Mature sacrococcygeal teratoma: A case report and literature review

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Summary

***Background*: Sacrococcygeal teratomas are derived from embryonic germ cell layers. They present mostly in infancy and are extremely rare in adults; with an associated risk of malignancy. Modern imaging technique may be helpful to delineate the extent of the mass but surgical excision is generally indicated at the time of detection.**

***Patients and methods:* A case report is presented with a review of literature utilising MEDLINE, Microsoft Net and Yahoo search engines.**

***Results:* A three day old female baby presented with a mature sacrococcygeal teratoma containing well-developed limb buds. She had surgical excision and primary repair with good results. A two-year follow up utilising serial serum alpha-fetoprotein assay and CT Scan revealed no evidence of tumour recurrence.**

***Conclusion:* Sacrococcygeal teratoma is a rare tumour that may be benign or malignant. Complete excision is the primary therapy and is adequate if the tumour is benign. Chemotherapy and radiotherapy are however indicated in malignant cases and in recurrence after previous excision.**

Keywords: *Sacrococcygeal teratoma*

Résumé

Introduction: Tératome sacrococygeal est d'origine des couches du gamète embroyonnaire. Le plus souvent, il arrive pendant l'enfance et il est extrêment rare chez des adultes, avec un risque de la malignité associée. La technique nouvelle d'imagerie pourrait être utile pour tracer le degré de masse mais l'excision chirurgicale est souvent opérée au moment de la détection.

Patients et méthodes: Il s'agit du rapport d'un cas avec un bilan d'une literature tout en utilisant MEDLINE, Microsoft Net et mécanismes d'opération du Yahoo.

Résultats: Un bébé du sexe féminine âgée du trio jours était atteinte du tératome sacrococcygeal grave qui contient bouton membre bien mûr. On l'avait opéré à travers l'excision chirurgicale et la chirurgie réparatrice premaire couronnée de success; examens de contrôles d'une durée de deux ans avec l'utilisation d'un serial sérum alpha-fetoprotein essay et CT scanneur n'a indiqué aucune preuve d'une récurrence de la tumeur.

Conclusion: Tératome sacrococcygeal est une tumeur rare qui peut être benigne ou maligne. L'excision complète est la thérapie primaire nécessaire si la tumeur est bengine. Toutefois, la chimiothérapie et la radiotherapie sont nécessaire pour des cas malins et dans le cas de la récurrence aprés une excision précédente.

Introduction

Sacrococcygeal teratoma (SCT) is a rare condition affecting 1 in 35,000 to 40,000 live births[1]. In spite of this rarity, it is the most common tumour of the newborn with a male: female ratio of about 1:4 [2]. Sacrococcygeal teratoma varies considerably in size and are composed of two or three germ layers and multiple tissue types. They are attached to the coccyx and are believed to arise from totipotent somatic cells that originate from the primitive knot (Hensen's node)[3]. Sacrococcygeal teratoma may be predominantly cystic, solid or both. They are usually not accompanied by chromosomal or other physical abnormalities and regardless of their large size, are rarely malignant[2,4]. Only about eleven percent (11%) recur after resection[5].

Modern ultrasound allows prenatal detection of SCT from the second trimester; pre-natally detected SCT has a sur-

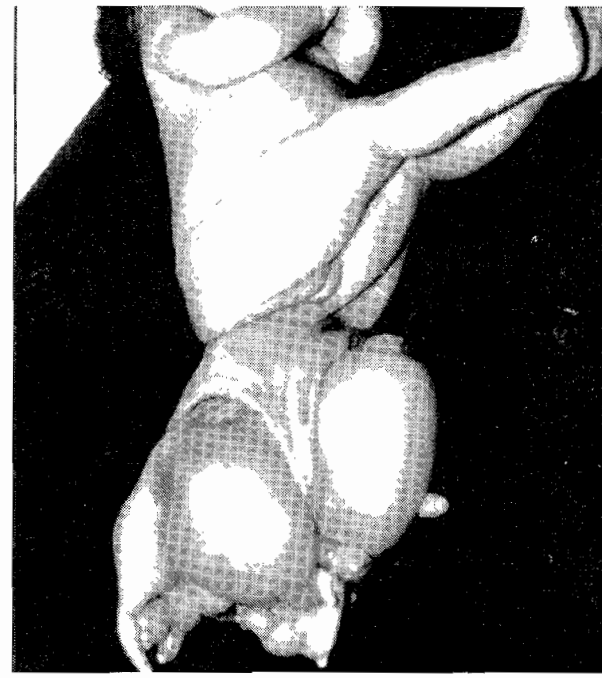

Fig 1 ***Sacrococcygeal teratoma with well-developed limb buds note the relative proportion of the SCT to the trunk of the baby.***

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vival rate of about fifty percent (50%) when associated with complications[1,6,7]. In the absence of complications, babies with prenatally detected SCT have a survival rate of about 88%, only a little lower than the postnatal SCT survival rate of about 95%.[6,8,9]. The commonly occurring prenatal complications are related to the location and mass effect of the tumour; these include obstruction of the ureters or urethra resulting in uropathy, hydronephrosis, and oligohydramnios. Others are hydramnios and placentomegaly resulting in pre-term labour. The most serious complication is high-output cardiac failure which results in hydrops and respiratory insufficiency[10].
We report this rare case of a huge, mature SCT with well developed limb buds, in a healthy baby safely delivered at home and successfully treated in our hospital.

Case report

The three-day-old female Mosotho baby had been born at term to a 26-year-old para 4 full-time housewife. Delivery was reportedly normal by the vagina route at home. There were allegedly no problems during pregnancy and delivery. The baby had been born with a huge swelling in the sacrococcygeal region, which had well-developed limb buds. This abnormal appearance was the reason for referral from the Berea district hospital to the Queen Elizabeth II National Referral Hospital, Maseru, Kingdom of Lesotho.

Clinical examination revealed a healthy looking neonate with a huge mass in the sacrococcygeal region, pink in room temperature with a body weight of 3.8kg and temperature, 37.0°c. The respiratory rate was 22/minute with a heart rate of 132/minute and occipito-frontal circumference of 35.5cm (reference range 35.0 – 37.0cm). There were no other anomalies except the sacrococcygeal mass. Rectal examination revealed normal anal sphincteric tone and a firm lump posteriorly. The huge sacrococcygeal swelling with well-developed limb buds (figures 1 and 2), measured 26cm x 30cm, with normal skin covering. It was partly soft and partly firm in some areas.

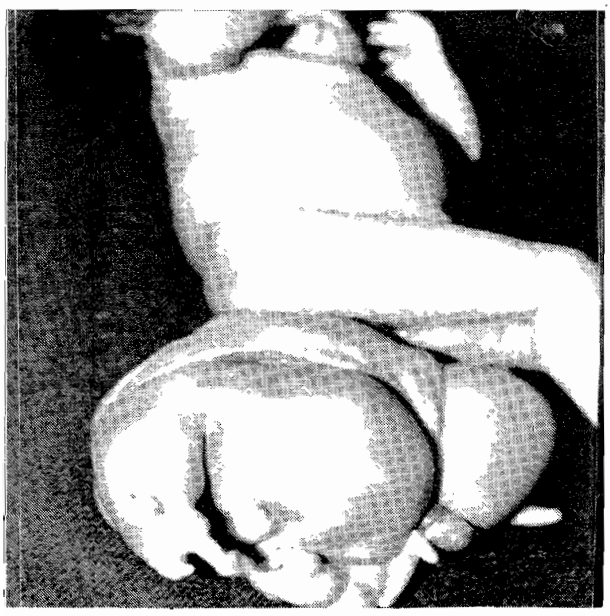

Fig. 2 ***Close-up view of same patient.***

The results of relevant investigations revealed a white blood cell count of 10.1 x 10^9/L (reference range 2.0 – 20.0 x 10^9/L), haemoglobin of 19.9 g/dl (reference range 15 – 22g/dl) with normochronic and normocytic red blood picture. The serum biochemistry values were within the reference range. The Venereal Disease Research Laboratory test (VDRL) was non-reactive and blood group was AB positive. The alpha - fetoprotein assay was normal. Computerized tomography scan (CT-Scan) showed a pre-sacral extension of the tumour. Intravenous urography was normal.

Complete surgical excision of the tumour was accomplished through an inverted chevron incision. The coccyx was excised to facilitate pre-sacral dissection. Routine closure of the wound was done over a penrose drain in the pararectal space. A skin necrosis was observed at the operation site on the third postoperative day. This was managed by daily warm sitzbath till the wound was clean with healthy granulation tissue. A modified Limberg flap was performed three weeks later to cover the defect. Partial breakdown of this flap was noticed on the fourth day postoperatively; the wound was managed conservatively with sitzbath and allevyn cavity dressing. This progressively healed with an acceptable scar and the patient was discharged home for subsequent follow up at the surgical out patient clinic.

Histopathology of the excised tumour confirmed sacrococcygeal teratoma with representative elements of the three germ layers; no malignant features were seen. Subsequent follow up showed normal growth and development by the child. She was continent of faeces and urine. There was no clinical evidence of tumour recurrence: repeat CT scan at the age of 6 months, 15 months and 24 months did not show tumour recurrence.

Discussion

Teratomas, by definition, are tumours that are composed of multiple tissues containing at least two germ-layer derivatives foreign to the part of the body in which they arise[11,12]. Sacrococcygeal teratoma (SCT) accounts for nearly 40% of all cases of teratomas seen in children[12]. In spite of this, SCT is rare, with a reported incidence of 1 in 35,000 to 40,000 live births[1] thus most reported series are small. Larger series, are only reported from large referral hospitals and usually over a long period[13,14]. Tapper and Lack[12] reported 254 patients with teratomas over a 54-year period, and 102 (40%) of these were SCT; this is less than two cases per year. Similarly, Schropp et al[14] reported 73 patients with SCT over a 40-year period, again barely two cases per year. The largest series of SCT was that reported by Altman et al [8] in 1974, this was a collective series of 400 cases by the Paediatric Surgeons of the American Academy of Paediatrics. As a result of this, Altman et al proposed the classification of SCT based on

its extent.

Embryologically, SCT arise from the pluripotent embryonic cells of the coccyx[15]. Though the majority of SCT occur sporadically, a familial occurrence has been reported in association with an autosomal dominant mode of transmission[15,16]. No chromosomal studies were done in the present case and we surmised that the tumour in this female child developed spontaneously in view of the absence of other congenital malformations in the baby and in other members of her family.

Our patient's mother had no pre-natal ultrasound examination and the delivery was at home. Sacrococcygeal teratoma may be diagnosed prenatally from 22 weeks gestation with a uterus larger than the gestational age and/or associated polyhydramnios[16]. Although early prenatal presentation is associated with increased foetal morbidity and mortality, presentation after 30 weeks gestation is a relatively good prognostic indicator for foetal survival[4,16]. Pre-natal diagnosis is important because the tumour may be large enough to cause dystocia. Rupture of the tumour with massive haemorrhage may occur during birth. Thus, it may be necessary to have the mother delivered by caesarian section in a centre where the baby can receive immediate surgical treatment. In view of the severe complications generally associated with SCT diagnosed early in pregnancy, attention is currently being focused on foetal surgery; the procedure of choice when the diagnosis is made early in gestation[4,17].

However, the majority of SCT are apparent at birth with about 75% of cases seen in females[12]; our patient was a female. The reason for this female preponderance is not known[18]. In SCT, the size of the tumour does not correlate with the likelihood of recurrence or poor outcome, but the age at diagnosis and treatment, as well as histological evaluation and stage at the time of resection, are important prognostic factors[8,19]. Complete excision is the primary therapy for all SCT and it is adequate if the tumour is benign. In cases of malignancy, however, recurrence can occur after excision and is associated with a high mortality[15]. Chemotherapy and radiotherapy are therefore also indicated in such cases[15,18]. During surgical resection, the importance of removing the coccyx can not be overemphasized. A high recurrence rate of up to 37% has been reported where the coccyx had not been excised during the initial procedure[20-22].

Alpha-fetoprotein, a well-known marker of teratoma, is valuable in differentiating between mature and malignant teratomas at presentation and during follow up of patients; it may be utilised to detect early occurrence of malignancy[18,23]. This is not only appropriate during the first three postoperative years when recurrence is likely, but also on a long-term basis, as a significant number of them suffer from deficient ano-rectal function and a diminished quality of life[18,24]. In the two-year follow-up period, our patient exhibited a normal growth and development pattern, and was continent of urine and faeces. Furthermore, there was no clinical, biochemical or radiological evidence of tumour recurrence. A longer follow up period is recommended.

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