

Review Article

Resecting the rigid and reconstructing the rest: a comprehensive review of chest wall sarcoma management

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Received: 31 July 2026

Accepted: 09 August 2026

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ABSTRACT

Primary chest wall sarcomas are rare malignancies, representing approximately 0.04% of new cancer diagnoses and requiring a complex multidisciplinary management strategy. A comprehensive Boolean search was conducted using PubMed/Medline, Google Scholar, Embase and Scopus to identify studies based on their relevance to the multidisciplinary management of adult patients with primary or secondary chest wall sarcomas. This review explores the clinical landscape of these heterogeneous tumors, differentiating between bone-derived and soft-tissue sarcomas. Diagnosis necessitates advanced imaging, including CT, MRI, and PET/CT, and precise biopsy techniques to facilitate accurate histopathological classification and surgical planning. Wide en bloc surgical resection achieving negative (R0) margins remains the primary curative modality, significantly impacting local control and overall survival. The integration of neoadjuvant and adjuvant systemic therapies and radiation is highly subtype-dependent, with chemosensitive tumors like Ewing sarcoma benefiting most from multimodality approaches. Furthermore, we detail the evolving science of chest wall reconstruction, where rigid prosthetic systems and advanced soft-tissue coverage are utilized to restore skeletal stability and respiratory function. Finally, this review highlights the emerging roles of targeted therapies and 3D intraoperative navigation. Ultimately, a coordinated multidisciplinary paradigm is essential to optimize oncological outcomes and preserve the quality of life for patients with these challenging thoracic neoplasms.

Keywords: Chest wall sarcoma, Chest wall resection, Chest wall reconstruction, Margin, Multidisciplinary management, Bone and soft tissue tumors

INTRODUCTION

Chest wall tumors are a heterogeneous group of neoplasms arising from osseous, cartilaginous, or soft-tissue elements of the thoracic cage.^{1,2} They are broadly categorized as primary (originating in the chest wall) or secondary (resulting from direct invasion or metastasis from adjacent organs like the breast or lung).³ Primary chest wall tumors are rare, representing less than 5% of all thoracic neoplasms.^{1,4} Primary sarcomas of the chest wall account for approximately 0.04% of all newly diagnosed cancers.⁵ There is a wide variation in age at

presentation. Older patients typically present with larger, more aggressive malignant tumors, while younger patients are more likely to have smaller, benign lesions.⁶ For instance, chondrosarcomas are most common in the third and fourth decades of life, whereas Ewing sarcoma is predominantly seen in children and young adults.^{7,8}

A significant driver for secondary or radiation-induced soft tissue sarcomas is prior radiation therapy, which typically manifests after a median latency period of 10 years.^{9,10} Genetic predispositions also play a role - conditions such as Li-Fraumeni syndrome, Gardner

syndrome, and familial adenomatous polyposis (associated with desmoid tumors) are known risk factors.^{11,12}

METHODS

To ensure a broad and relevant evidence base, a systematic search of major medical databases was conducted, including PubMed/Medline, Google Scholar, Embase and Scopus. There was no restriction based on the year. Key search terms utilized included - "chest wall sarcoma", "chest wall resection", "chest wall reconstruction", "primary chest wall tumors", and "chest wall prosthesis". Studies were selected based on their relevance to the multidisciplinary management of adult patients with primary or secondary chest wall sarcomas. The review prioritized full-text studies published in

English, including single and multicenter retrospective cohort studies, prospective observational studies, and established clinical practice guidelines. Technical articles with fewer than three patients (case reports), non-English publications, and studies focusing exclusively on pediatric populations were generally excluded to maintain a focus on adult oncological paradigms. Two independent authors assessed and extracted the data. Data extraction focused on several critical domains of care - diagnostic modalities, surgical outcomes, reconstructive materials and multidisciplinary therapy. The synthesis of this data follows a narrative approach, aimed at providing an overview for clinicians to guide personalized, rational decision-making within a multidisciplinary tumor board framework (Figure 1).

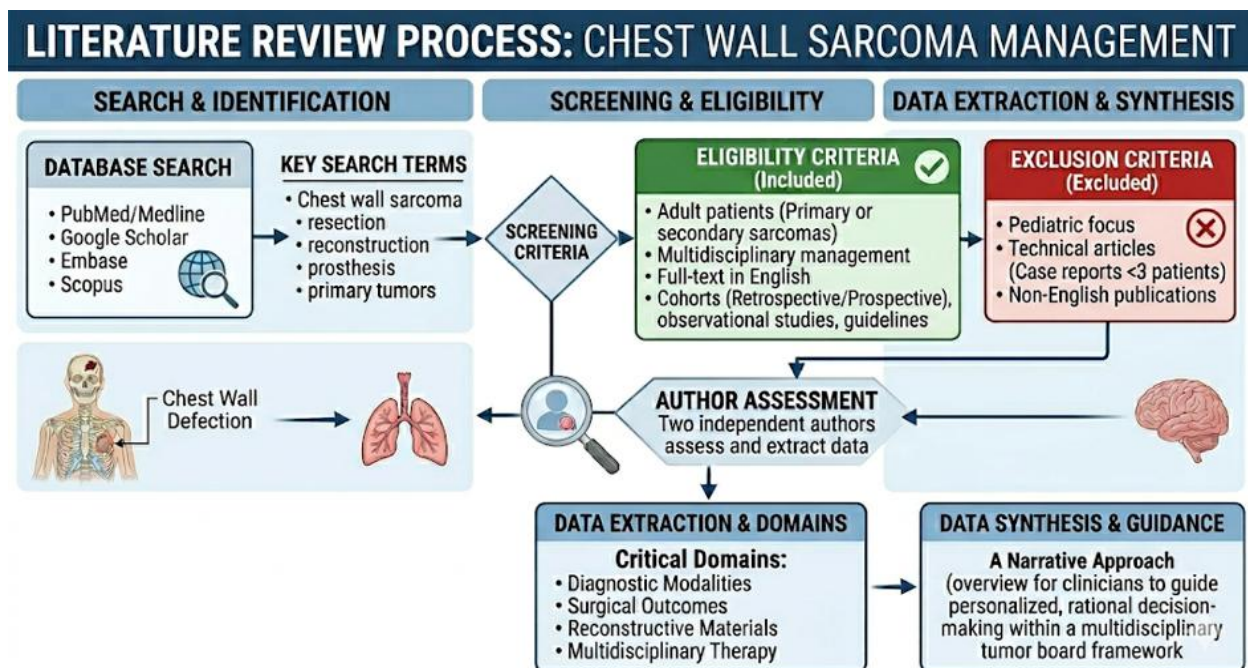


Figure 1: Search methodology.

HISTOPATHOLOGICAL DIVERSITY: THE SARCOMA SPECTRUM

Management pathways are largely dictated by whether a tumor originates in the bone/cartilage or soft tissue. Soft tissue sarcomas comprise about 45% of all chest wall sarcomas.^{2,3} The most frequent primary malignant bone tumor of the chest wall is chondrosarcoma, typically occurring in adults and characterized by "ring-and-arc" calcifications on imaging.⁷ Ewing sarcoma and osteosarcoma are more prevalent in pediatric and young adult populations.^{8,13}

Ewing sarcoma is a small round blue cell tumor, while osteosarcoma is characterized by malignant osteoid

production. Undifferentiated pleomorphic sarcoma (UPS) (formerly malignant fibrous histiocytoma) is a common soft tissue malignant entity with variable morphology, often appearing in elderly patients or following radiation.¹⁴ Other frequent soft tissue types include liposarcoma, fibrosarcoma, and locally aggressive desmoid tumors.

Modern diagnosis relies heavily on immunohistochemistry and molecular genetic testing. For example, the hallmark EWSR1-FLI1 translocation confirms a diagnosis of Ewing sarcoma.^{8,15} Molecular markers like MDM2 or SS18 rearrangements are essential for differentiating morphologically similar tumors, directly informing surgical and systemic therapy choices.^{16,17}

Table 1: Chest wall sarcoma spectrum.

Origins	Types of Sarcoma	Incidences	Clinical Presentations	Histopathologic and Molecular Characteristics	Radiographic and Imaging Features
Bone	Ewing sarcoma ^{8, 15}	Most common malignant diagnosis in children and adolescents	Painful, fast-growing mass; may be accompanied by fever and malaise in aggressive cases	Sheets of uniform small round blue cells with scant cytoplasm; characterized by the hallmark <i>EWSR1-FLI1</i> translocation	CT shows bone destruction and soft-tissue extension; MRI reveals heterogeneous marrow and soft-tissue involvement
Bone	Osteosarcoma ¹³	Makes up 10% to 15% of malignant chest wall tumors; occurs in young or elderly adults	Painful masses; metastatic disease is frequently present at the time of diagnosis	Malignant osteoid production by anaplastic spindle cells; may contain calcified bone matrix or hemorrhage	Lytic or sclerotic osteoid bone matrix within the mass; appears as a calcified mass on imaging
Cartilage	Chondrosarcoma ⁷	Most common primary malignant chest wall tumor; rare in patients <20 years, most common in 3rd and 4th decades	Painful, hard, slow-growing, fixed mass, typically located on the anterior chest wall	Lobulated architecture with atypical chondrocytes in lacunae and hyaline cartilage formation; graded by cellularity and nuclear atypia	CT shows "ring-and-arc" calcifications reflecting mineralization of the hyaline cartilage matrix
Adipose tissue	Liposarcoma ¹⁸	Common soft tissue sarcoma of the chest wall; most frequent in men aged 40 to 60 years	Usually presents as an enlarging painless palpable mass; can be large and associated with trauma	Variable morphology including dedifferentiated or myxoid subtypes; <i>MDM2</i> rearrangements are essential diagnostic tools	MRI is the preferred modality for characterizing soft-tissue extent and identifying necrosis
Vascular	Angiosarcoma / Hemangiosarcoma ¹⁹	Rare; can be radiation-induced (median 10-year latency) or associated with chronic lymphedema or chemical exposure	May present as a skin lesion or a mass 5-10 years post-radiation for breast cancer	Malignant vascular tumor; typically presents with a high tumor grade	MRI used to assess local recurrence; CT used to evaluate for pulmonary metastases
Fibrous tissue	Desmoid tumor (Aggressive Fibromatosis) ²⁰	Histologically benign but locally aggressive; common in patients <40 years and associated with <i>APC</i> gene mutations/FAP	Often present as a firm, fixed, nontender mass; can occur at sites of previous trauma, scarring, or radiation	Fibroblastic proliferations containing collagen and elastic fibers; frequently associated with <i>CTNNB1</i> mutations	MRI shows low-to-intermediate signal intensity on T1 and variable signal intensity on T2
Muscle	Rhabdomyosarcoma ²¹	More common in pediatric populations	Can present with noticeable swelling or localized pain	Malignant soft tissue tumor of muscle origin; often high grade	CT and MRI show tissue-resolving features and provide multiplanar characterization

CLINICAL PRESENTATION AND DIAGNOSTIC WORKUP

Patients most commonly present with a localized palpable mass, which may be asymptomatic or accompanied by pain, swelling, and impaired movement. Pain is the most frequent symptom in malignant cases, often indicating periosteal irritation or bony destruction. Large tumors can cause respiratory compromise, dyspnea, or neurovascular compression symptoms like paresthesia.^{1,6}

CT scan is primarily used to assess bony architecture, cortical destruction, and for pulmonary staging to detect metastases. MRI is the preferred modality for soft-tissue infiltration, delineating neurovascular involvement, and planning surgical margins due to its superior tissue-resolving features.^{22,23} PET/CT is increasingly utilized for metabolic characterization, grading, and screening for distant occult metastases.²⁴

Histologic confirmation is essential. Excisional biopsy is typically reserved for small lesions (<5 cm), while core needle or incisional biopsy is preferred for larger masses (>5 cm) to minimize contamination.²⁵⁻²⁷ Crucially, the biopsy tract must be planned carefully so it can be

completely resected during the definitive surgery to avoid recurrence.

THERAPEUTIC STRATEGIES: THE MULTIDISCIPLINARY PARADIGM

Wide en bloc surgical resection with negative (R0) margins is the cornerstone of curative treatment.²⁸ The definition of an adequate margin varies - typically 2 cm for low-grade or benign lesions and 4 cm for high-grade malignant tumors. Margin status is the most significant predictor of local recurrence and overall survival.^{29,30}

Chemotherapy effectiveness is highly subtype-dependent. Ewing sarcoma, osteosarcoma, and rhabdomyosarcoma are chemosensitive and often treated with neoadjuvant systemic therapy followed by surgery.^{21,31,32} Conversely, chondrosarcomas and many adult soft tissue sarcomas are notoriously chemo-resistant, making surgery the primary treatment.³³ Radiation serves as an integral multidisciplinary component. Preoperative radiation may facilitate margin-negative resection and tumor downstaging, while postoperative radiation is indicated for close or positive margins to improve local control.^{34,35} Advanced techniques like proton beam therapy are being investigated to minimize toxicity to the heart and lungs.³⁶

Table 2: Management strategies for chest wall sarcoma spectrum.

Tumour types	Malignancy Status	Typical Locations	First-line Management	5-Year Survival Rate	Reconstruction Methods
Chondrosarcoma ³⁷	Malignant	Anterior chest wall, ribs, sternum	Wide en bloc surgical resection (curative) with 4-cm margins; adjuvant radiotherapy for unresectable cases or positive margins	65%-90%	Prosthetic materials (titanium plates, methyl methacrylate sandwich), muscle flaps
Ewing Sarcoma ³¹	Malignant	Chest wall, often involving ribs	Neoadjuvant chemotherapy followed by wide local excision and adjuvant chemotherapy	30%-100% (lower with metastases)	Rigid prosthetic materials and soft tissue coverage
Osteosarcoma ³²	Malignant	Rib, scapula, clavicle	Neoadjuvant chemotherapy combined with wide local excision and adjuvant chemotherapy	15%-50% (dependent on metastases)	Thoracic wall stabilization with titanium bars or methyl methacrylate
Solitary Plasmacytoma ³⁸	Malignant	Bone (ribs) or soft tissue	Definitive radiotherapy (40 to 50 Gy); surgery reserved for diagnostic confirmation or mechanical instability	40%-60%	Skeletal stabilization if mechanical instability occurs after radiotherapy
Undifferentiated Pleomorphic Sarcoma (MFH) ³⁹	Malignant	Soft tissue of the chest wall	Wide local excision (standard) with neoadjuvant radiotherapy for high-grade cases	30%-40%	Composite implants (titanium mesh and muscle flaps)
Liposarcoma ⁵	Malignant	Soft tissue	Wide local surgical	60%	Muscle flaps

Continued.

Tumour types	Malignancy Status	Typical Locations	First-line Management	5-Year Survival Rate	Reconstruction Methods
		compartment of chest wall	resection; chemotherapy/radiotherapy often have limited roles		(latissimus dorsi, pectoralis major) for soft tissue coverage
Angiosarcoma⁴⁰	Malignant	Thoracic wall skin and soft tissue	Wide local excision; chemotherapy (paclitaxel) for advanced/systemic disease	10%-20%	Extensive soft tissue reconstruction with pedicled or free flaps (DIEP, ALT)
Desmoid tumor (Extra-abdominal desmoid)⁴¹	Benign but locally aggressive	Extra-abdominal chest wall, often at sites of prior trauma or thoracotomy	Active surveillance (initial strategy); surgical resection with 4 cm margins for progressive or symptomatic disease	High (generally favourable survival)	Rigid or nonrigid reconstruction based on defect size (>5 cm)
Fibrous Dysplasia⁴²	Benign	Lateral or posterior ribs	Observation or wide local excision for symptomatic relief and diagnostic confirmation	-	Usually not required for small rib resections
Osteochondroma⁴³	Benign	Ribs or scapula, often at costochondral junction	Surgical resection for symptomatic relief or to minimize risk of malignant transformation	-	Primary closure or simple mesh if defect is small

THE ART AND SCIENCE OF RECONSTRUCTION

Reconstruction is critical to maintain respiratory mechanics and protect internal organs when defects exceed 5 cm in diameter or involve more than three to four ribs.⁴⁴ Rigid materials include methylmethacrylate sandwiches (layered between mesh), titanium plate systems (e.g., MatrixRIB, STRATOS), and increasingly, 3D-printed custom implants that mimic native

biomechanical behavior (Figure 2). Soft materials including synthetic meshes (e.g., Marlex, Prolene, Gore-Tex) provide a barrier and a scaffold for tissue ingrowth.^{45,46} Adequate soft tissue coverage with viable tissue is essential to minimize infection and protect underlying prosthetics. Pedicled muscle flaps (e.g., Latissimus dorsi, Pectoralis major, Rectus abdominis) are workhorses in reconstruction. In complex cases where pedicled flaps are unavailable, free flaps such as the DIEP or ALT flap may be utilized.⁴⁷

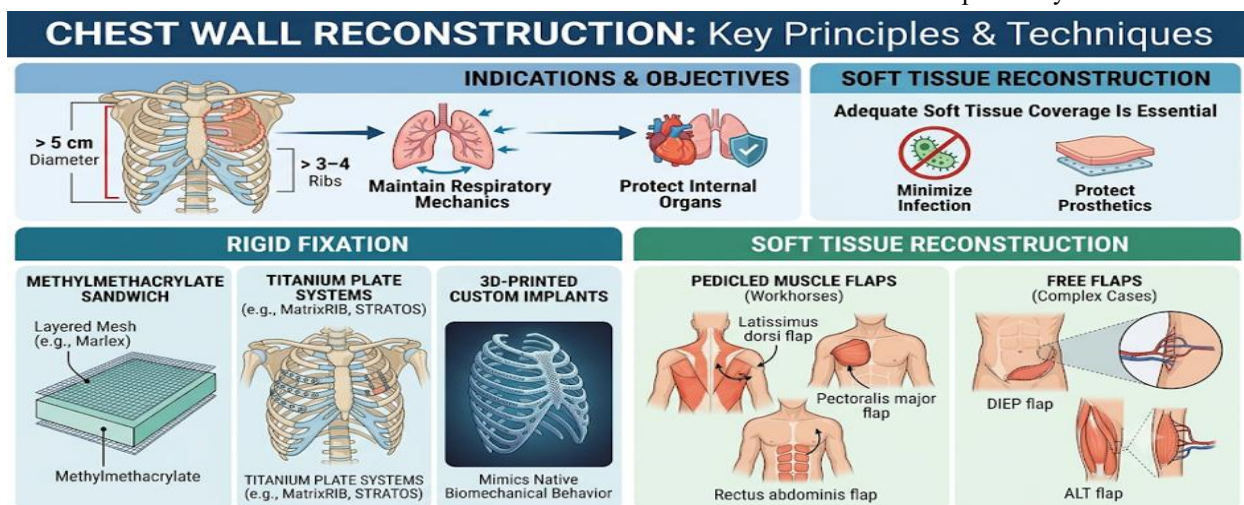


Figure 2. Options for reconstruction.

Table 3: Options for reconstruction.

Material Class	Specific Material	Indication/ Suitability	Advantages/ Pros	Disadvantages/ Cons
Synthetic / Alloplastic	Methylmethacrylate (Acrylic resin) ⁴⁸	Mandatory for anterior/lateral defects >4–5 cm; stabilization of sternum, ribs, and chest wall.	Excellent rigidity (+++); provides chest wall stability and organ protection; avoids paradoxical movement.	Exothermic reaction during molding; risk of seroma due to low permeability; excessive rigidity causes pain; risk of dislocation/rupture.
Synthetic / Alloplastic	Titanium ⁴⁹	Large full-thickness defects; stabilization after trauma; sternocostal reconstruction; suitable for 3D customization.	Highly biocompatible; biologically inert; diamagnetic; malleable (bars); excellent stability (+++) and organ protection.	High cost (3D implants); risk of bar fracture (0–11%); potential for screw/plate dislocation; difficult fixation for complex defects.
Synthetic / Alloplastic	Polypropylene (Marlex, Prolene) ⁵⁰	General chest wall reconstruction; often used in "sandwich" with methylmethacrylate.	Good integration with surrounding tissue; allows muscle reattachment; provides a barrier between pleural and subcutaneous spaces.	Does not provide rigid protection alone; risk of paradoxical movement in large defects; 10–25% infection rate reported.
Synthetic / Alloplastic	Polytetrafluoroethylene (PTFE / Dualmesh / Gore-Tex) ⁵¹	Full-thickness defects requiring reconstruction; watertight closure.	High customization potential (+++); excellent patient tolerance; prevents fluid/air movement.	Lack of skeletal rigidity; cannot replace normal chest curvature after major resection.
Synthetic / Alloplastic	Polyglactin (Vicryl) ⁵²	Absorbable mesh for patients at high risk of infection; small reconstructions.	Lower long-term infection risk; creates fibrotic scar for closure.	No protection of mediastinal viscera; temporary; insufficient for large defects.
Bioprosthesis	AlloDerm (Human acellular dermal matrix) ⁵³	Contaminated or irradiated fields; patients at high risk for mesh-related complications.	Supports cellular repopulation and revascularization; lower infection risk in compromised fields.	Poor maintenance of rigidity; risk of partial resorption or failure to integrate.
Bioprosthesis	Bovine Pericardium / Dermis ⁵⁴	Biological reconstruction; soft tissue support.	High biocompatibility; biomechanical properties similar to native tissue.	No inherent rigidity; restricted to soft-tissue-like applications.
Bioprosthesis	Cadaveric Bone Grafts ⁵⁵	Sternal reconstruction (sternectomy) due to infection or necrosis.	Provides structure for osteoprogenitor cells; restores structural integrity.	Scarce availability; logistical problems related to organ retrieval; limited amount for large defects.

ONCOLOGICAL OUTCOMES, PROGNOSTIC FACTORS AND SURVEILLANCE

Prognosis is highly variable and depends on histologic subtype, tumor grade, size, and margin status. Achieving an R0 resection significantly improves 5-year survival rates compared to incomplete (R1/R2) resections. High tumor grade remains a major predictor of disease recurrence and distant metastasis.^{28,29}

Failure typically occurs as local recurrence or distant hematogenous metastasis, predominantly to the lungs. Local recurrence rates are reportedly higher for chest wall sarcomas compared to those in the extremities.^{5,28}

Surveillance involves long-term imaging to detect local and pulmonary recurrence. A common practical approach for high-grade patients includes follow-up every 3–4 months for the first 2–3 years, then twice yearly until the fifth year, followed by annual visits.⁵⁶

FUTURE HORIZONS AND TARGETED THERAPIES

Beyond traditional chemotherapy, targeted therapies are emerging for specific subtypes. For example, Tyrosine Kinase Inhibitors (TKIs) like pazopanib may be used for solitary fibrous tumors, and taxanes (paclitaxel) are advised for advanced angiosarcomas.^{40,57}

Emerging technologies like 3D surgical simulation and navigation are being used to improve precision in achieving oncological margins during complex resections.⁵⁸ Furthermore, research into biomimetic materials and mesenchymal stem cells aims to enhance the bio-integration of grafts and promote tissue regeneration.

Table 4: Advancements in systemic therapy.

Therapy category	Specific agents or class	Target histologic subtype	Clinical application and evidence
Targeted Therapy (TKI)	Pazopanib ⁵⁷	Solitary fibrous tumor	Used for tumors that are refractory to traditional doxorubicin.
Targeted Therapy (TKI)	Tyrosine Kinase Inhibitors (General) ⁴¹	Desmoid tumor	Recommended when tumors are unresectable, rapidly growing, or fail conservative/active surveillance.
Histotype-Specific Cytotoxic	Trabectedin ⁵⁹	Leiomyosarcoma, Liposarcoma, Solitary fibrous tumor	Utilized for specific soft tissue subtypes including myxoid liposarcoma.
Systemic/Biologic	Paclitaxel (Taxanes) ⁴⁰	Angiosarcoma	Recommended for advanced disease because these tumors exhibit high sensitivity to taxanes.
Systemic/Biologic	Dacarbazine ⁶⁰	Solitary fibrous tumor	Treatment option for tumors refractory to standard chemotherapy.
Hormonal/Biologic	Hormonal therapy ⁶¹	Metastatic breast cancer	Utilized to control systemic disease in patients with secondary chest wall involvement.

Limitations

A primary limitation of this review is the rarity and extreme heterogeneity of chest wall sarcomas, which represent only about 0.04% of all newly diagnosed cancers. This scarcity leads to a general lack of substantial prospective and multicentric studies, forcing current management paradigms to rely heavily on retrospective, monocentric case series with small patient cohorts. Consequently, there is currently no internationally accepted standard or uniform surgical strategy for chest wall reconstruction, particularly regarding when to use rigid versus soft prostheses or which specific materials are optimal.

Many reconstructive materials lack long-term comparative data and are still considered experimental, remaining excluded from formal international guidelines. Furthermore, the review notes that systemic therapy outcomes are often extrapolated from more common extremity sarcomas because chest wall patients are underrepresented in prospective clinical trials. Finally, standard prognostic tools are often not specifically tailored for the chest wall, and the definitive diagnostic role of newer imaging modalities like PET/CT remains to be formally established due to a lack of large-scale evidence.

CONCLUSION

Managing chest wall sarcomas remains a formidable diagnostic and therapeutic challenge that necessitates a highly coordinated multidisciplinary paradigm involving thoracic surgeons, plastic surgeons, oncologists, and pathologists. Wide en bloc surgical resection achieving negative (R0) margins remains the definitive cornerstone of curative intent, as it is the most critical determinant of local control and long-term survival. While neoadjuvant and adjuvant therapies are integral, their application must be meticulously tailored to specific histologic subtypes and tumor grades, reflecting the significant variability in chemosensitivity between common entities such as Ewing sarcoma and chondrosarcoma. Modern reconstructive techniques have revolutionized the ability to restore skeletal stability and respiratory mechanics following extensive resections. Despite these technological strides, the field is currently limited by a lack of internationally accepted standards and high-quality prospective data to guide optimal material selection and reconstructive strategies. Future advancements in targeted molecular therapies and precise intraoperative navigation offer the potential to further enhance oncological precision and the overall quality of life for patients facing these rare and complex neoplasms.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Chanda S, Gupta D, Rahmani AQ. Resecting the rigid and reconstructing the rest: a comprehensive review of chest wall sarcoma management. *Int Surg J* 2026;13:2076-85.