

Limb-Sparing Resection of the Scapula and Reconstruction with a Constrained Total Scapula Prosthesis: A Case of Multicentric Epithelioid Hemangioendothelioma Involving the Scapula and Surrounding Soft Tissues

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ABSTRACT

Introduction. The scapula is a relatively uncommon site for primary bone sarcomas. Tumors arising from the scapula are often initially contained by the rotator cuff muscles that protect pertinent adjacent muscles as well as the brachial plexus and axillary blood vessels. Limb-sparing resection of a scapula sarcoma is technically complex and requires meticulous dissection and mobilization of the brachial plexus and axillary blood vessels. Several muscles must be capable of being preserved in order to reconstruct the scapula and shoulder girdle with a special customized total scapula replacement. The goal of reconstruction is to restore a stable shoulder girdle to preserve elbow and hand function.

Methods and Results. In the procedure demonstrated in this video, limb-sparing scapula resection and reconstruction was performed for a patient with a multicentric epithelioid hemangioendothelioma. The steps of the procedure are detailed along with accommodations made for the multicentric nature of the tumor. To allow for optimal postoperative function and maximum soft tissue coverage, a small constrained scapular prosthesis was utilized. The glenohumeral ligaments were reconstructed with a Gore-

Tex aortic graft. Multiple muscle rotation flaps were performed to cover and protect the prosthesis as well as restore shoulder girdle stability.

Conclusion. Limb-sparing surgery for scapula sarcomas and anatomic reconstruction with a constrained total scapula prosthesis is a reliable and safe technique for resecting selected sarcomas and reconstructing the shoulder girdle. A stable shoulder girdle can be restored for optimal hand and elbow function. A total scapula prosthetic reconstruction is the authors' procedure of choice when the deltoid, trapezius, rhomboid, latissimus, and serratus anterior muscles are capable of being preserved. The functional outcome is superior to a forequarter amputation and a flail (nonreconstructed) shoulder in which the extremity is left hanging by soft tissues (nonanatomic method).

A 49-year-old male patient presented in June 2007 with 4 months of worsening right shoulder pain and swelling. The patient had initially presented to a dermatologist with two "pimple-like" lesions growing from his skin overlying the posterior portion of the scapula. These skin lesions were removed and diagnosed as an epithelioid hemangioendothelioma. One month later, the patient began to experience pain in his right shoulder. These symptoms became progressively worse. Magnetic resonance imaging demonstrated multiple lesions involving his acromion, base of coracoid, supraspinatus, infraspinatus, teres minor, rhomboid, and trapezius muscles. He underwent an open biopsy of his right acromion that was consistent with epithelioid hemangioendothelioma. The patient was treated with a radical resection of the right scapula including the

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distal third of the clavicle, proximal humerus, rotator cuff, and portions of the deltoid, trapezius, and rhomboid muscles that were affected with neoplasm. The shoulder girdle was reconstructed with a constrained total scapula and proximal humerus tumor prosthesis.

Histologically the tumor consisted of plump, ovoid, and spindle cells with abundant acidophilic cytoplasm containing nuclei with frequent open chromatin and conspicuous nucleoli. Cellular atypia was mild and mitosis quite rare. The immunomarkers were positive for CD34, Cam 5.2, and CK5/6. Final pathology supported the diagnosis of epithelioid sarcoma-like hemangioendothelioma. All margins were free of disease.

The goal of the surgery is to resect the entire tumor without having to perform a forequarter amputation, and to subsequently restore a stable shoulder girdle in order to preserve hand and elbow function.

This patient is presently 1.5 years after surgical resection and reconstruction. There is no evidence of local or systemic disease. He has a stable shoulder girdle and full use of his hand and elbow. He has full flexion and extension of the elbow. He can protract and retract the scapula and shrug his shoulder. He experiences minor intermittent trapezius fatigue-type discomfort. A stable shoulder girdle was restored. Active motion is impaired but anticipated. He can abduct the shoulder approximately 20° and forward flex 20°. There have been no problems with either infection or dislocation. Adjuvant chemotherapy has not been shown to be of significant benefit for this type of tumor, and since there was no evidence of disease after surgery, chemotherapy was not recommended.

A limb-sparing resection of the scapula and reconstruction with total scapula endoprosthesis can be performed for most sarcomas arising from or involving the scapula, in lieu of a forequarter amputation. The trapezius, deltoid, latissimus, rhomboid, and serratus anterior muscles must be capable of being preserved in order to reconstruct the shoulder girdle with a total scapula replacement. A total scapula prosthesis provides an anatomic means of reconstructing the shoulder girdle as opposed to resecting the scapula and leaving the shoulder girdle flail and the extremity hanging. Contraindications include: tumor extension into the chest wall and the axilla, with

involvement of the neurovascular bundle; extreme involvement of surrounding pertinent periscapular muscles; complications from a prior biopsy with extensive contamination of surrounding tissues. Complications associated with limb-sparing surgeries for scapula tumors can include neurovascular injuries, tumor recurrence, infection, skin necrosis, and prosthetic dislocations. We have found these complications to be of low incidence when the procedure is performed as demonstrated in this video. Postoperatively, a stable shoulder girdle is restored, which is crucial for optimal hand and elbow function. Active shoulder motion is compromised. However, the functional outcome is superior to a forequarter amputation as well as to a flail shoulder that has not been reconstructed.

Wittig et al. in 2002, described their results of three cases of total scapula reconstruction following tumor resection. Patients scored 24 to 27 (80–90%) according to the Musculoskeletal Tumor Society Upper Extremity Functional Evaluation System. Six different categories are assessed with this system (pain, hand dexterity, emotional acceptance, lifting ability, hand positioning, and function and activities). The highest score per category is 5. A painless, stable shoulder girdle was restored in all patients. All patients retained a functional hand and elbow. Rotation below the shoulder level, which is required for most activities of daily living and personal hygiene, was preserved. Internal rotation, adduction, and extension strength were grade 5. Active shoulder abduction and forward flexion ranged from 25° to 45° and were grade 3. Each patient was capable of scapular protraction, retraction, and elevation, and the periscapular muscles contracted when carrying objects, thus assisting in upper extremity stabilization. Elbow flexion and extension strength were grade 5. Hand sensation and dexterity were normal, and grip strength was grade 5 for each patient. All patients could reach the top of their head, opposite shoulder and armpit, and perineal area with their hand. There were no limitations in activities of daily living, including feeding, dressing, and personal hygiene. Lifting ability was normal with the arm at the patient's side. Cosmesis was acceptable to all patients. The limitations were in activities that required raising the extremity above shoulder level.