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Unique CT Imaging Features of Congenital Mesoblastic Nephroma: Insights from a Case Series and Literature Review

Unnati Chouksey[1] Akshay Baheti[1] Sajid Qureshi[2] Poonam Panjawani[3] Suyash Kulkarni[1] Nitin Shetty[1] Kunal Gala[1] Mukta Ramadwar[3] Ejaj Md[1] Poorval Manoj Kshire[3] Vasundhara Patil[1]

[1] Department of Radiodiagnosis, Tata Memorial Centre, Homi Bhabha National Institute, Mumbai, Maharashtra, India
[2] Department of Pediatric Surgery, Tata Memorial Hospital, Homi Bhabha National Institute, Mumbai, Maharashtra, India
[3] Department of Pathology, Tata Memorial Hospital, Homi Bhabha National Institute, Mumbai, Maharashtra, India

Address for correspondence Vasundhara Patil, MD, DNB, Department of Radiodiagnosis, Tata Memorial Centre, Homi Bhabha National Institute, Mumbai, Maharashtra 400012, India (e-mail: vsmriti@yahoo.com).

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Abstract

Congenital mesoblastic nephroma (CMN) is a rare pediatric renal neoplasm, affecting primarily infants, accounting for 3 to 6% of pediatric renal tumors. It is subdivided into three variants: classic, cellular, and mixed, each demonstrating distinct imaging characteristics. Radiological diagnosis remains challenging due to its rarity and overlapping imaging features with other renal tumors.

In this study, computed tomography (CT) scan of three cases of CMN were retrospectively analyzed and an attempt was made to identify distinct CT imaging features that may aid in accurate diagnosis of CMN. Findings like discontinuous cortex sign, branching vessel encasement, and renal pelvis involvement may be potential diagnostic signs for CMN.

The abovementioned imaging findings were compared with other pediatric renal tumors, including Wilms' tumor, clear cell sarcoma, and rhabdoid tumor focusing on characteristics like vessel encasement, subcapsular extension, renal pelvis involvement, and presence of hemorrhage and necrosis within the mass.

Additionally, the significance of findings like multifocality, vascular invasion, and thrombosis and extrarenal features like nodal and distant metastasis has also been discussed as an essential aid in differentiating other renal tumors.

Keywords

- congenital mesoblastic nephroma
- pediatric renal neoplasm
- subcapsular extension
- renal vessel encasement
- centrally placed renal mass

Introduction

Congenital mesoblastic nephroma (CMN) is a rare pediatric renal neoplasm, predominantly affects infants and young children and constitutes about 3 to 6% of pediatric renal neoplasm.[1]

It is, however, the most common renal neoplasm identified within the first 6 months of life.[2,3] It is subdivided into three main variants—classic, cellular, and mixed types, which differ in their imaging appearance.[4–6]

Radiological diagnosis of CMN is challenging due to its rarity and different imaging characteristics of its subtypes,

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and requires a biopsy. In this study, we retrospectively analyzed the computed tomography (CT) imaging findings of three cases of CMN along with a review of the literature to understand the imaging pattern and identify specific radiological signs that may aid in accurate diagnosis of CMN.

Case Presentations

Demographic findings and histopathology reports along with major imaging features of the patients are summarized in ►**Table 1**. These patients were diagnosed with renal mass on ultrasound performed outside and were referred to our tertiary care center. On presentation, CT scan was performed according to the decision of the multidisciplinary tumor board. As per the institute protocol a noncontrast CT scan was performed followed by a venous phase taken at 70 to 80 seconds delay for imaging of renal masses. Sedation was given during imaging under the guidance of an expert anesthetist as per need basis.

Case 1

A 20-day-old male presented with an antenatally diagnosed left renal mass on ultrasound outside and was referred to our institute. A contrast-enhanced CT scan was performed at our institute and revealed a relatively well-defined $3.3 \times 3 \times 3.5$ cm sized solid mass in the interpolar region of the left kidney (►**Fig. 1**). It had relatively homogeneous enhancement (less than the cortex in the venous phase) and infiltrated the renal medulla and cortex with indistinct margins. The cortex showed focal intervening areas of involvement with extension of the tumor to the subcapsular region through it. There was no involvement or encasement of the renal vasculature or pelvicalyceal system. Radical nephrectomy was performed and histopathology was classic type CMN with scanty focal cellular areas. Renal capsule and renal sinus were not involved by the tumor.

Case 2

A 3-month-old male diagnosed with an abdominal mass on antenatal ultrasound at 8 to 9 months of intrauterine life was delivered by cesarean section due to meconium-stained liquor, and later referred to our institute

On presentation, contrast-enhanced CT scan was performed, which revealed a large $10 \times 8.5 \times 10.8$ cm sized solid cystic left renal mass crossing the midline with imaging features as described along with case 3 and in ►**Table 1** (►**Fig. 2**).

Case 3

A 10-day-old female presented with abdominal distension on day 7 of life. Contrast-enhanced CT scan at presentation revealed a large multiseptated $7.2 \times 6.4 \times 7$ cm sized solid cystic left renal mass.

Both renal lesions described in cases 2 and 3 were biopsied, and the histopathology was cellular variant of CMN. The lesions were large, multiseptated solid-cystic with heterogeneous enhancement of solid component (less than renal cortex). They were hypoattenuating on plain scan. Hyperdense intralesional hemorrhage and increased vascularity was seen in case 3. The margins with renal medulla and cortex were ill defined and focally involved the cortex with subcapsular extension and had intervening normal cortex (►**Fig. 3**).

Lesions were medially located, infiltrated the renal pelvis, and encased the left main renal artery, its segmental

Table 1 Summary of the clinical and imaging findings of the three cases

	Case 1	Case 2	Case 3
Age of presentation	20 days	3 months	10 days
Sex	Male	Male	Female
Antenatal USG	Left renal mass	Left renal mass	No reported anomaly
Presenting symptom	Asymptomatic	Cesarean section due to meconium stain liquor, with NICU stay	Abdominal distension on day 7
Laterality	Left	Left	Left
Size	$3.3 \times 3 \times 3.5$ cm	$10 \times 8.5 \times 10.8$ cm	$7.2 \times 6.4 \times 7$ cm
Epicenter in kidney	Medial	Medial	Medial
Margins with cortex and medulla	Ill-defined	Ill-defined	Ill-defined
Discontinuous cortex sign	Present	Present	Present
Subcapsular component	Present	Present	Present
Classic double-layered sign	Present	Present	Present
Renal vessel encasement	Absent (small lesion)	Present	Present
Histopathology	CMN, predominantly classic type with focal cellular areas	Cellular variant of CMN	Cellular variant of CMN

Abbreviations: CMN, congenital mesoblastic nephroma; NICU, neonatal intensive care unit; USG, ultrasound.

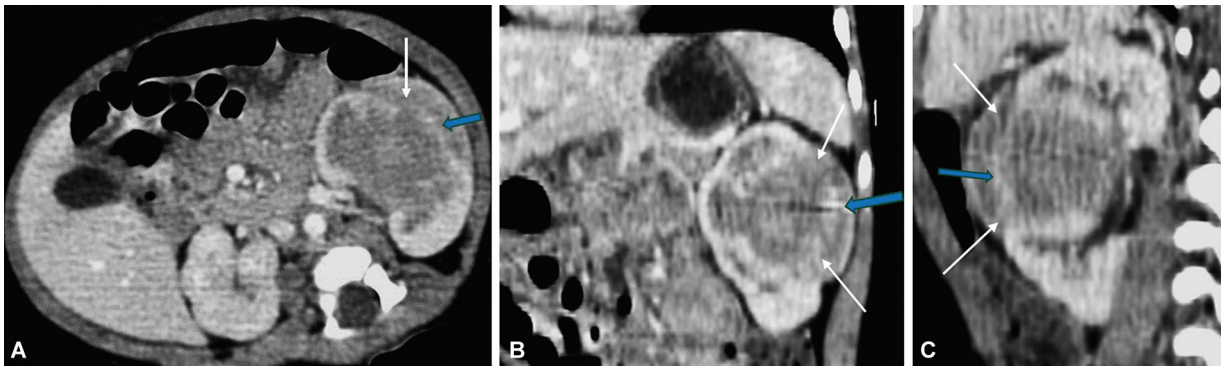

Fig. 1 (A–C) Contrast-enhanced axial, coronal, and sagittal images reveal a centrally located hypoenhancing mass (compared to cortex) with indistinct margins with medulla and cortex. It shows a focal hypodense extension to the cortex (white arrow) involving it and the intervening cortex (blue arrow)—"discontinuous cortex sign."

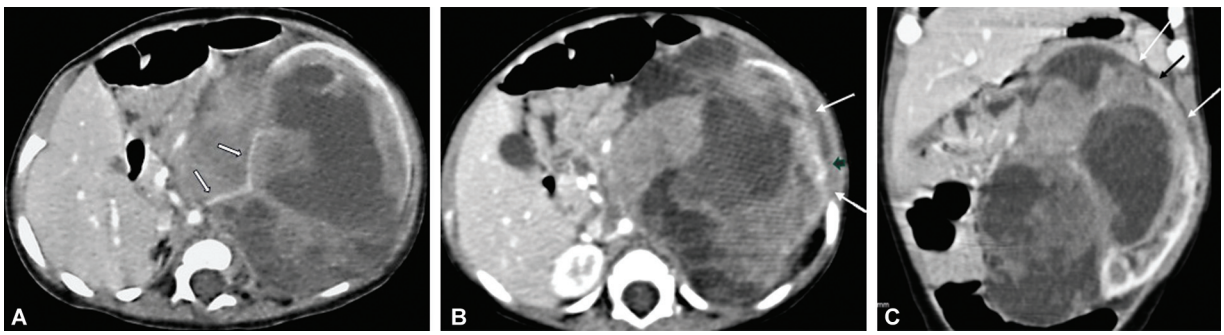

Fig. 2 (A–C) Contrast-enhanced axial (A, B) and coronal (C) images reveal a centrally located large solid cystic hypoenhancing mass (compared to cortex) with indistinct margins in the intra- and extrarenal locations. It encases the renal artery and its segmental branches (A, white arrow). Focal hypodense areas of cortical involvement (B, C, white arrow) are noted with normal intervening cortex (back arrow)—"discontinuous cortex sign."

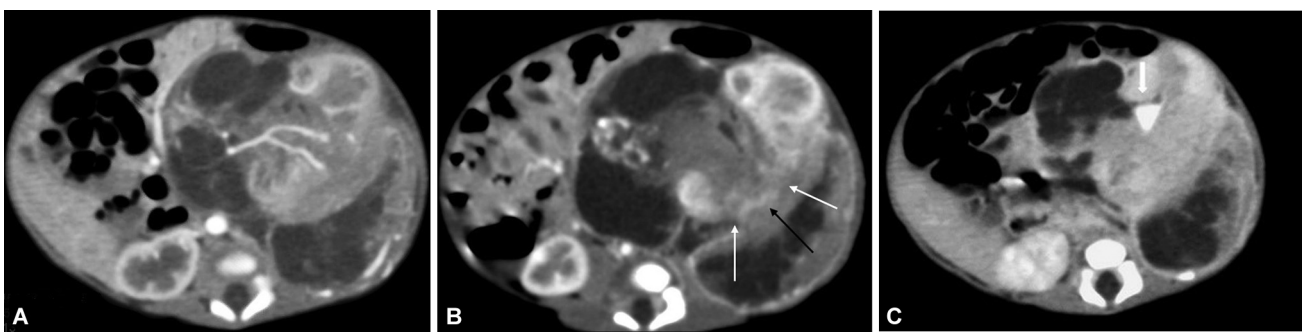

Fig. 3 (A–C) Contrast-enhanced axial images in the corticomedullary (A), nephrogenic (B), and delayed phase (C) reveal a centrally located large solid cystic hypoenhancing mass with indistinct margins with cortex and medulla and involves renal hilum with encasement of the renal artery and its segmental branches (A, white arrow). Focal hypodense projections extending laterally to involve the cortex (B, C, white arrow) and normal intervening cortex (back arrow)—"discontinuous cortex sign" seen. Encasement of the renal pelvis was seen on the delayed scan.

branches, and the accompanying veins with no tumor thrombus.

They crossed the midline and displaced the aorta and inferior vena cava (IVC) to the right without infiltration and showed perinephric fat stranding. None of these cases had calcifications, suspicious adenopathy, distant metastasis, or contralateral renal involvement.

Both these patients underwent radical nephrectomy and the histopathology revealed cellular variety of mesoblastic nephroma, with high mitotic index, with areas of hemorrhage and cystic degeneration within. The renal sinus, renal capsule, and the perinephric fat was involved (►**Fig. 4**). The renal vessels and the cut margin of the ureter were free of tumor. The hilar, interaortocaval, and the para-aortic nodes were dissected as well, and were free of tumor.

All three patients followed up at our institute for a year with 3 monthly ultrasounds, followed by 6 monthly ultrasound for the second year, and yearly ultrasounds thereafter. All three patients are currently doing well and are asymptomatic.

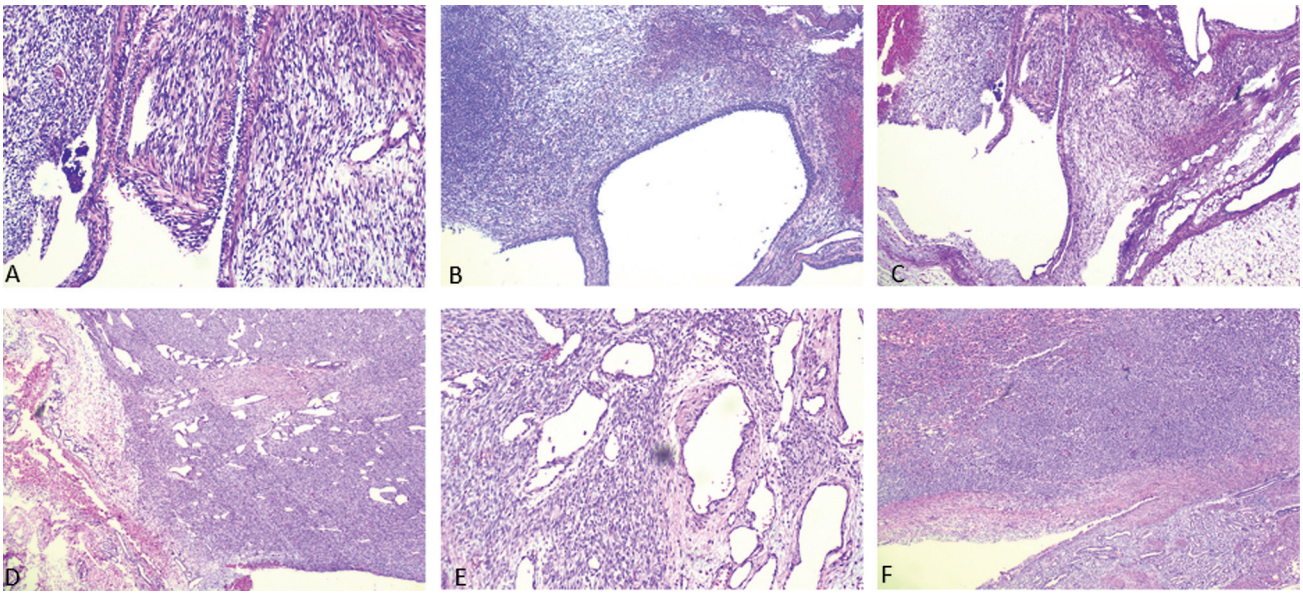

Fig. 4 Histology of case 3 shows tumor involving the cortex (**A** and **B**), tumor in the subcapsular location (**C**), tumor infiltrating the perinephric fat (**D**), and in renal sinus (**E, F**).

Discussion

CMN are rare renal neoplasms of infancy, distinctive for their occurrence within the first few months of life, the majority being diagnosed before 9 months of age.[1,2,5] It may be diagnosed during intrauterine life as early as the mid-second trimester and associations with polyhydramnios, hydrops fetalis, and prematurity are known.[4]

It shows male predilection with a male-to-female ratio of 1.5:1.[4] It usually presents as a palpable abdominal mass.[4] Multifocality or bilaterally is unknown, and it rarely metastasizes.

CMN probably arises from early nephrogenic mesenchymal tissue and are low-grade mesenchymal/myofibroblastic tumors.[3] Three histological subtypes of CMN are identified, namely, classical (30%), cellular (50–60%), and mixed (10–20%). The cellular variety tends to be more aggressive, may have local invasion and recurrence, and tends to occur in slightly older age, more than 3 months.[3,4,7,8]

Pathological findings: Both the classic and cellular varieties are unencapsulated and infiltrative with ill-defined margins and often invade the renal hilum and perinephric fat. Histologically, its edges have finger-like projections extending far into the renal tissue to entrap renal elements and encircle tubules and glomeruli.[4] The cellular variety is multiseptated with cystic/necrotic areas and hemorrhage within.[7–9]

Imaging Findings on CT Scan

Classic CMN is usually small, solid, and relatively homogeneously enhancing (less than the renal cortex). The cellular variety is larger, heterogeneous, and multiseptated, with cystic, necrotic, hemorrhagic areas, or myxoid stroma.[2] CMN is hypoattenuating on the plain scan, and solid areas reveal gradual increasing enhancement (less than the cortex) with more intense enhancement at the periphery, probably due to functioning renal elements at the periphery.[2–4] It infiltrates the perirenal fat and renal sinus. It extends far into the renal parenchyma and has ill-defined interface, hence, nephrectomy is recommended.[8,10]

All three cases showed focal hypodense projections extending laterally to involve the cortex with subcapsular extension of the tumor and normal intervening cortex giving a "discontinuous cortex sign" (►**Figs. 1A–C,** –**2B, C**, and **3B**). The renal capsule was elevated by the subcapsular component and the cortex and renal capsule formed the "classical double-layered sign" described by Li et al.[11]

Moreover, encasement of the main renal artery and its segmental branches were seen in both cellular variants—which can be described as a "branching vessel encasement sign" (►**Figs. 2A** and **3A**). This finding, however, is not pathognomonic and can be seen in central tumor like rhabdoid.

The renal pelvis was displaced and encased in two cases of cellular CMN, as described by Li et al as "intratumor pelvis signs" (►**Fig. 3C**).[11]

The differential diagnosis of CMN based on the age of presentation is Wilms tumor, clear cell sarcoma, rhabdoid tumors, and neuroblastoma.[2]

However, Wilms tumors are rare in this age and are usually well-defined masses with distinct interface with renal parenchyma and adjacent structures due to its pseudocapsule, unlike CMN. They may have calcification and fat and do not encase the renal artery until it grows large, unlike CMN.[3] Renal vein and IVC thrombus, nodal metastasis, and bilateral/multifocal involvement are known in Wilms tumor, which are not seen in CMN.

Clear cell sarcoma, rare before 6 months of age, usually occurs in 2 to 3 years. They are solid, heterogeneous usually well-circumscribed, with areas of hemorrhage and necrosis

and enhances more than the renal cortex in the corticomedullary phase.[3,10] It tends to permeate and has extracapsular extension in 70%. It infiltrates the perirenal lymphovascular system with nodal metastasis in 30% and bony metastasis at presentation, not seen in CMN.[4,10] Renal vein thrombosis is seen in 5% and may have subcapsular hemorrhage.[4,10]

Renal rhabdoid tumors are highly aggressive and approximately 60% occur below 1 year of age.[2] It tends to be large, heterogeneous, and multilobulated with hypodense areas of hemorrhage and necrosis separating the lobules.[4,10] Like CMN, it may be centrally located, involve the renal hilum, encase the renal vessels, and, invade the renal parenchyma with indistinct margins. Subcapsular components are usually fluid unlike CMN and may have rim-like enhancement.[12] It has calcifications (66%), vascular thrombus, and nodal and distant metastasis to the lungs, liver, nodes, brain, or bones not seen in CMN.

CMN are treated by nephrectomy and have a good prognosis, better with classic CMN. Follow-up with ultrasound is recommended in cellular CMN to detect early recurrence.[6] This study included only three cases of CMN, which limits the generalizability of the findings, and larger studies are required to validate the observed imaging patterns and their diagnostic significance. Moreover, a direct comparison of imaging findings with other renal tumors was not done, and incorporating magnetic resonance imaging findings may have added benefits.

Conclusion

The distinguishing imaging features of CMN are less known due to its rarity. This study describes specific imaging features observed in these three cases, and not common in other renal tumors. The spectrum of imaging findings comprising of centrally placed lesions invading the hilum, with branching vessel encasement, and renal pelvis encasement, discontinuous cortex sign, and subcapsular component, in an infiltrative renal mass in infants may help to identify CMN. Further validation of these imaging findings is needed in a larger cohort, to accurately identify CMN.

Patients' Consent

Patient's consent has been obtained.

Funding

None.

Conflict of Interest

None declared.

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