High Grade Sarcomas Arising from the Shoulder Girdle

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General Information

- Shoulder Girdle is the 3rd most frequent site to be affected by a sarcoma
- Proximal humerus is more commonly affected than the scapula
- Proximal humerus is the third most frequent site for an osteosarcoma (15% of all osteosarcomas)
- Clavicle is very rare site for developing a sarcoma
General Information

- **Proximal humerus**: osteosarcoma, chondrosarcoma, Ewing’s sarcoma
- **Scapula**: chondrosarcoma, Ewing’s Sarcoma, osteosarcoma, metastatic renal cell carcinoma
- Most (90%-95%) high grade sarcomas arise from the metaphysis of the proximal humerus or scapula and present as extracompartmental tumors (extend beyond the bony cortices of the proximal humerus or scapula)

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Metaphyseal Origin and Extraosseous Extension
Metaphyseal Origin and Extraosseous Extension
**Limb Salvage**

- Historically a forequarter amputation was performed for high grade sarcomas of the proximal humerus and scapula.

- Early 1970s Marcove et al initiated limb sparing surgery and published their results in 1977; Local tumor control was the same as that achieved with a forequarter and a functional hand and elbow were preserved.
Today, 95% of high grade shoulder girdle sarcomas are treated with limb sparing surgery. Improving use of preoperative (induction) chemotherapy and radiotherapy. Improvements in surgical techniques and prosthetic designs. Advanced imaging modalities (CT, MRI). Better understanding of the local growth and biological behavior of sarcomas.
Earliest treatment until the 1970s was a forequarter amputation. This was debilitating and disfiguring, and chronic phantom limb pain was also experienced.
Historical

- 1977 Marcove et al published their results with limb sparing surgery for 17 patients

- **Standard Tikhoff -Linberg** resection for scapula tumors (Extraarticular total scapula resection, lateral clavicle, rotator cuff, deltoïd, trapezius, rhomboids, portion of latissimus)

- **Extended-Tikhoff Linberg** for proximal humerus tumors
Tikhoff-Linberg

Limb-Sparing Resection

- Tikhoff-Linberg Type Resection (extraarticular total scapulectomy)

Ewing’s Sarcoma

Clavicle

Glenoid

Humeral Head

Classical Tikhoff-Linberg

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Tikhoff-Linberg

Inferior Angle of Scapula

Deltoid
Extended Tikhoff-Linberg

Deltoid Overlying Proximal Humerus Tumor
Pathological study of specimens revealed that it was safe to perform an osteotomy medial to the coracoid process.

- Resections of smaller magnitude
- Body of scapula remained to facilitate reconstruction
Early Reconstruction Options

- Proximal humerus stabilized to clavicle or rib (earliest)
- Flail shoulder
- Poor strength and stability
- Traction neuropraxia (brace or sling for support)
- Poor cosmesis
Early Results
Reconstruction Options

- Intramedullary rod stabilized to clavicle or rib
- Hardware failure
- Painful unstable shoulder
- Frequent wound complications
- Traction neuropraxia
- Poor cosmesis
Wound Complications from IM Rod
Reconstruction Options
Other Reconstruction Options

- **Free vascularized fibulas for fusions:** prolonged immobilization, fractures, infections, high complication rates, if succeed lose rotation below shoulder level.

- **Allografts and allo-prosthetic composites:** abandoned, high infection and fracture rates (performed for intraarticular resections---high local recurrence rates); function not better than prostheses despite an intraarticular resection.
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Deltoid Overlies Proximal Humerus

Metaphysis of Proximal Humerus

Rotator Cuff

Capsule

Metaphysis of Scapula
Local Growth of Sarcomas

- Sarcomas grow locally in a *centripetal manner* and form ball-like masses.
- Obey fascial borders and grow along the path of least resistance.
- Investing fascial layers of muscles form *compartmental borders* and form a barrier to tumor penetration; sarcomas rarely penetrate beyond adjacent fascial borders (compartmental borders).
- Adjacent muscles and their fascial layers are compressed into a *pseudocapsule* that contains microscopic tumor nodules (satellite nodules).
Compartments of the Shoulder Girdle

- A compartment refers to a fascial boundary to tumor extension (investing fascial layers of muscles that immediately surround a bone)

- Space that is bound by fascial borders

- **Functional Anatomic Compartment** exists around the proximal humerus and scapula
Compartments of the Shoulder Girdle

- **Proximal humerus**: deltoid, lateral subscapularis and lateral portion of the remaining rotator cuff, coracobrachialis, axillary nerve and circumflex vessels

- **Scapula**: Rotator cuff muscles

- The glenoid and proximal humerus reside within the same functional compartment

- **The subscapularis is a crucial boundary**: protects the axillary vessels and brachial plexus from tumor involvement along with the axillary sheath

- The muscles that form the compartmental borders also form the pseudocapsule of the tumor. Resection of these muscles with the tumor essentially confers a compartmental resection of the tumor

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Local Growth of Proximal Humerus Sarcomas

- Tumor arising from proximal humerus
- Deltoid
- Axillary nerve and circumflex vessels
- Subscapularis
- Latissimus dorsi
LOCAL SPREAD OF HEMATOMA SECONDARY TO PATHOLOGICAL FRACTURE

- Body of scapular
- Infraspinatus
- Deltoid
- Serratus anterior
- Subscapularis
- Short head of biceps
- Axillary vessels and brachial plexus
- Fracture Hematoma
- Osteosarcoma of proximal humerus
- Coracobrachialis
- Pectoralis major
Local Growth of Scapular Sarcomas
Extraarticular vs Intraarticular Resection

- High grade shoulder girdle sarcomas (extracompartamental) routinely contaminate the glenohumeral joint (grossly and microscopically) and readily spread to the apposing articular surface.

- **Proximal humerus**: deltoid and overlying rotator cuff form the pseudocapsule (satellite nodules) and must be resected for an adequate margin (compartmental resection).

- Proximal humerus: axillary nerve involved by tumor and must be removed.

- Retention of the glenoid confers no functional benefit with axillary nerve and abductor muscle involvement.

- Extraarticular resection permits medialization, stabilization, and soft tissue coverage.
Mechanisms of Local Tumor Spread for Sarcomas of the Shoulder

Pericapsular
Intra-articular structures (biceps tendon)
Fracture hematoma
Direct articular spread
Subsynovial extension
Spread along Biceps

Joint Contamination

Metaphyseal Origin and Centripetal Growth
Spread Along Rotator Cuff

Intracapsular

Fracture
Satellite Nodule in Deltoid Fracture and Joint Contamination

Deltoid = Pseudocapsule

Satellite Nodule in Deltoid

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Soft Tissue Component Across Joint
Soft Tissue Component Across Joint

Subscapularis Muscle

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Deltoid Involved

Across Joint

Tumor Crossing Joint
Classification of Shoulder Girdle Resections

- Based on local growth of sarcomas
- Biological behavior and grade
- Response to adjuvants
- Tumor extent
Goals of Resection

- Oncologically safe procedure

- Minimal risk of local recurrence (local recurrence in this region is usually treated with a forequarter amputation and local recurrence may adversely effect survival)
SURGICAL CLASSIFICATION OF
SHOULDER GIRDLE RESECTIONS

TYPE I
Intra-articular proximal humeral resection
A. Abductors retained (shown)
B. Abductors resected

TYPE IV
Extra-articular scapular and humeral head resection
A. Abductors retained
B. Abductors resected (shown)

TYPE II
Partial scapulectomy
A. Abductors retained (shown)
B. Abductors resected

TYPE V
Extra-articular humeral and glenoid resection
A. Abductors retained
B. Abductors resected (shown)

TYPE III
Intra-articular total scapulectomy
A. Abductors retained (shown)
B. Abductors resected

TYPE VI
Extra-articular humeral and total scapula resection
A. Abductors retained
B. Abductors resected (shown)
Classical Tikhoff-Linberg

TYPE IV

Extra-articular scapular and humeral head resection

A. Abductors retained
B. Abductors resected (shown)
Modified Tikhoff-Linberg

TYPE V

Extra-articular humeral and glenoid resection

A. Abductors retained
B. Abductors resected (shown)
Extended Tikhoff-Linberg

TYPE VI

Extra-articular humeral and total scapula resection

A. Abductors retained
B. Abductors resected (shown)
LITERATURE REVIEW OF INTRA VS EXTRA-ARTICULAR RESECTION OF THE PROXIMAL HUMERUS FOR STAGE IIB BONE SARCOMAS
Analysis of 156 cases

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Goals of Reconstruction

- Restore shoulder girdle stability
- Painless shoulder
- Preserve a functional hand and elbow
- Maintain motion (rotation) below shoulder level where most activities of daily living are performed
- A reliable means of reconstruction that will permit prompt resumption of chemotherapy and allow early return to activity/functional use
Methods of Reconstruction

- **Bony Reconstruction**
  - Modular Segmental Proximal Humerus Prosthesis
  - Total Scapula Prosthesis (if specific muscles preserved)
    - Nonconstrained
    - Constrained

- **Soft Tissue Reconstruction**
  - Static and Dynamic Methods of Soft Tissue Reconstruction
TIKHOFF-LINBERG RESECTION AND TOTAL SCAPULAR REPLACEMENT

Goretex sleeve

Dacron tape

fig. 3
Radiological Imaging

- Plain Radiograph
- MRI
- CT
- Angiogram
- Venogram
- Bone Scan
- Thallium Scan
- CT of Chest
Estimating Response to Induction Chemotherapy

- Plain Radiograph
- Arteriogram (“Gold Standard”)
- CT scan
- Quantitative Thallium Scan
- Quantitative Bone Scan
Estimating Resectability

- Clinical Triad for an Unresectable Tumor
  - Intractable Neurogenic Pain
  - Motor Loss
  - Venogram demonstrating an obliterated axillary vein

- Final Decision made after intraoperative Exploration!!!
Resectable Tumor

BRACHIAL PLEXUS
(TUMOR RESECTABLE)

Axillary vein patent
Sheath compressed
Unresectable Tumor

BRACHIAL PLEXUS
(TUMOR UNRESECTABLE)

Sheath infiltrated

Axillary artery patent
Axillary vein occluded

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Biopsy

Inappropriately Performed Biopsies are Leading Cause for Amputations!!!!!
Proximal Humerus Resection and Reconstruction
Coracoid

Deltoid

Subscapularis fibers

Pectoralis minor

Pectoralis major

Conjoin tendon (biceps short head and coracobrahialis)

Vessel sheath
Pectoralis Major

Pectoralis Minor

Deltoid Overlying Tumor

Biceps Short Head

Neurovascular structures in Axillary Sheath
Ligation of Circumflex Vessels and Axillary Nerve

Axillary Vessels and Brachial Plexus

Tumor Deep to Subscapularis and Deltoid
Spine of scapula
MODULAR REPLACEMENT SYSTEM: PROXIMAL HUMERUS
Pectoralis major and minor

Clavicle

Trapezius

Supraspinatus

Infraspinatus

Teres minor

Latissimus dorsi

teres major

Triceps

Biceps long head

Biceps short head
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Proximal Humerus Resection with Prosthetic Reconstruction: 3 yr Follow-up
Scapulectomy and Total Scapula Reconstruction
Ewing's Post Chemo CT Complete Response
CT Post Chemo
Undifferentiated Sarcoma

Post Chemo MR
Undiff Sarcoma

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Local Recurrence
S/P Intra-artic Resection
Telangiectatic OS

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Anterior incision for extra-articular resection

Teres minor and major released

Evaluation of subscapular area and posterior chest wall

Humeral osteotomy

Deltoid retracted

Supraspinatus

Levator scapulae released

Trapezius

Rhomboids released
A = Intra-articular resection
B = Extra-articular resection

Axillary nerve posterior branch
Posterior joint capsule
Extra-articular resection
Intra-articular resection
Ligated suprascapular artery and vein
A Total Scapula Reconstruction is recommended if the axillary nerve and specific periscapular muscles can be preserved.

- Deltoid
- Trapezius
- Serratus Anterior
- Rhomboids
- Latissimus

These are essential for soft tissue coverage, stabilizing and suspending the prosthesis and for providing the necessary muscle force couples to power the prosthesis.
TYPES OF SCAPULAR RECONSTRUCTION
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Scapular Design

- Non-Constrained Components (Earlier Versions)
- Gore-tex aortic graft for capsular reconstruction

Modular Proximal Humerus

Dacron Tape

Superior Border

Axillary Border

Non-Constrained Components

Capsular Reconstruction
Gore-tex aortic graft

Gore-tex Aortic Graft
Sutured to Scapula Neck
Constrained Total Scapula Prosthesis

- Facilitate intraoperative attachment
- Rotator cuff substituting (fixed fulcrum; passively stabilize humeral head in glenoid; improve active motion)
- Enhance stability
Constrained Components

**Body**
- Down-sized compared to normal
- Holes for Myodesis
- Vacant area—scarring of muscles

**Glenoid**
- Bipolar hip
- Captured polyethylene liner
Prevents Superior Humeral Migration!
Motion

Holes for Myodesis of Periscapular Muscles
Soft Tissue Reconstruction

Cranial portion of trapezius

Deltoids

Caudal portion of trapezius

Triceps

Rhomboids rotated to cover prosthesis

Latissimus dorsi
Soft Tissue Reconstruction

The entire prosthesis is covered!
Post-op Axillary Radiograph

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Non-Constrained
10 yrs post-op

Non-Constrained
10 years post-op
Excellent elbow flexion
13 y.o.
6 mos post op
Normal elbow flexion
Some FF of Shoulder
1 yr post-op
Cosmesis Good
Trap Function Good
Strong Biceps
Hercules!!!
Periscapular Muscles Contracting
Pectoralis Major Contracts
Shoulder Abducts
Results

- 89 Patients with high grade sarcomas arising from the shoulder girdle who underwent prosthetic reconstruction
  - 74 proximal humerus
  - 15 scapula
  - Follow-up 2-20 years (median: 10 years)
  - Most common dx: osteosarcoma, chondrosarcoma and Ewing’s sarcoma
Results

- Overall Local Recurrence: <5%
- No patient required a forequarter amputation
- Subgroup of patients with osteosarcomas (n=43): No local Recurrences
- 10 patients with pathological fractures: No local recurrence
- 65% are prolonged survivors
MSTS Scoring System

- Pain (5=No Pain)
- Emotional Acceptance (5=Cosmetically acceptable)
- Function (3-4: All ADLs but can not participate in high level athletic activities)
- Hand positioning (3-4: Not unlimited but can position above shoulder)
- Dexterity (5: Normal Hand Dexterity)
- Lifting Ability (3-4: Virtually Normal)
- Score: 24-27/30 points
Results

- All survivors are pain free with a stable shoulder
- All can carry out ADLs with operative extremity
- No braces required
- Virtually normal hand and elbow function; Biceps strength: Grade 4+
- MSTS score of 24-27 (80%-90%)
- Abd/FF: 30°-60°
- IR: Normal; ER: -15° to Neutral  (Improved with latissimus dorsi transfer)
- Kaplan-Meier Survival at 10 years: 95%-100%
Complications

- Transient Nerve Palsy: 12% (All in patients who received preoperative chemotherapy)
- Skin Necrosis and Wound Infection: 2% (No prosthesis required removal)
- Aseptic loosening: 1%-2%
- 1 glenohumeral dislocation of a total scapula
- No instability with proximal humerus reconstructions
- No traction neuropraxia
Summary

- Extraarticular resection including the muscles that form the pseudocapsular layer is a reliable method of resection for high grade shoulder girdle tumors that present with an extraosseous component. It provides an oncologically safe margin.

- Reconstruction with proximal humerus and total scapular prostheses and with static and dynamic methods of soft tissue reconstruction provides a durable method of reconstruction and restores a functional, pain free and stable extremity.
Thank You!