A rare pediatric renal tumor: classic congenital mesoblastic nephroma: two cases and review of literature

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Background **Mesoblastic nephroma (Boland's tumor) is the most common benign renal tumor occurring in infants and neonates. The most common presentation is abdominal mass, but it can have varied presentations because of associated paraneoplastic syndromes. Majority of these tumors can be cured with surgical excision alone; however, long-term follow-up is required for recurrence or metastasis. Histopathologically, it is divided into two subtypes: classic and cellular. The less common cellular congenital mesoblastic nephromas have cellular elements in them and tend to have a more malignant potential.**

Patients and methods **Two patients were studied over a period of 2 years from June 2010 to June 2012.**

Results **By reporting two patients presenting in the infantile period with classic congenital mesoblastic nephromas, an attempt is made in this paper to characterize the clinical behavior of this variety of renal tumors. Hypertension and paraneoplastic syndromes can be associated with this tumor. Herein we compare our experience with other similar cases reported in the literature.**

Conclusion **When renal tumors occur in infancy or at neonatal age, mesoblastic nephroma should be kept in mind. Association of hypertension and paraneoplastic syndromes should be looked for. Surgery is usually curative and postoperative follow-up for recurrence is required, more so in cellular variety.** ***Ann Pediatr Surg*** **10:112–114 © 2014 Annals of Pediatric Surgery.**

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Introduction

Congenital mesoblastic nephroma (CMN) is a rare renal tumor, with an incidence of 3.5% of all renal tumors [1]. Glick *et al.* [2] studied 101 renal tumors, of which 11 occurred at the age of 6 months or less, with CMN occurring in seven cases. In 1967, Bolande *et al.* [3] described CMN as a separate entity from congenital Wilms' tumor and emphasized on its benign nature [4]. The CMN generally presents as an asymptomatic abdominal mass, sometimes accompanied by hematuria. It can also have varied presentation because of paraneoplastic syndromes. With the advancement in technology, antenatal diagnosis is not unknown when it can lead to polyhydramnios (71% of the gestations associated with the tumor), hydrops, and premature delivery, in addition to hypertension and hypercalcemia (owing to the tumor's secretion of a substance similar to parathormone) [5,6]. Nephrectomy is the treatment of choice and is alone sufficient for good survival.

Patients and methods

Case 1

A 2-month-old male child presented with progressively increasing abdominal mass. An abdominal ultrasound (USG) scan revealed an echogenic mass in the upper pole of the right kidney, with its outer part showing concentric hyperechoic and hypoechoic layering, and the center was of slightly lower echogenicity (ring sign). The lower pole of the right kidney, left kidney, and adrenals were normal. Computed tomographic (CT) scan of the abdomen confirmed the presence of a low attenuation mass in the upper pole of the right kidney, measuring $9.3 \times 3 \times 3$ cm (Fig. 1). The right renal artery was stretched around the inferior part of the mass. CT scan of the chest was normal.

The hemogram, serum electrolytes, coagulation profile, and creatinine were normal. Ionized calcium was borderline high, initially at 1.39 mmol/l (0.9–1.3 mmol/l), but repeat samples were within normal range. A right nephrectomy with hilar and caval node excision was performed.

The pathological findings were consistent with classic CMN. The patient is now 2 years old and is asymptomatic. Follow-up USG of these two patients showed no recurrence.

Case 2

A 3-month-old male child presented with incidentally detected abdominal lump and hypertension. On examination, the child was hypertensive and routine blood investigations were normal. USG and CT scan revealed mass arising from the upper pole of the right kidney. Blood pressure was controlled and the child underwent right nephrectomy (Fig. 2). On gross, the tumor was ill-defined with no obvious capsule (Fig. 3). Microscopy showed spindle cells and no atypia and was suggestive of classic CMN (Fig. 4). Postoperative antihypertensive medication was required for 2 weeks. The child is now doing well at 2-year follow-up.

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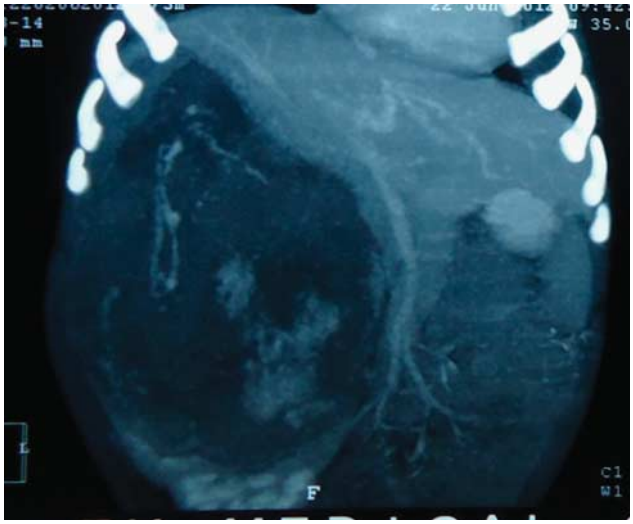
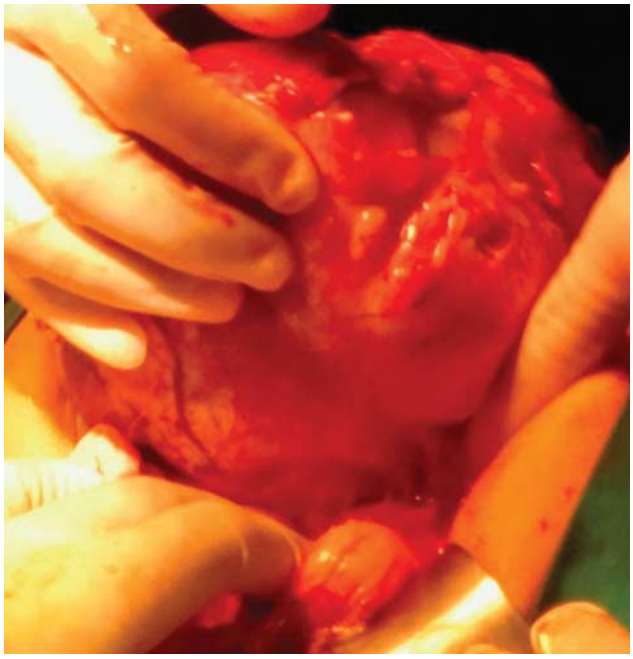
Fig. 1



Computed tomographic scan showing huge mass.

Fig. 2



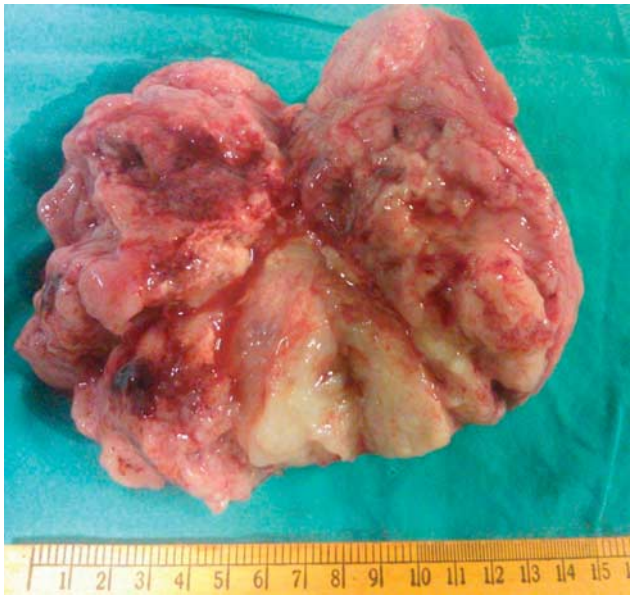
Intraoperative removal.

Follow-up protocol at our institute for renal tumors was carried out by pediatric oncologists. Close surveillance was done in the first year by 3-monthly USG, and later on by 6–12-monthly USG. CT scan was repeated only when USG showed recurrence. Regular blood pressure was monitored for tapering and stopping antihypertensive medication.

Discussion

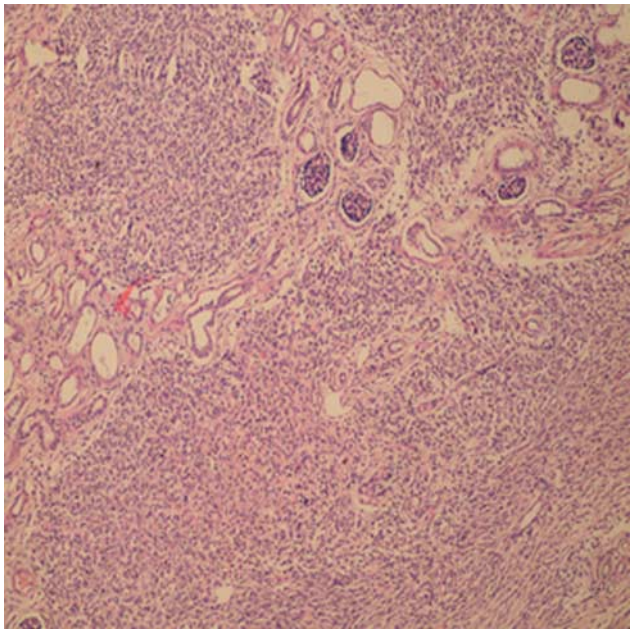
The vast majority of renal tumors encountered in infancy are benign, representing mesenchymal hamartomatous maldevelopment of the fetal kidney. In the older

Fig. 3



Gross appearance with no identified capsule.

Fig. 4



Microscopy showing classic congenital mesoblastic nephroma.

literature (before 1975), leiomyomatous hamartoma and fetal renal hamartomas were used synonymously with CMN [7].

Although CMN is a rare tumor, it is the most common renal tumor under 6 months of age and constitutes 3.4% of all renal tumors [8]. Of the cases with CMN, 67% have hypertension at presentation. Hypertension in CMN has been explained on the basis of hyperreninemia, which is seen in 80% of the cases of CMN. Overproduction of renin may be primary, that is, it is secreted in the tumor,

or secondary because of ischemia of the renal tissue from compression by the tumor, which is more likely in the leiomyomatous type [9–11].

Although the most common presentation is an asymptomatic abdominal mass, other clinical features include hematuria, proteinuria, polyuria, vomiting, hypercalcemia, jaundice, dehydration, azotemia, and electrolyte disturbances [11]. None of these were seen in our patient association of Von Willebrand disease, which has also been reported [12].

Imaging plays an important role not only in the detection of renal tumors but also in the process of differential diagnosis. Cystic renal masses are well delineated by ultrasonography; however, solid lesions may require further evaluation by CT or MRI [13]. On USG imaging, CMN appears typically as a well-defined, solid, homogeneous lesion of a fine-to-medium coarse basic texture. There may be some heterogeneity because of the hypoechoic areas of hemorrhage or necrosis, which were not seen in our case. A distinctive 'ring sign' may be seen in typical intrarenal CMN [14], as also seen in our patient. Although most cases of CMN have been diagnosed postnatally, with advances in the imaging technology, an increasing number of cases are being detected on antenatal scans [15]. Irsutti *et al*. [16] have discussed the antenatal USG and MRI features of CMN.

Childhood renal tumors were classified according to the 'Stockholm classification', in which CMN is included in low-risk group and staged as per other renal tumors into ST1-5 (SIOP staging).

Cystic change with hemorrhage and necrosis has been reported to be of significance in determining the prognosis of mesoblastic nephromas. Whereas Garbyal *et al*. [17] consider it to be of adverse significance, the cystic CMN reported by Murthi *et al*. [18] had a good outcome. Both patients in our series had cystic change within the tumor. Both of them with small cysts had difficult excision, but no intraoperative tumor spillage. Therefore, apart from contributing adversely to the tumor dissection, the presence of cysts and hemorrhage most probably does not have any prognostic value.

Recent studies have shown that the classic and cellular variants have genetic differences. Only the cellular variant shows translocation (12; 15) (p13; q25), which leads to the *ETV6-NTRK3* gene fusion. There have been varied findings in mixed subtypes [19].

Surgery is the primary modality of treatment. The role of chemotherapy for the treatment of CMNs is controversial. It is well appreciated that the definitive management for a CMN is a surgical excision, with disease-free margins. In the event of a recurrence or with an inoperable tumor, chemotherapy may be beneficial. Kulkarni *et al*. [20] have stated the role of diagnostic cytopathology in differentiating mesoblastic nephroma from other renal tumors.

Conclusion

CMN, though rare, should be considered in differential diagnosis of renal mass occurring in infancy. Hypertension should be looked for and stabilized before surgical intervention for best outcome. Diagnosis is made best by CT/MRI. Surgery is curative, but long-term follow-up is needed.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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