Abstract # 11322 - Characteristics and Oncologic Outcomes of Patients with Ewing Sarcoma of the Shoulder

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Background: Ewing sarcoma is a malignant tumor of mesenchymal origin that is the second most common bone tumor of childhood, accounting for 16% of all primary bone tumors.[1] Ewing sarcoma arises in the lower extremities, pelvis, or chest wall as the three most common locations, respectively. By contrast, Ewing sarcomas arising from other locations in the body are rare, and those arising from the shoulder (clavicle and scapula) comprise fewer than 5% of all Ewing sarcomas.[2] As such, there is a paucity of literature on Ewing sarcomas affecting this location, with only several small case reports published. In addition, very little is known about the demographics, tumor characteristics, or oncologic outcomes of patients with Ewing sarcomas of the shoulder. Here, we present a retrospective case series identifying the demographic data, tumor characteristics, and oncologic outcomes of six patients with Ewing sarcomas of the clavicle or scapula, and compare these patients to those with Ewing sarcoma described in the general literature.

Questions/Purposes: We sought to identify 1) the demographic characteristics, 2) the tumor characteristics, and 3) the oncologic outcomes of patients with Ewing sarcomas of the scapula or clavicle compared with those of patients with Ewing sarcoma described in the general literature.

Patients and Methods: This is a retrospective case series of six patients who were treated at a large, urban, adult hospital between 1993 and 2010 for Ewing sarcomas affecting the clavicle or scapula. Patients were identified from a database and patient data was collected from paper and electronic medical records. All patients were de-identified and assigned a study number for purposes of statistical analyses. Given the small number of the patients in this study, no *a priori* power analysis was used and no inferential statistical analyses were performed.

Results: Patients in our case series were 33% male and 67% female compared with 60% male and 40% female as reported from the Surveillance, Epidemiology, and End Results (SEER) database.[3] 100% of patients in our series who reported ethnicity were white compared to 92% from SEER.[3] They had an average age at diagnosis of 23 compared to an average age at diagnosis of 15 for Ewing's sarcoma in general.[1] In our case series, 33% of patients had a tumor size >8 cm in largest dimension at diagnosis compared with 56% of patients at time of diagnosis in the SEER database.[3] 100% of patients with known data in our series had the t(11;22) translocation present compared with 92% in published literature.[4] 83% and 67% of tumors in the case series expressed vimentin and CD99, respectively, compared with 74% and 90% as reported in prior studies.[5–8] In addition, 50% of tumors in the case series expressed neuron-specific enolase (NSE) compared with 58% reported in prior literature.[9] Tumors in this case series also expressed Leu-7 and S100 both at a rate of 18%, compared with 58% and 25.4% in published literature, respectively.[9,10] Patients in our series had a 5-year survival rate of 67% compared to an overall 5-year survival rate of 55% for Ewing sarcoma from SEER.[3] 50% of patients in our series had metastatic disease compared with 27% in the general literature.[11] Details concerning each patient from our case series are included in Table 1.

Conclusions: To date, there are few descriptive studies of Ewing sarcomas affecting the shoulder. Here, we have described a retrospective case series of six patients with Ewing sarcomas of the shoulder and examined their demographic characteristics, tumor characteristics, and oncologic outcomes. Ewing sarcomas of the shoulder more commonly occur in females in contrast to Ewing sarcomas in general. Patients with Ewing sarcomas of the shoulder are also older at time of diagnosis. Ewing sarcomas of the shoulder were associated with Caucasian ethnicity with similar predilection as Ewing sarcomas reported in the general literature. The proportion of patients with a tumor size >8 cm at time of diagnosis was smaller than that of patients with Ewing sarcoma in general; they also had improved 5-year rates of survival. Ewing sarcomas of the shoulder had a higher occurrence of the t(11;22) translocation and expression of vimentin than reported for Ewing sarcomas in other locations. By contrast, there were lower rates of CD99, NSE, Leu-7, and S100 positivity in Ewing sarcomas of the shoulder than reported for Ewing sarcomas in previous investigations. Finally, patients with Ewing sarcomas affecting the clavicle or scapula more commonly had metastatic disease compared with patients with Ewing sarcomas in the published literature.

Table 1:

	Patient 1	**Patient 2**	**Patient 3**	**Patient 4**	**Patient 5**	**Patient 6**
Gender	F	M	F	F	F	M
Ethnicity	White	Not reported	Not reported	White	White	White
Age at diagnosis	51	21	10	16	13	26
Prior history of malignancy	None	None	None	None	None	None
Location	Left clavicle	Left scapula	Right scapula	Right scapula	Right scapula	Left scapula
Treatment	Surgery, chemotherapy	Surgery, chemotherapy, XRT	Surgery, chemotherapy, XRT	Surgery, chemotherapy, XRT	Surgery, chemotherapy, XRT	Surgery, chemotherapy
Tumor size in largest dimension (cm)	4	15	14	5	2.1	6.5
Viable tumor (%)	40	80	35	Not reported	Not reported	0
Metastases	None	Chest, Lung, Retroperitoneum	None	Lung	None	None
Chromosomal translocation	t(11;22)	Not performed	Not performed	t(11;22)	t(11;22)	t(11;22)
Tumor Markers Present	Vimentin, CD99	Vimentin	Vimentin, NSE, Leu-7	Vimentin, CD99, NSE, S100	Vimentin, CD99, NSE	CD99
5-Year Survival	Deceased	Deceased	Alive	Alive	Alive	Alive

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