Journal of Taibah University Medical Sciences (2021) 16(1), 77–85

Taibah University

Journal of Taibah University Medical Sciences

www.sciencedirect.com

Original Article

Primary bone sarcomas in KSA: A Saudi tumor registry review

Wazzan S. Aljuhani, MD [a], Abdullah M. Alanazi, MBBS [b] and Mohammad A. Alghafees, MBBS [b,*]

[a] *Department of Orthopedic Surgery, National Guard Hospital, Riyadh, KSA*
[b] *College of Medicine, King Saud bin Abdulaziz University for Health Sciences, Riyadh, KSA*

Received 29 June 2020; revised 30 October 2020; accepted 4 November 2020; Available online 1 December 2020

الملخص

أهداف البحث: يكشف لنا معدل الانتشار الجغرافي للأورام تأثير البيئة والعرق والمستوى الثقافي على السرطانات. وتهدف هذه الدراسة على سرطان العظام إلى تحديد معدل الحدوث والاختلافات في نمط الورم، وموقع نشأته، ومعدل وفياته باختلاف الجنس، والمناطق، والعمر، والاختلاف في الخصائص بين أنماط الورم.

طرق البحث: شملت هذه الدراسة المقارنة الارتجاعية جميع المرضى الذين تم تشخيص إصابتهم بسرطانات عظام أولية بدءا من ١ كانون الثاني/يناير ٢٠١٣ وحتى ٣١ كانون الأول/ديسمبر ٢٠١٧. وتم توليد التكرارات والنسب المئوية حسب المتغيرات الفئوية. تم حساب الوسط الحسابي، والانحراف المعياري للمتغيرات الكمّيّة. وتم حساب معدل الحدوث لكل ورم.

النتائج: من بين ٤٥١ مريضا وُجد أن ٢٤٨ مريضا (٥٥٪) يعانون من سرطانات عظمية، وأن ١٦٠ مريضا (٣٥.٥٪) يعانون من سرطان أيوينغ، وأن ٤٣ مريضا (٩.٥٪) يعانون من سرطانات غضروفية. وكان معدل الإصابة هو ١.٥٦ حالة من كل مليون شخص سنويا في السرطانات العظمية، و٠.٩٥ حالة من كل مليون شخص سنويا في سرطان أيوينغ، و٠.٢٧ حالة من كل مليون شخص سنويا في السرطانات الغضروفية. وكان معدل البقيا لمدة ٣ سنوات حوالي ٨٢.٣٪. تم تحديد اختلافات ملحوظة في نمط الورم، وموقع نشأته، ومعدل البقيا لـ٣ سنوات باختلاف العمر والجنس. كما تم أيضا تحديد وجود اختلافات ملحوظة في موقع نشوء الورم، ودرجته، وأساس تشخيصه، وموضعه في أحد الجانبين باختلاف أنماط الأورام.

الاستنتاجات: لقد استنتجنا من المنشورات الطبية أن نسب حدوث سرطان العظام الملحوظة كانت أقل بكثير مما تم تحديده عالميا. وإذا فهمنا النمط السلوكي للورم في المنطقة فإننا نستطيع تطوير إستراتيجيات معالجة تعتمد على الخطورة وعلى الاستجابة، وتسمح باتخاذ القرارات بشكل باكر.

الكلمات المفتاحية: سرطان العظام؛ الوبائية؛ معدل الحدوث؛ علم الأورام؛ المملكة العربية السعودية

Abstract

Objectives: The geographical incidence of tumours is usually influenced by the environment, race, and culture. This study aimed to report the incidence and differences in tumour type, site of origin, and mortality across gender, regions, age, and the different characteristics of tumour types.

Methods: This retrospective cohort study included all patients diagnosed with primary bone sarcomas from January 1, 2013, to December 31, 2017. Frequencies and percentages were generated for categorical variables. Means and standard deviations were calculated for quantitative variables. A chi-squared test was used to detect differences among categorical variables. Student-t, ANOVA, and Tukey tests were used to detect differences among quantitative variables. Lastly, we calculated the incidence of each tumour type.

Results: Of 451 patients, 248 (55%) had osteosarcomas; 160 (35.5%) had Ewing's sarcoma, and 43 (9.5%) had chondrosarcoma. The incidence was 1.56 cases per 1,000,000 per year for osteosarcoma, 0.95 cases per 1,000,000 per year for Ewing's sarcoma, and 0.27 cases per million per year for chondrosarcoma. The three-year survival rate was 82.30%. Significant differences in tumour type, origin site, and three-year survival across age and gender were detected. Similarly, significant differences were also noted in origin site, grade, basis of diagnosis, and lateralisation across tumour types.

Conclusions: In our study, the observed bone sarcoma incidence rates were lower than the ones reported

* Corresponding address: College of Medicine, King Saud bin Abdulaziz University for Health Sciences, Uzzam, AlManar, Riyadh 14222, KSA.
E-mail: Alghafees687@gmail.com (M.A. Alghafees)
Peer review under responsibility of Taibah University.

Production and hosting by Elsevier

 https://doi.org/10.1016/j.jtumed.2020.11.001

worldwide. Understanding the pattern of tumour behaviour in the region will help develop a risk and response-based treatment plan for early decision-making.

Keywords: Bone sarcoma; Epidemiology; Incidence; KSA; Oncology

Introduction

A sarcoma is a heterogeneous group of neoplasms that arises form mesenchymal cells and can affect any anatomical site, whether it is soft tissue or bone. Primary bone sarcomas have a variety of histologic subtypes, mainly osteosarcoma, chondrosarcoma, Ewing's sarcoma, and other less common bone sarcomas.

Bone Sarcomas are among the rarest malignancies that affect humans. According to the American Cancer Society, primary bone cancers account for less than 0.2% of all cancers. Osteosarcoma is the most common subtype, accounting for almost 35% of the bone sarcoma cases, followed by chondrosarcoma (25%) and Ewing's sarcoma (16%).[1] In Europe, the 2012 RARECARE Project had collected data from 1995 until 2002 and stated that the incidence of all sarcomas was 5.6 per 100,000 per year, of which soft tissue sarcoma makes up 84% (4.7 per 100,000) and bone sarcoma makes up 15% (0.8 per 100,000).[25]

The anatomical site distribution of each type is quite different. Multiple studies have shown that osteosarcoma shows a profound propensity for the involvement of the long bones of the appendicular skeleton; in particular, the distal femur, proximal tibia, and proximal humerus, while Ewing's sarcoma favoured sites such as ribs, tibia, hip bone, and vertebrae. For chondrosarcoma, pelvic bones (ileum being the most frequent), the femur, the humerus, and the ribs are the most frequent sites for growth.[2–4] In addition, a European study showed that bone sarcomas had a bimodal age pattern (15–24 and > 65) with peaks at the age of 15–24 for osteosarcomas and Ewing's sarcoma, and above 65 for chondrogenic sarcomas.[25]

The geographic incidence of tumours usually uncovers the influence that the environment, race, and culture exert upon the prevalence of cancers. Therefore, due to the absence of local studies, this study of bone sarcomas is presented to highlight the incidence and detect differences based on tumour type, site of origin, and mortality across gender, regions, and age, and differences in the characteristics of tumour types between January 1, 2013, and December 31, 2017. Such studies would help develop selective screening protocols and facilitate treatment strategies and surveillance treatment pathways.

Materials and Methods

Study design and participants

This retrospective cohort study included all the patients that were diagnosed with primary bone sarcomas from January 1, 2013, to December 31, 2017. Patients who were diagnosed with soft tissue sarcomas or metastatic bone sarcomas were excluded from the study.

Data collection method

The data were collected from the Saudi Cancer Registry (SCR). The SCR collects tumour data from all private, military, and health ministry hospitals in KSA through five regional offices, and data analysis and periodic reporting occur at the main office in Riyadh. All of the patients who were diagnosed with bone sarcomas in the specified period except the ones that did not meet the inclusion criteria were enrolled. The included variables were gender, age, patient's region, tumour's site of origin, tumour's histological subtype, tumour's behaviour, tumour's grade, tumour's extent, tumour's laterality, the basis of diagnosis, and survival status.

Statistical analysis

All the data were managed using Microsoft Excel 2019 (Microsoft Ltd., WA, USA) and analysed using SPSS version 23.0 (IBM Corporation, NY, USA). Frequencies and percentages were generated for categorical variables. Means and standard deviations were calculated for quantitative variables. A chi-squared test was used to test the presence of significant differences among categorical variables. T-test and ANOVA test were utilised to identify significant differences among quantitative variables. The Tukey post-hoc test was used to detect where the exact differences lay. The incidence rate was calculated for each type of tumour per million for each year separately and then the average over the five years was calculated and reported. The country's population size for each year was acquired from the National Authority for Statistics in KSA and was used to calculate the incidence rate.

Results

The total incidence of bone sarcomas in KSA between 2013 and 2017 was 451. Figure 1 shows the tumour type-specific incidence for each year. The incidence rate was 7.82 cases per 1,000,000 per five years for osteosarcoma, 4.75 cases per 1,000,000 per five years for Ewing sarcoma, and 1.35 cases per 1,000,000 per five years for chondrosarcoma. The yearly incidence rate was 1.56 cases per 1,000,000 per year for osteosarcoma, 0.95 cases per 1,000,000 per year for Ewing sarcoma, and 0.27 cases per 1,000,000 per year for chondrosarcoma. No significant differences were observed within the incidence rates of tumours across the years.

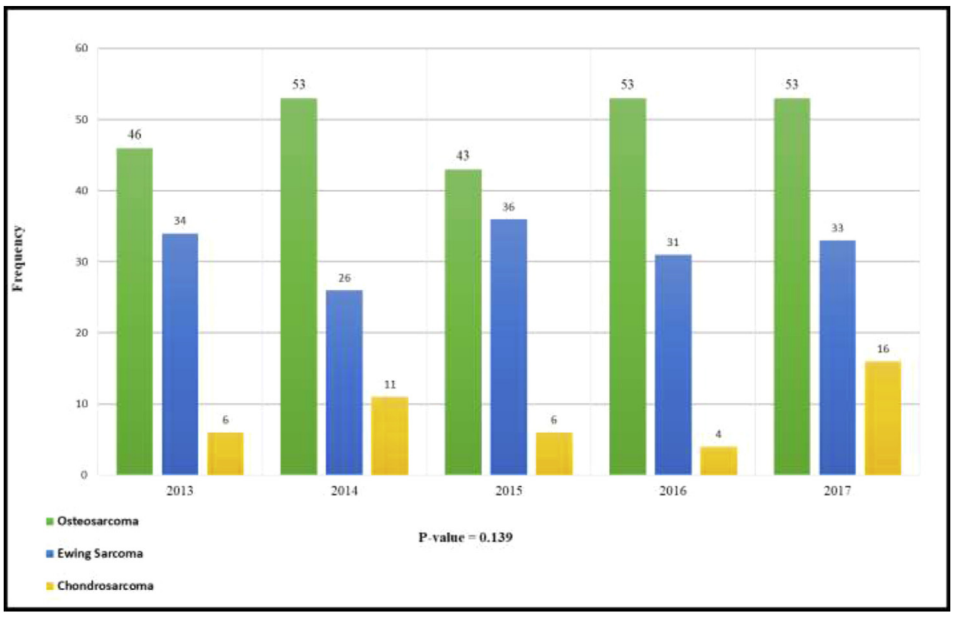

Figure 1: Incidence of bone sarcoma in KSA between 2013 and 2017. * Significant at level ≤ 0.05.

Table 1 shows the demographical data of the patients. The mean age of the patients was 22.34 ± 16.85. Of these, 274 (60.8%) patients were aged 18 years and younger, 53 (11.8%) were aged between 19 and 24 years, 92 (20.4%) were aged between 25 and 59 years, and 32 (7.1%) were aged 60 years and older. Males constituted 256 (56.80%) of the participants and 195 (43.20%) were female. The largest proportion of patients was from the central region 143 (31.70%); the least proportion was from the northern region (39 [8.60%]). Nine (2%) of the patients did not have a registered place of residency. Among the 451 patients, 371 (82.30%) survived for more than three years, and 80 (17.70%) did not.

Table 1: The Demographic Profile of The Participants (n = 451).

Gender	**n**	**%**
Male	256	56.80
Female	195	43.20
Age	**Mean**	**SD**
	22.34	16.85
Age Groups	**n**	**%**
18 and younger	274	60.8
19–24 years	53	11.8
25–59 years	92	20.4
60 and older	32	7.1
Place of Residency	**n**	**%**
Central Region	143	31.70
Eastern Region	67	14.90
Northern Region	39	8.60
Western Region	138	30.60
Southern Region	55	12.20
Unknown Place of Residency	9	2.00
3-year survival	**n**	**%**
Dead	80	17.70
Alive	371	82.30

Table 2 demonstrates the details of bone sarcoma characteristics. From the 451 patients, 248 (55%) had osteosarcomas, 160 (35.5%) had Ewing sarcoma, and 43 (9.5%) had chondrosarcoma. In terms of the origin site of the tumour, long bones of lower limbs were the most common site, with 234 (51.9%), followed by bones of the skull and face and pelvic bones, sacrum, and coccyx, with both categories having a frequency of 52 (11.5%). When assessing the grade of tumours, it was revealed that 265 (58.8%) did not have a record of the grade. The data on tumour degree of extension showed that most of the patients had localised tumours [239 (53%)], and 97 (21.5%) had distant metastasis. The lateralisation state of tumours displayed that 197 (43.70%) had left lateralisation, and 164 (36.40%) had right lateralisation. With regard to the basis of diagnosis, the vast majority was diagnosed based on the histology of the primary tumour 423 (93.8%).

Table 3A compares the type of tumour, site of origin, and three years' survival, across gender. A significant difference ($p = 0.034$) between males and females was present only in the type of tumour. More males were affected by osteosarcoma, and more females were affected by Ewing sarcoma. Table 3B displays the differences between type of tumour, site of origin, and three years' survival among patients from different places of residency; no significant difference was found. Table 3C describes the relationship

Table 2: Tumor Details (n = 451).

	n	%
Type of Tumor		
Osteosarcoma	248	55
Ewing sarcoma	160	35.5
Chondrosarcoma	43	9.5
Origin Site of Tumor		
Long bones of upper limb	55	12.2
Short bones of upper limb	2	0.4
Long bones of lower limb	234	51.9
Short bones of lower limb	13	2.9
Bones of the skull and face	52	11.5
Vertebral column	22	4.9
Rib, Sternum, and Clavicle	18	4
Pelvic bones, Sacrum, and Coccyx	52	11.5
Unknown primary site	3	0.7
Grade		
Grade I	14	3.10
Grade II	25	5.50
Grade III	74	16.40
Grade IV	73	16.20
Unknown	265	58.8
TNM Extension		
In Situ	1	0.2
Localized	239	53
Regional: Direct Extension	67	14.9
Regional: Lymph Node	5	1.1
Regional: Direct Extension & Lymph Node	5	1.1
Distant Metastasis	97	21.5
Unknown	37	8.2
Lateralization		
Not paired	81	18.00
Right	164	36.40
Left	197	43.70
Bilateral Involvement	1	0.20
Paired at site of origin	8	1.8
Bases of diagnosis		
Unknown	14	3.10
Medical Imaging	7	1.60
Cytology and hematology	3	0.70
Histology of metastases	4	0.90
Histology of primary tumor	423	93.8

between age and type of tumour, site of origin, and three years' survival. A significant difference in age was present in these characteristics ($p < 0.001$ in type of tumour, $p = 0.002$ in site of origin, and $p = 0.001$ in three years' survival). The Tukey post-hoc test revealed that in terms of the types of tumour groups, there was a significant difference in the mean age between osteosarcomas and Ewing sarcoma ($p = 0.03$), osteosarcoma and chondrosarcoma ($p < 0.001$), and Ewing sarcoma and chondrosarcoma ($p < 0.001$). As for the difference in age between the different sites of origin, the Tukey post-hoc test revealed a significant difference in the mean age only between the long bones of the lower limbs and bones of the skull and face ($p = 0.01$).

Table 4 demonstrates the differences of characteristics among the types of tumours. A significant difference in the site of origin between the three types of tumours was present ($p < 0.001$). Although in all types, the most common site was the long bones of lower limbs. Ewing sarcomas showed considerably higher rates of occurrence in the vertebral column and pelvic bones, sacrum, and coccyx. Chondrosarcomas have shown considerably higher occurrence in the bones of the skull and face, rib, sternum, and clavicle. A significant difference between the three types was also found in the grade of the tumour ($p < 0.001$). The majority of chondrosarcoma patients had first- and second-grade tumours, while the majority of both osteosarcoma and Ewing sarcoma patients had third- and fourth-grade tumours. A significant difference between the types of tumours was also observed in terms of the base of diagnosis and lateralisation ($p < 0.001$ and $p = 0.014$), respectively. Higher rates of diagnosing chondrosarcoma through medical imaging were observed. The different patterns of lateralisation were present among the three types of tumours. No significant difference was found among the types of tumours in terms of extension or three years' survival.

Table 3: A. Gender-Based Comparison of Type of Tumor, Site of Origin and Mortality. B. Site of Residency Based Comparison of Type of Tumor, Site of Origin and Mortality.C. Age-Based Comparison of Type of Tumor, Site of Origin and Mortality.

Variable	Gender				*P*-Value
	Male (n = 256)		Female (n = 195)		
	n	%	n	%	
Type of Tumor					
Osteosarcoma	130	51.20	118	60.80	0.034 *
Ewing sarcoma	102	40.20	55	28.40	
Chondrosarcoma	22	8.70	21	10.80	
Origin Site of Tumor					
Long bones of upper limb	28	11.00	27	13.90	0.068
Short bones of upper limb	2	0.80	0	0.00	
Long bones of lower limb	126	49.60	108	55.70	
Short bones of lower limb	8	3.10	5	2.60	
Bones of skull and face	28	11.00	24	12.40	
Vertebral column	19	7.50	3	1.50	
Rib, Sternum, and Clavicle	9	3.50	9	4.60	
Pelvic bones, Sacrum, and Coccyx	34	13.40	18	9.30	
3 Year Survival					
Dead	42	16.50	37	19.10	0.485
Alive	212	19.10	157	80.90	

Variable	Cite of Residency										*P*-Value
	Central Region (n = 143)		Eastern Region (n = 67)		Northern Region (n = 39)		Western Region (n = 138)		Southern Region (n = 55)		
	n	%	n	%	n	%	n	%	n	%	
Type of Tumor											
Osteosarcoma	83	58.00	31	47.00	20	51.30	83	60.10	25	47.20	0.554
Ewing sarcoma	48	33.60	27	40.90	16	41.00	44	31.90	20	37.70	
Chondrosarcoma	12	8.40	8	7.70	3	7.70	11	8.00	8	15.10	
Origin Site of Tumor											
Long bones of upper limb	17	11.90	8	12.10	4	10.30	10	7.20	14	26.40	0.154
Short bones of upper limb	2	1.40	0	0.00	0	0.00	0	0.00	0	0.00	
Long bones of lower limb	78	54.50	28	42.40	21	53.80	82	59.40	19	35.80	
Short bones of lower limb	4	2.80	5	7.60	1	2.60	2	1.40	1	1.90	
Bones of skull and face	14	9.80	11	16.70	5	12.80	15	10.90	7	13.20	
Vertebral column	6	4.20	4	6.10	2	5.10	8	5.80	2	3.80	
Rib, Sternum, and Clavicle	8	5.60	3	4.50	0	0.00	6	4.30	1	1.90	
Pelvic bones, Sacrum, and Coccyx	14	9.80	7	10.60	6	5.40	15	10.90	9	17.00	
3 Year Survival											
Dead	33	23.10	11	16.70	8	20.50	21	15.20	3	5.70	0.059
Alive	110	76.90	55	83.30	31	79.50	117	84.80	50	94.30	

Variable	Age		*P*-Value
	Mean	SD	
Type of Tumor			
Osteosarcoma	21.3	15.70	< 0.001 *
Ewing sarcoma	17.46	12.14	
Chondrosarcoma	46.51	18.38	
Origin Site of Tumor			
Long bones of upper limb	20.35	15.05	0.002 *
Short bones of upper limb	12.5	12.02	
Long bones of lower limb	20.35	15.96	
Short bones of lower limb	17.31	7.53	
Bones of skull and face	29.38	19.31	
Vertebral column	27	21.93	
Rib, Sternum, and Clavicle	31.72	24.62	
Pelvic bones, Sacrum, and Coccyx	23.25	13.47	
3 Year Survival			
Dead	28.25	21.70	0.001 *
Alive	21.07	15.35	

* Significant at level ≤ 0.05.

Table 4: Tumor Type Based Comparison of Tumor Characteristics.

Variable	Type of Tumor						*P*-Value
	Osteosarcoma (n = 248)		Ewing Sarcoma (n = 160)		Chondrosarcoma (n = 43)		
	n	%	n	%	n	%	
Origin Site of Tumor							
Long bones of upper limb	26	10.50	23	14.60	6	14.00	< 0.001*
Short bones of upper limb	1	0.40	1	0.60	0	0.00	
Long bones of lower limb	165	66.50	56	35.70	13	30.20	
Short bones of lower limb	5	2.00	7	4.50	1	2.30	
Bones of skull and face	28	11.30	15	9.60	9	20.90	
Vertebral column	5	2.00	15	9.60	2	4.70	
Rib, Sternum, and Clavicle	3	1.20	7	4.50	8	18.60	
Pelvic bones, Sacrum, and Coccyx	15	6.00	33	21.00	4	9.30	
Grade							
Grade I	5	3.20	0	0.00	9	34.60	< 0.001*
Grade II	9	5.80	1	16.70	15	57.70	
Grade III	71	46.10	3	50.00	0	0.00	
Grade IV	69	44.80	2	33.30	2	7.70	
TNM Extension							
In Situ	1	0.40	0	0.00	0	0.00	0.546
Localized	127	55.70	88	58.30	24	68.60	
Regional: Direct Extension	43	18.90	22	14.60	2	5.70	
Regional: Lymph Node	4	1.80	1	0.70	0	0.00	
Regional: Direct Extension & Lymph Node	4	1.80	1	0.70	0	0.00	
Distant Metastasis	49	21.50	39	25.80	9	25.70	
Bases of Diagnosis							
Medical Imaging	2	0.80	1	0.60	4	9.80	< 0.001*
Cytology/Hematological	1	0.40	2	1.30	0	0.00	
Histology of metastases	0	0.00	4	2.50	0	0.00	
Histology of primary	235	97.10	152	95.00	36	87.80	
Lateralization							
Not paired	33	13.30	35	21.90	13	30.20	0.014 *
Right	99	39.90	54	33.80	11	25.60	
Left	112	45.20	69	43.10	16	37.20	
Bilateral Involvement	1	0.40	0	0.00	0	0.00	
Paired at site of origin	3	1.20	2	1.30	3	7.00	
3 Year Survival							
Dead	49	19.80	24	15.00	7	16.30	0.454
Alive	199	80.20	136	85.00	36	83.70	

* Significant at level ≤ 0.05.

Discussion

The study aimed to highlight the descriptive epidemiology of primary bone sarcomas in KSA. The incidence rate was 7.82 cases per 1,000,000 per five years for osteosarcoma, 4.75 cases per 1,000,000 per five years for Ewing sarcoma, and 1.35 cases per 1,000,000 per five years for chondrosarcoma. The yearly incidence rate was 1.56 cases per 1,000,000 per year for osteosarcoma, 0.95 cases per 1,000,000 per year for Ewing sarcoma, and 0.27 cases per 1,000,000 per year for chondrosarcoma. A significant difference between males and females was present only in the type of tumour. A significant difference in age was present among types of tumours, sites of origin, and three years' survival. A significant difference among the three types of tumours in terms of site of origin, grade, base of diagnosis, and lateralisation was also present.

Due to a plethora of factors, the geographical incidence of bone tumours differs from one region to another. Collectively, the osteosarcoma, chondrosarcoma, and Ewing sarcoma yearly incidence rates detected were lower than the ones detected globally. It is thought that the reasons behind the lower incidence rate include underreporting by hospitals to SCR, lower population, and risk factors that need to be assessed locally. In comparison to our results, a German study that included 671 patients diagnosed with bone sarcoma assessed an age-standardised cumulative incidence of 2.1 per 100,000.[5] In addition, a British study found the incidence rate to be 8.1–9.3 per 1,000,000 in males and 5.4 to 6.8 in females.[6]

Moreover, a French study found that the incidence of bone sarcoma was 0.6 per 100,000.[7] In the Netherlands, a study found that the incidence of chondrosarcoma was 0.15 per 100,000 the osteosarcoma incidence was 0.25 per 100,000 and the Ewing sarcoma incidence was 0.15 per 100,000.[8] In Brazil, the median incidence rate of bone cancer was 5.74 per 1,000,000 among children and 11.25 per 1,000,000 among adolescents.[9] In Taiwan, an overall

incidence of 6.70 per 1,000,000 person-years was detected for bone sarcomas.[14] In Finland, a nationwide study detected an annual incidence of 1.8 osteosarcomas per 1,000,000.[16] In Croatia, the annual incidence rate was 1.68 cases per 1,000,000 for osteosarcoma and 0.76 per 1,000,000 for chondrosarcoma. However, the yearly incidence rate for Ewing sarcoma was 0.79 per 1,000,000, which is lower than the one observed in our study.[18] In Belgium, the overall annual incidence of bone sarcomas was 0.91 per 100,000.[19] In Kuwait, the annual incidence of bone sarcomas was 3.3 cases per 1,000,000 inhabitants. In addition, unlike the presented study, which found osteosarcoma to be the most common tumour observed, the most common tumour detected was Ewing sarcoma.[20] In India, the annual incidence of Ewing sarcoma was 1.6 per 1,000,000 in males and 1.0 per 1,000,000 in females.[21]

When it comes to the patterns of tumour behaviour, a Nigerian study found that different types of bone tumours have a difference in their geographical incidence, which contradicts our findings.[22] Another study assessing the incidence of bone sarcomas in both the United States and the United Kingdom found that chondrosarcoma varied significantly among regions, which also contradicts the negative findings in the present study. However, the same study also found a significant difference between the types of tumours across ages, which is consistent with our findings.[10] Another American study also found a significant difference between the types of tumours across ages, which is also consistent with our findings.[11] Furthermore, a recent Japanese study that investigated the epidemiology and outcomes of bone sarcomas found variations of tumour types across ages.[12] In terms of gender, a European study found that bone sarcomas were more prevalent in males, agreeing with the present study, which found a higher percentage of males diagnosed with bone sarcoma than females.[13] A Japanese study found a slight male preponderance in bone sarcomas, which is consistent with our findings.[14] Furthermore, agreeing with our findings, a Taiwanese study found a significant difference in tumour types across genders.[15] However, a Chinese study found that there was no significant difference across genders when it comes to any type of bone sarcomas, which is inconsistent with our findings.[17]

Understanding the tumour behaviour in the region can aid in detecting the prognostic factors for survival in bone sarcoma. This would aid in the development of risk and response-based treatment strategies that allow for early decision-making. For instance, a Dutch study that aimed to develop a prognostic model for surviving bone sarcoma found that metastasis at diagnosis, large tumours (volume $\geq$ 200 ml or largest diameter $\geq$ 8 cm), primary tumours located in the axial skeleton, and histological response of less than 100% were all associated with survival.[23] A British study also found that the critical factor for adverse prognosis was metastases at diagnosis. Among the group that had metastases, the ones with lung involvement had better survival chances. The risk analysis also demonstrated that site, age-group, and the period of diagnosis had a significant influence on risk-free survival chances.[24]

This study had some limitations. First, due to the retrospective nature of the study, the research team could not directly observe the cases and had to rely on other individuals for accurate recordkeeping. Consequently, the data registered in the Saudi Cancer Registry may be incomplete. Second, the data in the registry only spans from 2013, the year it was founded, to 2017. In terms of the current study's strengths, first, the study is the first of its kind in the Middle East and would be a good initiative for conducting more extensive studies that investigate factors affecting the prognosis quality and the mortality rates of bone sarcomas. Second, the Saudi Cancer Registry includes cases from all hospitals in the country, whether they are private, military, or health ministry hospitals. Third, the study took raw collected data from the Saudi Cancer Registry and applied statistical methods to detect major epidemiological differences across tumour types and compare them with the ones detected in other studies.

Conclusion

To our knowledge, this study is the first of its kind in our region, that is, KSA, giving a baseline to be compared with other regions globally. It is thought that underreporting to the SCR, lower population, and risk factors for developing bone sarcomas might be the reasons for lower incidence rates; therefore, it is recommended that hospitals should investigate the barriers to reporting tumour cases to the SCR as soon as possible. Significant differences in tumour type, origin site, and three-year survival across age and gender were detected. Significant differences of origin site, grade, the base of diagnosis, and lateralisation across tumour types were also detected. These data are useful for health leaders to develop strategies such as selective screening and to facilitate treatment strategies ranging from the traditional operative approach to newer approaches that include non-operative and surveillance treatment pathways. In addition, understanding the tumour behaviour pattern in the region will help to develop tailored risk and response-based treatment strategies that allow for early decision-making.

Recommendations

In KSA, there has been an absence of studies that focus on improving the clinical outcomes of bone sarcoma, possibly because few studies focus on such relatively rare malignancies. Future studies are recommended to focus on risk factors associated with poor outcomes and developing response-based treatment strategies that allow for early decision-making.

Source of funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Conflict of interest

The authors have no conflict of interest to declare.

Ethical approval

The study was approved by the Institutional Review Board of King Abdullah International Medical Research Center, Ministry of National Guard Health Affairs, Riyadh, KSA, (approval number RC16/223/R, dated 12 February 2017). Patients' confidentiality was ensured. The patients' data were collected and used by the research team only. Serial numbers were used instead of medical record numbers to ensure anonymity. Due to the retrospective nature of the study and the use of anonymised patient data, the requirements for informed consent were waived.

Authors' contribution

WSJ, AMA, and MAG formulated the idea, designed the methods, validated the study, and provided research materials. MAG curated and analysed the data and interpreted and presented the results. WSJ, AMA, and MAG wrote the original draft of the article. WSJ critically revised the original draft for important intellectual content, supervised, and administered the project. All authors approved the final draft and are responsible for the accuracy and integrity of the work and the similarity index of the manuscript.

Acknowledgment

The authors would like to thank the Saudi Cancer Registry for providing access to the data for research purposes.

References

1. Dorfman HD, Czerniak B, Kotz R, Vanel Y, Park K. WHO classi cation of bone tumors. In: InFletcher DM, Unni KK, Mertens F, editors. *World health organization classi cation of tumors. Pathology and genetics. Tumors of soft tissue and bone*. Lyon: IARC Press; 2002. pp. 225–232.
2. Ushigome S, Machinami R, Sorensen PH. Ewing sarcoma/primitive neuroectodermal tumor (PNET). In: Fletcher DM, Unni KK, Mertens F, editors. *World health organization classification of tumors. Pathology and genetics. Tumors of soft tissue and bone*. Lyon: IARC Press; 2002. pp. 298–300.
3. Randall RL. Approach to the diagnosis of bone and soft tissue tumors – clinical, radiologic, and classification aspects. **Bone Soft Tissue Pathol 2010**: 3–17. https://doi.org/10.1016/b978-0-443-06688-7.00002-x.
4. Qureshi A, Ahmad Z, Azam M, Idrees R. Epidemiological data for common bone sarcomas. **Asian Pac J Cancer Prev APJCP 2010**; 11: 393–395.
5. Trautmann F, Schuler M, Schmitt J. Burden of soft-tissue and bone sarcoma in routine care. **Canc Epidemiol 2015**; 39(3): 440–446.
6. Whelan J, McTiernan A, Cooper N, Wong Y, Francis M, Vernon S, et al. Incidence and survival of malignant bone sarcomas in England 1979-2007. **Int J Canc 2011**; 131(4): E508–E517.
7. Ducimetière F, Lurkin A, Ranchère-Vince D, Decouvelaere A, Péoc'h M, Istier L, et al. Incidence of sarcoma histotypes and molecular subtypes in a prospective epidemiological study with central pathology review and molecular testing. **PloS One 2011**; 6(8):e20294.
8. Goedhart L, Ho V, Dijkstra P, Schreuder H, Schaap G, Ploegmakers J, et al. Bone sarcoma incidence in The Netherlands. **Canc Epidemiol 2019**; 60: 31–38.
9. Balmant N, Reis R, Santos M, Maschietto M, Camargo B. Incidence and mortality of bone cancer among children, adolescents and young adults of Brazil. **Clinics 2019**; 74.
10. Valery P, Laversanne M, Bray F. Bone cancer incidence by morphological subtype: a global assessment. **Cancer Causes Control 2015**; 26(8): 1127–1139.
11. Dorfman HD, Czerniak B. Bone cancers. **Cancer 1995**; 75: 203–210. https://doi.org/10.1002/1097-0142(19950101)75:1<203::aid-cncr2820751308>3.0.co;2-v.
12. Fukushima T, Ogura K, Akiyama T, Takeshita K, Kawai A. Descriptive epidemiology and outcomes of bone sarcomas in adolescent and young adult patients in Japan. **BMC Muscoskel Disord 2018**; 19(1).
13. Stiller C, Bielack S, Jundt G, Steliarova-Foucher E. Bone tumours in European children and adolescents, 1978–1997. Report from the automated childhood cancer information system project. **Eur J Canc 2006**; 42(13): 2124–2135.
14. Ogura K, Higashi T, Kawai A. Statistics of bone sarcoma in Japan: report from the bone and soft tissue tumor registry in Japan. **J Orthop Sci 2017**; 22: 133–143. https://doi.org/10.1016/j.jos.2016.10.006.
15. Hung G-Y, Horng J-L, Yen H-J, Yen C-C, Chen W-M, Chen PC-H, et al. Incidence patterns of primary bone cancer in taiwan (2003–2010): a population-based study. **Ann Surg Oncol 2014**; 21: 2490–3008. https://doi.org/10.1245/s10434-014-3697-3.
16. Sampo M, Koivikko M, Taskinen M, Kallio P, Kivioja A, Tarkkanen M, et al. Incidence, epidemiology and treatment results of osteosarcoma in Finland – a nationwide population-based study. **Acta Oncol 2011**; 50: 1206–1214. https://doi.org/10.3109/0284186x.2011.615339.
17. Pingping Bao, Yuhong Z, Weiqi L, Chunxiao W, Chunfang W, Yuanjue S, et al. Incidence and mortality of sarcomas in shanghai, China, during 2002–2014. **Front Oncol 2019**; 9. https://doi.org/10.3389/fonc.2019.00662.
18. Bergovec Pingping Bao M, Kubat O, Smerdelj M, Seiwerth S, Bonevski A, Orlic D, et al. Epidemiology of musculoskeletal tumors in a national referral orthopedic department. A study of 3482 cases. **Canc Epidemiol 2015**; 39(3): 298–302. https://doi.org/10.1016/j.canep.2015.01.015.
19. Rothermundt C, Klenke F, Bode B, Baumhoer D, Arndt V, Feller A, et al. Incidence, mortality, and survival trends of soft tissue and bone sarcoma in Switzerland between 1996 and 2015. **Canc Epidemiol 2019**; 63: 101596. https://doi.org/10.1016/j.canep.2019.101596.
20. Katchy KC, Ziad F, Alexander S, Gad H, Abdel Mota'al M. Malignant bone tumors in Kuwait. **Int Orthop 2005**; 29(6): 406–411. https://doi.org/10.1007/s00264-005-0014-6.
21. Chakraborty D, Rangamani S, Kulothungan V, Chaturvedi M, Stephen S, Priyanka Das, et al. Trends in incidence of ewing sarcoma of bone in India – evidence from the national cancer registry programme (1982–2011). **J Bone Oncol 2018**; 12: 49–53. https://doi.org/10.1016/j.jbo.2018.04.002.
22. Oyemade Gaa, Abioye Aa. A clinicopathologic review of benign giant cell tumors of tendon sheaths in ibadan, Nigeria. **Am J Surg 1977**; 134(3): 392–395. https://doi.org/10.1016/0002-9610(77)90413-5.
23. Bosma S, Ayu O, Fiocco M, Gelderblom H, Dijkstra P. Prognostic factors for survival in Ewing sarcoma: a systematic review. **Surg Oncol 2018**; 27: 603–610. https://doi.org/10.1016/j.suronc.2018.07.016.

24. Cotterill S, Ahrens S, Paulussen M, Jürgens H, Voûte P, Gadner H, et al. Prognostic factors in ewing's tumor of bone: analysis of 975 patients from the European intergroup cooperative ewing's sarcoma study group. **J Clin Oncol 2000**; 18: 3108–3114. https://doi.org/10.1200/jco.2000.18.17.3108.
25. Stiller CA, Trama A, Serraino D, Rossi S, Navarro C, Chirlaque MD, et al. Descriptive epidemiology of sarcomas in Europe: report from the RARECARE project. **Eur J Canc 2013**; 49: 684–695.

How to cite this article: Aljuhani WS, Alanazi AM, Alghafees MA. Primary bone sarcomas in KSA: A Saudi tumor registry review. J Taibah Univ Med Sc 2021;16(1):77–85.