# Neonate with congenital mesoblastic nephroma surviving respiratory failure and pulmonary embolism at laparotomy

R. Ranawaka[1], A. Vithanage[1], P. Rathnayake[2], K. Hettiarachchi[1], N. Jayakody[1]
1. Sirimavo Bandaranayake Specialised Children's Hospital, Peradeniya, Sri Lanka
2.Teaching hospital Kandy, Sri Lanka

**Keywords:** Congenital mesoblastic nephroma, pulmonary embolism, disseminated intravascular coagulation

## Introduction

Congenital mesoblastic nephroma (CMN) first described by Bolande et al in 1967 [1] is the commonest neonatal tumour of renal origin comprising of 3-10% of all childhood tumours [2]. Common modes of presentation include abdominal mass, antenatal diagnosis and features of para-neoplastic syndrome [3]. There are two main histological varieties; classic type where nephroureterectomy is curative and atypical (cellular) type with unpredictable natural history [2,4].

We report a two day old developing severe cardio-respiratory failure secondary to CMN, surviving emergency tumour debulking laparotomy that was complicated by pulmonary embolism and disseminated intravascular coagulation (DIC). Subsequent nephroureterectomy with total tumour clearance was performed. To our knowledge there are no reports of CMN with similar presentation in the world literature.

## Case

An eighteen hour old baby with antenatal diagnosis of "intraabdominal tumour" was transferred on developing laboured breathing. Within hours she required ventilatory support and cardiac inotropes.

Ultra sound scan detected a large intra-abdominal mass displacing other viscera. The left kidney was not identified. Computerised tomography (CT) scan was not accessible.

During emergency laparotomy a bosselated encapsulated tumour occupying almost the whole abdomen with the consistency of an "oily sponge cake" seen originating from left kidney. The liver, bladder, uterus and Fallopian tubes were normal.

Suddenly, generalised bleeding and bradycardia developed suggesting pulmonary air embolism leading to DIC. Blood components were transfused and, the operative field was filled with isotonic saline to facilitate evacuation of pulmonary embolus. Surgery was terminated and tumour remnant over-sutured. Resected tumour mass weighed 350grams. Approximately the same volume of tumour was left in-situ.

Tumour markers were done as advised by the oncologist (serum βHCG: <0.1miu/ml(<1miu/ml), α fetoprotein: 776.1iu/ml(<25iu/ml). Histology revealed degenerated tumour tissue and undifferentiated spindle cells with no blastemal or epithelial elements. Immunohistochemistry was focally positive for vimentin and, negative for SMA,PCK,CD10 and MyoD1. The final diagnosis of CMN was made(4).

Subsequent CT scan of abdomen showed tumour remnant compressing the aorta and inferior vena cava (Figure 1).

At re-laparotomy, well encapsulated tumour arising from the lower pole of left kidney with firm and fibrotic consistency found. Rest of the viscera were normal. Left nephroureterectomy with total tumour resection was performed. Tumour weighed 400grams (Figure2).

Histology confirmed classic type CMN confined within excision margins. She recovered completely and remains well three months after surgery. Surveillance USS is planned in three month's time.

Correspondence: R.Ranawaka
E-mail: rravibindu@yahoo.com

Figure 1. CT scan of abdomen after initial tumour debulking laparotomy

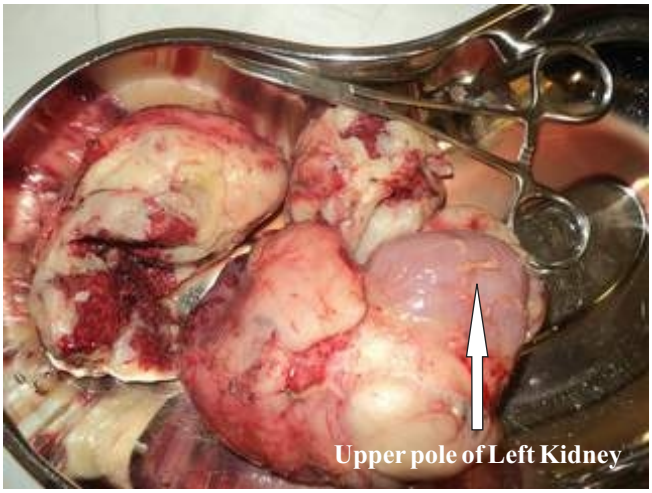


Figure 2. Specimen after total resection

## Discussion

Pulmonary embolism could occur in surgery during exposure of a large vascular surface. The manifestations range from trivial reduction in platelet count and prolongation of clotting time to fulminant DIC [5]. The management includes correction of DIC with transfusion of blood components and, evacuation of pulmonary air embolus by filling the surgical field with isotonic saline while continuing ventilation.

Initial surgery leaving a residual tumour bulk could theoretically be considered as tumour upstaging. Adjuvant therapy has a place in metastatic CMN but is not described for classic type. Since complete tumour clearance was confirmed histologically, surveillance for tumour recurrence with regular imaging was planned.

The rare experience of successful tumour excision after complicated surgery in a neonate with no proper pre-operative imaging and histology would be informative to surgeons who experience similar surgical emergencies.

**References**

1. Bolande PP, Brough AJ, Izant RJ, Congenital mesoblastic nephroma of infancy. Pediatrics 1967;40: 272–279

2. dos Santos LG, de Sousa J, de Carvalho R et al, Cellular congenital mesoblastic nephroma: case report. Jornal Brasileiro de Nefrologoa 2011;33:88-90

3. Minesh K, Horacio O, Michael AS, Congenital Mesoblastic Nephroma Presenting with Neonatal Hypertension. Journal of Perinatology 2005; 25:433–435

4. Argani P, Fritsch M, Kadkol S et al, Detection of the ETV6-NTRK3 chimeric RNA of infantile fibrosarcoma/cellular congenital mesoblastic nephroma in paraffin- embedded tissue: application to challenging pediatric renal stromal tumors. Modern Pathology 2000; 13:29-36

5. Levi M, Ten Cate H, Disseminated intravascular coagulation. New England Journal of Medicine 1999; 341:586–92.

**Key points:**

- Pulmonary embolism could occur in surgery during exposure of large vascular surfaces.
- Management includes filling the surgical field with isotonic saline while continuing ventilation and correction of DIC.