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**Primary vascular tumours of the kidney**

Omiyale AO. Primary vascular tumours of the kidney

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**Abstract**

Primary vascular tumours of the kidney are rare and may pose diagnostic difficulties because of their similar clinical, morphological, and immunohistochemical features. This article summarizes the clinical and pathological features of primary renal angiosarcoma and anastomosing haemangioma of the kidney including epidemiology, genetics, and prognosis. Renal anastomosing haemangiomas are benign neoplasms characterized by anastomosing capillary-sized vascular channels. These tumours are rare, with about 75 cases reported in the literature. Most anastomosing haemangiomas are found incidentally on ultrasound, computed tomography, or magnetic resonance imaging. Common symptoms include abdominal pain, haematuria, and abdominal mass. Renal anastomosing haemangiomas are characterized by recurrent mutations in *GNAQ* and *GNA14* genes. The prognosis of anastomosing haemangioma is excellent.

Primary renal angiosarcomas are malignant tumours showing endothelial differentiation. To date, 76 cases have been described in the literature. Primary renal angiosarcomas are frequently symptomatic. The clinical features of renal angiosarcomas are similar to those of renal anastomosing haemangiomas, including abdominal pain, haematuria, and abdominal mass. Angiogenesis-related genes and vascular-specific receptor tyrosine kinases such as KDR, TIE1, SNRK, TEK*,* andFLT1 are upregulated in angiosarcomas. Primary renal angiosarcomas are highly aggressive neoplasms with a poor prognosis despite surgical treatment, chemotherapy, radiotherapy, or targeted therapy.

**Key Words:** Kidney; Renal tumours; Angiosarcoma; Haemangioma; Anastomosing haemangioma of the kidney; Vascular tumours

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**Core Tip:** Primary vascular tumours of the kidney are extremely rare. This article summarizes the clinical and pathological features of primary renal angiosarcoma and anastomosing haemangioma of the kidney.

**INTRODUCTION**

Although vascular tumours are relatively common in the skin and soft tissue, they are extremely rare in the kidney, ranging from benign to malignant neoplasms that may be diagnostically challenging because of the overlapping clinical, morphological and immunohistochemical features.

These tumours include renal angiosarcomas and renal haemangiomas. Various subtypes of haemangioma have been described in the kidney including cavernous, capillary, and anastomosing haemangiomas[1-4]. However, the most common subtype is anastomosing haemangioma[1,2,5].

This article provides an overview of the clinical and pathological features of anastomosing haemangioma of the kidney and primary angiosarcoma of the kidney, and discusses the epidemiology, genetics, and prognosis.

**ANASTOMOSING HAEMANGIOMA OF THE KIDNEY**

Renal anastomosing haemangiomas are benign neoplasms characterized by anastomosing capillary-sized vascular channels. These tumours are exceptionally rare with about 75 anastomosing haemangiomas reported in the literature[5-9]. These tumours occur in a wide age range from 10 to 83 years (mean, 49 years) with a male-to-female ratio of 2:1[10].

The aetiology and risk factors for renal anastomosing haemangiomas are unknown. Some cases have been reported in the setting of end stage renal disease[11,12].

The vast majority of anastomosing haemangiomas are found incidentally on radiological evaluation for other purposes. Common symptoms include abdominal pain, haematuria, and abdominal mass[5,10].

The imaging findings are non-specific. On computed tomography, these tumours are often circumscribed, hyperdense, and heterogeneous due to fatty or non-enhancing hypodense areas and show post-contrast enhancement[13].

Renal anastomosing haemangiomas are characterized by recurrent mutations in *GNAQ* and *GNA14* genes[14,15]. *GNAQ* gene encodes guanine nucleotide-binding protein G (q) subunit alpha (Gαq protein) that activates signalling pathways that regulates cell proliferation, survival, development, and function of blood vessels[14-16].

Grossly, anastomosing haemangiomas are typically small ranging from 0.1 cm to 12 cm (mean, 2.2 cm)[5,10,17]. These tumours are often well-demarcated spongy mahogany brown masses[5]. They are usually unilateral and solitary tumours; however, a few cases of bilateral[18] and multifocal[11] tumours have been described.

Histologically, anastomosing haemangiomas consist of anastomosing capillary-sized blood vessels, reminiscent of splenic sinusoids. The blood vessels are lined by bland endothelial cells. Typically, these tumours lack endothelial cell multilayering, papillary tufting, cytologic atypia, necrosis, and prominent mitotic figures. They may show extramedullary haematopoiesis, hyaline globules, and mild lymphocytic infiltrate[1,2,5,10,18].

Rarely, renal anastomosing haemangiomas may infiltrate perinephric fat, renal sinus fat[2,12], and the renal vein or its segmental branches[2,12,18,19]. The neoplastic cells are immunoreactive for CD31, CD34, ERG, FLI1, and factor VIII-related antigen (now rarely used)[5].

Renal anastomosing haemangiomas may co-exist with other renal neoplasms such as metanephric adenoma, papillary adenoma, papillary renal cell carcinoma, acquired cystic disease-associated renal cell carcinoma, and clear cell renal cell carcinoma[11,12].

Most patients with renal anastomosing haemangioma, described in the literature, were treated with radical nephrectomy, reflecting a tendency to overtreat these patients, probably because of inaccurate preoperative diagnosis (Table 1).

The prognosis of anastomosing haemangioma is excellent with no evidence of recurrence, metastasis or tumour-related death at an average follow-up of 24.8 mo (range, < 1-156 mo) (Table 1).

**PRIMARY ANGIOSARCOMA OF THE KIDNEY**

Renal angiosarcomas are malignant tumours showing endothelial differentiation. These tumours are very rare, with about 76 cases described in the literature as case series and reports[20-24]. There is a male-to-female ratio of 6:1, with patient age ranging from 24 years to 95 years (median, 62 years).

Although angiosarcomas arising in other anatomical sites have been associated with risk factors such as exposure to thorium dioxide, arsenic-based pesticides, polyvinyl chloride, and radiation therapy particularly for breast, endometrial and prostate cancers[25-27], no specific aetiology or risk factors have been established for primary angiosarcoma of the kidney[20,28].

The clinical features of renal angiosarcomas are identical to those of renal anastomosing haemangiomas. Common symptoms include abdominal pain, haematuria, abdominal mass, and weight loss. A rare case of spontaneous tumour rupture with retroperitoneal haematoma has been described[29].

Computed tomography imaging shows large masses with heterogeneous enhancement and hypervascularity[30].

Angiogenesis-related genes and vascular-specific receptor tyrosine kinases such as KDR, TIE1, SNRK, TEK*,* andFLT1, are upregulated in angiosarcomas[31]. High-level MYC gene amplifications are seen in most radiation-induced and chronic lymphoedema-associated angiosarcomas[32]. A subset of cases is characterized by PLCG1, KDR, and PTPRB mutations[33,34].

Some primary angiosarcomas, typically in young adults, have recurrent CIC gene rearrangements, with or without concurrent CIC mutations, and are characterized by upregulation of CIC-target genes including *ETV1, ETV4,* and *ETV5*[34]. Angiosarcomas with CIC gene abnormalities are associated with an inferior disease-free survival[34].

Primary renal angiosarcomas are usually large ranging from 3.6 cm to 30 cm (mean, 13 cm). Typically, they are ill-defined haemorrhagic spongy masses with necrosis[1,20].

Microscopically, these tumours range from well-formed vasoformative areas to areas with solid morphology showing sparse vasoformation. These patterns are often mixed within the same tumour. Vasoformative areas are composed of small to medium-sized anastomosing blood vessels, lined by epithelioid and/or spindled endothelial cells showing nuclear pleomorphism, endothelial papillary tufting, multilayering, intraluminal budding, and hobnailing[1,20,28]. Solid areas consist of sheets of malignant epithelioid and/or spindled cells with subtle vasoformation, cytologic atypia, and mitotic figures. Necrosis may be present. Angiosarcomas usually have a haemorrhagic background and extravasated red blood cells are seen within the tumour[1,20,28].

Epithelioid angiosarcomas are composed of sheets of large atypical polygonal or epithelioid cells with nuclear pleomorphism, high nuclear to cytoplasmic ratio, prominent central nucleoli, mitotic figures, and moderate amounts of cytoplasm. Epithelioid angiosarcomas may be mistaken for carcinoma, melanoma, or lymphoma[20,28,35-37].

The neoplastic cells are positive for CD31, ERG, FLI1, CD34, and factor VIII-related antigen[20,28,38,39]. Epithelioid angiosarcomas may be positive for epithelial markers including CK7, Cam5.2, AE1/AE3, and EMA, which may lead to a misdiagnosis of carcinoma[20].

Similar to angiosarcomas arising at other locations, renal angiosarcomas have a tendency for widespread metastasis at diagnosis or afterwards in the course of the disease. Approximately 66% of patients develop metastases, most commonly to the lung and liver. Other sites of metastasis include bone, lymph nodes, peritoneum, small bowel, soft tissue, and skin. Currently, there are no specific standardized treatment guidelines for primary renal angiosarcomas. These tumours are treated with radical nephrectomy, chemotherapy, radiotherapy, or targeted therapy (Table 2).

Primary renal angiosarcomas are highly aggressive neoplasms with 76.3% of patients dying of tumour within 1 mo to 24 mo (mean, 7.5 mo), despite surgical and adjuvant therapy (Table 2). Poor prognostic factors for angiosarcomas occurring at other anatomic sites include age > 69 years, tumour size ≥ 5 cm, regional disease (*vs* localized disease), non-surgical treatment, and distant metastasis[40].

**CONCLUSION**

Primary vascular tumours of the kidney are rare neoplasms. Unlike primary renal angiosarcoma, the prognosis of renal anastomosing haemangioma is excellent with no evidence of recurrence or metastasis. These tumours share similar clinical, morphological and immunohistochemical features, and must be distinguished from each other. Features that favour angiosarcomas include the presence of malignant spindled and/or epithelioid cells with a variable degree of vasoformation, cytologic atypia, prominent mitotic figures, endothelial multilayering, papillary tufting, and necrosis.

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**Footnotes**

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**Table 1 Treatment, follow-up, and outcome of patients with anastomosing haemangioma of the kidney**

|  |  |  |  |  |
| --- | --- | --- | --- | --- |
| **Author** |  | **Treatment** | **Follow-up (mo)** | **Outcome** |
| Bean *et al*[14] |  | Nephrectomy | 9 | NED |
| Bean *et al*[14] |  | Nephrectomy | 84 | NED |
| Bean *et al*[14] |  | Nephrectomy | 107 | NED |
| Memmedoğlu and Musayev[41] |  | Nephrectomy | 12 | NED |
| Memmedoğlu and Musayev[41] |  | Nephrectomy | 12 | NED |
| Tahir and Folwell[42]  |  | Nephrectomy | 1 | NED |
| Pantelides *et al*[6] |  | Nephrectomy | 6 | NED |
| Downes *et al*[7] |  | Nephrectomy | NA | NA |
| Downes *et al*[7] |  | Biopsy | NA | NA |
| Chandran *et al*[8] |  | Nephrectomy | NA | NA |
| Cha *et al*[9] |  | Nephrectomy | 5 | NED |
| Montgomery and Epstein[2]  |  | Nephrectomy | 12 | NED |
| Montgomery and Epstein[2]  |  | Nephrectomy | 36 | NED |
| Montgomery and Epstein[2]  |  | Nephrectomy | NA | NA |
| Montgomery and Epstein[2]  |  | Excision | 8 | NED |
| Heidegger *et al*[43]  |  | Nephrectomy | 156 | NED |
| Kryvenko *et al*[12] |  | Nephrectomy | NA | NA |
| Kryvenko *et al*[12] |  | Nephrectomy | NA | NA |
| Kryvenko *et al*[12] |  | Nephrectomy | NA | NA |
| Kryvenko *et al*[12] |  | Nephrectomy | NA | NA |
| Kryvenko *et al*[12] |  | Nephrectomy | NA | NA |
| Kryvenko *et al*[12] |  | Nephrectomy | NA | NA |
| Kryvenko *et al*[12] |  | Nephrectomy | NA | NA |
| Kryvenko *et al*[12] |  | Nephrectomy | NA | NA |
| Kryvenko *et al*[12] |  | Nephrectomy | NA | NA |
| Kryvenko *et al*[12] |  | Nephrectomy | NA | NA |
| Kryvenko *et al*[12] |  | Nephrectomy | NA | NA |
| Kryvenko *et al*[12] |  | Nephrectomy | NA | NA |
| Al-Maghrabi and Al-Rashed[44] |  | Partial nephrectomy | 12 | NED |
| Caballes *et al*[17] |  | Nephrectomy | 18 | NED |
| Büttner *et al*[11] |  | Nephrectomy | NA | NA |
| Büttner *et al*[11] |  | Nephrectomy | NA | NA |
| Büttner *et al*[11] |  | Nephrectomy | NA | NA |
| Büttner *et al*[11] |  | Nephrectomy | NA | NA |
| Büttner *et al*[11] |  | Nephrectomy | NA | NA |
| Büttner *et al*[11] |  | Nephrectomy | NA | NA |
| Büttner *et al*[11] |  | Nephrectomy | NA | NA |
| Büttner *et al*[11] |  | Nephrectomy | NA | NA |
| Lee *et al*[45] |  | Nephrectomy | NA | NA |
| Zhao *et al*[46]  |  | Nephrectomy | 12 | NED |
| Kryvenko *et al*[18]. |  | Nephrectomy | 7 | NED |
| Kryvenko *et al*[18]. |  | Nephrectomy | 6 | NED |
| Kryvenko *et al*[18]. |  | Nephrectomy | 3 | NED |
| Kryvenko *et al*[18]. |  | Nephrectomy | 122 | NED |
| Tao *et al*[47] |  | Nephrectomy | 21 | NED |
| Abboudi *et al*[48] |  | Nephrectomy | <1 | NED |
| Silva *et al*[49] |  |  Resection | NA | NA |
| Berker *et al*[50] |  | Partial nephrectomy | 10 | NED |
| Berker *et al*[50] |  | Nephrectomy | 4 | NED |
| O'Neill *et al*[13] |  | NA | NA | NA |
| O'Neill *et al*[13] |  | NA | NA | NA |
| O'Neill *et al*[13] |  | NA | NA | NA |
| O'Neill *et al*[13] |  | NA | NA | NA |
| O'Neill *et al*[13] |  | NA | NA | NA |
| O'Neill *et al*[13] |  | NA | NA | NA |
| O'Neill *et al*[13] |  | NA | NA | NA |
| Brown *et al*[1]  |  | Nephrectomy | 72 | NED |
| Brown *et al*[1]  |  | Nephrectomy | 24 | NED |
| Brown *et al*[1]  |  | Partial nephrectomy | NA | NA |
| Brown *et al*[1]  |  | Nephrectomy | 24 | NED |
| Brown *et al*[1]  |  | Nephrectomy | NA | NA |
| Perdiki *et al*[51] |  | Partial nephrectomy | 25 | NED |
| Perdiki *et al*[51] |  | Nephrectomy | 14 | NED |
| Wetherell *et al*[52] |  | Nephrectomy | 1 | DFUD |
| Manohar *et al*[53] |  | Nephrectomy | 24 | NED |
| Omiyale *et al*[19] |  | Nephrectomy | 10 | NED |
| Johnstone *et al*[54] |  | Nephrectomy | NA | NA |
| Mehta *et al*[4] |  | Nephrectomy | 3 | NED |
| Mehta *et al*[4] |  | Nephrectomy | 12 | NED |
| Mehta *et al*[4] |  | Nephrectomy | 3 | NED |
| Tran and Pernicone[55]  |  | Nephrectomy | NA | NA |
| Zhang *et al*[56] |  | Partial nephrectomy | 16 | NED |
| Cheon *et al*[57] |  | Nephrectomy | 6 | NED |
| Chou *et al*[58] |  | Nephrectomy | 8 | NED |
| Chou *et al*[58] |  | Nephrectomy | 14 | NED |

DFUD: Died from unrelated disease; NED: No evidence of disease; NA: Not available.

**Table 2 Treatment, follow-up, and outcome of patients with primary angiosarcoma of the kidney**

|  |  |  |  |  |
| --- | --- | --- | --- | --- |
| **Ref.** |  | **Treatment** | **Follow-up (mo)** | **Outcome** |
| Costero-Barrios *et al*[59]  |  | Nephrectomy, Chemo, RT | 12 | AWD |
| Peters *et al*[60] |  | Nephrectomy | 2 | DOD |
| Singh *et al*[35] |  | NA | NA | NA |
| Kern *et al*[61] |  | Nephrectomy | 3 | DOD |
| Kern *et al*[61] |  | Nephrectomy | 1.5 | DOD |
| Aydogdu *et al*[62] |  | Nephrectomy | NA | NA |
| Akkad *et al*[63] |  | Nephrectomy | 30 | NED |
| Witczak *et al*[64] |  | nephrectomy | NA | NA |
| Chaabouni *et al*[38] |  | Nephrectomy | 1 | DOD |
| Johnson *et al*[65] |  | Rapid deterioration | NA | DOD |
| Zenico *et al*[66] |  | Nephrectomy | 4 | DOD |
| Nguyen *et al*[67] |  | Nephrectomy, Chemo | 18 | DOD |
| Terris *et al*[68] |  | Nephrectomy, RT | 10 | DOD |
| Matter *et al*[69] |  | Nephrectomy, Chemo, RT | 18 | DOD |
| Yoshida *et al*[70] |  | Nephrectomy, Recombinant IL-2  | 13 | DOD |
| Pauli and Strutton[71]  |  | Nephrectomy, RT | 2 | DOD |
| Martínez-Piñeiro *et al*[72] |  | Nephrectomy, S | 4 | DOD |
| Bernstein *et al*[73] |  | NA | NA | NA |
| Liu *et al*[36] |  | Nephrectomy, RT | 6 | NED |
| Yau *et al*[74] |  | Nephrectomy, Chemo, RT | 3 | DOD |
| Carnero López *et al*[75] |  | Nephrectomy, Chemo | 5 | DOD |
| Kazaz *et al*[76] |  | Nephrectomy, Chemo | NA | NA |
| Souza *et al*[77] |  | Nephrectomy | 1 | DFUD |
| Detorakis *et al*[78] |  | Nephrectomy, Chemo | 11 | DOD |
| Komoto *et al*[79] |  | Nephrectomy | 9.2 | DOD |
| Boni *et al*[80] |  | Nephrectomy, Chemo  | 15 | DOD |
| Chang *et al*[81] |  | Nephrectomy, Chemo, RT | NA | NA |
| Iannaci *et al*[82] |  | Nephrectomy | NA | DOD |
| Subramanian *et al*[83] |  | Nephrectomy | NA | NA |
| Waqas *et al*[84] |  | Nephrectomy, Chemo | NA | NA |
| Gourley *et al*[85] |  | Nephrectomy | NA | DOD |
| Su[86]  |  | Nephrectomy, Chemo | NA | DOD |
| López Cubillana *et al*[87] |  | Nephrectomy, Chemo | 5 | DOD |
| Juan *et al*[88] |  | Nephrectomy, Chemo, RT | 9 | DOD |
| Prince[21]  |  | Nephrectomy, RT | NA | A and W |
| Sesar *et al*[22] |  | Nephroureterectomy | NA | NA |
| Testa *et al*[23] |  | Nephrectomy | 27 | DFUD |
| Xuan[24]  |  | Nephrectomy | NA | NA |
| Brown *et al*[1] |  | NA | 6 | DOD |
| Brown *et al*[1] |  | NA | 11 | DOD |
| Brown *et al*[1] |  | NA | 1 | DOD |
| Brown *et al*[1] |  | NA | NA | NA |
| Brown *et al*[1] |  | NA | 1 | DOD |
| Brown *et al*[1] |  | Nephrectomy | NA | NA |
| Brown *et al*[1] |  | Nephrectomy | NA | NA |
| Brown *et al*[1] |  | Nephrectomy | 2 | DFUD |
| Hiratsuka *et al*[89] |  | Nephrectomy | 29 | NED |
| Adjiman *et al*[90] |  | Nephrectomy | NA | DOD |
| Limmer *et al*[91] |  | Nephrectomy | 1 | DOD |
| Darlington *et al*[92] |  | Nephrectomy, Chemo | 12 | NED |
| Allred *et al*[93] |  | Nephrectomy, Chemo | 3 | DOD |
| Fukunaga *et al*[94] |  | Nephrectomy | 13 | DOD |
| Desai *et al*[95] |  | Nephrectomy, Chemo | 4 | DOD |
| Sabharwal *et al*[96] |  | Nephrectomy, Chemo | >1 | NA |
| Aksoy *et al*[29] |  | Nephrectomy, S | 3 | DOD |
| Heo *et al*[97] |  | Nephrectomy | NA | NA |
| Mordkin *et al*[98] |  | Nephrectomy, Chemo, S | NA | NA |
| Berretta *et al*[99] |  | Nephrectomy, Chemo | 8 | DOD |
| Lodhi *et al*[100]  |  | Chemo | NA | AWD |
| Cason *et al*[101] |  | Nephrectomy, RT | 10 | DOD |
| Askari *et al*[102] |  | Nephrectomy | 4 | DOD |
| Guan *et al*[103] |  | Nephrectomy, Chemo | 4 | DOD |
| Papadimitriou *et al*[104] |  | Nephrectomy | NA | A and W |
| Celebi *et al*[105] |  | Nephrectomy, Chemo, TKI, VEGF Inhibitor | 13 | DOD |
| Rüb *et al*[106] |  | Nephrectomy, Chemo | 12 | AWD |
| Zhang *et al*[107] |  | Nephrectomy | NA | NA |
| Tsuda *et al*[108]  |  | Nephrectomy | 21 | DOD |
| Grapsa *et al*[109] |  | NA | NA | NA |
| Li *et al*[37] |  | NA | NA | NA |
| Qayyum *et al*[110] |  | Palliative (patient's decision) | NA | NA |
| Leggio *et al*[30] |  | Nephrectomy | 8 | DOD |
| Garmendia *et al*[111]  |  | Nephrectomy | NA | NA |
| Sanyal *et al*[112] |  | Nephrectomy, RT | 24 | DOD |
| Cerilli *et al*[113] |  | Nephrectomy, RT | 6 | DOD |
| Douard *et al*[114] |  | Nephrectomy | 3 | DOD |
| Yamamoto *et al*[115] |  | Nephrectomy, RT | 19 | NED |

RT: Radiotherapy; Chemo: Chemotherapy; TKI: Tyrosine kinase inhibitor; S: Splenectomy; DOD: Died of disease; AWD: Alive with disease; A and W: Alive and well; DFUD: Died from unrelated disease; NED: No evidence of disease; NA: Not available.