Managing Sacrococcygeal Teratoma in a New Born of a Psychopathic Widow: Case Report

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SUMMARY

Background: Sacrococcygeal tumors are composite embryonal tumours reflecting any one or more of embryonal/foetal remnants such as germinoma, embryonal carcinoma, teratoma, choriocarcinoma and yolk sac tumors. Teratomas are the commonest variety of these tumours encountered in clinical practice. Sacrococcygeal tumors are most commonly found in females. Male presentations tend to carry high risk of malignancy. Clinical management of sacrococcygeal tumours in males therefore requires more meticulous attention to the details of surgery and follow-ups.

Study design: This is a clinical case report of a huge sacrococcygeal tumour highly valued by a mentally deranged mother, which was excised from a three (3) month old baby boy under general anaesthesia in prone position over a three and half hour period. The purpose of the report is to highlight the special challenges in the management of the case not only on account of the sheer size of the tumour but also the unusual psychopathic attachment of the mother to her baby's tumour. The accompanying literature review was by both manual and Medline searches.

Result: Surgery and postoperative recovery were uneventful. Histology showed tumour variety to be sacrococcygeal teratoma. Patient showed steady progress with each follow-up visit.

Conclusion: Sacrococcygeal tumours are rare, but are the commonest tumours of the newborn. Early surgery avoids tendency to malignant transformation and a good follow-up program is necessary for a guarded prognosis.

Niger Med J. Vol. 50, No. 3, July – Sept., 2009: 74 – 76.

Key words: sacrococcygeal teratoma, germ cell, malignancy, male neonate.

INTRODUCTION

The Sacrococcygeum is the most common location for extragonadal germ cell tumors. This makes Sacrococcygeal tumors to be the most common tumors in the new born.

Germ cell tumours in general are relatively rare, constituting only about 3% of neoplasm in children and adolescents[1]. Sacrococcygeal tumors per se, occur only in I:35,000 births worldwide[2] yet remain the most common tumors in the new born period.[3] The incidence of these tumor in Nigeria is not known with certainty but Nwako and Eziashi (1974) reported seeing about one or two cases per year at the University of Nigeria Teaching Hospital Enugu[4]. Since the inception of Ebonyi State University Teaching Hospital Abakaliki in the year 2000, this is the third case of Sacrococcygeal tumor recorded[5].

CASE REPORT

Baby O.A was a 4-day old male that was born with a tumour at the lower back weighing 3.9kg (baby and tumour). The mother was a 30-yr old psychopathic widow (Gravida[4] Para[4]), a rural farmer by occupation. She had no formal antenatal care, rather took a lot of herbal concoctions during the course of the pregnancy. The antenatal period however was uneventful and, the pregnancy which lasted till term did not at any stage look bigger than date. Family history did not reveal multiple pregnancies. She had a safe delivery at home assisted by traditional birth attendants and the labour was neither unduly prolonged nor difficult.

The baby cried immediately at birth, passed meconium and urine soon after delivery. The presence of this huge tumour at the lower back caused a stir and confusion until it was decided 4-days later, to present the baby to and seek the opinion of the Ebonyi State University Teaching Hospital Abakaliki. At presentation, the male baby was generally healthy and agile, moving all limbs effectively and had no respiratory difficulty and the fontanelles were flat and normotensive. The massive sacrococcygeal tumour at the lower back measured 35cm X 26cm, with cystic and solid areas, and displaced the anus anteriorly (Figure 1).

Rectal examination showed normal sphincter tone and there were no evidence of intra-pelvic extension of the tumour. The mother resisted any interventions initially, but after psychiatric consultations, allowed some investigations to be done. Lumbosacral x-rays showed the tumour as a homogenous soft tissue lesion with no calcification, and all the sacral bones were normal. Other specific investigations including serum alfa feto protein and abdomino-pelvic ultrasonography showed no abnormality. Surgery was delayed for three (3) months because separation from the mother with personality disorder was impossible before that date.

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Patient was assessed preoperatively and found to have Anaesthetic risk ASA3+. One unit of blood was cross-matched for the procedure. Intravenous line was secured through a cut-down on a cubital vein and patient hydrated with 0.18% saline in 4.3% dextrose using a haemoset. With the patient positioned prone under general anesthesia, relaxant technique, surgery was carried out via a "chevron incision". The mass was found to be well encapsulated, with no intra-pelvic extension. It was then excised intact with the coccyx in continuity (figure 2 & 3). Haemorrhage was reduced to the barest minimum by gaining an early control of the pre-sacral artery and vein. However patient received all the packed cells from a unit of whole blood during the operation.

The tumour weighed 2.1kg. Histology reported the tumour as sacro-coccygeal teratoma with mature adipocytes, skin and neural tissue." There were no features of malignancy and hence no indication for chemo- or radio-therapy. Patient was discharged in satisfactory conditions for joint follow up with the psychiatrists.

Figure 1: Patient on induction of anaesthesia.

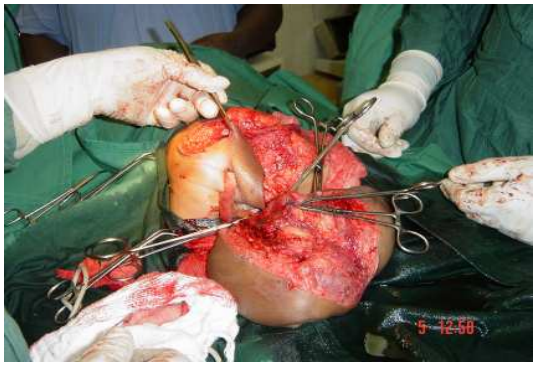

Figure 2: Tumour dissection

Figure 3: Completion of procedure

DISCUSSION

Germ Cell Tumors (GCTs) arise from primitive (primordial) totipotential (or pluripotent) cells featuring embryonic development. They therefore contain cells at different stages of maturity6. 50% of Germ Cell Tumors are found in the gonads with about 45% of these located in the ovary. The other 50% are located at extra-gonadal sites within the soft tissues of the midline. The sacrococcygeal region accounts for about 35% of these cases 7 -as seen in this case. The reason for this predilection for the sacrococcygeum has been a source of many hypotheses:- Ahfeld in the late 1800 propounded the "included twin theory" which felt that sacrococcygeal tumors are results of "failed twins[8] or some form of "fetus in-fetu"[9,10]. Actually there appears to be a higher incidence of these tumors among twins[11,12] a history that the mother denied in this case. Bosaren and Steinman believed that sacrococcygeal tumors are pathologic development of individual cells in the region of the primitive knot (or Hensen's node - the caudal part of the primitive streak)[13]. This may explain why there is a 30% recurrence risk if the coccyx is not excised in continuity with the tumor[14] a precaution that was taken in this case.

Calbet, Bremmer and Willis accept that sacrococcygeal tumors are embryonic tissues in proximity to the primitive streak and notochord but have escaped from the hormonal and enzymatic control of these organs and assumed an uninhibited, unorganized differentiation. This they do by altering their cellular membrane chemistry,[15,16,17]. This may explain why they develop as huge exophytic protruding mass between the coccyx and anus displacing the later anteriorly as seen in this case and also the reason why about 25% of them are malignant[18]. They can obstruct labour hence the recommendation that such women be delivered by caesarean section to avoid both maternal and fetal trauma and, possibly their demise[19,20] -a risk this woman and her baby escaped. Most recently however, Schwalbe postulated that extra gonadal germ cell tumours are from germ cells displaced during embryogenesis and ontogeny and which miss their target - the genital ridge, to reside somewhere else along the midline. [21] About 75% - 90% of sacrococcygeal tumors occur in females, with a male to female ratio of about 1:3[22]. Even here at Ebonyi State University teaching Hospital Abakaliki, this patient was the only male neonate with sacrococcygeal teratoma. Also 75% - 90% sacrococcygeal tumor are benign, but malignant varities are commoner in male neonates[23,24] hence the need for more cautious management of sacrococcygeal tumors in males.

Moreover, before the age of one month, the risk of a sacrococcygeal tumor being malignant in only 5% - but by one year, this risk has risen to 60% and, in children older than one year, the risk of their sacrocoocygeal tumor being malignant is about 75%[25]. The incidence of tumor malignancy is therefore significantly related to the age of the patient and this increase in frequency parallels the higher Altman (American Association of paediatricians AAP) classification[21,1]. This child stood that risk as the surgery was being delayed due to the mother's morbid psychiatric attachment to him which delayed, denied and frustrated many efforts at timely intervention.

As embryonal tumors, sacrococcygeal tumors can posses

any of the germ cell types with the yolk sac elements being the most common malignant variety while teratomas are the most common benign histological variant[26] the presence of such malignant elements is commonly associated with elevation of the baby's serum Alfa feto protein levels[27] hence the necessity to monitor the level of this marker in this child. Also this child will be followed up for at least 3years[28] or even up to adulthood[29] by regular digital rectal examination, plain pelvic x-rays, and USG in the absence of C.T Scan and MRI.

Similarly, the mother has been counseled as to possible recurrence of this tumor in future pregnancies since an isochrome mutation of the short arm of chromosome 12-(i2p) has been shown to be the first hit phenomenon that predisposes to extra gonadal germ cell tumours and sacrococcygeal tumors in particular[30].

CONCLUSION

Sacrococcygeal tumors are rare tumours but they are the commonest tumors in the newborn. 75–90% of them are benign. They are common in females but males harbor more of the malignant varieties. The older the child, the more risk of malignant change in the tumour, hence the need to excise these tumors in the neonatal periods. Therefore when both odds weigh against the patient (a male patient, in the post neonatal age group), the risk for malignancy is heightened.

We have presented the case of a male neonate, who, on account of his mother's psychiatric condition, had his sacrococcygeal tumor grown to a weight of 2.1kg and, the child up to the age of 3 months before the tumour was excised. The histology confirmed the tumor to be a mature sacrococcygeal teratoma. The child and his mother are still being followed up.

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