*Case report*

A Rare Case of Mature Cystic Neonatal Sacrococcygeal Teratoma

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ABSTRACT

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| **Introduction:** Sacrococcygeal teratoma (SCT) is a rare germ cell tumour but the commonest neoplasm during neonatal periods.  **Aim:** We report a rare case of neonatal pure mature cystic sacrococcygeal teratoma (SCT) Type III, who underwent excision of tumour and coccygectomy, which was the first to be done in Malacca Hospital (a tertiary state hospital in Malacca, Malaysia), since the starting of paediatric surgery service in year 2019.  **Discussion:** Postoperatively, the child develops voiding dysfunction which was one of the common functional sequelae of SCT which was managed by clean intermittent self-catheterization and subsequently resolved spontaneously. Histopathological examination revealed that the cyst wall contained all three germ cell lineages, consistent with a pure mature cystic teratoma.  **Conclusion:** Pure mature cystic SCT is rare, but benign in nature if diagnosed before age of 6 months of life and has a good surgical outcome with manageable post-treatment sequelae such as neurogenic bladder and post operative distinct scar. Surveillance up to at least 5 years is crucial as it possess the risk of recurrence with potential malignant features. |

*Keywords: Sacrococcygeal, teratoma, neonate, excision, coccygectomy, sequelae*

1. INTRODUCTION

A sacrococcygeal teratoma (SCT) is a neoplasm arising from the caudal end of the spine, usually protruding from the inferior end of the infant’s spinal column, displacing the anus forward (Pringle 2017). It is a rare germ cell tumour with an incidence of 1:40,000 live births (Hambraeus 2016), but made up of 70% of all childhood teratomas with girls to boy ratio of 4:1 (Swamy 2008). They arise from the totipotent cells of Hensen’s node and contain tissue derived from more than one germ layer either from endoderm, mesoderm and ectoderm, which contain neural elements, squamous and intestinal epithelium, skin appendages, teeth, and sometimes calcium (Srivastava 2010). Hensen’s node is also known as primitive knot, is an enlarged group of cells located in the anterior portion of the primitive streak where gastrulation, formation of the three germ cell layers begins, and plays a major role in the anterior-posterior axis specification, development of chordamesoderm and later notochord development (Doty 2011). While totipotent cells are embryonic stem cells that are present during the first few cell divisions post fertilization and can form any of the different types of cell in the body. These cells normally degenerate and disappear; however, if they escape the control of embryonic inductors and organizers, a teratoma results (Kainer 2018).

2. PRESENTATION OF CASE

A newborn baby girl with antenatal scan showing bladder exstrophy was referred to paediatric surgery team. She was born term at 37 weeks via emergency caesarean section. Post-natal examination showed no signs of dysmorphism, however noted a large gluteal cystic mass extending anteriorly, causing mass effect to the anal opening to be located anteriorly with transillumination test positive of the mass. Subsequently, an ultrasound of the mass was done which showed a sacrococcygeal cystic mass with intrapelvic extension suggestive of a SCT. However, there was a diagnostic dilemma on whether the cystic mass is truly a SCT (which is quite rare for pure mature cystic SCT) or an anterior sacral myelomeningocele.

Thus, an MRI pelvis and whole spine was done on Day 3 of life, revealing a huge dumbbell shaped sacrococcygeal multiseptated cystic mass measuring 13.8cm in total cranio-caudal length, with intra-abdominal component measuring 4.0cm (AP) x 5.2cm (CC) and extra-abdominal components measuring 5.4cm (AP) x 8.8cm (CC), with no enhancing or solid component identified. The mass causes bilateral hydronephrosis and hydroureter with mild mass effect to urinary bladder and mild herniation into the lower sacral foramina, appearing as a SCT type III (*Figure 1*). Blood investigations otherwise showed greatly raised serum Alpha Fetoprotein (AFP) of 13434.2 IU/ml (0-6.6), raised serum lactate dehydrogenase (LDH) of 305 (120-246) and normal B-hCG <2U/L (0-10).

A screenshot of a computer

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*Figure 1: MRI Pelvis and Whole Spine on Day 3 of life*

On day 7 of life, we proceeded with excision of SCT Type III and coccygectomy in prone position via perineal approach (*Figure 2*). Post excision of tumour, excessive skin was reconstructed and closed in a ‘Mercedes Benz sign’ fashion with drain insertion. Drain was removed 4 days after the surgery.

A collage of a baby

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*Figure 2: Intraoperative images during excision of SCT Type III (image 1-5); Wound inspection 4 days after surgery (image 6).*

Post operatively, it was found that the child developed urinary retention with stress incontinence after removal of urinary catheter. In view of difficulty for the parents to perform clean intermittent self-catheterisation (CISC) for the child, decision was made for temporary urinary catheter, and she was later discharged home 16 days after surgery with an indwelling urinary catheter which was changed every 2 weeks regularly. During her clinic review 3 months after the surgery, she was thriving well with no other post operative complication other than voiding dysfunction due to neurogenic bladder. Parents were able to get a grasp on CISC and indwelling urinary catheter was removed. After few months on CISC, she was able to void as usual.

From the histopathology examination, the section of the cyst wall showed component of three germ cells lineages including the brain, choroid plexus, squamous epithelium, anucleate keratin flakes, mature adipocytes, blood vessels, skeletal muscle bundles and glands lined by mucinous type epithelium. Scattered psammoma bodies was observed with inapparent mitosis seen, no immature component or evidence of malignancy present. These features were consistent with a pure mature cystic teratoma, and no malignant tissue was seen from coccygectomy specimen as well. Repeated serum AFP also has dropped to 95.2 (0-6.6) from previous 13, 000.



*Figure 3: Surgical scar during recent clinic visits at age of 1 year 10 months old*

3. discussion

SCT are rarely malignant (2%) if diagnosed before the age of 6 months of life, but if diagnosed after 6 months of life will have a malignancy rate of up to 65%. Altman classification is the most used classification for SCT, according to anatomical presentation of the lesion (Altman 1974).

Table 1: Altman Classification of SCT

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| Altman classification of sacrococcygeal teratomas 6 . | | |
| **Type** | **Description** | **Malignant risk** |
| Type I (46.7%) | Tumours predominantly external (sacrococcygeal) with only a minimal presacral component | 8% |
| Type II (34.7%) | Tumours presenting externally but with a significant intrapelvic extension | 29% |
| Type III (8.8%) | Tumours were apparent externally, but the predominant mass was pelvic and extended into the abdomen | 34% |
| Type IV (9.8%) | Presacral with no external presentation | 38% |

With the advancement in diagnostic interventions, SCTs are being diagnosed earlier during the prenatal period in the first trimester via detailed ultrasound scan (Lee MY 2011), comprising of 25% of the pregnancies. Several risk factors for an adverse outcome of prenatally diagnosed SCT includes tumour size, growth rate, vascularity, solid morphology and the presence of polyhydramnios, cardiac decompensation, large size placenta or hydrops fetalis (Hambraeus 2016). Tumoral appearance on ultrasound may be solid (most frequently), cystic or mixed. In 15% of cases, it can be purely cystic (Woodward PJ 2005). Our case is one of the example of a rare type of purely cystic SCT Type III as compared to solid or mixed solid-cystic SCT.

SCT can be classified into mature teratomas (MT), immature teratomas (IT) or malignant teratomas (features of yolk sac tumour with choriocarcinoma, embryonal carcinoma or rarer form sarcomas or neuroblastomas). MT and IT usually demonstrate benign clinical behaviour, however, may recur with malignant features (Lo Curto M 2007). 1

20% of the SCTs are associated with congenital abnormalities (polydactyly, microtia, atrial septal defect, cleft lip, hypospadias, club foot, hydronephrosis, hip dysplasia, vertebral abnormalities, Trisomy 13), which in this case the child was born with bilateral hydronephrosis and hydroureter (Rattan KN 2021, Lahdenne P 1991, Lubala TK 2015). High serum AFP level is attributed to the production by the yolk sac tumour (i.e., SCT), foetal liver and foetal gastrointestinal tract. After excision of SCT, AFP has a progressive decrease up to 9 months mean time to normalization and should not be confused with tumoral relapse (Barreto MW 2006). Thus, serial monitoring of AFP 3 monthly may be helpful in detection of recurrence for instance in this case who showed a 99% reduction of serum AFP in 5 months’ time. Recurrence is also commonly associated with incomplete resection of coccyx or inadequate treatment of malignant cases either due to loss to follow-up or lack of neoadjuvant or adjuvant treatment whenever indicated (Lo Curto M 2007). It was recommended for SCT patients to be followed up for at least 5 years with no maximum time for follow up to monitor for recurrence and management of post-treatment sequelae (Santos VDN 2022).

Functional sequelae of SCT includes urinary tract (commonest) and bowel dysfunction, owing to prenatal compression of surrounding pelvic structures and surgical trauma. Other postoperative complications include surgical site infection and/or wound dehiscence which is justifiable by the proximity of surgical site to the anus with colonic bacteria and partially associated with lack of local hygiene and systematization in surgical dressings (Santos VDN 2022). Children after SCT excision often has a distinct scar and irregular buttock contour as well caused by tumour’s fragmentation of gluteal musculature. However, they exhibit a good overall health-related quality of life (HRQoL) during childhood (Hambreus M 2020).

4. Conclusion

In conclusion, pure mature cystic SCT is rare, comprising only 15% of the SCT cases. Mature SCT is usually benign and has a good surgical outcome with manageable post-treatment sequelae such as neurogenic bladder and post operative distinct scar. However, surveillance is crucial as it still possess the risk of recurrence with potential malignant features for at least 5 years with no maximum limit of time for follow up.

Consent

Authors declare that written informed consent was obtained from the patient’s parents for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editorial Board members of this journal.

Ethical approval

Not Applicable

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