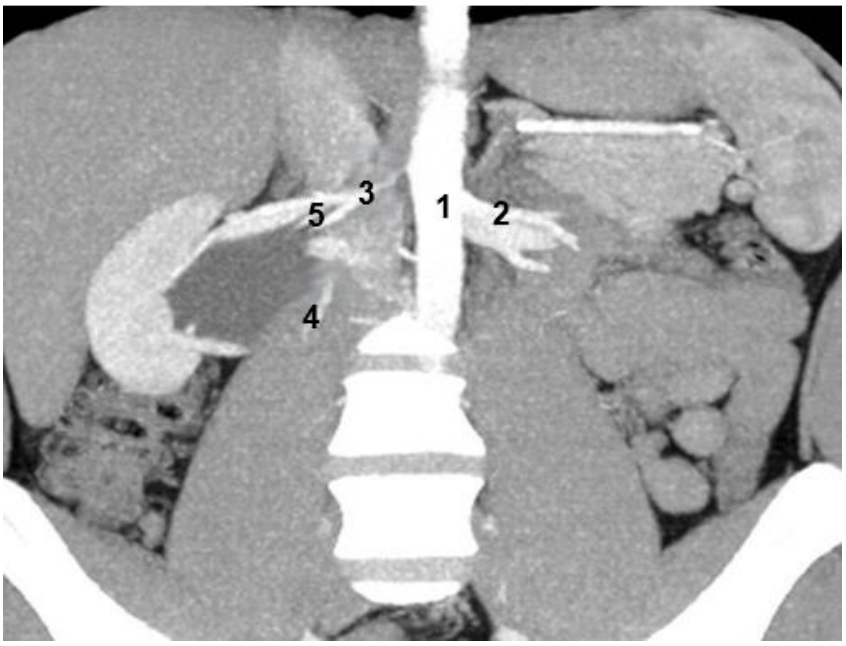

Fig.2. Renal and supernumerary renal arteries of the right kidney: 1 - aorta abdominalis; 2 – a. renalis sinistra; 3 – a. renalis dextra; 4 – a. polaris inferioris renis, 5 – a. segmenti inferioris renis (angio-CT).

For the first time, obstruction of the renal collecting system caused by the intersection of the branches of the renal artery, defined as an asymptomatic vascular malformation, was described by Fraley E. (1966), hence it was named Fraley syndrome (FS) [4]. As reported by Benz G., et al. (1977), this form of intrarenal vascular obstruction clinically is manifested by nephralgia or hematuria. The authors noted the obstruction of the distal segment of the renal collecting system caused by the abnormal course of the renal vessels, more frequently of the superior segmental artery, which causes filling defect of the infundibulum of the renal pelvis, and as a consequence, the superior calyceal group is distended and presents a delay in emptying, increasing the risk of urinary tract infection or stones formation [3].

Cerqueira TB., et al. (2007) studied Fraley's syndrome according to laterality, establishing its unilateral presence, more frequently on the right side [4].

Rare presence of FH was mentioned by Antonio D'Amico et al. (2000), who performed an imaging study over a 20-year period (1976-1996), identifying it in only 6 patients, the diagnosis being made by intravenous pyelography and renal arteriography. In 5 patients this syndrome was detected on the right side, and in the 6th patient - it was bilaterally diagnosed [6].

Considering that this vascular anomaly more frequently is asymptomatic, it can be revealed very late, already at an adult age, and often its diagnosis is missed [6].

Thanks to modern imaging methods, Fraley's syndrome can be identified, a fact confirmed by many authors [1, 3, 4], but in the specialized literature these cases are rarely reported [2].

Conclusion. Fraley's syndrome is a rare cause of obstruction of the intrarenal collecting system, and the present case highlights the importance of computed tomography in its identification. Knowledge of the specific radiologic aspects of Fraley's syndrome is necessary for the optimal choice of the surgical tactics to avoid medical errors.

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