

Case Report*Open Access, Volume 3***Sacrococcygeal teratoma in Nigerians: A case report and clinico-pathologic review****Ukachukwu AK^{1,2*}; Aghahowa ME^{3,4}; Ezike KN^{5,6}; NwokorieRM⁷; Nwaribe EE⁸; Anaeto SI¹**¹Neurosurgery Unit, Department of Surgery, Asokoro District Hospital, Abuja, Nigeria.²Duke Global Neurosurgery and Neurology, Durham, North Carolina, USA.³General Surgery Unit, Department of Surgery, Asokoro District Hospital, Abuja, Nigeria.⁴Department of Surgery, College of Health Sciences, Nile University of Nigeria, Abuja, Nigeria.⁵Department of Pathology, Asokoro District Hospital, Abuja, Nigeria.⁶Department of Anatomic Pathology, College of Health Sciences, Nile University of Nigeria, Abuja, Nigeria.⁷Department of Anesthesia, Asokoro District Hospital, Asokoro, Abuja, Nigeria.⁸Neurosurgery Unit, Department of Surgery, National Hospital, Abuja, Nigeria.***Corresponding Author: Alvan Ukachukwu**Consultant Neurosurgeon, Asokoro District Hospital,
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Abstract

Introduction: Sacrococcygeal Teratomas (SCTs) are rare childhood tumors, commonly seen in the neonatal period. The mainstay of their diagnosis is both clinical and radiological. Early gross-total surgical excision has the best prognostic outcome, especially for benign tumors.

Case report: We present the case of a 12-year old girl with recurrent infected right gluteal benign SCT who was successfully managed in a secondary-level health facility in Nigeria. She has remained symptom-free with no recurrence four years post-surgery. We also reviewed the medical literature on SCT in Nigeria.

Discussion: Our case report and others from Nigeria are consistent with the global trend on SCTs. They mostly occur within the first 15 years of life, with a bimodal peak in the first 4 years and at 8-11 years. 75-90% are benign, and the tumors often contain tissues from the three germ layers. Complete surgical excision is the treatment of choice.

Conclusion: SCTs are rare tumors, but require early diagnosis and complete surgical excision to prevent malignant transformation. The case report draws attention to the need for a multi-institutional tumor registry in Nigeria and other sub-Saharan African countries.

Keywords: sacrococcygeal teratoma; surgical excision; nigeria; literature review.

Abbreviations: CT: Computed Tomography; MRI: Magnetic Resonance Imaging; SCT: Sacrococcygeal Teratoma.

Introduction

Teratomas are tumors that arise from the proliferation of pluripotent stem cells which can differentiate into ectodermal, endodermal, and mesodermal derivatives in embryonic life [1,2]. These tumors occur in various gonadal and extragonadal regions, with Sacrococcygeal Teratomas (SCTs) being the most common extragonadal teratoma [2,3]. SCTs are rare childhood tumors [1-4]. They are, however, the most common tumor in the neonatal period [5-7]. SCTs account for up to 30% of all childhood teratomas [1] and usually present as benign or malignant forms. SCTs are very rare in adults, with less than 100 cases reported in Teratomas are tumors that arise from the proliferation of pluripotent stem cells which can differentiate into ectodermal, endodermal, and mesodermal derivatives in embryonic life [1,2]. These tumors occur in various gonadal and extragonadal regions, with Sacrococcygeal Teratomas (SCTs) being the most common extragonadal teratoma [2,3]. SCTs are rare childhood tumors [1-4]. They are, however, the most common tumor in the neonatal period [5-7]. SCTs account for up to 30% of all childhood teratomas [1] and usually present as benign or malignant forms. SCTs are very rare in adults, with less than 100 cases reported in the literature [8].

Several theories have been adduced to the origin of SCTs. The Budde-Willis hypothesis suggests that teratomas arise from cells released from the normal developmental control of the primary embryonic organizer [1]. The germ cell theory postulates an origin from primordial germ cells either within the gonadal anlage or in extragonadal sites following arrest in germ cell migration from the yolk sac to the gonadal anlage in embryonic life [1,8]. Other theories suggest derivation from multipotent cells in Henson's node which migrate caudally to the coccygeal region; wandering germ cells derived from a non-parthenogenetic source, abandoned during the migration of embryonic germ cells from the yolk sac to the gonad; and origin in totipotential or pluripotential embryonic cells [3,8,9].

We report the successful management of a huge infected recurrent sacrococcygeal teratoma in a pre-teen Nigerian girl.

Case report

A 12-year-old girl presented with a huge lump in the right gluteal region from birth. Surgery was done at 3 months of age in a rural health center, but it soon recurred and progressively increased in size. There was associated recurrent, intermittent, occasionally colicky pain that was relieved with simple analgesics in the last 5 years which progressively increased in its frequency and duration, and was worse on defecation. There was significant wasting of the right lower limb, with associated weakness, tingling, and numbness. She had recurrent difficulty with micturition and defecation. The girl was the 8th of 15 children in a polygamous home. She had been living in an internally displaced persons camp for 3 years following the destruction of her community by the Boko Haram insurgents. The cost of her care was covered by donations from a philanthropic religious organization and waiver of some fees by the hospital management.

She was ambulant with the aid of walking crutches and had a large multilobulated mass measuring 13 X 12 X 11 cm in the right lower lumbar and sacral regions, extending across the

midline to the left, with an irregular oblique hypertrophic scar over its mid aspect. The mass was soft and slightly tender, with hyperpigmented skin having multiple draining sinuses. The patient also had significant atrophy of the right lower limb with reduced muscle power (grade 3 on the right, grade 5 on the left) and hypoesthesia. Our clinical diagnosis was an infected sacrococcygeal teratoma.

Plain lumbosacral spine radiography did not show any abnormality. Plain pelvic radiography showed a right-sided hemipelvic soft tissue mass, with pelvic asymmetry evidenced by a small right hip and femur. Magnetic Resonance Imaging (MRI) of the lumbosacral spine (Figure 1) showed a well-defined oval-shaped sacrococcygeal mass measuring 12.4 X 13.5 X 12.7 cm, extending from the right lower lumbar region, across the midline, to the S3 presacral space. The mass was heterogeneous on T1W, T1 contrast, and T2W sequences, with multiple hypointense non-enhancing regions, divided by enhancing septations, suggesting areas of necrosis or loculated abscess collections. There was associated elevation/splaying of the adjacent sacrococcygeal bone, with mild straightening (but preserved alignment) of the lumbar spine. The other visualized vertebrae and intervertebral discs were normal in morphology and signal intensity. The urinary bladder had significant residual urine volume.

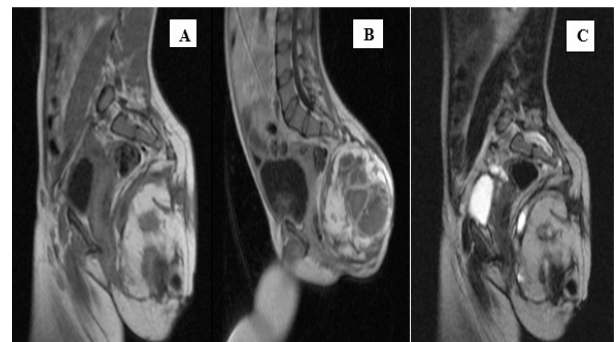


Figure 1: Shows T1W (A), T1 contrast (B), and T2W (C) MRI sequences of the lumbosacral and gluteal region.

She was worked up for surgery and had pre-operative bowel preparation, with additional low rectal washout with soap and water enema on the morning of surgery. Intra-operatively, she received antibiotic cover and had gross-total en-bloc excision of the mass via a sacral approach done by combined neurosurgery and general surgery team. The intra-operative findings included an oblique hypertrophic scar over the mass and gluteal region, multiple discharging sinuses, and well-circumscribed encapsulated thick-walled mass with cystic portions containing purulent fluid and solid portions having bone and cartilaginous tissue in situ (Figure 2). There was no bowel involvement.

Postoperatively, she received culture-sensitive parenteral antibiotics for 5 days, and subsequent oral antibiotics for 14 days. She had transient urinary and fecal incontinence which resolved within 3 days, and superficial wound dehiscence which was managed with topical honey-impregnated dressings. The patient was discharged home on the 24th postoperative day for subsequent outpatient follow-up. She has remained symptom-free with no recurrence for four years.

Macroscopically, the mass was 12 X 12 X 11 cm weighing 649

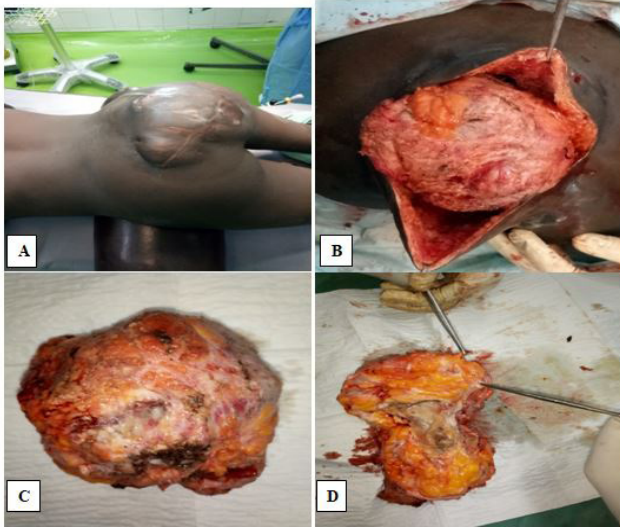


Figure 2: A: The patient is in prone position. The mass in the lumbosacral/gluteal region is irregular and multilobulated, with an oblique hypertrophic scar on the skin and multiple discharging sinuses. Atrophy of the right lower limb can also be seen. B: An elliptical skin flap has been raised showing a well encapsulated fleshy mass. C: The mass has been dissected out en-bloc, and has multiple sinuses with purulent exudate. D: Incised mass shows a central cyst sac with cartilaginous tissue in the thickened wall.

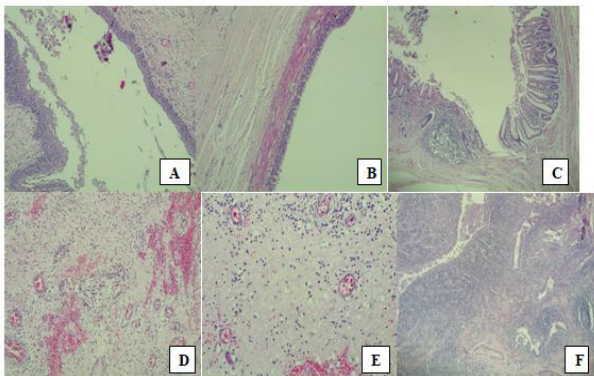


Figure 3: Microscopic sections showing cystic structures/tissue lined by squamous epithelium (A), respiratory epithelium (B), and intestinal epithelium (C). Sections also show immature cartilage (D and E), and dense inflammatory infiltrates (F).

g, with 2 accompanying skin tissue flaps measuring 14 X 12 cm and weighing 7 g each. Cut section through the mass showed it to be partly solid and partly cystic, and greyish-yellow in color. The cystic cavities were filled with whitish gelatinous materials. Microscopic sections (Figure 3) showed tissue covered by skin which exhibited hyperkeratosis and acanthosis of the epidermis which overlay a germ cell neoplasm composed of cystic structures and tissues from the 3 germ layers: mature skin with adnexal structures from ectoderm; pseudostratified ciliated respiratory epithelium and intestinal-type epithelium with goblet cells from endoderm; and immature cartilage from mesoderm. The stroma was densely infiltrated by mixed inflammatory cells including neutrophils, lymphocytes, plasma cells, and macrophages. Multiple foci of abscess collection were also seen. The histological diagnosis was Grade 1 Immature Teratoma.

Discussion

There is an increased interest in SCTs because of a rising hospital incidence [3,10]; estimated at 1 in 35,000 – 40,000 live births [4,6,8]. SCTs occur in patients between 1 day to 15 years of age in most case series [1-4], with a bimodal peak in the first 4 years of life and between 8 and 11 years [1,2]. A retrospective review of 74 cases from the pathology specimen register in Ibadan, South-West Nigeria, showed an average age of 4.3 years for all teratomas, with a range of 1 day to 15 years [1,2]. Our index patient (a 12-year old girl) falls within this age range. SCTs are, however, rare in Nigerian adults, as only a few reports document their presence in the adult population [8,12-14]. All the reports note that teratomas, including SCTs, are more common in females, with a female to male ratio of 5-18:1 [1-5,8,15].

Pathologically, SCTs have been classified as benign or malignant; cystic, solid, or mixed; and mature, immature, or malignant [1-3]. 75-90% of SCTs are benign, and males account for more of the malignant cases [7]. The malignant types have been further classified as embryonal carcinomas or yolk sac tumors, including endodermal sinus tumors [1-3]. Our patient had a benign mixed immature teratoma, similar to other Nigerian reports [1,2,5,12,13,16].

SCTs contain a variety of tissue types derived from the 3 germ cell layers – ectoderm, mesoderm, and endoderm [1]. Our patient's histology showed tissue from all 3 germ layers: mature skin with adnexal structures from ectoderm; pseudostratified ciliated respiratory epithelium and intestinal-type epithelium with goblet cells from endoderm; and immature cartilage from mesoderm. A case report from Enugu, South-East Nigeria, noted the tumor contents to be mature adipocytes, skin and neural tissue [7], while another case series reported the presence of cardiac muscle in some of their excised SCT specimens [9]. In the pathology specimen series from Ibadan, derivatives of all 3 germ layers were seen, including breast tissue, salivary gland, hair, sebaceous material, and pancreatic tissue showing both islets of Langerhans and exocrine acini [1,2].

The clinical presentation of SCTs includes gluteal or sacral mass, sacrococcygeal pain, bladder dysfunction/acute urinary retention, and chronic constipation or fecal obstruction [3,4,8]. Other rare modes of presentation are neurologic symptoms (including intractable sciatica, loss of anal reflex, and lower limb weakness), abdominal and perineal pain, anemia, elevated urea, sacral skin discoloration or hairy nevus, tumor surface ulceration, inguinal lymphadenopathy, pulmonary metastasis, and absent anal opening [3,8]. In Sokoto, North-West Nigeria, Legbo et al reported the case of a newborn presenting with a fully formed third lower limb in the sacrococcygeal region [6]. The patient in our report presented with an infected gluteal mass associated with intermittent pain, bisphincteric incontinence, right lower limb wasting, and weakness.

SCTs may be associated with other congenital anomalies such as polydactyly, talipes calcaneovarus, encephalocele, ano-rectal malformation, omphalocele, sacral agenesis, syndactyly, and renal and cardiac malformations [4,9,17,18]. The differential diagnoses of SCTs are sacral myelomeningocele or meningocele, fibro-lipoma, embryonal rhabdomyosarcoma, anterior meningocele, neurofibroma, cystic hemangioma or lymphangioma, rectal duplication cysts, anal gland cysts, intracanalicular epidermoid tumors, dermal sinus stalks, hydromyelia, external Wilms tumors, retrorectal hamartomas, neuroblastomas, and

pacinomas [4,8,9,17,19].

Clinically, Altman et al. classified SCTs as follows [20]:

- Type I: predominantly external with minimal presacral component
- Type II: present externally but with significant intrapelvic extension
- Type III: external but predominant parts are intrapelvic with abdominal extension
- Type IV: presacral with no external presentation.

Type I tumors are the most common worldwide and in Nigeria [3,4,10,11], just as in our patient.

Clinical assessment and radiologic imaging are the mainstay of diagnosis of SCTs [1,3,4,10,20]. Current investigation modalities include the use of Computed Tomography (CT) scans and MRI [8,17,21]. While CT scans delineate the lumbosacral and pelvic bony architecture well, MRI is better at delineating the soft tissue architecture. Pelvic and lumbosacral radiography, barium enema, and abdominopelvic ultrasonography are also routinely used, especially where the more current imaging modalities are unavailable or unaffordable [4,7,8,11,14]. Our patient's presentation and treatment were delayed because of financial constraints; a philanthropic organization covered the costs of her investigations.

SCTs are diagnosed prenatally using routine 2-dimensional, advanced 3-dimensional and doppler ultrasonography, as well as ultrafast MRI [6,15,17,21], thus enabling anticipated delivery and early commencement of appropriate treatment. Treatment options include intra-uterine decompression of cystic teratomas [22] and planned early operative delivery of the fetus [17,21]. Early diagnosis and treatment of SCTs are important to prevent malignant transformation, estimated at 5% before one month of age, 60% by one year, and 75% after one year [5,7]. Our patient presented at 12 years of age without malignant tumor transformation.

Complete surgical excision is the treatment of choice for SCTs [3,4,6-9,15], and this can be done with excision of the coccyx to remove all nidi of pluripotent cells that could increase the risk of recurrence [10]. The approach could be through trans-abdominal, trans-perineal, trans-sacral, or combined abdomino-sacral routes [3,4,8,15]. Adjuvant or neoadjuvant radiotherapy or chemotherapy are known adjuncts to surgical excision, especially in malignant tumors [3-5,15,23]. Our patient had gross total en-bloc excision of the mass and overlying skin via a trans-sacral approach. The underlying defect was covered with mobilized muscle and skin flaps from the gluteal region. We did not consider adjuvant chemotherapy or radiotherapy because the histology was benign. Our approach is similar to procedures reported from other parts of Nigeria [3].

The outcome of the treatment of SCTs is good for benign tumors that are diagnosed and resected early [3,4,9]. Our patient's histological diagnosis of type 1 immature teratoma is benign, and she has been symptom-free, with no recurrence, in four years of follow-up. In the series from Lagos, all 19 patients with benign tumors were alive at 2 years or more of follow-up, while all 11 patients with malignant tumors had tumor recurrence within 3 months and died within 12 months [3].

The postoperative complications include wound infection, wound dehiscence, post-operative diarrhea, post-operative hemorrhage, intraoperative rectal injury, and malignant recurrence [4,15], with a post-surgical mortality rate of 18% in neonates, and up to 55% in the post-neonatal period [9]. Our patient had pre-operative bowel preparation, with additional low rectal washout with soap and water enema on the morning of surgery as a precaution in case the tumor involved the rectum. This would prevent fecal contamination of the surgical wound and enable primary repair of any involved part of the rectum. However, she had transient urinary and fecal incontinence which resolved over time, and wound dehiscence which was managed with topical honey-impregnated dressings. As a consequence, her discharge from the hospital was delayed until 4 weeks post-surgery.

Conclusion

Sacrococcygeal teratomas are rare tumors but are the most common types of teratomas in childhood. Early diagnosis and prompt complete surgical excision prevent malignant transformation in benign tumors. This paper has discussed the case of a 12-year old girl with SCT who was successfully managed with complete en-bloc surgical excision in a secondary health facility in Nigeria. The case report shows that gross-total surgical excision of this rare tumor has an excellent prognosis for benign tumors, and can be successfully performed in a resource-limited setting. The paper is limited by the fact that it is a single-patient case report and does not represent the full clinicopathological and outcome profile of this tumor in the Nigerian population. This highlights the need for a multi-institutional tumor registry in Nigeria through which detailed epidemiological, clinicopathological, management, and outcome studies on rare tumors, like SCT, can be conducted.

Disclosure: The authors declare no conflict of interest.

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