RARE TERATOMA IN A DOWN SYNDROME NEONATE
A CASE REPORT AND LITERATURE REVIEW

Nadesin Muniswaran[1]; Yuganeswary[1]; Pigineswaran[1];Tan Shung Ken[1] ;Tammy Teoh Han Qi[1]
Pon Kah Min[1] ;Mohan Arunasalam[2];Nik Fatimah Malek[3];Lim Choo Hau[4]
Hospital Pulau Pinang[1];Pantai Hospital Ipoh[2];Hospital Sultanah Bahiyah[3];Hospital Seberang Jaya[4]

Abstract
Trisomy-21 is known to be associated with neoplasm, commonly involving haematological malignancy with a lower incidence of solid organ tumour. Retroperitoneal teratomas were reported as extremely rare and most of it diagnosed antenatally are known to be benign. A down syndrome baby was antenatally diagnosed to have polycystic left kidney, however at birth there was significant abdominal distention with a large left abdominal mass.Ultrasound and subsequent CECT abdomen pelvis done confirmed a large left retroperitoneal mass crossing midline which could represent a retroperitoneal teratoma. Surgery was performed and a large left retroperitoneal mass was removed. HPE reported as 98% immature teratoma and 2% yolk sac tumour. Most common extragonadal teratomas in has been reported as sacrococcygeal in origin, however more recent literature shows increasing incidence of retroperitoneal teratoma instead especially in Down syndrome. Treatment remains primary surgical resection.However immature teratoma with persistent elevated Alpha Fetoprotein(AFP) level post surgery or malignant teratomas requires chemotherapy.

Keywords:Retroperitoneal teratoma,Down Syndrome,Neonate.

INTRODUCTION

Neoplasm in Down Syndrome carries a unique profile, with higher incidence of leukaemia and lower incidence of solid organ tumours. Decreased incidence of solid tumours among them is reported to be associated with chromosome 21 genes.[1]Neonatal solid organ tumours are very rare and germ cell tumour is the commonest.[1]Retroperitoneal teratomas constitute 3.5-4% of all germ cell tumours in children and 1-11% of primary retroperitoneal tumours.[3]Retroperiteneal teratomas were reported as extremely rare and most of it diagnosed antenatally are benign. In our centre we encountered a rare case of malignant retroperitoneal teratoma in an infant with Down Syndrome.

CASE SUMMARY

A 2 months old baby girl was born premature at 36 weeks and 2 days with a birth weight of 2.38kg via SVD in Hospital Seberang Jaya.Mother was 45 years old with gestational diabetis mellitus.Detailed scan at 33 weeks showed skeletal dysplasia and left polycystic kidney. Amniocentesis and karyotyping done showed Trisomy 21.

A huge abdominal mass was palpable over the left side crossing midline upon birth.She was otherwise stable on room air and able to tolerate feeding with expressed breast milk.Her vital signs were stable. Alpha-Feto-Protein(AFP) taken at birth was 33658 IU/mL. Ultrasound abdomen showed a large heterogenous mixed solid cystic mass with foci of calcification occupying the left abdominal region crossing the midline raising a suspicion of retroperitoneal tumour.CECT abdomen pelvis confirmed a large left retroperitoneal tumour representing teratoma radiologically. She was planned for surgery and her nutritional status was optimised to achieve desired weight gain via orogastric tube feeding. Another great challenge was severe congenital hypothyroidism which was also optimised prior to surgery requiring an unusual dosage up to 100mcg of L-Thyroxine daily managed by endocrine team and despite ne

Figure 1:Pre operative

Figure 3:Coronal View CECT

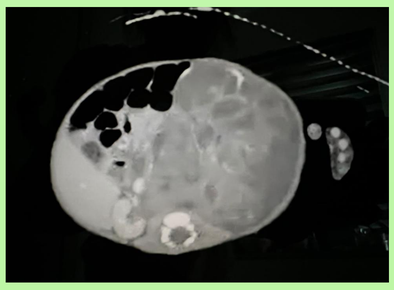

Figure 2:Axial View CECT

Figure 2:Post Operative

Apart from that she also suffered from a transient abnormal myelopoiesis which caused her to have persistent thrombocytopenia with blast cells.

Child developed hypertension subsequently and was controlled with propranolol and nifedipine.While waiting for surgery she unfortunately developed respiratory distress due to rapidly increasing abdominal distention.Repeat ultrasound showed tumour occupying most of the abdominal cavity with ascites. A repeat AFP went up to 54178 IU/mL. She was electively intubated and transferred to PICU HPP for stabilisation and emergency surgery.Immunoglobulin was administered by the paediatric hemato-oncology team to improve her thrombocytopenia.Ultrasound guided Pig tail drainage of the ascites was performed to facilitate her ventilation.

After adequate resuscitation midline laparotomy and excision of tumour was performed at D51 of life. Intra operatively a large left retroperitoneal tumour was excised with minimal rupture and spillage of the contents.Her left kidney was significantly displaced and compressed by the tumour but we manage to salvage it.Other retroperitoneal organs and structures adhered to the tumour was safely preserved and tumour was completely excised.The tumour measured 13cm x 14cm and weigh 400Gm.Histopathology report showed mixed grem cell tumour with immature teratoma (98%) and yolk sac tumour (2%).

Post operatively she had DIVC responded to blood products.She recovered over time as she needed prolonged oxygen support. Child is currently doing well 4 months post surgery with no evidence of recurrence from ultrasound. She did not receive chemotherapy till da s beer d d whic

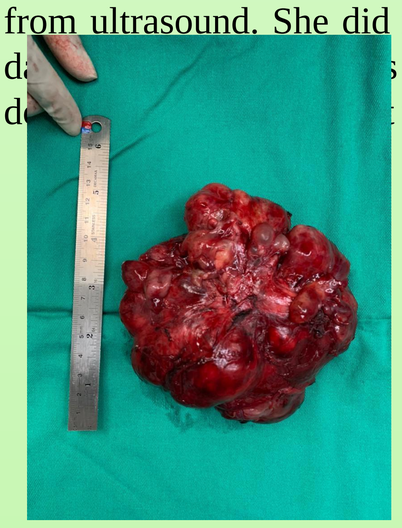

Figure 4:Tumour Image 1

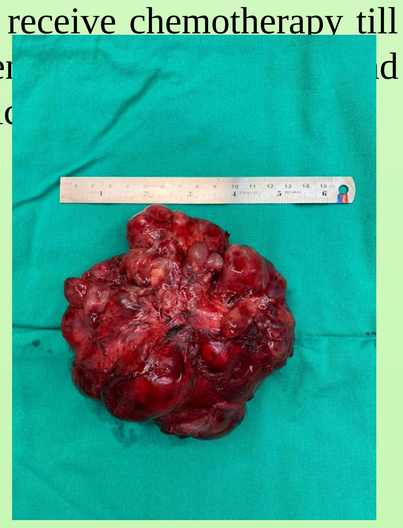

Figure 4:Tumour Image 2

DISCUSSION

From literature reviews teratoma has been reported to be most common histologic subtype of childhood germ cell tumours(GCT) arising from gonadal and extragonadal locations.[1,7] The overall percentages of patients with Sacrococcygeal teratoma(SCT) and retroperitoneal(RP) teratomas were 41.7% and 4.4%, respectively, in a German study and 88% and 12%, respectively, in an Italian study.[1]However more advanced search identified that out of 11 infants with Down syndrome,8 was with RP teratoma, 3 with cranial teratomas and none with SCT.[1]Increased incidence of RP teratomas in infants with DS may be due to the pathologic association between RP teratomas and fetus-in-fetu.[1]

Lack of SCT in DS may be due to the activation of genes specifically in patients with DS.[1] Serum AFP is often used as a tumour marker for malignant recurrences of sacrococcygeal teratoma.It can also be helpful during follow up.[4]Apart from that AFP level is also useful in distinguishing between benign and malignant GCT.[5]Complete surgical excision remains as the mainstay treatment in RP teratomas. Experienced paediatric surgeons have reported the operations for retroperitoneal teratomas to be difficult and hazardous as it is associated with severe life threatening complications especially bleeding.[5]

Overall long term prognosis is excellent as most of them with benign retroperitoneal teratomas had no recurrence at 12 years of follow up.Patients with malignant tumours were disease free at 6 months and 5 years post adjuvant chemotheraphy.[5]Multi centre report describes excellent outcome with up to 90%,6 years survival with cisplatin based chemotheraphy along with surgical resection of the RP teratomas.[6]

Our patient did not receive chemotherapy due to persistent thrombocytopenia and blast cells in the peripheral blood film which resolved over time post operatively. However upon waiting repeated AFP levels were reducing in trend and remained within normal range thus decision was made not for chemotheraphy and to continue surveillance with AFP level and ultrasound abdomen to look for recurrence.We postulate that the resolution of hypertension and hypothyroidism in our patient post surgery is associated with the removal of the tumour though exact mechanisms are unknown.

CONCLUSION

In conclusion, retroperitoneal teratoma in a neonate is a challenging tumour to deal with. It has also shown to be more common in Down syndrome children.However with early detection and complete surgical excision it helps in overall prognosis and improve survival despite having malignant component.

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Contact
Email:muniswarannadesin@gmail.com
Phone number:+0163116943
Surgical Department
Hospital Pulau Pinang