CASE REPORT

Perinatal neuroblastoma: a hidden bullet in the chest

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SUMMARY

A neonate with antenatally diagnosed intrathoracic mass by ultrasound scan was delivered uneventfully at 35 weeks gestation by caesarean section due to pre-eclampsia and fluctuating hypertension in the mother. The intrathoracic mass was echogenic and the diagnosis was inconclusive. At 12 h of life the baby deteriorated acutely, in terms of increased oxygen requirement, ventilatory care, heart rate fluctuation, hypotension requiring inotropic support and died despite intensive care support. The parents were counselled that an autopsy would be invaluable in providing a diagnosis given the antenatal finding of an intrathoracic mass. The final diagnosis of neuroblastoma was performed at postmortem.

BACKGROUND

Intrathoracic variety of neuroblastoma is a very rare tumour and usually presents with respiratory symptoms. In developing countries like India where antenatal diagnosis is not always possible, one should have a suspicion of neuroblastoma as a differential in the context of intrathoracic mass, mediastinal widening on chest X-ray, unexplained sudden cardiorespiratory deterioration and refractory shock. The importance of autopsy is highlighted for confirmation of the diagnosis.

CASE PRESENTATION

A newborn baby, late preterm (35 weeks gestation) was delivered by emergency lower segment caesarean section for maternal indication fluctuating blood pressure. Antenatally the mother was managed in another hospital and was referred to our hospital for safe confinement. The mother was admitted in view of fluctuating blood pressure 2 days before the delivery. A third trimester fetal ultrasound scan carried out by the in-house fetal medicine team revealed the possibility of an echogenic mass in the thorax, likely to have sequestered lung, cystic adenomatoid malformation or neuroblastoma. Parents were counselled about these possibilities and the effect of these on the infant at birth and after. At birth, the baby did not require resuscitation or respiratory support. Baby's weight was 1.9 kg, length 45 cm and head circumference 33 cm. The baby was appropriately grown for gestational age and the head circumference was above 10th centile. Haemodynamically baby was stable, eumorphic and euglycaemic. Baby was started on feeds at the outset but later received intravenous fluids on deterioration. At 10 h of age, the baby started deteriorating in the form of desaturations,

Figure 1 X-ray showing widening of the mediastinum.

variable heart rate and hypotension. The baby was put on ventilator and started on inotropes. Chest X-ray carried out before intubation showed mediastinal widening ([figure 1](#)). Echo and cranial ultrasonography were essentially normal. Despite active resuscitation the baby succumbed to death. Parents were counselled and the option of autopsy was discussed with a view to pinpoint the diagnosis. After appropriate parental consent autopsy was conducted. A mass was detected in the posterior mediastinum and immunohistology and histopathology confirmed it to be neuroblastoma. The postmortem cultures did not reveal any bacterial growth. Histopathology and immunohistochemistry confirmed the diagnosis as thoracic neuroblastoma. The mother remained well in the postpartum period. The X-ray of the chest ([figure 1](#)) showing widening of the mediastinum, autopsy showing thoracic neuroblastoma ([figures 2 and 3](#)), H&E staining and immunohistology of the specimen ([figures 4–7](#)) favouring the diagnosis of neuroblastoma are depicted.

INVESTIGATIONS

- ▶ Complete blood cell count was normal.
- ▶ Septic screen was also carried out which was negative.
- ▶ Chest X-ray showed mediastinal widening.
- ▶ An autopsy and biopsy of the mediastinal mass was carried out and sent for histopathological examination.

DIFFERENTIAL DIAGNOSIS

1. Congenital cystic adenomatous malformations
2. Lung sequestration
3. Neuroblastoma

CrossMark

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Figure 2 Autopsy: a grey-brown irregular mass is seen in the mid-thorax in the midline attached to the vertebral column measuring about 3×3 cm, firm in consistency.

TREATMENT

Intensive care support: intravenous fluids, inotropes, intravenous antibiotics and ventilatory care.

OUTCOME AND FOLLOW-UP

Expired on day 1 despite intensive care support.

DISCUSSION

Neuroblastoma is the most common solid tumour in infancy, but the acute, catastrophic, neonatal presentation of cardiorespiratory

Figure 4 Histopathology: H&E stain. Areas of small, uniform round cells seen in sheets as well as rosettes pattern. Areas of necrosis and calcification are seen over wide areas. The rosettes are seen to have eosinophils.

failure and death of the intrathoracic variety is very rare and so far has not been reported. They usually present with respiratory symptoms and require respiratory support.^{1 2} There is one case report which states thoracic neuroblastoma presented with severe pulmonary hypertension.³ The cause of pulmonary hypertension may be due to variable presence of pulmonary hypoplasia. In our case, the baby was stable for almost 10 h, then developed signs of respiratory and cardiovascular symptoms. The course of

Figure 3 Autopsy: a grey-brown irregular mass is seen in the mid-thorax in the midline attached to the vertebral column measuring about 3×3 cm, firm in consistency.

Figure 5 Histopathology: H&E stain. Areas of small, uniform round cells seen in sheets as well as rosettes pattern. Areas of necrosis and calcification are seen over wide areas. The rosettes are seen to have eosinophils.

Figure 6 Immunohistochemistry (IHC) report: chromogranin A-IHC single marker tissue; positive in 50% of area of well-preserved tumour cells. IHC single marker for neuron-specific enolase tissue; positive in 50% of area of well-preserved tumour cells. IHC single marker for S100 protein tissue; negative.

Figure 7 Immunohistochemistry (IHC) report: chromogranin A-IHC single marker tissue; positive in 50% of area of well-preserved tumour cells. IHC single marker for neuron-specific enolase tissue; positive in 50% of area of well-preserved tumour cells. IHC single marker for S100 protein tissue; negative.

deterioration was very rapid. There was no persistent pulmonary hypertension of the newborn in our index baby. In a baby with known neuroblastoma, rapid deterioration at 10 h may be due to a catastrophic hormonal surge (epinephrine), leading to arrhythmia and sudden death.⁴ It is possible to measure metabolites of epinephrine (vanillyl mandelic acid, VMA) in the urine; however, urinary VMA analysis could not be performed as the baby was

anuric. Autopsy and histopathology gave us the ultimate diagnosis of thoracic neuroblastoma. This case highlights the importance of autopsy in determining histopathological diagnoses. The approach for the management of intrathoracic neuroblastoma should be aggressive and include a multidisciplinary team of neurosurgeons, paediatric surgeons, radiologists and neonatologists. Parents should be counselled that death is a possible outcome.⁵

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Learning points

- ▶ Intrathoracic neuroblastoma, although rare, can present with a catastrophic event even on day 1 of life.
- ▶ Aggressive management should be carried out though the mortality is very high.
- ▶ Multidisciplinary team should be involved for the management.
- ▶ Urinary vinyl medelic acid can be carried out for screening but biopsy and histopathological examination are appropriate to pinpoint the diagnosis.

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