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Diagnosis, Treatment, and Prognosis of Chest Wall Sarcomas: A Systematic Review

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BACKGROUND: Sarcomas are rare mesenchymal neoplasms comprising over eighty different histiologic subtypes and occur over a wide range of ages and locations. Prognosis for these neoplasms reflect the heterogeneity of sarcomatous pathology and their variable response to current therapeutic options. The thoracic wall is a particularly rare location for sarcomas to arise. The central location of thoracic tumors to important structures adds to the challenges of treating these tumors, often requiring a multidisciplinary approach to surgical treatment. Given the rarity of both a diagnosis of a sarcoma and sarcomas to arise from the thoracic wall, in addition to variability of treatment response for different sarcomatous pathologies, limited consensus exists on the treatment and prognosis of primary chest wall sarcomas. The current literature on this topic consists mostly of case reports with single institution retrospective studies, and database studies comprising a small percentage of the studies found

PURPOSE: The purpose of this systematic review is to contribute to the literature on this particularly rare topic. In this review, we aim to summarize the current understanding of diagnosis modalities, treatment, and prognosis of primary chest wall sarcomas.

METHODS: A search using PubMed and Embase search engines was performed to identify studies relevant studies reporting on the treatment and prognosis of chest wall sarcomas. Articles were selected based on the PRISMA guidelines. Publications with non-human subjects, pediatric patients, non-thoracic primary tumor locations, non-english texts, no mention of treatment and follow-up, and case reports/ series were excluded. Of the 1,572 studies published between January 1, 2011 and December 31, 2021, 17 original articles met inclusion criteria, and focused on patients with primary chest wall tumors and their treatment options.

RESULTS: 1,798 patients (1,058 males, 740 females) were included in the 17 original retrospective articles reviewed in this paper. Primary soft tissue sarcomas comprised 31.3% (562/1798) of tumors reported in this review. Bony sarcomas comprised 65.6% (1180/1798). Only 5 papers (29.4%) of papers included patients who had metastatic disease on presentation. Chondrosarcoma was the most commonly reported bone sarcoma, mentioned in 8 papers, comprising 974 patients. The second most common was Ewing Sarcoma with 6 papers, comprising 50 patients. The most common reported soft tissue sarcoma was liposarcoma, mentioned in 6 papers, comprising 58 patients with the undifferentiated pleomorphic sarcoma (4 papers, 42 patients) being the second most common. 10 papers reported on imaging use, 8 papers reported using CT for diagnostic imaging, while 7 and 2 papers reported MRI and X-Ray use, respectively. High grade tumors comprised 32.3% (588) of graded tumors and intermediate/low grade tumors comprised 53.5% (973) of graded tumors. 1720 (95.7%) patients underwent surgical resection and if applicable reconstruction and 90.4% (745/824) had negative margins. 333 patients (18.8%) received either neoadjuvant or adjuvant chemotherapy. 501 out of 129 soft tissue tumors (25.7%) received chemotherapy. 125 out of 498 soft tissue tumors (25.1%) received radiation. 339 patients (19.6%) received neoadjuvant or adjuvant radiation therapy. No paper reported use of immunotherapy. The median follow-up of 44.6 months. The 5-year overall survival and disease-free survival for the study was 68.9% (1140/1654) and 43.1% (283/658). The reported mortality rate was 37.9% (513/1355).

CONCLUSIONS: This review summarized the current understanding of diagnostic modalities, treatment and prognosis for primary thoracic sarcomas. Limited sample sizes and the exclusion of patients who presented with distant metastases or had previous resections, limit the conclusions we can draw on the best course of action for treatment of thoracic wall sarcomas and their prognosis. It does seem clear that typical prognostic factors of sarcomas, including tumor grade, size of tumor, and resection margin continue to be important in overall survival and disease free survival of chest wall sarcomas. The review highlights the heterogeneity of sarcomatous pathology arising from the thoracic wall, and that current treatment modalities account for their variable responses to treatment options. It is evident that a multidisciplinary approach to treatment is needed. Further

research is needed for a defined consensus on prognosis and treatment paradigms for the treatment of chest wall sarcomas.

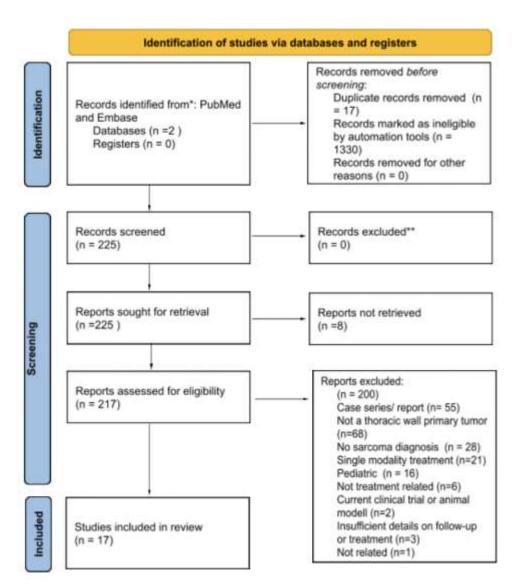


Figure 1. Primsa Flow Diagram