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Outcome of management of complicated extragonadal teratoma in a resource poor setting

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ABSTRACT

Background: Extragonadal teratomas (EXGTs) are ubiquitous in the human body; hence, they have varied presentation. In underdeveloped areas presentation and management are affected by socio-economic, cultural and health facilities factors. The aim of this study was to review the outcome of management of complicated EXGT in a tertiary health centre. **Materials and Methods:** A review data of paediatric patients with EXGT was done between January 1999 and December 2012. Variables reviewed were bio-data, mode of presentation and site of tumour, comorbidity, treatments and outcome. The data was analysed with Statistical Package for Social Sciences (SPSS(R)) version 16.0. **Results:** There were 21 complicated EXGT (77.8%) among 27 children, age ranges from 4 days to 16 years (median = 2 years). Male:Female ratio of 1:2. The complications per region of the body at presentation were cervical 4 (66.7%), mediastinal 2 (100%), abdominal 3 (75%) and sacrococcygeal 12 (75%). The complications were respiratory distress 6, intestinal obstruction 5, faecal incontinence 2, bladder outlet obstruction 3, malignant transformation 5, ruptured sacrococcygeal teratoma 2, ulcerated tumour 2, anaemia 3 and malnutrition 3. There were 5 (23.8%) progressive disease post-excision outside our facility. Excision biopsy was successful in 19 (85%) patients two of which had neoadjuvant cytotoxic therapy. Overall mortality was 5 (23.8%) (septicaemia, anaemia, respiratory distress, renal failure) and post-excision mortality was 11.8% (endotracheal tube blockage and progressive disease). **Conclusion:** Delay presentation (due to local belief, ignorance and poverty) malnutrition, sepsis, malignant transformation characterised presentation of children in this study and the lack of paediatric intensive care unit facility and intensivists compromised survival of children with EXGT.

Key words: education, morbidity, supportive care, teratomas, health facility

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INTRODUCTION

Extragonadal teratomas (EXGTs) are rare, accounting for only 1-5% of germ cell tumours.[1] They have a ubiquitous distribution in the human body, hence their presentation varies. In addition, they sometimes show unusual, distinctive features unlike those commonly described. This may be in the form of unusual location, acute presentation as an emergency or an unusual phenotypic appearance.[2] In our environment, late presentation is common for many reasons sometimes from social and economic factors; hence, they usually present when complications have set in.[3]

This study aims to review the outcome of management of complicated EXGT in a tertiary health centre.

MATERIALS AND METHODS

A review of 14 years data on paediatric patients with EXGT managed in our institution between January 1999 and December 2012 was done. The variables reviewed were bio-data, mode of presentation, site of tumour, comorbidity, treatments, histology and outcome. The data was analysed with Statistical Package for Social Sciences (SPSS(R)) software version 16.0.

RESULTS

There were 28 patients managed for EXGT out of which 22 had complicated EXGT. The age of the

patients ranges from 4 days to 16 years (median = 2 years). Male:Female ratio of 1:2. The distribution of the complications per region at presentation were cervical 5 (71.4%), mediastinal 2 (100%), abdominal 3 (75%) and sacrococcygeal 12 (75%) [Table 1]. 5 (22.7%) complications were progressive disease post-excision outside our institution. This was due to incomplete excision perhaps they were malignant ab initio or had malignant transformation. The complications noted were as shown in Table 2. Excision biopsy was successful in 19 (86%) patients two of which had neo-adjuvant cytotoxic therapy after biopsy of the tumour mass. The histologic types are shown in Table 3 with mature teratoma being the most common (59.1%). Post-excision 2 (10.5%) had respiratory stridor and only 1 (5.3%) had recurrence [Table 4].

Post-excision mortality was 3 (15.8%) one died due to endotracheal tube blockage in the intensive care unit (ICU) and two from progressive growth of tumour. Overall mortality was 5 (22.7%) this was due to septicaemia, anaemia, respiratory distress or renal failure.

Table 1: Distribution of complications of EXGT by region of the body

Region	Patients (total)	% per region
Cervical	5 (7)	71.4
Mediastinal	2 (2)	100
Abdominal	3 (4)	75
Sacrococcygeal	12 (15)	80
Total	22 (28)	78.6

EXGT: Extragonadal teratoma

Table 2: complications seen in EXGT at presentation

Complications	Number of patients
Respiratory distress	7
Dysphagia	2
Intestinal obstruction	5
Faecal incontinence	2
Bladder outlet obstruction	3
Malignant transformation	5
Rupture	2
Ulceration	2
Anaemia	3
Malnutrition	3
Renal failure	1

EXGT: Extragonadal teratoma

Table 3: Histologic types of excised EXGTs

Histologic type	Number of patients	%
Mature	13	59.1
Immature	4	18.2
Endodermal sinus tumour	3	13.6
Undetermined	2	9.1
Total	22	100

EXGTs: Extragonadal teratomas

Table 4: post-excision complications

Outcome	Number of patients
Faecal incontinence	0
Respiratory stridor	2
Urinary stress incontinence	1
Limb paralysis/paresis	0
Recurrence	1

DISCUSSION

EXGT is rare tumour in neonates and infants and arise as a result of aberrant migration of the progenitor germ cells.[4] Despite this common origin, this tumour demonstrate diverse clinical and morphological variations. It at times show unusual features owing to its location and can present acutely as an emergency.[2] Teratoma can occur in almost any region of the body and in any organ, but they are commonly observed in the paraxial and midline location.[5] The distribution of teratomas includes ovaries, testes, anterior mediastinum, retroperitoneum, presacral and coccygeal areas, pineal and other intracranial sites and neck.[6,7] Williams *et al*.[8] showed that the neck is the second most common site after the sacrococcygeal region in Nigerian children. This is in keeping with what we found in this study. The sacrococcygeal region is the most common site of location in this tumour, which should give away diagnosis and make parents present their wards, but this is not the case in many of these patients. Rather both hidden (Altman type IV) and the obvious tumours (types I-III) were presented late in these patients. Some even had treatment by the untrained 'surgeons' that did incomplete excision of the tumour, which led to malignant recurrence in three of patients with sacrococcygeal teratoma (SCT). One female child presented at our centre with type III SCT at 7 months of age with pelvic organ compression causing bladder outlet obstruction, constipation and progressive bilateral lower limbs oedema from venous compression [Figure 1]. She had diverting loop colostomy, suprapubic cystotomy and emergency excision of the tumour through combined abdominal and sacrococcygeal approach. EXGT can present with urological emergency when located in the presacral or abdominal region as they can obstruct the bladder outlet.

Most of the patients in our series presented late (median age of 2-years) with complications (78.6%); most of which needed to be treated as an emergency. The occurrence of malignant transformation is common

in patient with hidden tumours or those with delay presentation.[9,10] The female preponderance is still consistent with reports in other series.[5-7]

The late presentation is as a result of inability to afford interventions offered at delivery, refusal of surgery because of lack of conviction of surgical intervention in neonates, prior excision by the untrained who did not even send specimen for histology and hidden tumour in some patients.

The complications seen varied with the location of the tumour as 71.4% of those in the cervical region presented with acute respiratory obstruction necessitating emergency excision in the neonatal period [Figure 2]. One of these patients died on 2nd day post-excision from blocked endotracheal tube while being electively ventilated for persistent tracheomalacia in the ICU. This experience made us to be more careful with subsequent patients and we never recorded any mortality again. Our earlier experience was uneventful in one neonate who had successful excision of cervical teratoma.[11] All two patients that presented with mediastinal teratoma were detected late while being investigated for causes of respiratory obstruction and dysphagia. Mediastinal teratoma are known to be discovered accidentally when the patient is being investigated for other symptoms such as cough, dyspnoea, chest pain and other symptoms of mediastinal structure compression.[12] Our patients had uneventful mediastinal tumour excision.

About 80% of EXCT are benign while 20% are malignant.[5] EXGT histologically can be mature (benign), immature or endodermal sinus tumour. This has a bearing on the type of treatment. Most of the EXGT in our study were benign, which agreed with other previous study on teratoma in Nigeria[5] and would have benefited from early and complete removal as those that had malignant transformation might have been helped with a cure.

The treatment of choice has ever been a complete surgical excision.[13,14] This usually suffice, but in most of these complicated EXGT there was a need for extra intervention to take care of the complication, i.e., colostomy, suprapubic cystostomy and neoadjuvant chemotherapy for malignancy. One of the patient who had renal failure would have also benefitted from dialysis, which was not possible because the parents could not afford the cost and patient was not fit for upper tract diversion of the ureters [Figure 3].

Post-excision event free survival ranges from 92.2% to 85% in the UK.[15] In another series in Italy, it was



Figure 1: Transverse loop colostomy and suprapubic cystostomy in a 7-month-old girl with Altman Type III sacrococcygeal teratoma which caused intestinal obstruction and urinary retention. See the oedematous lower limbs and perineum from pelvic vessels compression



Figure 2: Tracheal compression from a cervical teratoma in a neonate. See the central and peripheral cyanosis in the baby



Figure 3: Massive vulva and perineal oedema extending to the lower limb in a patient with recurrent SCT 1 year post-excision. Tumour extended into the retrorectal space and pelvis causing anal incontinence and bladder neck compression

90.4%.[16] In our series where patients were presented with complications we achieved 84.2% survival.

CONCLUSION

Delay presentation due to local belief, ignorance and poverty, malnutrition, sepsis, malignant transformation

characterised presentation of children in this study and the lack of paediatric ICU facility and intensivists compromised survival of children with EXGT. The untrained surgeons in the community need to be educated to know their limits and refer patients accordingly.

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