SJIF Impact Factor: 3.458

WORLD JOURNAL OF ADVANCE HEALTHCARE RESEARCH

ISSN: 2457-0400
Volume: 3.
Issue: 3.
Page N. 90-93
Year: 2019

Original Article www.wjahr.com

PROFILE OF PAEDIATRIC CHEST WALL TUMOURS AND MANAGEMENT OUTCOME IN A TERTIARY HEALTH FACILITY IN SOUTHERN NIGERIA

Okoro Philemon E.*[1], Gbobo Isesoma[1] and Obiora Christopher[2]

[1]Department of Surgery, University of Port Harcourt Teaching Hospital, Port Harcourt, Nigeria.
[2]Department of Pathology, University of Port Harcourt Teaching Hospital, Port Harcourt, Nigeria.

Received date: 16 April 2019 **Revised date:** 06 May 2019 **Accepted date:** 27 May 2019

***Corresponding author: Okoro Philemon E.**

Department of Surgery, University of Port Harcourt Teaching Hospital, Port Harcourt, Nigeria.

ABSTRACT

Aim: To report our experience of chest wall tumours in children in our region and to highlight the peculiar features and management outcome. **Settings and Design:** This is a 5 year retrospective study of consecutive cases of chest wall tumours in children 15 years and below seen in our service between August 2013 and July 2018. **Patients and Methods:** We reviewed the medical records of patients who met our inclusion criteria. Data retrieved included: age, gender, duration before presentation, presentation feature, previous interventions, treatment offered, histology, follow-up period, outcome. Data was subjected to simple statistical analysis. **Results:** A total of 41 children with chest wall tumours were seen during the period under review but 37 had adequate records to be included in the review. Histological reports showed benign neoplasm in 16 cases and malignancy in 21 cases. There were 6 mortalities and 5 recurrences. **Conclusion:** Chest wall tumours are among the surgical problems encountered in children in our region. These tumours are often presented late and a significant percentage malignant. Surgical removal is imperative but can pose technical challenges. There is need to enlighten the public to encourage early presentation and treatment.

KEYWORDS: Chest wall; Tumours; Paediatric; Pattern; Management Outcome.

INTRODUCTION

Chest wall tumours (CWT) in children are heterogenous comprising both benign and malignant ones. The location of these tumours gives them peculiar attributes regarding their morbidity, treatment and outcome. This is also the reason they are considered together as a group.[1,2,3] Their mere presence on the chest wall in a child can impair the respiratory mechanism of the thoracic region leading to hypoxia or even respiratory failure. Closure of the skin following excision of large chest wall tumours can be challenging requiring flap rotation or even thoracic cage reconstruction.[4] Small tumours which could have been ordinarily considered resectable on the limbs may be inoperable due to direct invasion of the chest cavity and underlying viscera. In addition, neonates and other younger children may have difficulty with spontaneous respiration, and require respiratory support following surgical removal of large chest wall tumours. These attributes are among the reasons paediatric chest wall tumours and their management remain a challenge and objects of debate among surgeons. Paediatric chest wall tumours requiring extensive thoracic excision and chest wall reconstruction place significant demand on the skills of the surgeon and the appropriate diagnostic infrastructure.(Fig 1) Reports on paediatric chest wall tumours are generally scanty. This paucity of data on the pattern of these CWT, their management and outcomes makes it difficult to assess improvements or otherwise in the care of children with CWT. This study was therefore conducted to report the experience with these tumours in children in our region and to highlight the peculiar features and management challenges. We hope that this report will provide a pedestal upon which other studies on this subject can build and pave way for improvements in the quality of care for children with CWT.

MATERIALS AND METHODS

This is a 5 year retrospective study of consecutive cases of chest wall tumours in children 15 years and below seen in our service between August 2013 and July 2018. Inclusion criteria were: tumour in any part of the thoracic wall in a patient 15 years and below. Exclusion criteria were: non neoplastic swellings like infective, inflammatory, or traumatic chest wall swellings; age above 15 years; and incomplete data. Data obtained

included gender, age, prior interventions, histological type of tumour, treatment, follow up duration, and outcome. Data was subjected to simple statistical analysis using the microsoft excel softwareversion 2007.

RESULTS

A total of 41 children with chest wall tumours were seen during the period under review but 37 of them met the inclusion criteria. These comprised 24 males and 13 females. The age range was 2 weeks to 14 years with a mean of 6 (±0.5) years (Table 1). The duration of swelling was variable from 2weeks to 3 years (Table 2). Diagnosis was established based on reports from open biopsy. FNAC was used in 5 cases which we considered inoperable or required neoadjuvant chemotherapy. Histological reports showed benign lesion in 16 cases and malignant lesion in 21 cases. (Table 3) Eighteen (48.6%) patients had received some form of treatment from natives and charlatans before presentation. Unorthodox interventions included drinking of native concoctions, skin scarifications, massage, and skin application of concoctions. Procedures and treatment given in our centre included: incisional biopsy, simple, wide, or partial excision with or without flap rotation or skin grafting, and cytotoxic chemotherapy.(Table 4) There were 6 mortalities, 2 of whom had evidence of metastasis at the time of presentation. There were 5 recurrences after an average follow up period of one year.(Table 5).

Table 1: Age of patients.

Age of patients (Years)	Number of patients (n=37)
<2	10 (27.0 %)
2-5	8 (21.6%)
5-10	14 (37.8%)
10-15	5 (13.5%)

Table 2: Duration of symptoms of CWT before presentation.

Duration before presentation	Patients (n=37)(%)	Prior non orthodox treatmentn(%)
< 1 month	10(27.0)	2 (20.0)
1-3 months	7(18.9)	3 (42.9)
3-6 months	12(32.4)	6 (50.0)
6-12 months	5(13.5)	4 (80.0)
1 year and above	3(8.1)	3 (100.0)

Table 3: Histology of Chest Wall Tumours.

Benign	Lipoma	6	43.2%
	Osteochondroma	1	
	Teratoma	5	
	Neurofibroma	4	
Malignant	Rhabdomyosarcoma	10	56.8%
	Ewing's tumour	3	
	Dermatofibrosarcoma	4	
	Neurofibrosarcoma	3	
	Neuroblastoma (metastatic)	1	

Table 4: Treatments offered.

Treatment	Patients
Simple excision + simple skin closure	11
Simple excision + flap wound closure	8
Wide excision + flap closure + adjuvant chemotherapy	8
Neo adjuvant chemotherapy + wide excision + Chemotherapy	6
Chemotherapy only	4

Table 5: Outcome on one year follow up.

Outcome	Benign (n=16)	Malignant (n=21)
Disease free	14 (87.5%)	9 (42.9%)
Partial response	-	3 (14.2%)
Recurrence	2 (12.5%)	3 (14.2%)
Mortality	-	6 (28.6%)

Figure 1: Rhabdomyosarcoma of the anterior chest wall in a toddler.

DISCUSSION

A wide variety of chest wall tumours (CWT) are seen in children in our practice. The disease process, symptomatology, treatment, and outcomes are also a wide range. Delay in presentation is still a problem in the care of these children. The long duration of some of the cases which turned out to be malignant suggests that they may have been initially benign (or malignant but less aggressive) but later became more aggressive (or transformed to malignancy). Hence, the duration of the swelling is not a helpful feature in distinguishing benign from malignant tumours in this series. This difficulty was also noted by other authors,[5,6] None of the patients in our series was diagnosed prenatally, however one of our patients who was seen at one week of age, was born with the chest mass, which was later confirmed to be a teratoma. A literature search also shows that congenital CWT are rare and so prenatal diagnosis is rare.[7] In this series, only 45.9% presented within 3 months of the onset of the swelling. Almost 50% of patients had received prior intervention from unorthodox sources. The impacts of such interventions by untrained hands on these patients included false reassurance, delay or failure of presentation to trained hands and orthodox medicine. The negative role of charlatans in the care of children with surgical conditions in our environment had been reported by Okoro et al.[8] Hence many of the patients were seen when the mass was already extensive and requiring flap rotation after excision. Most of the CWT in our study arose from soft tissues. This is in keeping with the study by Burt but at variance with another study which saw more of the CWT arising from the bones and cartilages.[9,10] The histological diagnosis of our patients tumours depended on the results of excision/ incision biopsies in most of the patients. FNAC was only used in the minority of cases. This is contrary to what was reported by Goyal et al who used FNAC as the bastion for their diagnosis.[-] In our practice, we preferred the open biopsy technique which gave the histological diagnosis, and often resulted in complete or partial removal of the mass. Many other authors have also reported preference for open biopsy or trucut biopsy rather than FNAC.[5] In addition, many of the malignant ones required neo adjuvant chemotherapy or were considered inoperable. The fact that more than 42% of all chest wall tumours encountered in this series were malignant, is a pointer to the fact that childhood chest wall swellings should always be treated every seriousness and urgency to reduce the morbidity and or mortality that delay can cause. The important role of contemporary imaging modalities of Magnetic Resonance Imaging and CT scan in diagnosing and staging these CWT have been reported. Imaging characteristics and features have enabled distinguishing of one histological type from another.[12,13,14] Paucity of these imaging techniques and the requisite manpower are among the challenges encountered in our region. The most common imaging we used for these patients is plain chest radiograph. Whereas, some other studies reported Ewing's sarcoma to be the commonest CWT, our series points to rhabdomyosarcoma as the commonest malignant chest wall tumour.[15,16,17] The small size of our series and the relatively short follow up period makes it difficult to objectively compare the outcome of our management of CWT with some other similar studies from other parts of the globe. However within the period of follow up in this study, only 42.8% of patients with malignant disease were disease free. This result is poorer than the results from other reports despite longer follow up in those reports.[19] This lower cure rate is related to

the lateness of presentation and the pathological type of CWT encountered in this series. Whereas stage of the tumour and the histological type were the major prognostic indicators in our series, studies elsewhere identified the tumour grade and differentiation as the major indicators.[18,19] The presence of metastatic deposits was also a major prognostic indicator in our series and this is in line with reports from other centres.[4]

CONCLUSIONS

Chest wall tumours are therefore, among the surgical problems encountered in children in our region. These CWT patients are often presented late and a significant percentage of the CWT are malignant. Proper evaluation is often impeded by paucity of contemporary imaging tools. Surgical removal is often tasking due to direct spread of the malignant lesions into the chest cavity and underlying viscera and difficulty of skin closure following excision even in benign tumours. Efforts should be directed at more public enlightenment and to cub the activities of unorthodox medicine practitioners who often delay patients' presentation to the rightful attending specialists. Early presentation and treatment are key to improved outcome of treatment in children with chest wall tumours.

REFERENCES

1. Penfield FL, Somers J, Templeton AC. Chest wall tumors. Curr Probl Surg, 1995; 8: 663-747.
2. Weisenburger TH, Roth JA, Ruckdeschel JC. Overview of malignancies of the chest wall and pleura. 2nd ed., Philadelphia, PAW.B. Saunders, 1995.
3. Athanassiadi K, Kalavrouziotis G, Rondogianni D, Loutsidis A, Hatzimichalis A, Bellenis I Primary chest wall tumors: early and long-term results of surgical treatment. Eur J Cardiothorac Surg, 2001; 19(5): 589-93.
4. Dang NC, Siegel SE, Phillips JD. Malignant chest wall tumors in children and young adults. J Pediatr Surg, 1999; 34(12): 1773-78. https://doi.org/10.1016/S0022-3468(99)90310.
5. Sabanathan S, Shah R, Mearns AJ. Surgical treatment of primary malignant chest wall tumors, Eur J Cardio-thorac Surg, 1999; 11: 1011-1016.
6. Ala-Kulju K, Ketonen P, Jarvinen A, Salo J, Luosto R. Primary tumours of the ribs. Scand J Thorac Cardiovasc Surg, 1988; 22: 97-100.
7. Jozaghi Y, Emil S, Albuquerque P, Klam S, Blumenkrantz M. Prenatal and postnatal features of mesenchymal hamartoma of the chest wall: case report and literature review. Pediatr Surg Int., 2013; 29(7): 735-40.
8. Okoro PE, Anyaeze CM, Gbobo I. Exposure of children to surgical charlatans in rural Nigerian communities. Nig J Surg Sci, 2010; 20: 22-29.
9. Burt M. Primary malignant tumors of the chest wall: the Memorial Sloan-Kettering Cancer Center experience. Chest Surg Clin N Am 1994; 49: 363-369.
10. Pass HI, Roth JA, Ruckdeschel JC, Weisenburger TH. Primary and metastatic chest wall tumors Thoracic oncology. 2nd ed., Philadelphia, PA W.B. Saunders:1995.
11. Goyal S, Sharma S, Bahl N. Role of FNAC in palpable chest wall lesions in developing country. Diagn Cytopathol, 2014; 42(8): 653-9. doi: 10.1002/dc.23101.
12. Winer-Muram HT, Kauffman WM, Gronemeyer SA, Jennings SG. Primitive neuroectodermal tumors of the chest wall (Askin tumors): CT and MR findings. Am J Roentgen, 1993; 161: 265-268. 10.2214/ajr.161.2.8392786.
13. Martini N, Huvos AG, Burt ME, Heelan RT, Bains MS, McCormack PM, et al. Predictors of survival in malignant tumors of the sternum J Thorac Cardiovasc Surg, 1996; 111: 96-106.
14. Anderson BO, Burt ME. Chest wall neoplasms and their management. Ann Thorac Surg, 1994; 58: 1774-81.
15. Shamberger RC , Grier HE. Chest wall tumors in infants and children. Semin Pediatr Surg, 1994; 3(4): 267-76.
16. Franken Jr EA, Smith JA, Smith WL. Tumors of the chest wall in infants and children. Paediatr Radiol, 1977; 6(1): 13-18.
17. Denbo JW, Shannon OW, Wu Y, Wu J, Billups CA, Navid F et al. Timing of surgery and the role of adjuvant radiotherapy in ewing sarcoma of the chest wall: a single-institution experience. Ann Surg Oncol, 2012; 19(12): 3809-15.
18. Greager JA, Patel MK, Briele HA, Walker MJ, Wood DK, Das Gupta TK. Soft tissue sarcomas of the adult thoracic wall. Cancer, 1987; 59: 370-373.
19. Gordon MS, Haidu SI, Bains MS, Burt ME. Soft tissue sarcomas of the chest wall: results of surgical resection. J Thorac Cardiovasc Surg, 1991; 101: 843-854.