

Management of Scapular Tumors



Matthew T. Houdek, MD^{a,*}, Benjamin K. Wilke, MD^b, Jonathan D. Barlow, MD^a

KEYWORDS

• Scapulectomy • Tikhoff-Linberg • Shoulder girdle

KEY POINTS

- Limb salvage surgery
- Upper extremity sarcoma
- Proximal humerus sarcoma

INTRODUCTION

The scapula forms the posterior shoulder girdle and is essential for function, acting as the origin or insertion for 17 different muscles.^{1,2} It is responsible for 6 types of motion, allowing for a functional upper extremity including elevation, depression, upward and downward rotation, protraction, and retraction.^{1,2} Because of the critical function of the scapula, limb salvage procedures are preferred as an alternative to forequarter amputation.

Scapular resections have advanced since the first reported partial scapulectomy performed in 1819 by Liston.³ Scapular resections were then limited to small series and case reports, and in 1909 De Nancrede concluded that anything less than a forequarter amputation for a shoulder girdle tumor was inadequate.⁴ Interscapulothoracic resection offered a limb salvage approach to patients with scapular tumors, and was first described by Bauman in Russian and then Tikhoff and Linberg in English;^{5,6} however, these resections were felt to potentially put the patient at high risk of recurrence secondary to the proximity of the brachial plexus and axillary vessels. Because of this, forequarter amputations were performed for many patients with large tumors involving the scapula until the 1970s. With advances in medical imaging, neoadjuvant treatments, and surgical techniques,

limb salvage surgery has become the preferred surgical technique, allowing for an oncologic margin while preserving hand function.⁷⁻⁹

PATIENT EVALUATION

Like other types of sarcomas, patients with a primary bone sarcoma of the scapula often present with a mass and pain, while patients with a soft-tissue sarcoma often present with a painless, enlarging mass. Because the scapula is surrounded by multiple muscles, masses can get quite large before presentation. In the scapula, the most common bony sarcomas are chondrosarcomas in adults and Ewing sarcoma in children and adolescents.¹⁰

A limb salvage surgery is contraindicated if the brachial plexus is involved and resection of the portions of the brachial plexus would lead to a nonfunctional hand that is not amenable to nerve or muscle transfer. Clinically, involvement of the brachial plexus can be suspected if there is intractable pain and motor deficits. In the initial work up of patients involves plain film radiographs and cross-sectional imaging consisting of an MRI with contrast and computed tomography (CT) scan of the chest. It is important to obtain an MRI that includes the entire chest, axilla, glenohumeral joint, and neck to evaluate the brachial plexus for tumor extension. In addition, a CT-angiogram and

^a Department of Orthopedic Surgery, Mayo Clinic, Rochester, MN, USA; ^b Department of Orthopedic Surgery, Mayo Clinic, Jacksonville, FL, USA

* Corresponding author. 200 First Street Southwest, Rochester, MN 55905.

E-mail address: houdek.matthew@mayo.edu

venogram are important to evaluate the axillary and subclavian vein, as obliteration of the vein can indicate involvement of the axillary sheath and subsequent involvement of the brachial plexus.

BIOPSY

If a sarcoma is suspected, a carefully planned needle biopsy is essential. It is important that the biopsy is performed under the guidance of the team that will ultimately be treating the patient, as an inappropriately placed biopsy could lead to an amputation.¹¹ Since scapular sarcomas often present with a large soft-tissue mass; a needle biopsy is often able to obtain lesional tissue. The biopsy should be performed posteriorly in line with the planned incision. Anterior biopsies should be avoided, if possible, to avoid contamination of the neurovascular structures. If the tumor is located in the scapular neck or glenoid, the biopsy should be placed through the posterior deltoid and teres minor.

Staging of Patients

Once a sarcoma is diagnosed patients should be staged based on tumor histology. For soft-tissue sarcomas, the patient requires a CT scan of the chest to evaluate for metastatic disease. For bone sarcomas, patients require a CT scan of the chest and either a whole-body bone scan or PET-CT scan for osteosarcomas or Ewing sarcomas. A bone scan or PET-CT is not required for chondrosarcoma unless a de-differentiated chondrosarcoma is diagnosed on the preoperative biopsy.^{12,13} In addition a bone-marrow biopsy is not required in patients with a Ewing sarcoma if there are no signs metastatic disease on other staging studies.^{14,15}

Once the patient has been staged, referral to medical oncology and radiation oncology is essential for the treatment of patients in a multidisciplinary fashion. Treatment is individualized and based on tumor histology. For patients with a Ewing sarcoma, interval compressed chemotherapy consisting of vincristine-doxorubicin-cyclophosphamide and ifosfamide-etoposide (VDC/IE) combined with surgical resection has improved survival.^{16,17} For patients with osteosarcoma, the standard treatment involves neoadjuvant chemotherapy, surgical resection, and adjuvant chemotherapy with the chemotherapy backbone including methotrexate, doxorubicin, and cisplatin (MAP).¹⁸ For patients with chondrosarcoma, surgery is the only option for management, as these tumors are resistant to chemotherapy and

radiotherapy.¹⁸ Because patients with soft-tissue sarcoma involving the scapula often present with deep, large, high-grade tumors, a combination of radiotherapy and surgical resection has become the standard treatment.¹⁹ Chemotherapy is typically not combined for treatment of patients with soft-tissue sarcomas, as the use of chemotherapy has not been shown to improve survival when combined with radiotherapy and surgery.²⁰

Surgical Resection

With the adoption of limb salvage surgery for patients with malignancies involving the shoulder girdle, different classification systems were developed to quantify the magnitude of the resection and the preservation of critical soft tissue structures. Currently the Malawer classification system and Musculoskeletal Tumor Society (MSTS) Classification of Skeletal Resections (Fig. 1) are the most utilized classification systems.^{5,21} Both systems stratify by location of the resection and the status of the deltoid and the rotator cuff, as preservation of the glenohumeral joint has been found to be most impactful on functional outcomes.^{5,21}

Patients with sarcomas of the scapula are managed with the patient in a rolling lateral position, to allow access to the posterior and anterior shoulder girdle. This should include the

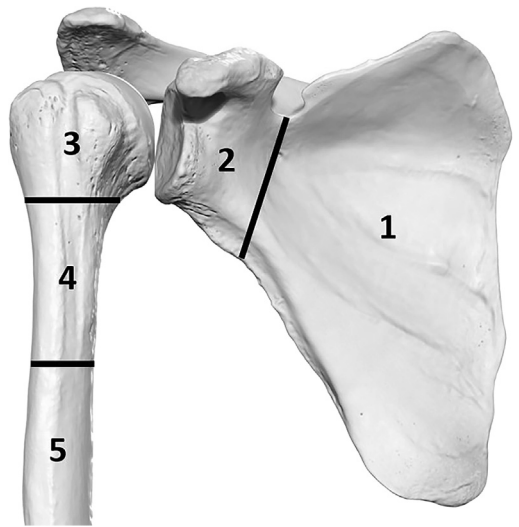


Fig. 1. Anatomic breakdown of shoulder girdle resections based on the MSTS system. The Tikhoff-Linberg classification is based on similar anatomic structures and would include Type 1: (3, 4, 5); Type 2: (1); Type 3: (1, 2); Type IV: (1, 2, 3); Type V: (2, 3, 4, 5); Type IV: (1, 2, 3, 4, 5). Both systems also include the status of the deltoid and rotator cuff, as either being intact (A) or deficient (B).

ipsilateral arm and neck, and the prep should extend past midline on the chest and back.

TOTAL SCAPULAR RESECTIONS

For patients who require resection of the entire scapula, (Fig. 2) or if the resection includes the proximal humerus, a utilitarian incision is utilized. This incision starts as an extended deltopectoral approach and extends proximally over the clavicle, curving posterior along the lateral border of the latissimus and then turning toward the inferior portion of the scapula and midline. The anterior portion of the incision should be explored first to evaluate if the brachial plexus and axillary vessels can be freed of the tumor. The pectoralis major is released from the humerus and retracted medial. The conjoint tendon and pectoralis minor are released from the coracoid, keeping a cuff of tendon on the coracoid. This allows exposure of the brachial plexus and vessels. The short head of the biceps is released distal to the subscapularis tendon at the level of the musculotendinous junction of the short-head of the biceps. It is important to carefully evaluate the preoperative MRI for tumor extension down the biceps tendon. This allows the biceps to be reflected medial, pulling the musculocutaneous nerve, allowing it to be protected. The shoulder can then be externally rotated, and the latissimus and teres major are reflected off the proximal humerus to expose the radial nerve. The anterior circumflex vessels are ligated, allowing the axillary artery and vein to fall medial. Often

the axillary nerve can be salvaged and can be freed off the inferior joint capsule anteriorly, and then the dissection is completed during the posterior exposure.

Based on the tumor extent, the deltoid can be released off the clavicle, acromion and scapular spine. In the process of doing this posteriorly, the axillary nerve and posterior circumflex vessels can be visualized. If the deltoid can be preserved, the nerve and vessels are kept in continuity with the muscle belly by freeing them from the underlying capsule, in addition by ligating the branches headed to the teres minor. The incision is then turned posterior and along the lateral boarder of the latissimus, curving toward the inferior angle of the scapula. Large skin flaps are created, and the trapezius and triceps are released from the scapula. It is important to evaluate the extent of the tumor in relation to the chest wall, as a portion of the serratus anterior muscle can often be preserved; however, if there is concern for tumor involvement, the serratus anterior should be included in the resection. The shoulder can then be internally rotated and the patient's hand placed behind the iliac crest in a figure-4 fashion. This pushes the tip of the scapula away from the chest wall, allowing for medial periscapular muscles to be reflected off the scapula. This is carried from an inferior to superior fashion, allowing the scapula to be everted away from the chest wall. Once this is completed, the clavicle can be osteotomized or the acromioclavicular joint is disarticulated,

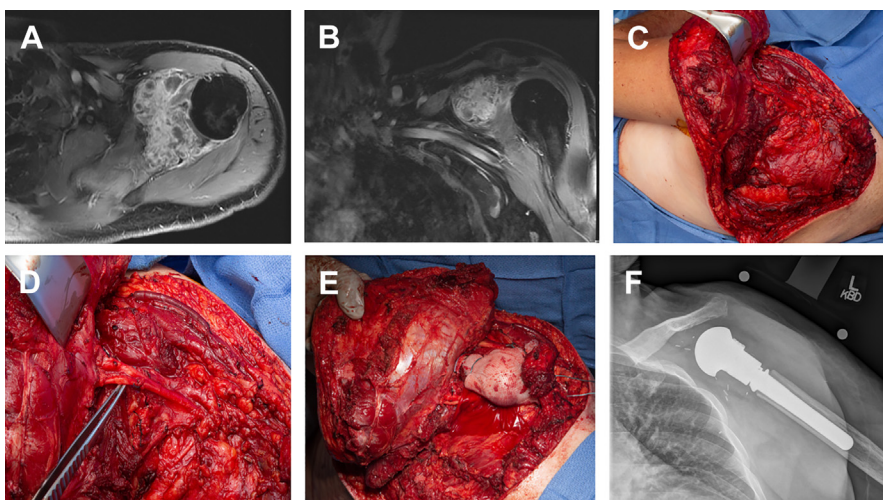


Fig. 2. Axial (A) and coronal (B) MRI with contrast showing a chondrosarcoma involving the scapula and glenoid. The patient underwent an extra-articular resection of the proximal humerus and scapula through a utilitarian incision with the posterior limb of the incision (C) shown following scapular resection with preservation of axillary nerve (D). The proximal humerus was reconstructed using an endoprosthesis wrapped in mesh (E), which was suspended from the clavicle (F).

allowing for mobilization of the specimen. If there is a large anterior soft tissue mass, the scapula can be lifted off the chest wall posteriorly, exposing the axillary vessels from the back and allowing for easy ligation of the suprascapular vessels and a neurectomy of the suprascapular nerve. Once the soft tissues of the scapula are fully released, the proximal humerus can be cut distal based on the extent of the tumor in the setting of an extra-articular resection, or the rotator cuff can be sectioned at the insertion of the humeral head circumferentially in the setting of an intra-articular resection. If there is difficulty removing the specimen following the humeral cut, it is often secondary to the preservation of the coracoclavicular ligaments based on the level of the clavicular osteotomy.

The residual deltoid and trapezius are repaired to any residual clavicle and can be tenodesed if there is no residual bone. It is important to use transosseous sutures if there is remaining clavicle. Based on the type of bony reconstruction, the residual soft tissues can be used to assist with closure, but also to restore function. The latissimus can be used to assist with external rotation by mobilizing it to a lateral, anterosuperior location on the residual humerus or implant.²² Similarly, the lower trapezius can be mobilized to the lateral, posterosuperior residual humerus, or implant to assist with external rotation.²³ The wound should be closed in multiple layers over large suction drains. The patient should be kept in an abduction shoulder immobilizer for soft-tissue healing. Patients are allowed active, active-assisted, and passive range of motion of the elbow, hand, and wrist immediately following surgery; however, shoulder motion should avoided until 6 weeks postoperatively. At that time, patients are allowed to begin stretching and deltoid retraining exercises.

PARTIAL SCAPULECTOMY

Partial scapular resections are typically performed from a posterior approach (Fig. 3), unless a portion of the glenoid needs to be resected along with a proximal humeral resection. For posterior approaches to the scapula, the patient is positioned in a rolling lateral position with the arm and neck included in the sterile field. The approach to the scapula is performed utilizing the posterior portion of the utilitarian approach used for total scapular resections. The incision can either be a "J" incision starting from approximately a hand's breath from the

lateral tip of the acromion and extending distally along the border of the latissimus before curving toward the tip of the scapula, or a diagonal incision extending from the tip of the scapula to the glenohumeral joint. The authors' preferred incision is a "J" incision, as it can be extended anteriorly in case the anterior neurovascular structures need to be explored.

Large skin flaps are elevated during exposure, and often the trapezius can be preserved. The deltoid is reflected off the scapular spine, exposing the underlying scapula. The latissimus is pulled inferior, exposing the serratus insertion. The shoulder can be internally rotated, and the patient's hand placed behind the iliac crest in a Fig. 4 fashion. This pushes the tip of the scapula away from the chest wall, allowing for the medial periscapular muscles to be reflected off the scapula. This is carried from an inferior to superior fashion, allowing the scapula to be everted off the chest wall. The lateral border of the scapula is then exposed by reflecting the teres and triceps, exposing the underlying neurovascular bundles. Branches entering the tumor are ligated, allowing the axillary nerve and posterior circumflex vessels to fall lateral. Once the scapula is completely exposed, the scapular osteotomy can be performed based on the tumor extent. The infraspinatus and the supraspinatus tendons can be released directly off the humerus, keeping a cuff of muscle on the tumor specimen. The supraspinatus will need to be released from a posterior to anterior fashion. The serratus is then released from the under surface of the scapula, allowing the surgeon to be able to pass his or her hand under the entire scapula, allowing for a tactile sense of where to create the osteotomy. The authors' preference is to utilize a harmonic scalpel to cut from a posterior to anterior fashion. The soft tissues of the subscapularis are then divided. This allows for the specimen to be rotated off the chest wall, exposing any neurovascular structures entering the specimen that can then be carefully ligated allowing for the specimen to be delivered from the field.

A soft-tissue only repair is frequently performed by suturing the residual deltoid and trapezius to any residual scapula, or these muscles may be tenodesed if there is no residual scapula. Any residual rotator cuff should also be sutured together, and the teres can be tenodesed to the latissimus to assist with rotation. The wound is closed in multiple layers over large suction drains, and the patient is kept in a shoulder immobilizer for soft-tissue healing. Patients

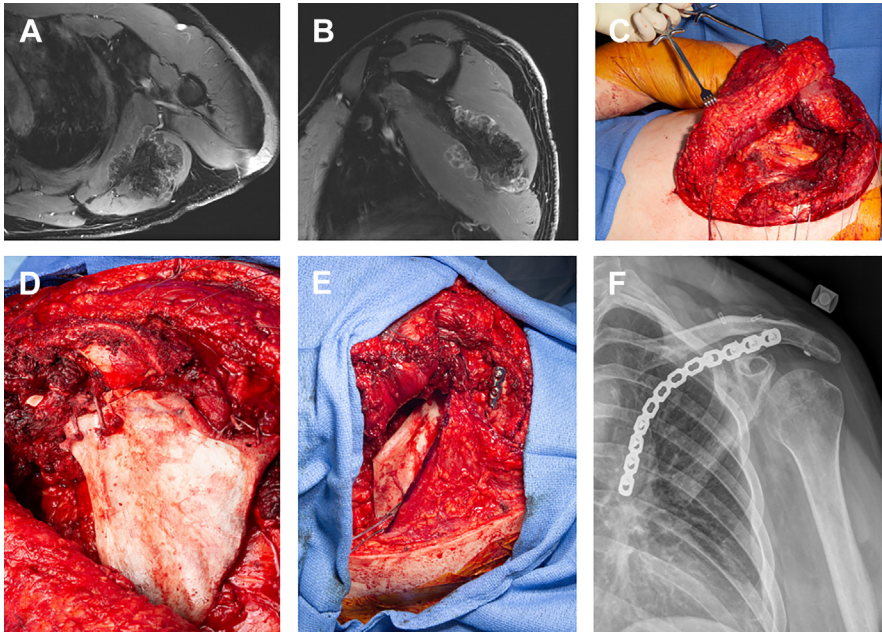


Fig. 3. Axial (A) and sagittal (B) MRI with contrast showing a secondary chondrosarcoma involving the inferior aspect of the scapula. The patient underwent a partial scapulectomy from a posterior approach (C). The scapula was reconstructed with an allograft (D) and soft-tissue reconstruction was performed with a lower trapezial (E) transfer with healing of allograft (F) at most recent follow-up.

are allowed to be active, active assisted, and passive range of motion of the elbow, hand, and wrist immediately following surgery; however, shoulder motion is avoided until 4 weeks postoperatively. At that time patients are allowed to begin stretching and deltoid retraining exercises.

CURETTAGE

Extended intralesional curettage should only be used in the setting of a biopsy-confirmed benign aggressive tumors or in the setting of symptomatic metastatic disease on a case-by-case basis. In this setting, a combined approach of curettage and adjuvants including thermal, mechanical, and chemical, provides local tumor control. The authors' preferred technique is to use a high-speed burr, hydrogen peroxide, and an argon beam electrocoagulator.

Reconstruction

The necessity of reconstruction following scapulectomy is a debated topic. For patients with partial scapulectomy, only a soft-tissue closure is necessary, and the patient should expect near normal function. If the glenoid is resected, the functional outcomes are inferior compared to if the glenoid and deltoid are preserved.^{24–26} If the glenoid is resected, often reconstruction can be performed with an augmented reverse

total shoulder base plate, combined proximal humerus allograft, and free vascularized fibula for arthrodesis, or suspension of the proximal humerus based on the extent of bony and soft-tissue resection.²⁷ Following partial scapular resection, complications are associated with the extent of soft tissue resection and involve mainly wound complications and subluxation of an implant if one is utilized.^{24–27}

For patients who have had a total scapular resection, in addition to the proximal humerus, series have shown the use of a scapular prosthesis, or an allograft scapula combined with a scapular thoracic arthrodesis, can return some shoulder function in selected patients.^{28–30} In limited series, the use of a scapular endoprosthesis could provide improvements in shoulder range of motion and functional outcomes; however, similar to partial scapular resection, functional outcomes are based on the amount of soft tissue preserved following tumor resection. If there is not sufficient soft tissue, a functional spacer of the proximal humerus suspended from the chest wall or residual clavicle can allow for a stable platform for elbow, wrist, and hand motion.³¹ Complications remain high following reconstruction for total scapular resection and are most commonly secondary to wound complications and related to the use of neoadjuvant radiotherapy.^{29,31} In addition to wound

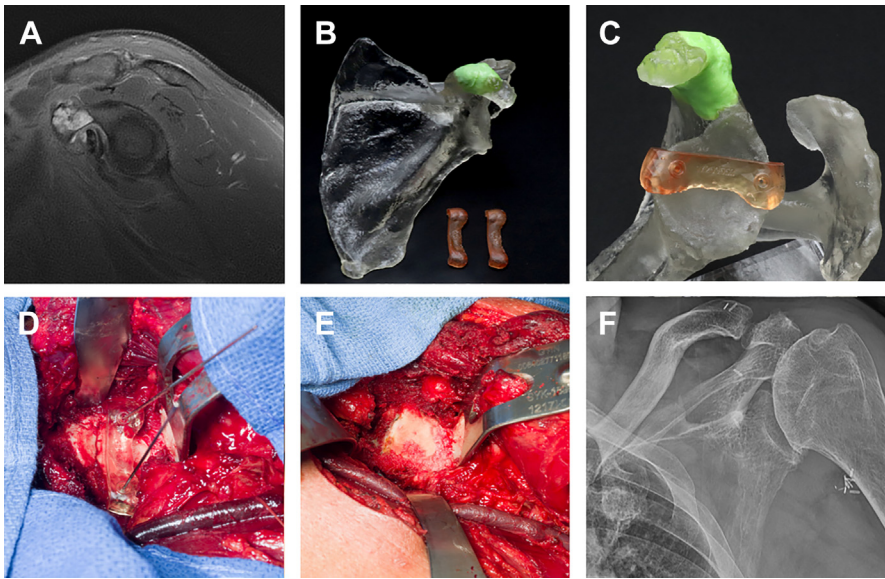


Fig. 4. Sagittal T2 (A) MRI showing a low-grade chondrosarcoma involving the coracoid. In order to reduce the size of resection, a 3-dimensional model and patient-specific guides were fabricated (B) to allow for an osteotomy of the coracoid from the glenoid (C). The guide was placed intraoperatively (D), and the cut (E) allowed for a negative margin excision and preservation of most of the glenoid (F).

complications, prosthetic complications associated with a humeral suspension include subluxation of the implant and stress shielding.³²

New Developments

Similar to other areas in orthopedics, the use of custom implants for reconstruction of complex oncologic resections has become a new option for surgeons following scapular resection.^{33–35} Although there is promise that these patient-specific implants could allow for functional recovery following scapulectomy, the results of these studies are limited to small series and case reports. In addition to custom implants, 3-dimensional printed cutting guides and anatomic models are allowing for smaller resections (see Fig. 4), and a partial scapular resection where previously larger resections were necessary to provide an appropriate oncologic margin.³⁶

Patient Follow-Up

Following oncologic resection, patients should follow-up routinely with the orthopedic and medical oncology teams based on the tumor histology. Follow-up involves physical examination and routine imaging. Radiographic imaging includes plain radiographs of the shoulder and pulmonary imaging consisting of a CT scan of the lungs, or a chest radiograph. The routine use of cross-sectional imaging of the operative site is debated, as a local recurrence can often

be detected by clinical examination alone.^{37,38}

At our institution for high-grade tumors, patients are followed every 3 to 4 months for the first 2 years postoperatively, every 6 months for years 2 to 5, and then annually for years 5 to 10. For low-grade tumors, follow-up is every 4 months for the first 2 years, every 6 months for year 3, and then annually for years 4 to 5. Following the completion of follow-up with the orthopedic oncology team for low- or high-grade tumors, it is recommended the patient follow-up with his or her primary care provider for periodic chest radiographs, as long-term metastatic disease has been reported.

Following oncologic resection, patients with scapular sarcomas have been thought to have improved oncologic outcomes compared with bone sarcomas located in other areas. In patients with scapular chondrosarcoma, previous series have demonstrated a low rate of recurrence;^{26,39} however, these were limited because of their lack of patients with high-grade tumors. In patients with a greater portion of high-grade tumors involving the scapula, 10-year survival is between 50% to 80%, and is strongly associated with the grade of the tumor.^{28,40,41} An important factor when treating patients with a scapular chondrosarcoma is to avoid any intralesional procedure. All scapular chondrosarcomas should be treated with wide local excision. Prior series have shown that patients who undergo an intralesional procedure are at increased risk of local

recurrence, and that local tumor recurrence is associated with metastatic disease.²⁸

SUMMARY

Scapular resections are large oncologic undertakings. Patient outcomes are strongly tied to the extent of resection, with the goal of surgery to provide a stable platform for elbow, hand, and wrist motion. Even though intralesional procedures have the potential to allow for a smaller resection, they should be avoided secondary to the risk of local recurrence and subsequent metastatic disease. Although there are options for scapular reconstruction, the use of endoprosthetic or allograft reconstruction of the scapula is limited to small case series with limited follow-up; as such, recommendations for reconstruction in all patients cannot be given.

CLINICS CARE POINTS

- Careful preoperative evaluation of the brachial plexus and axillary vessels is essential to determine if a limb salvage procedure can be performed. If two of the main nerves to the hand (median, radial, or ulnar) are involved and need to be resected, the patient's hand would be nonfunctional, and as such, a forequarter should be performed.
- In patients undergoing total scapular resection, the anterior limb of the incision should be performed first to evaluate the involvement of the brachial plexus and axillary vessels.
- Following oncologic resection, reconstructive options are limited in the setting of a total scapular resection. As such, the goals for reconstruction remain providing a stable platform for elbow, wrist, and hand use.

DISCLOSURE

The authors have no disclosures.

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