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Original contribution

Congenital capillary proliferation of the kidney: a distinctive renal vascular lesion of childhood

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Summary Renal vascular lesions (RVL) are rare, and their morphological spectrum remains largely unknown, particularly in children. In this study, we characterize the clinicopathological features of RVL in a cohort of 12 children. Seven lesions were classified as previously recognized entities: vascular malformations (4), papillary endothelial hyperplasia (2), and pyogenic granuloma (lobular capillary hemangioma; 1). An eighth lesion showed nonspecific findings, which were interpreted as reactive during our review. The remaining 4 cases presented either prenatally, at birth, or shortly after birth and were morphologically similar. These were characterized by a peculiar pattern of capillary proliferation with entrapment of native renal structures, variable amounts of extramedullary hematopoiesis and reactive lymphocytes, foci of infarction and hemorrhage, and the presence of feeding and draining vessels at their periphery. To our knowledge, this represents a previously undescribed congenital vascular lesion involving the kidney, which we have descriptively and provisionally termed congenital capillary proliferation of the kidney (CCPK). While it is unclear whether CCPK represents a malformation or neoplastic proliferation, it shows overlapping features with congenital hemangioma of the liver (solitary congenital hepatic hemangioma) and congenital nonprogressive hemangioma (CNH) of the skin and soft tissue, suggesting a possible common pathogenesis among these 3 entities.

1. Introduction

Vascular lesions are among the most common mesenchymal lesions of the skin and soft tissue and have been extensively characterized in the literature, as reflected in a recent 2014 classification system proposed by the International Society for the Study of Vascular Anomalies (ISSVA) [1,2]. In

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contrast, the literature on visceral vascular lesions remains scant and limited by the use of overlapping nomenclature, which often does not reflect the terminology used by ISSVA. Among visceral vascular lesions, renal vascular lesions (RVL) are particularly uncommon, and their morphological spectrum has not been fully explored. Although many isolated case reports of RVL occurring in adults can be found in the literature [3,4], only a limited number of cases has been systematically studied [5,6], resulting in a few well-characterized entities. In contrast to adult RVL, the morphological spectrum of pediatric RVL remains largely unknown. In an attempt to characterize these lesions, we provide an evaluation of unselected RVL submitted to the Renal Tumor Pathology Center in a 21-year period, and describe an unrecognized vascular lesion occurring in the infant kidney.

2. Materials and methods

The study was approved by the Ann & Robert H. Lurie Children's Hospital Internal Review Board. We identified all pediatric RVL in the Renal Tumor Pathology Center files from 1994 to 2015. A full set of hematoxylin and eosin (H&E) slides, the institutional pathology report, and available immunohistochemical and other ancillary studies were reviewed for all cases. Unstained formalin-fixed, paraffin-embedded (FFPE) slides and/or paraffin blocks were available for some of the cases. Additional immunohistochemical stains were performed on select cases. Most cases were received in consultation, and only limited clinical history was available for a subset of these cases.

3. Results

Twelve cases of RVL were identified, corresponding to 7 females and 5 males with ages ranging from 1 week to 14 years. The clinicopathological features of these 12 cases are summarized in the Table. Seven cases (1–7) corresponded to lesions that had been previously characterized in the literature, and an eighth case showed nonspecific changes interpreted as reactive. In addition, 4 cases (9–12) had unique morphological features that did not fit into any previously described entity in the kidney.

3.1. Cases 1 to 4

These 4 cases were classified as vascular malformations and subtyped according to the 2014 ISSVA classification [1]. Cases 1 and 2 (27 months and 14 years, respectively) were well-circumscribed lymphatic malformations composed of numerous small lymphatic vessels (showing CD31 and faint D2-40 endothelial immunohistochemical expression) located in the interstitium and permeating the renal parenchyma (Fig. 1A). Case 3 (22 months), a capillary venous malformation, was entirely surrounded by a fibrous pseudocapsule and composed of capillary and venous channels with areas of sclerosis and edema and occasional thrombi (Fig. 1B). Several small red-blue lesions were identified in the bowel serosa at the time of nephrectomy in case 3, which showed identical morphological features to the renal lesion. A lymphatic component was not evident upon histopathological examination of both the kidney and bowel serosal lesions, and no unstained FFPE slides were available for immunohistochemistry. Case 4 (8 years), an arteriovenous malformation, was an ill-defined proliferation of dilated, aberrantly shaped arteries and veins involving both the native renal parenchyma and the renal sinus soft tissue (Fig. 1C). Frequent permeation of renal tubules and glomeruli by the aberrant vessels was appreciated. The renal parenchyma showed marked chronic inflammation and scarring, as well as architectural disorganization with features of renal dysplasia/maldevelopment.

3.2. Cases 5 and 6

These lesions consisted of delicate fibrovascular structures containing organized fibrin and showing variable hyalinization

Table Clinicopathological features of 12 RVL occurring in children

Case	Age*	Gender	Laterality	Procedure	Size (cm)	Diagnosis
1	27 mo	M	Left	Total nephrectomy	N/A	Lymphatic malformation
2	14 y	F	Right	Total nephrectomy	3.8	Lymphatic malformation
3	22 mo	F	Left	Total nephrectomy	6.5	Capillary venous malformation
4	8 y	F	Left	Total nephrectomy	N/A	Arteriovenous malformation with lymphatic component
5	14 mo	F	Right	Excisional biopsy	1.5	Organizing hematoma (papillary endothelial hyperplasia)
6	7 wk	M	Left	Total nephrectomy	3.0	Organizing thrombus (papillary endothelial hyperplasia)
7	6 wk	M	Left	Total nephrectomy	N/A	Favor pyogenic granuloma (lobular capillary hemangioma)
8	11 y	F	Left	Total nephrectomy	2.1	Suggestive of reactive vascular proliferation
9	7 wk	F	Right	Total nephrectomy	4.5	Congenital capillary proliferation of the kidney
10	13 wk	M	Right	Partial nephrectomy	3.8	Congenital capillary proliferation of the kidney
11	2 wk	F	Left	Total nephrectomy	2.8	Congenital capillary proliferation of the kidney
12	1 wk	M	Left	Total nephrectomy	1.5	Congenital capillary proliferation of the kidney

Abbreviation: N/A, not available.
* At the time of nephrectomy.