

Case Report

Extra-Ordinary Sacrococcygeal Teratoma. A case report and review of the literature

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Abstract:

Sacrococcygeal teratoma is the most common congenital neoplasm in neonates. It is more common in females and is associated with a higher incidence of congenital anomalies. This tumour is often large, vascular, and carries malignant potentials. Management consists of complete tumour excision and removal of the coccyx. We report a one day old female newborn, part of twin born prematurely at 34 weeks of gestation as a product of caesarean section due to pre labor rupture of membranes at King Hussein Medical Center, presented with a very large soft, cystic, pendulous mass over the sacrococcygeal area, with a circumference of about 30 cm. No neurological deficit was elicited and radiography showed no involvement of the underlying bone. Surgery was performed three days after delivery. The tumour was completely excised enblock with coccyx and sent for histopathological examination. Post-operative period was uneventful and the child was discharged in a good health, and is followed up till now, 4 years after surgery and no any signs of recurrence.

Keywords: congenital anomalies, neoplasm, neonates.

Sacrococcygeal teratoma (SCT) is the most common congenital neoplasm of the fetus. This tumor contains derivatives of more than one of the three embryonic germ cell layers and usually arises as a mass in the sacrococcygeal region. Several theories have been proposed to explain the origin of the tumor which occurs in newborn infants and contains cells from all germ layers¹. Bonet believed that the teratoma represented a displaced fertilized ovum while other investigators attribute its origin to incomplete twinning or so-called fetus-in-fetu. However, according to the currently accepted theory, the sacrococcygeal teratoma is probably derived from the totipotent cells of the primitive knot (Hensen's node)^{2, 3}. As the somites develop, the primitive knot migrates in a cephalad direction and locates at the tip of the tail in the six-week-old embryo.

As the tail retracts, the primitive knot comes back to rest at the tip of the coccyx in the ten-week-old fetus. It seems probable that some of the totipotent cells from Hensen's node then develop independently from the rest of the fetus, thus forming the teratoma^{3, 4}.

It is rare disease, incidence of 1 per 40,000 live births. Girls are affected 4 times as often as boys^{5, 6}. The risk of malignancy increases with age, reported rates reaching 10% in neonates and rising to 67% from the 2nd month of life onward⁷⁻⁹. The exact etiology of most SCTs is unknown. Fetuses with this malformation may have associated morbidity and mortality related to prematurity, dystocia, traumatic delivery and intratumoral hemorrhage. The possibility of malignant recurrences after teratomas that were classified as benign on primary resection and the ensuing necessity of radical removal of these tumors has been pointed out frequently⁸⁻¹¹. Management consists of complete tumour excision and removal of the coccyx with particular attention to a good functional and aesthetic result. SCT series are relatively small, the follow-up is limited in time, and the

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studies have usually concentrated on the outcome of malignant tumours^{12, 13}. With regard to ano-rectal function in patients with benign SCT, the follow-up is usually limited to whether or not patients are incontinent^{7, 14}. We are reporting a case of neonate who was operated for a giant sacrococcygeal teratoma (SCT), had intra-operative bleeding into the tumour, followed by cardiac arrest, and was resuscitated successfully.

Case report

A one day old female newborn, part of twin born prematurely on 34 weeks of gestation as a product of caesarean section due to pre labor rupture of membranes at King Hussein Medical Center. On the 20th weeks gestation u/s showed big sacral mass consist of both solid and cystic component which is highly suggestive of sacrococcygeal teratoma. Follow up by u/s was done weekly till delivery which showed no other anomalies. Both twins were breech presentation. After delivery the newborn was healthy, pink, and has mild respiratory distress, and huge sacro-coccygeal mass with approximately (30×25×15) centimeters extending to both buttocks (Fig 1 and 2).



Fig 1: The mass is really giant actually it is larger than the baby.

Tumour was generally cystic in consistency but firm at places with breaching epithelium underlying hemorrhage and echymosis and elements of haemangiomas. Her weight was 2.300 kgm.



Fig 2: The mass is huge and highly vascular

Preoperative investigations, included, CBC, serum electrolytes, abdominal and renal U/S, cardiac Echo. B-HCG. and Alfa-fetoprotein. Lab. Results were within normal limits U/S showed big sacro-coccygeal mass containing both cystic and solid tissue, normal kidneys and other intra- abdominal organs, and no associated anomalies. Cardiac assessment and echocardiogram showed small patent ductus arteriosus with left to right shunt, small atrial septal defect secundum, and normal ventricular function. Alfa-fetoprotein was 950ng/ml, and B-HCG was 0.46 IU/L. Surgery was performed three days after delivery. Inhalational induction was done with Sevoflurane, Atracurium were used as muscle relaxant and Fentanyl as analgesic. Intraoperative monitoring included ECG, heart rate, noninvasive blood pressure, Pulse oximetry and temperature.

Surgery lasted for about two and half hours, the patient was in prone position. (Fig3).

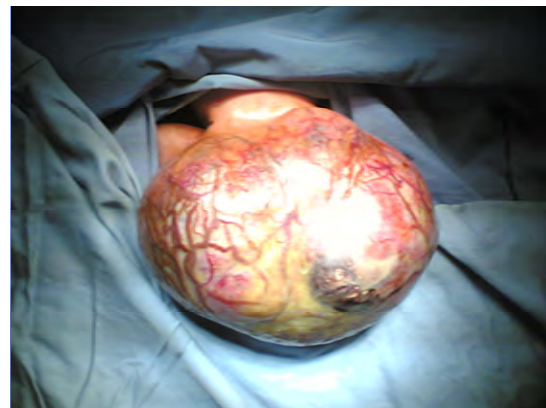


Fig 3: The high vascularity of the tumour

Complete excision of the mass was performed including the coccyx without rupture of the tumor, (Fig 4).

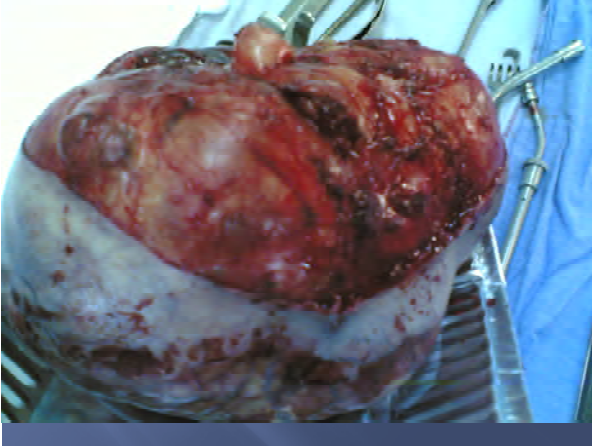


Fig 4: The tumour has different components as can be seen clearly.

Intra-operative blood loss was replaced with 120 ml. of packed cells and 50 ml. of FFP. Near the end of surgery, the patient developed cardiac arrest, the situation was managed immediately by turning the neonate to supine position and external cardiac massaged was started. During CPR, she received adrenaline, bolus of normal saline and calcium gluconate. Normal sinus rhythm with good cardiac output was re-established within 5 minutes. She was electively ventilated in the Intensive Care Unit (ICU) (Fig 5), for 4 days and then extubated successfully.



Fig 5: At the end of the operation.

Post extubation the patient remained stable with no obvious neurological deficit, but she developed wound infection, which was treated by antibiotics and dressings.

She was transfer to the ward. Histopathological report showed immature teratoma grade 3 mixed with mature tissue, containing both solid and cystic component. She was discharged home on the 10th postoperative day. Follow-up at four years revealed that the child had normal developmental milestones, a well-healed scar, and no clinical or biochemical evidence of recurrence.

Discussion

Teratomas are said to arise from primordial cells or totipotent (capable of producing cells representing all three germ cell layers). These primitive cells could come directly from the primitive knot, which rests in the coccygeal region in a fully developed fetus, causing sacrococcygeal teratoma^{15, 16}. Billmire and Grosfeld found that sacrococcygeal teratomas are the most frequently observed teratomas in children¹⁷. The increased incidence of associated congenital anomalies with sacrococcygeal teratomas is well recognized. Most of these are skeleto-muscular and not life threatening. Birch et al. have suggested that there may be common aetiological factors, perhaps genetically determined, which are responsible for the development of germ cell tumors, neural tube defects, and other congenital anomalies⁸.

There have been many studies providing detailed accounts of clinical presentation, pathology, and surgical treatment of sacrococcygeal teratomas¹⁴⁻¹⁶. However, few attempts have been made to analyze the morbidity and mortality of this condition. The main concerns regarding sacrococcygeal teratomas are death prior to transfer to a neonatal surgical centre, intra-operative death commonly related to heavy blood loss, and

malignant change related to delayed treatment or incomplete excision.

An increasing number of these tumours are now being diagnosed antenatally^{10, 18}. Fetal sacrococcygeal teratoma may lead to polyhydramnios and a uterus larger than gestational age, which are also indications for ultrasound scanning in pregnancy¹⁰. Since 2005, antenatal diagnosis in 4 of our patients has led to early referral to the tertiary centre and may have contributed to improved survival. It is notable that there have been no deaths related to sacrococcygeal tumour in our centre in the last 5 years. However, recent reports^{10, 18} suggest that there may be an overall increase in mortality due to referral of increasing numbers of stillbirths, premature babies with large tumours, hydrops, and cardiac failure with high mortality if the presentation is before 30 weeks of gestation¹⁸. Nevertheless, we feel that antenatal diagnosis should at least lead to salvage of neonates such as the one shown in Fig. 1. Antenatal diagnosis and referral to a centre where surgical and anesthetic expertise is available would have avoided this mortality. It is possible that antenatal ultrasound will also detect a larger number of intra-pelvic teratomas, possibly avoiding delay in the treatment of these hidden tumours.

One of the commonest causes of operative mortality in this condition has been hypovolaemic shock from excessive blood loss. Smith et al. found that the main blood supply to sacrococcygeal teratomas arises from the median sacral vessels and recommend early ligation of these vessels to reduce bleeding during operation¹⁹. It has not been our practice to perform early ligation, but we lay great emphasis on extensive haemostasis throughout the operation. Since this tumour is encapsulated, dissection can be performed in a relatively avascular plane between the tumour and the surrounding compressed normal tissues, these measures, together with adequate venous access and blood volume replacement intra- and postoperatively, have eliminated operative mortality.

Another important factor in reducing morbidity and mortality is prevention of tumour recurrence and malignancy. This is dependent upon early and complete excision and long term follow-up as reported by others¹⁴. Because of their obvious nature, types 1 and 2 are generally diagnosed and treated early. However, there is often delay in presentation and diagnosis with type 3 and 4 tumours. The most common presenting symptoms in this group are constipation or urinary dribbling because of bladder and rectal outlet compression by the large tumour.

Rectal examination and serial ultrasound scans have been supplemented by the use of serum alpha-fetoprotein as an index of tumour recurrence. In interpreting these results, it is important to recognise that elevated values are normal in newborns and then decline to adult values by 9 months of age. Persistent elevation or a rise in alpha-fetoprotein from previous normal levels strongly suggest malignancy¹⁷.

In conclusion, mortality and morbidity are generally low in patients with sacrococcygeal teratomas. These results can be further improved by the use of antenatal ultrasound and expert neonatal anesthetic and surgical care, which should be routinely extended to include careful reconstruction of the levator ani and buttocks.

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