Virchows Arch (2000) 436:398–400

CASE REPORT

**Hyung-Seok Kim · Eun-Jung Park · Jae-Hyuk Lee
Jong-Hee Nam · Min Cheol Lee · Chang-Soo Park
Sang-Woo Juhng · Chan Choi**

Intravascular papillary endothelial hyperplasia in the kidney of a child

Received: 29 September 1999 / Accepted: 28 October 1999

Abstract Intravascular papillary endothelial hyperplasia (IPEH) is a benign vascular lesion which is thought to represent an unusual form of organizing thrombus. A case of IPEH in the kidney of a 7-year-old girl is described. She suffered from intermittent flank pain and gross hematuria for 6 months. On radiological examinations, well-defined hypoechoic lesions were identified in the medullary portion of the left kidney. A well-demarcated, sponge-like mass was noted on gross examination. It was an intravascular mass lined by a fibrous capsule of various thicknesses. It was characterized by papillary fronds lined with benign endothelial cells. This is the first description of a renal IPEH in a child.

Key words Intravascular papillary endothelial hyperplasia · Kidney · Child · Masson's hemangioma

Introduction

Intravascular papillary endothelial hyperplasia (IPEH) is a common vascular lesion of soft tissue, which was first described by Masson in 1923 [11]. This lesion is interpreted as the result of reactive endothelial proliferation rather than a neoplasm [7]. Clearkin and Enzinger first used the term IPEH in 1976 [3]. It mimics a malignant vascular tumor morphologically and is referred to by several terms: hemangioendotheliome végétant intra-vasculaire [11], intravascular angiomatosis [13], and Masson's pesudoangiosarcoma [10]. It is commonly located within veins in the head, neck, fingers and trunk [3, 6, 10, 13], where it appears as a small, firm, superficial mass. Hashimoto [6] classified it into three types by sites and associated lesions. The pure form is characterized by lesions within a dilated vascular space, and the mixed form, by a focal area of intravascular papillary endothelial hyperplasia in hemangioma. The third form belongs to neither of the first two. There have been over 200 well-documented cases of IPEH. However, visceral IPEH is rare [2, 4, 5, 8, 9, 14]. The visceral IPEH reported include three cases of renal IPEH [5, 9, 14]. Interestingly, all previous cases of renal IPEH have occurred in adults, and it has never been reported in a child before now. This report documents a new case of a renal IPEH found in a child and briefly reviews the relevant literature.

H.-S. Kim · E.-J. Park · J.-H. Lee · J.-H. Nam · M. C. Lee
C.-S. Park · S.-W. Juhng · C. Choi
Department of Pathology,
Chonnam National University Medical School,
Chonnam University Research Institute of Medical Science,
Kwangju, Republic of Korea

C. Choi (✉)
#5 Hakdong, Dong ku, Kwangju, Republic of Korea, 501–190
e-mail: cchoi@chonnam.chonnam.ac.kr
Tel.: +82-62-2321246, Fax: +82-62-2273429

Clinical history

A 7-year-old girl was referred from the pediatric clinic for investigation of intermittent abdominal pain and gross hematuria that had been present for 6 months. Physical examination did not reveal a flank mass. Routine laboratory tests were within normal limits except that hematuria was obvious on urine analysis. A renal ultrasonography revealed a well-circumscribed echogenic mass in the mid-portion of the left kidney. A CT revealed a low attenuation mass 1.8×1.6 cm in size and without a fat component in the pelvic region of the left kidney (Fig. 1). On admission, vanillylmandelic acid, homovanillic acid, and neuron-specific enolase in the 24-h urine were within normal limits. A left nephrectomy was done as a renal tumor, such as Wilms' tumor, neuroblastoma, or angiomyolipoma, was suspected.

Materials and methods

Formalin-fixed paraffin-embedded tissue sections were stained with hematoxylin and eosin for routine examination. Immunohistochemical staining was done with the Microprobe Immuno/DNA stainer (Fisher Scientific, Canada). Antibodies for factor VIII-related antigen (Zymed USA), CD34 (DAKO, USA), and type-IV collagen (DAKO, USA) were used.

Pathological findings

The left kidney was 10×6×3 cm in size. External examination was nonspecific. When bisected, a well-circum-

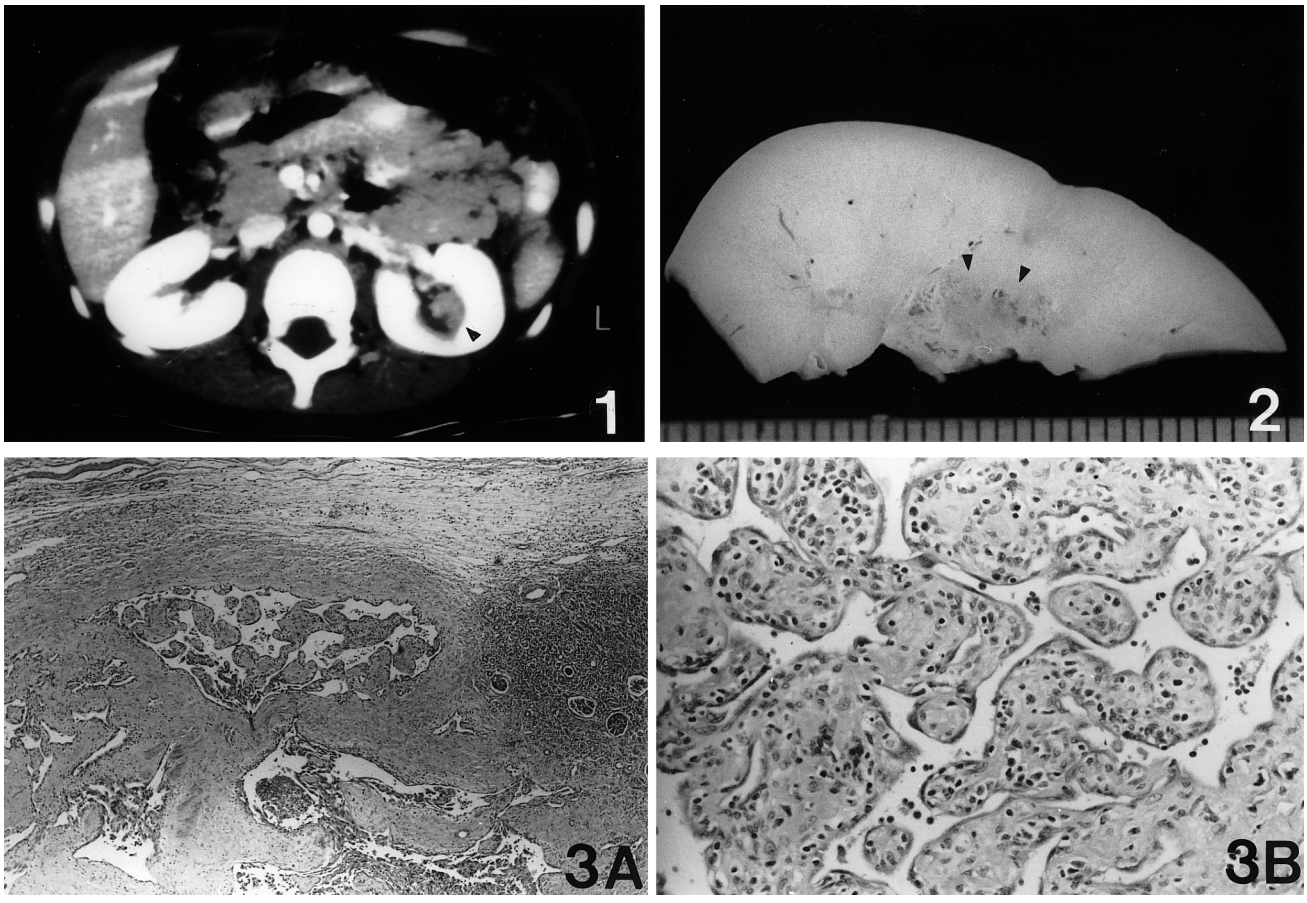

Fig. 1 CT reveals a round mass with homogeneous low density, measuring 1.8×1.6 cm (*arrowhead*)

Fig. 2 The cut surface of left kidney reveals an ill-defined, sponge-like lesion in the medullary portion (*arrowhead*)

Fig. 3 **A** The low magnification shows an intravascular papillary mass within a fibrous capsule. (Hematoxylin-eosin, original magnification ×40). **B** Some papillae appear as free-floating islands within the vascular space. One layer of swollen or plump endothelial cells line the papillary structures. Cellular pleomorphism or mitotic figures are not noted. (Hematoxylin-eosin, original magnification ×200)

scribed, sponge-like lesion 1.8×1.6 cm in size was noted in the medullary portion (Fig. 2).

Histological preparations showed a vascular mass lined by a fibrous capsule of variable thickness separating it from the surrounding renal tissue. The lesion was characterized by papillary structures lined by a layer of endothelial cells (Fig. 3A). Some papillae appeared as free-floating islands within a vascular space. The papillary structures were covered with a single layer of plump endothelial cells. Cellular pleomorphism, stratification or mitotic figures were not noted. Endothelial cells have one nucleus, and occasionally a small indistinct nucleolus. Foci of hemangioma or lymphangioma were not found. These light microscopic findings closely matched the reports of other authors who describe IPEH in the kidney and other sites [12–14].

We observed a positive granular or diffuse staining reaction for factor VIII-related antigen in the cytoplasms of the endothelial cells covering the papillary fronds. Type-IV collagen showed positive reactivity along the cytoplasm of the endothelial cells. Immunohistochemistry for CD 34 was negative in the cytoplasms of the endothelial cells covering the papillary fronds of the IPEH.

Discussion

Intravascular papillary endothelial hyperplasia is characterized by exuberant intravascular endothelial proliferation. Although IPEH of soft tissue is commonly found, visceral IPEH is rare [2, 4, 5, 8, 9, 14]. Three cases of renal IPEH have been reported, and all of them had developed in adults [5, 9, 14]. This report is the first description of renal IPEH in a child.

Previously reported renal IPEHs in adults have been in the medullary portion and have had overall histological features similar to those of IPEHs in other sites. Cutaneous and soft-tissue lesions are classified into three forms: a pure form that occurs within a dilated vascular space, a mixed form that appears as focal changes in hemangioma, and a third form that belongs to neither of the first two [6]. The present case of IPEH has the pure form. Although the pathogenesis of IPEH is still a matter of speculation, many authors agree with the opinion of Clearkin

[3] and Salyer [13] that it is probably a peculiar morphological feature of a thrombus undergoing organization.

Some benign and malignant vascular tumors of the kidney should be excluded in the differential diagnosis of IPEH. Renal hemangioma usually presents with intermittent gross hematuria. It can be hypervascular or normal on angiography [12]. Microscopically, most renal hemangiomas are cavernous, with large blood-filled vascular spaces lined by a layer of cytologically benign, nonpapillary endothelial cells [5]. The differentiation of IPEH from angiosarcoma [1] is difficult in some cases. The findings which favor an IPEH over an angiosarcoma are as follows: (1) the papillary endothelial proliferation is entirely confined to intravascular spaces that otherwise have normal endothelial linings; (2) most of the papillary structures are associated with thrombi; (3) the endothelial cells in the papillary structures show only slight atypia and occasional mitotic figures; (4) a "piling up" of the endothelial cells is unusual; and (5) tumor necrosis is rare [6].

The space-occupying lesions of the kidney in childhood are Wilms' tumor, mesoblastic nephroma, rhabdoid tumor, and clear cell sarcoma. In order to avoid an unnecessary surgical procedure, benign vascular tumors should be included in the category of the space-occupying lesions of the kidney. Needle biopsy should be avoided because of the risk of bleeding.

Our report of a case of renal IPEH during childhood and the description of its pathological features and differential diagnoses led us to the following conclusions. Papillary endothelial hyperplasia should be included in the differential diagnosis of vascular lesions in this site. Preoperative diagnosis of renal IPEH is difficult because there are no particular symptoms and no general agreement on the radiological findings. However, no case of metastasis or malignant transformation has been reported with IPEH. Intra-operative tissue examination should be considered if operative treatment is chosen.

References

1. Cerilli LA, Huffman HT, Anand A (1998) Primary renal angiosarcoma: a case report with immunohistochemical, ultrastructural, and cytogenetic features and review of the literature. Arch Pathol Lab Med 122:929–935
2. Chen TJ, Kuo T (1984) Giant intracranial Masson's hemangioma. Report of a fatal case. Arch Pathol Lab Med 108:555–556
3. Clerkin KP, Enzinger FM (1976) Intravascular papillary endothelial hyperplasia. Arch Pathol Lab Med 100:441–444
4. Duong DH, Scoones DJ, Bates D, Sengupta RP (1997) Multiple intracerebral intravascular papillary endothelial hyperplasia. Acta Neurochir (Wien) 139:883-886
5. Garber BB, Prestipino AJ, Pollack HM, Levine SR, Whitmore KE (1990) Masson's tumor of the kidney: a new renal lesion. J Urol 143:344–346
6. Hashimoto H, Daimaru Y, Enjoji M (1983) Intravascular papillary endothelial hyperplasia. A clinicopathological study of 91 cases. Am J Dermatopathol 5:539–546
7. Henschen F. L'endovasculite proliferante thrombopoietique dans la lesion vasculaire locale. Ann Anat Pathol (Paris) 9: 113–121
8. Izukawa D, Lach B, Benoit B (1987) Intravascular papillary endothelial hyperplasia in an intracranial cavernous hemangioma. Neurosurgery 21:939–941
9. Johraku A, Miyanaga N, Sekido N, Ikeda H, Michishita N, Saida Y, Fujiwara M, Noguchi M, Shimazui T, Akai H (1997) A case of intravascular papillary endothelial hyperplasia (Masson's tumor) arising from renal sinus. Jpn J Clin Oncol 27: 433–436
10. Kuo T, Sayers CP, Rosai J (1976) Masson's "vegetant intravascular hemagioendothelioma": a lesion often mistaken for angiosarcoma: study of seventeen cases located in the skin and soft tissues. Cancer 38:1227–1236
11. Masson P (1923) Hemangioendotheliome végétant intra-vasculaire. Bull Soc Anat (Paris) 95:517–532
12. Petersen NE, Thompson HT (1971) Renal hemangioma. J Urol 105:27–31
13. Salyer WR, Salyer DC (1975) Intravascular angiomatosis: development and distinction from angiosarcoma. Cancer 36: 995–1001
14. Steffee CH, Iskandar SS (1996) Intravascular papillary endothelial hyperplasia in a thrombosed renal allograft vein. Hum Pathol 27:986–98