CONGENITAL MESOBLASTIC NEPHROMA: REPORT OF 4 CASES

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ABSTRACT
Congenital mesoblastic nephroma (CMN) is a rare renal tumour commonly seen during infancy. They are benign tumours, but occasionally exhibit aggressive behavior. We present four infants who presented with CMN. Three of them are doing well following nephrectomy and have been on follow up. The 4th patient had an aggressive course and recurred within six months of nephrectomy and died soon after.

KEYWORDS: Congenital mesoblastic nephroma; renal tumours; infants.

INTRODUCTION
Congenital mesoblastic nephroma (CMN), although rare, is the commonest neonatal renal tumour.[6,7] These tumours are often diagnosed in a prenatal scan and account for about 3-6% of all renal neoplasms in children.[3,4] Approximately 50% occur during the neonatal period and 80% of cases are reported within the first month of life. Although a benign tumour, CMN can behave aggressively with catastrophic results.[5,7] We report 4 cases of CMN occurring during infancy.

CASE REPORTS
Four infants, 2 males and 2 females aged 3 months, 4 months, 5½ months and 11 months respectively presented with abdominal lumps. 3 had right sided tumours and 1 had the tumour in the left kidney. The tumour arose from the upper pole in 2, middle part in 1 and lower pole in 1 patient. CECT abdomen confirmed the location of the tumour with heterogenous mass showing cortical enhancements (Fig.1). None of the patients had any paraneoplastic syndromes. Nephrectomy of the affected side was done in all 4 patients (Fig.2). The excised specimens weighed 45 gms, 60 gms, 50 gms and 85 gms respectively. None of the tumours showed evidence of tumour necrosis or hemorrhages. All the patients had uneventful recovery. The first 3 patients were doing well at last follow up (median 57 months). The 4th patient had cellular variety on histology and recurred within 6 months of surgery. The patient did not respond to a course of chemotherapy and died soon after.

DISCUSSION
CMN is a mesenchymal tumour. They usually present as an abdominal lump. Other rare renal tumour which occurs during infancy are Wilms’ tumour, rhabdoid tumour, clear cell sarcoma of the kidney and renal cell carcinoma.[1,2] There are two main subtypes of CMN. The classic type, which is the more common, is of a benign nature and has a better prognosis. The second, the cellular (atypical) variant, is less common and runs an unpredictable course.[8] A mixed type has also been reported.[9] Three of our patients presented before 6 months of age and were of benign type. The fourth patient presented at 11 months of age with cellular type of histology. This patient had loco-regional recurrence of the tumour 6 months following nephrectomy and demonstrated aggressive behavior inspite of chemotherapy and died soon after.

Fig.1: CT scan showing themass arising from the lower pole of the left kidney
Macroscopically CMN is a solid un-encapsulated mass which often occurs near the renal hilum. It tends to invade the surrounding structures and renal parenchyma. Haemorrhage and necrosis are infrequent. The classic cytological description of the lesion is that of cellular clusters of spindle cells, mild nuclear pleomorphism, mitotic activity and no blastema. Histologically, it is typically composed of connective tissue growing between nephrons, usually replacing most of the renal parenchyma (Fig. 3). Some CMNs are associated with paraneoplastic syndromes such as hypertension (due to hyperreninemia) and hypercalcemia (due to prostaglandin secretion from the tumor cells). \[8,10\]

None of our patients had such associations. Nephrectomy usually suffice for the common type of CMN, but CMN of cellular variety often exhibit aggressive behaviour with invasion of surrounding structures and metastasis. Although one of our patient had cellular histology, there was no metastasis at the time of presentation and there was only loco-regional recurrence.

REFERENCE