The Egyptian Journal of Hospital Medicine (April 2023) Vol. 91, Page 3807-3812

Clinicopathological Study and Management of Primary Malignant Scapular Tumors; 10 Years National Cancer Institute Experience

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ABSTRACT

Introduction: Primary malignant scapular tumors are very rare. Little is written about flat bone sarcomas in the literature, and not much is known about the oncological outcome. **Objective:** The aim of the current work is reviewing clinicopathological features and surgical management of cases with primary malignant scapular tumors.

Patients and methods: Patients with primary malignant scapular tumours who visited the National Cancer Institute (NCI) at Cairo University, Egypt, between January 2009 and December 2019 were subjected to a retrospective descriptive analysis.

Results: The study included 25 patients with a mean age of 33.4 years old ranging from 9 to 86 years. Pathology types were chondrosarcoma in 14 patients (56%), Ewing/PNET in 7 patients (28%), osteosarcoma in 2 patients (8%), spindle cell sarcoma in 1 patient (4%), and aneurysmal bone cyst in 1 patient (4%). Twenty patients (80%) had surgery, and 5 patients(20%) had no surgical intervention for being either with a poor general condition or having metastatic disease. Overall survival at 6 months was 92% and was 88%, 75.4%, and 65.4% in the first year, 3rd year, and 5th year respectively. Disease-free survival at 6 months was 90.5%; and was 85.7%, 75.4%, and 42.7% in the first year, 3rd year, and 5th year respectively.

Conclusion: Primary malignant scapular tumors are rare. Early diagnosis is very important, as surgical excision with a wide negative margin (limb-sparing surgery) is the main line of treatment in most cases and tends to decrease the recurrence rate, otherwise, palliative treatment and forequarter amputation may be indicated.

Keywords: Scapular tumors, Scapulectomy, Limb-sparing surgery, Retrospective descriptive analysis, National Cancer Institute, Cairo University.

INTRODUCTION

Although the shoulder girdle (proximal humerus, scapula, and clavicle) is the third most frequent location for tumours of bone and soft tissues, malignant tumours of the scapula are uncommon [1,2]. Children and adolescents with Ewing's sarcoma have the most common malignancies affecting the scapula [2].

Before being identified, scapular tumours frequently become enormously large. As the disease progresses, the tumour may also spread to the chest wall, the proximal humerus, the rotator cuff, or the neurovascular bundle [3]. Limb-sparing resections for malignant tumours of the scapula were regarded as high-risk procedures due to the probable danger of local recurrence. So these malignancies frequently required a full or partial scapulectomy [4]. Syme conducted and documented the first total scapulectomy in 1856 [5]. Since then, other updates and suggestions have appeared in various series [1-3]. We aimed to assess primary malignant scapular tumors regarding clinicopathological features, surgical management, and oncological outcome (disease-free survival, and overall survival).

PATIENTS AND METHODS

This is a retrospective descriptive study involving all patients with primary malignant or borderline scapular tumors who received treatment at National Cancer Institute (NCI), Cairo University over 10 years from January 2009 to December 2019. We excluded those with benign tumors, metastatic tumors to the scapula from another primary, or soft tissue sarcomas invading the scapula.We collected patients' files and obtained the following data: age, sex, preoperative pathology, lines of treatment (neoadjuvant chemoradiation, adjuvant chemoradiation, surgery, and palliative treatment), surgical management data (types of surgical resection, postoperative complications, and postoperative pathological data (Disease-free survival (DFS), and overallsurvival (OS).

Malignant tumor staging was according to Enneking staging [6]. Classification of the type of surgical resection was based on what has been described by Malawer and colleagues [7].

Ethical Approval:

Patients received all the information they require regarding the research objectives and methodology after the study was given the go-ahead by Cairo University's Ethics Committee. Each study participant provided their written consent after receiving full information. The Declaration of Helsinki, the code of ethics of the World Medical Association, was followed when conducting this research on humans.

Statistical Analysis

IBM SPSS advanced statistics (Statistical Package for Social Sciences), version 24, was used to examine the data (SPSS Inc., Chicago, IL). Quantitative data were described as number and percentage, whereas numerical data were described as median and range or mean and standard deviation (SD), as appropriate. The suitable

Received: 09/11/2023
Accepted: 10/01/2023

method for examining the relationship between qualitative variables was the chi-square (Fisher's exact) test. By computing the hazard ratio and its 95% confidence interval, logistic regression analysis was used to quantify risk and eliminate the impact of confounding. The Kaplan-Meier method was used to do a survival study. Using the log-rank test, two survival curves were compared. By computing the hazard ratio and its 95% confidence interval, the Cox regression model underwent multivariate analysis to examine the independent prognostic impact of statistically significant factors on a univariate level. From the date of diagnosis until the date of death or last follow-up, overall survival (OS) was calculated. From the time of complete remission to the time of death, recurrence, or last follow-up, disease survival (DFS) was determined.

RESULTS

The study included 25 patients. The mean age of the patients was 33.4 (SD 17.4) years old (ranging from 9 to 86 years). Patients' and tumors' characteristics are illustrated in **Table 1**. Preoperative biopsy was done for all cases to include primary malignant cases, and exclude scapular secondaries, and benign cases. One case showed to be an aneurysmal bone cyst (borderline tumor), and it was included in our study, however, its postoperative pathology was osteosarcoma. Other pathological details are shown in *Table 1*.

Surgical approach and type of resection: Twenty patients out of 25 underwent surgical resection whether upfront (14 cases) or post-neoadjuvant (6 cases). The main approach for scapular resection is utilitarian shoulder girdle resection via posterior approach in 19 patients and dual approach (posterior plus anterior incisions) in one patient.

Postoperative pathology and surgical margins: Eighteen patients (90%) had negative margins while 2 patients (10%) had positive margins. Those were high-grade chondrosarcoma, one of them had positive bony and soft tissue margin, and the other case had only positive soft tissue margin.

Adjuvant treatment: Seven patients (35%) received adjuvant Chemotherapy (3 patient's osteosarcoma and 4 patients Ewing/PNET), 3 patients (15%) received adjuvant chemoradiotherapy (two cases were high-grade chondrosarcoma with positive margins and one case was high-grade osteosarcoma with close margin).

Recurrence: Twenty patients out of the 25 underwent surgical resection and, one patient did not do surgery due to elevated liver enzymes before the scheduled day of surgery and received definitive chemoradiotherapy with near complete response. So we have 21 patients who received definitive treatment. Three patients (14.2%) out of 21 patients who received definitive treatment had local recurrence and 5 patients (23.8%) developed distant metastases. Of the three patients who developed local recurrence, two patients had high-grade chondrosarcoma and underwent surgical resection, and one patient with high-grade osteosarcoma developed irresectable local recurrence and received chemoradiotherapy.

Table 1: Patients' and tumors' characteristics.

Characteristic	Number	Percentage %
Gender:		
Male	14	56%
Female	11	44%
Complaint:		
Pain	20	80%
Swelling	5	20%
Initially metastatic:		
Yes (only to lungs)	4	16%
No	21	84%
Tumor location in the scapula:	23	92%
Body	1	4%
Coracoid process	1	4%
Glenoid + Acromion		
Preoperative pathology:		
Chondrosarcoma	14	56%
Ewing/PNET	7	28%
Osteosarcoma	2	8%
Spindle cell sarcoma	1	4%
Aneurysmal bone cyst	1	4%
Treatment modality:		
Upfront surgery	14	56%
Neoadjuvant treatment then surgery	6	24%
Definitive chemoradiotherapy	1	4%
Palliation	4	16%
Disease staging:		
Stage I	10	40%
Stage II	11	44%
Stage III	4	16%
Type of surgical resection:		
Subtotal scapulectomy (IIA)	13	65%
Total scapulectomy (IIIB)	6	30%
Tikhoff-Lineberg Resection (IVB)	1	5%
Postoperative pathology:		
Chondrosarcoma	13	65%
Ewing/PNET	4	20%
Osteosarcoma	3	15%
Surgical margins:		
Negative	18	90%
Positive	2	10%
Postoperative complications:	3	15%
Seroma	2	10%
Wound infection	15	75%
No complications		
Recurrence / Metastasis:		
No	13	61.9%
Local recurrence	3	14.2%
Distant metastasis	5	23.8%

Of the five patients who developed distant lung metastases, one patient underwent right lung metastasectomy, and the other four received palliative chemotherapy.

Overall survival: Overall survival for all patients at 6 months was 92% and was 88%, 75.4%, and 65.4% in the first year, 3rd year, and 5th year respectively. For stage I patients overall survival at 6 months, first year, and 3rd year was 100% and decreased to 87% at 5th year. For stage II patients overall survival at 6 months was 90.9% and was 81.8%, 72.7%, and 62.3% in the first year, 3rd year, and 5th year respectively. For stage III patients overall survival at 6 months was 75% and was 50%, in the first year as shown in ***Figure 1***.

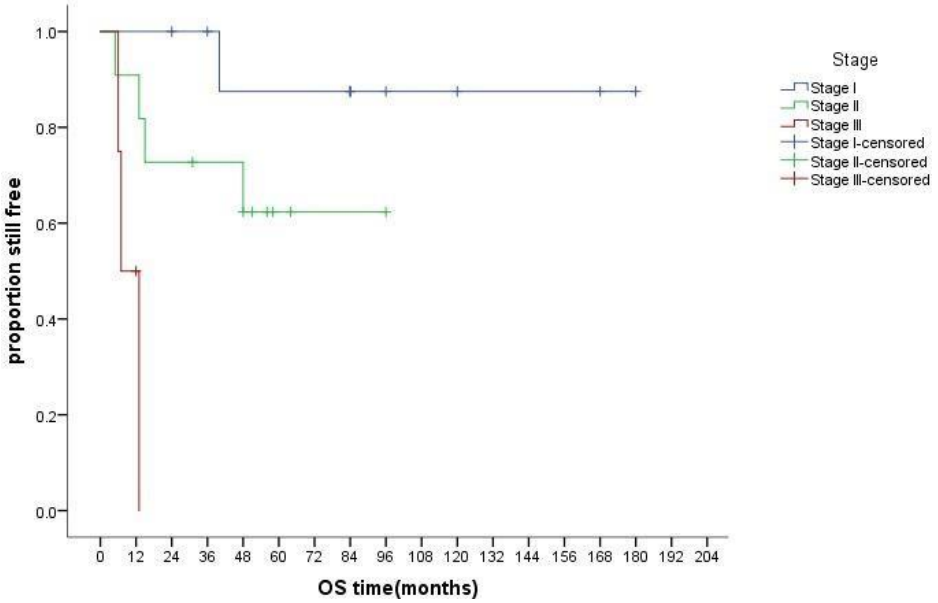

Figure 1: Overall Survival (OS)

Disease-free survival (DFS): Disease-free survival of the whole cohort in the first 6 months was 90.5%; and was 85.7%, 75.4%, and 42.7% in the first year, 3rd year, and 5th year respectively. For stage I patient's disease-free survival at 6 months, first year and 3rd year was 100% and decreased to 72% in 5th year. For stage II, and III patients disease-free survival at 6 months was 80% and was 70%, 60%, and 20% in the first year, 3rd year, and 5th year respectively as shown in ***Figure 2***.

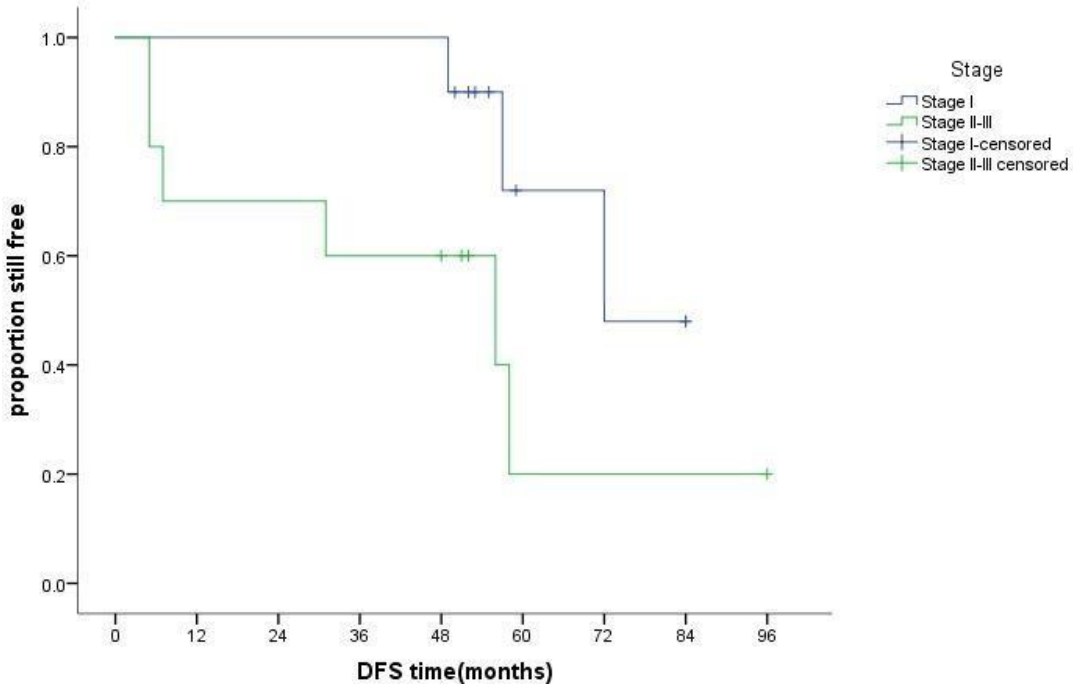

Figure 2: Disease-Free Survival (DFS)

DISCUSSION

Prior to 1970, many patients with high-grade sarcomas around the scapula nearly always required amputation of the forequarter [8]. The Tikhoff-Linberg treatment was promoted as having tumour control and survival rates comparable to amputation in the late 1970s [9]. Following this, limb-saving surgery for these malignancies has become the norm [9].

When Linberg first reported the interscapulothoracic excision in 1928, the proximal humerus was only briefly secured to the clavicle with sutures or wires, leaving the suspended humerus relatively stable. However, most of the patients had a flail joint with a limited range of motion and dismal cosmetic outcomes [4].

The traditional resection involved taking off the entire scapula, clavicle, and proximal humerus in one piece. A significant change involved removing the lateral section of the scapula and then prosthetically reattaching the proximal humerus to the remaining scapula or the thoracic wall [4]. The range of available treatments for bone cancers near the scapula has now been significantly expanded thanks to new imaging techniques and adjuvant therapies. However, conserving the glenohumeral joint remains the key aspect in achieving a satisfactory functional outcome, as only resection of the scapular wing can be satisfactorily replaced by muscular and fascial reconstruction [10].

After limb-saving resections, several techniques for scapular reconstruction have developed over time, including humeral suspension, allograft transplantation, arthrodesis, and prosthetic implants. With the exception of prosthetic implants, none of them have offered adequate function and stability; we owe this to 3D printing technology [2]. A clinically practical scapular prosthesis emerged as a result of endoprosthetic reconstructive experience at other sites and current, novel prosthetic design. As a result, replacing the scapula surgically with a modular or custom-made endoprosthesis has emerged as a viable option and, in some situations, shows promising outcomes in terms of enhancing glenohumeral stability. It has been established that endoprosthetic reconstruction offers good oncological results and functionality [10].

A biological method of reconstruction has also arisen, that is the extracorporeal irradiation and reimplantation of the scapula after tumor removal. It has shown good long-term functional and oncological outcomes [11].

Our institute (The National Cancer Institute-Cairo University, Egypt.) is a highly specialized tertiary center in oncology. It received 43 cases diagnosed as scapular neoplasms (25 were primary malignant scapular tumors, 16 cases were metastases from another primary, and two cases were benign osteochondroma). Our study included the primary malignant and borderline cases only (25 patients), of whom 20 patients underwent scapular resection.

The Mean age of our patients was 33.4years (9 - 86) with male predominance by 14 male patients (56%), and 11 female patients (44%). Similarly, in a study conducted by **Kaiser *et al.*** [12] male predominance was reported among the included 74 patients, 46 (62.1%) were males and 28 (37.8%) were females.

The commonest presenting symptom was upper back (shoulder pain), and swelling was much less common in most series as **Kaiser *et al.*** [12] 67 (90.5%) patients presented with pain while only 7 (9.5%) patients presented with swelling. This is comparable to what we had in our study, where we had 20 (80%) patients complained of upper back (shoulder) pain while 5 (20%) patients presented with upper back swelling (20%).

Chondrosarcoma is the most prevalent primary malignant tumor of the scapula, constituting 32% of all malignant lesions in a study [13], and 45% of cases in another study [14]. Similarly, in our study, we found that chondrosarcoma was the commonest pathological type in 13 (52%) patients.

The body of the scapula is a commonly involved site in the scapula. In our study, We had 23 (92%) patients presented to us with scapular lesions at the body of the scapula, 1 (4%) patient presented with a lesion at the coracoid process, and another 1 (4%) patient affected the glenoid/acromion. similarly, in a study of 74 patients conducted by **Kaiser *et al.*** **[12]**, they revealed scapular body lesions in 24 (32.43%) patients, 10 (13.5%) patients at glenoid, and 3 (4%) patients at the acromion.

The surgical resection technique varied between researches. For instance, 24 patients had limb-sparing resection, according to Puchner et al. [3] report. Type IIA subtotal scapular resection was performed on 7 (29.16%) patients. A whole scapula resection of type IIIB was performed on 8 (33.3%) patients, along with the removal of the majority of the abductor muscles.

Type IVB interscapulothoracic resection was performed on 7 (29.16%) individuals (Tikhoff-Linberg). Two (8.3%) patients underwent a total scapulectomy of type IIIA. In our study, only 1 (5%) patient had type IVB resection, while 13 (65%) patients received type IIA resection, 6 (30%) patients received type IIIB resection, and 13 (65%) patients received type IIA resection.

Puchner et al. [3] wide resection margins were attained in 18 (74%) patients, marginal margins in 3 (13%) patients, and intralesional margins in 3 (13%) patients.

In our analysis, 2 (10%) patients had positive margins, while 18 (90%) patients had large resections with negative margins. One patient had issues with wound healing and one patient had a dislocation following scapular endoprostheses and shoulder suspensions, according to Tang's study on 10 patients. [10]. In another study conducted by **Puchner *et al.*** **[3]** 24

patients had limb-sparing surgery, and 10 (41.7%) cases had complications. Of those 6 patients had wound infection (2 required surgical debridement, and 4 resolved by antibiotics and repeated dressings only), one patient had prosthesis dislocation, one had thrombosis, and 2 had radial nerve impairment was needed, and the other 4 patients **(3)**. In our study, we had only 5 (25%) patients with wound healing problems recovered smoothly by antibiotics and repeated dressings. According to a series of studies on adjuvant therapy, 14 (58.3%) patients underwent chemotherapy in accordance with the actual protocols specified by the underlying disease, while 13 (54.2%) patients underwent irradiation therapy, and six patients underwent both [(3)].

In our analysis, only 3 (15%) patients had adjuvant chemoradiotherapy while 7 (35%) patients received adjuvant chemotherapy (3 osteosarcoma patients and 4 Ewing/PNET patients) (two cases were high-grade chondrosarcoma with positive margins and one case was high-grade osteosarcoma with close margin). Our study's local recurrence rate of 14.28% was similar to that reported by Puchner et al. [(3)] who recorded a recurrence rate of 13%. A rate of 24% for distant metastases was reported by Pant et al. [(15)] in a different study, Puchner et al. [(3)] discovered that at the time of admission, three patients without surgical therapy and six out of 21 (26%) patients with localised illness had distant metastases [(3)].

Out of 21 patients in our study with localised disease who underwent surgical therapy, 5 (23.8%) patients later acquired distant metastases, and four patients initially reported to us with such metastases. Concerningly, there is a significant risk of metastases following scapula excision.

After all, rather than the surgical method, it reflects the biology of the disease [(16)]. Puchner et al. [(3)] examined the oncological outcomes of 14 (48%) patients who died from their main disease (ten of whom underwent surgery), 10 (34%) patients who were alive but had no signs of the disease, and 1 (4%) patient, who was alive but had the disease. Overall survival for all 29 patients was 70% at one year and 47% at five years, according to their findings. In our study, we discovered that while 12 (48%) patients were alive with no recurrence, 8 (32%) patients died of their primary disease (three of them underwent surgical resection then experienced recurrence, four patients were metastatic at the time of diagnosis, and one patient received definitive chemoradiotherapy without surgery). Due to a lack of information, it was impossible to determine if 5 further patients were still alive.

As described by **Puchner *et al.*** [(3)] disease-free survival of all 29 patients was 91 % after one year and 83 % after five years by **Puchner *et al.* (3)**. In our study, disease-free survival of the 25 patients in the first 6 months was 90.5% and was 85.7%,75.4%, and 42.7% in the first year, 3rd year, and 5th year respectively.

Puchner *et al.* [(3)] found that the overall survival of their 29 patients was 70 % after one year and 47 % after five years [(6)]. In our study, overall survival at 6 months was 92% and was 88%, 75.4%, and 65.4% in the first year, 3rd year, and 5th year respectively.

CONCLUSION

Primary malignant scapular tumors are rare. Early diagnosis is very important, as surgical excision with a wide negative margin (limb-sparing surgery) is the main line of treatment in most cases, otherwise, palliative treatment and forequarter amputation may be indicated. Postoperative rehabilitation is mandatory to return to an independent level of function and regain partial independence in daily activities with improved quality of life.

Acknowledgment: Great thanks and warmest gratitude to all surgery department members in our institute who helped and provided us with all that we needed to work on the study cases effectively.

Author Contributions: All authors contributed equally to the different steps of our study: data collection and analysis, literature research, statistical analysis, manuscript conceptualization, preparation, reviewing, and editing.

Data Availability: The datasets used +and/or analyzed during the current study are available from the corresponding author upon reasonable request.

Conflicts of Interest: No conflicts of interest.

Funding Support: No fund was obtained.

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