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Challenge in diagnosing fetal neuroblastoma

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Introduction

Neuroblastoma is a subtype of neuroblastic tumour along with ganglioneuroblastoma and ganglioneuroma, derived from primordial neural crest cells which subsequently develop into sympathetic ganglia and the adrenal medulla. The occurrence is age dependent with median onset of 2 years and a peak of 18 months. However there is also fetal neuroblastoma which diagnosed during the third trimester of pregnancy. Here we report a case of a fetal neuroblastoma in asymptomatic mother and foetus, initially thought as sacrococcygeal teratoma.

Case Description

A full term baby boy, accidentally was screened to have abdominal mass via ultrasound during 3rd trimester, 2 days prior to his delivery. Born spontaneous vaginally, on palpation there is pelvic mass extending up from pelvis to just below the umbilicus, measuring 5cm x 4cm, fixed, firm to hard in consistency. Abdominal ultrasound revealed huge pelvic mass with gross left hydronephrosis. CT scan performed on day 4 of life, revealed huge pelvic-abdominal solid mass with intralesional calcification, bilateral hydronephrosis due to external compression to the distal ureters by the mass. No major vessels encasement. Laparotomy performed on day 9 of life, there are solid tumour at the pelvic region severely adhered to urinary bladder neck, prostate, lower rectum and to the coccyx with measuring 7cm x 8cm. Highly vascular and bleeding upon dissection, lead to massive blood loss requiring massive blood transfusion. Coccyx was removed through another midline incision at the back. Clinically tumour behave as sacrocoxygeal teratoma however histophatologically revealed features of neuroblastoma. Chemotherapy initiated. CT Scan post cycles of chemotherapy showed regression of the tumor and subsequently repeated bone marrow and trephine biopsy after completed chemotherapy showing no more residual tumor invasion.

Fig 1. Preoperative surface marking before incision

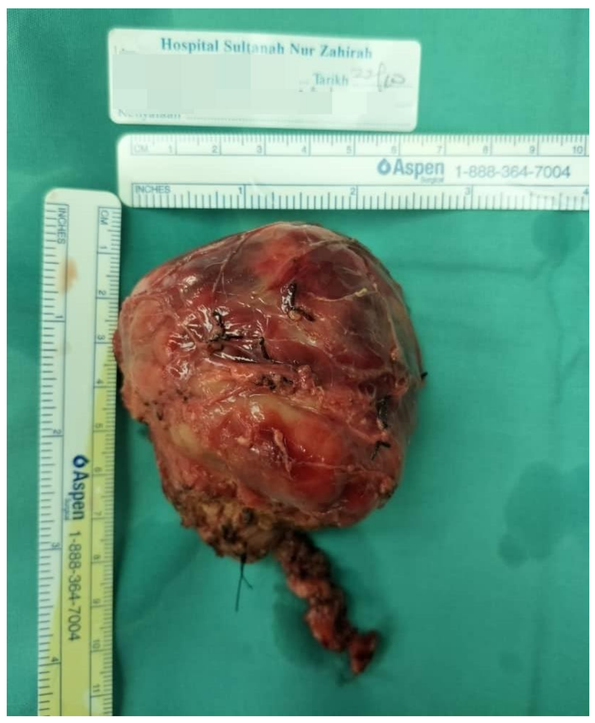

Fig 2. Macroscopic appearance of the tumor after surgical resection

Discussion

Neuroblastoma can occur anywhere along the sympathetic chain. The frequency of adrenal tumours is higher in children (40%) compared to infants (25%) [4]. Around half of newborns have fetal adrenal rest which usually involute in the first few weeks of life and which may be adrenal or extra-adrenal, reflecting migration pathways in embryonic development [1]. Those that persist into postnatal life may present as congenital neuroblastoma. Obstetric ultrasound scanning in the 3rd trimester may detect a 'fetal neuroblastoma', usually seen in the adrenal glands varying between 2 and 10cm in size. They may be solid or cystic, sometimes with foci of calcification [2]. Fetal neuroblastoma is typically an incidental finding in the third trimester but can be visualized as early as 23rd weeks with careful sonography [5]. It is common for the solid masses to increase in size on serial antenatal ultrasound exams, whereas the cystic and complex masses may grow or shrink during fetal life [5]. In neonates where the possibility of neuroblastoma has been raised antenatally but where there is no family history, urinary catecholamines (VMA/HVA) should be checked around 2-3 days of age with the referrence ranges as describe by Pussard et al [3]. When elevated, urinary catecholamines are helpful in confirming the diagnosis, however, they will be normal in two-thirds of patients with prenatally diagnosed tumor, with a negative predictive power around 70% [2]. Furthermore, urinary catecholamines are increased in only approximately 33% of prenatally diagnosed neuroblastomas, compared with 85-90% of postnatally diagnosed patients [5]. Infants are more likely to have thoracic and cervical primary tumours. In order of decreasing frequency, primary tumour occur in adrenal, paravertebral retroperitoneum, posterior mediastinum, pelvis, neck, with an unknown primary in 1% of case [4]. Adrenal haemorrhage, the most common cause of an adrenal mass in newborns, usually occurs at birth or during the first postnatal days, and is unsual in the fetus [5]. In most cases of prenatally diagnosed neuroblastomas, the mother is asymptomatic. However, there are occasional reports of maternal and/or fetal symptoms and signs that appear to be either direct or indirect consequences of the presences of the presence of a neuroblastic tumor [2]. Metastatic involvement of the placenta is associated with fetal hydrops and maternal preeclampsia, possibly caused by vascular compromise leading to placental insufficiency [6].

Conclusion

Where antenatal findings indicate an anomaly which could indicate the presence of neuroblastoma or any other intraabdominal mass, delivery should occur in a tertiary center where access to multidisciplinary approach such as paediatric oncology, paediatric surgery and paediatric anaesthesiology is readily available. Moreover, with presence of asymptomatic mother and foetus, the diagnosing fetal or prenatal neuroblastoma remain a challenge . We wish to report a case of fetal neuroblastoma in hoping to provide better insight in future research.

References

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