Surgical Excision of Mycosis Fungoides Using Thumb-Sparing Reconstruction

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Abstract

Background: The differential for soft tissue tumors of the hand and upper limb is broad. Hematologic malignancy remains quite low on the differential for soft tissue tumors involving the hand, and there is little in the literature describing surgical management of such cutaneous manifestations. When the tumor is large or involves the thumb, careful consideration of reconstructive options is required. **Methods:** We present a rare case of an aggressively enlarging mycosis fungoides, a cutaneous T-cell lymphoma tumor, involving the thumb. This tumor had a history of multiple failed treatment attempts, including radiation and chemotherapy. **Results:** Our surgical plan was a reverse radial forearm osteocutaneous flap. **Conclusion:** A reverse radial osteocutaneous forearm flap was successfully used to avoid thumb amputation and preserve thumb function.

Keywords: mycosis fungoides, thumb reconstruction, hand tumor, radial forearm flap, upper extremity

Case Report

A 78-year-old African-Canadian female with a history of mycosis fungoides (MF), which was first diagnosed in 1985 and referred to our center in 2005. Many different modalities had been used on her MF previously. These included interferon, psoralens plus ultraviolet light, targretin, nitrogen mustard, total skin electron beam therapy, and radiation. Despite this, by 2007, the MF progressed to a tumor on her right hand thenar eminence. This tumor responded to nonsurgical intervention using topical Aldara and radiation. She remained in remission until July 2010, when a new tumor arose on her left thumb. Biopsy of this lesion found Pautrier microabscesses, epidermotropism, and atypical lymphoid cells (all typical findings for MF) with some evidence of transformation estimated at 50% to 60% of large-cell atypical forms. The patient underwent multiple cycles of radiation therapy resulting in a 2-year cumulative dose near 6000 cGy to this region of the thumb. It remained resistant to radiation, and she was treated with chlorambucil and intralesional methotrexate injections. These methods also failed to halt tumor progression after a 1-month trial.

She was referred for thumb amputation in August 2012 (Figure 1). Pathology reports at this time showed an increase in large lymphocytes to 80% of the infiltrate indicating further transformation to large-cell MF. The patient remained well with no systemic illness or palpable lymphadenopathy. Magnetic resonance imaging (MRI) showed a soft tissue mass that measured $4.4 \times 4 \times 7.1$ cm in the

Figure 1. Appearance at presentation.

thenar eminence with extension into the first metacarpal, flexor pollicis longus, and extension along the radial aspect of the flexor retinaculum (Figures 2a, 2b, 2c, and 2d). The large defect after en bloc resection was predicted to require a flap of vascularized soft tissue for coverage of the defect. Regional options included the reverse radial forearm flap

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Figure 2. Magnetic resonance imaging of hand tumor. (a) T1 coronal view of arm; (b) T1 axial view of hand; (c) T1 sagittal view of hand; and (d) T1 coronal view of hand.

and posterior interosseous flap. Free flap options included free radial forearm, anterior lateral thigh flap, and latissimus dorsi free flap. The most likely flap based on imaging and senior surgeon (P.B.) experience was the reverse radial forearm flap. Informed consent was obtained from the patient by explaining the risk and benefits of the proposed surgery. Risks specific to this procedure included the risk of partial or complete flap loss, large forearm scar, and potential poor hand function outcome.

The left thumb was marked with a 1-cm margin around any obvious cutaneous manifestation of the tumor. The en bloc resection contained left thumb metacarpal, thenar musculature, motor branch of median nerve to thenar musculature, radial digital nerve to thumb, and the extensor pollicis brevis muscle. The resection resulted in a bony defect that measured approximately 5 cm from the trapezium to the proximal phalanx (Figure 3). The skin paddle was approximately 12 × 7 cm. Using reverse planning, a template of the skin defect was marked out on the forearm over the longitudinal axis of the reverse radial forearm flap. The location of the skin paddle was marked out in consideration of the planned bone flap and the distal perforators which supply the bone. The flap was elevated proximal to distal as is typical for a reverse radial forearm flap preserving the perforators to the bone. With the skin portion elevated and exposure

Figure 3. En bloc resection of tumor with 1-cm margins.

of the radius, the bone was marked out and elevated first cutting the cortex with an oscillating saw and then a series of osteotomes. The length of the radius harvested was 7 cm, and it was beveled at both ends. The pedicle and the bone flap were mobilized to allow for elevation and turned 180° into the defect (Figure 4).

The proximal phalanx and trapezium were beveled to match the cuts on the radius and achieve exposed matching cancellous surfaces. The proximal and distal ends were secured using standard lag screw technique. Two

Figure 4. A reverse radial forearm osteocutaneous flap harvested with bone length of 7 cm.

Figure 5. Fluoroscopic intraoperative image demonstrating bony fixation of the radial bone graft to recipient site.

2.0-mm screws were used proximally and two 2.0-mm screws distally. Fluoroscopy was used to guide fixation of the graft and positioning (Figure 5). During fixation, the thumb pulp was positioned for pinch rather than abduction because this was felt to be the most useful position in this 78-year-old patient. The flexor pollicis longus tendon was salvaged so that thumb interphalangeal flexion could be preserved. A split-thickness skin graft was harvested from the left thigh and was placed as a sheet graft over the forearm (Figure 6). The patient was discharged on postoperative day 1. The reconstructed thumb and donor forearm were protected with a short-arm thumb spica cast for 6 weeks. After that, range of motion and opposition exercises were initiated. Heavy lifting and weight bearing greater than 5 lbs were avoided for 6 weeks.

Discussion

The differential for soft tissue tumors of the hand and upper limb is broad (Table 1). In all cases of suspicious

Figure 6. Appearance of flap and scars at 6-month follow-up.

soft tissue masses in the hand, the workup should include a thorough history and physical exam, and appropriate imaging (x-ray film, MRI, ultrasound, and tissue biopsy if concerning features are present). Mycosis fungoides appears quite low on this differential due to its very low incidence in the hand. Subsequently, there is a paucity of information regarding its surgical management. Mycosis fungoides is a type of extranodal non-Hodgkin lymphoma that was named in 1806 after reports of skin lesions that evolved into mushroom-like tumors. Mycosis fungoides is the most common type of cutaneous T-cell lymphoma (CTCL) tumor and accounts for almost 50% of all primary cutaneous lymphomas with an incidence reported to be 0.29 per 100 000 people per year.^{9,10} Histologically, MF is characterized by a proliferation of small- to medium-sized T lymphocytes with cerebriform nuclei.¹⁰ The mean age of diagnosis has been reported to be around 55 to 60 years of age. Mycosis fungoides is 2.2 times more common in males than in females and has an ethnic predilection for African Americans.¹⁰ The typical clinical course for MF has a slow disease progression ranging from several years to several decades. The disease process initially becomes clinically evident as macules erupting on non-sun-exposed areas of the skin. The next stage is the patch or plaque phase which resembles eczema or psoriasis in appearance. The final advanced stage is large skin plaques with tumors. Isolated tumor lesions without a history of plaques or patches should prompt reconsideration of the diagnosis, as this progression of stages is the hallmark of MF. The advanced-stage tumors often become complicated by ulceration and secondary infection with resulting loss of function. Patients with advanced clinical stages, particularly those with tumor-stage disease, have a high incidence of large-cell transformation.² This transformation is reported to occur in 8% to 23% of patients with MF.^{2,7} Large-cell transformation is associated with poorer prognosis and is identified by the presence of lymphocytes greater than 4 times the size of a small lymphocyte in more

Table 1. Differential Diagnosis of Soft Tissue Lesions of the Hand.

Soft tissue lesion	Defining clinical feature
Benign	
Ganglion	Cystic subcutaneous mass affixed to underlying structures
Giant-cell tumor of tendon sheath	Nodular subcutaneous firm tumor arising from tendon sheath
Epidermal inclusion cyst	Globular firm mass in dermis/epidermis.
Lipoma	Rubbery subcutaneous mass
Schwannoma	Mobile subcutaneous mass $\pm$ Tinel sign
Nodular fasciitis	Rapid growing firm fixed mass (mimics sarcoma)
Pyogenic granuloma	Hemorrhagic friable lesion that occurs in response to trauma
Glomus tumor	Paroxysmal pinpoint pain and cold sensitivity
Malignant	
Squamous cell carcinoma	Most common cutaneous tumor, presents as nonhealing wound
Basal cell carcinoma	On skin-exposed areas, slow growing
Melanoma	Subungual pigmented lesion
Sweat gland and adnexal tumors	Slowly growing painful mass
Metastatic tumor	Variable findings, often present with pain, redness, swelling
Epithelioid sarcoma	Slow-growing firm lesion may ulcerate, associated with lymph node metastasis
Synovial sarcoma	Slow-growing painless mass
Undifferentiated pleomorphic Sarcoma	Painless mass, rapid growth after trauma
Mycosis fungoides	Slow progressive course from skin lesions to tumor

than 25% of the infiltrate.² One study reported median survival of 8.3 years for patients with large-cell transformation as compared with 18.3 years for all patients with MF.¹ This study also identified additional factors associated with poor prognosis, including advanced clinical stage, increased age, male sex, and increased lactate dehydrogenase.

Treatment for CTCL ranges from local to systemic approaches depending on the extent of disease. Goals of care are largely directed toward local symptomatic relief, as a curative regimen outside of an allogeneic or autologous bone marrow transplantation has not yet been elucidated. When MF is limited to the skin, local treatment includes psoralens plus ultraviolet light (a photoactivated compound that inhibits DNA and RNA replication), topical chemotherapy (such as nitrogen mustard), and radiation therapy.^{2,10} If nodes and other visceral organs, or skin lesions that are refractory to local therapy, are involved, systemic chemotherapy may be considered.¹⁰ Surgical excision is not considered curative for MF at this time, and no recommendations regarding margins for excision to result in reduced incidence of local recurrence have been established in the literature.

While overall survival is unaltered for a limb-sparing approach versus amputation for the resection of soft tissue sarcomas, evidence for the resection of cutaneous hematologic malignancy is deficient. Simple resection of primary cutaneous B-cell lymphomas has been described using 5-mm margins in a group of 9 patients.^{4,6} All 9 cases were disease-free after 5 years with no recurrence and no complications.

In the group which received radiation treatment, 14 out of the 16 individuals experienced skin changes (itch, erythema, and swelling) that lasted an average of 1.6 years, and 2 had chronic nonhealing ulcers. Based on this observation, it was suggested that surgical resection be considered as a first-line treatment option for primary cutaneous B-cell lymphomas.⁴ A case report of ocular CTCL described simple surgical resection without specific margins and reported a 3.5-year recurrence-free follow-up.⁸ Other rare subtypes of ocular cutaneous lymphoma have been treated effectively with surgery alone.³ We elected to use a 1-cm margin for surgical resection to maximally reduce locoregional recurrence in the setting of our complex thumb reconstruction. Additional long-term studies and standardized approaches with well-defined surgical margins would be beneficial in further delineating the success of surgical management as defined by functional recovery and rates of local recurrence. Due to the relatively small number of patients presenting at any particular center with MF, the ability to conduct larger studies yielding adequate statistical power is extremely limited.

The reverse radial forearm flap is a fasciocutaneous flap based on retrograde flow from the distal radial artery.^{5,11} Based on the preoperative Allen test and MRI imaging, it was predicted that the radial artery would be patent and available for this case. The reverse radial forearm flap is a popular flap because no microvascular anastomoses are required, regional tissue is used with a good match to recipient site, and the tissue is thin and pliable which is best for optimizing hand functional outcome.⁵ Furthermore, the reverse radial forearm flap can be harvested with bone

and sensory nerves to achieve a 1-stage reconstruction with all necessary parts in 1 flap. Drawbacks include sacrifice of the radial artery, large donor site defect, and weakening of the radial bone. We elected not to prophylactically plate the radius as the amount of bone remaining after harvest appeared adequate to maintain radius stability. Although the sample size was small, previous authors have placed an iliac crest bone graft in the donor radius bone.

Other important considerations in the surgical management of CTCL include the cumulative dose of local radiation to the soft tissues, which could hinder reconstructive outcomes and the stage and transformation of the CTCL, and may impact long-term survival. In current practice guidelines, CTCL is treated with radiation therapy and chemotherapy as first-line agents. We have presented a case of MF with a history of multiple failed treatment attempts, including radiation and chemotherapy, followed by surgical excision and successful reconstruction. The risk of recurrence is expected to be low with 1-cm surgical margins. A reverse radial osteocutaneous forearm flap was used to avoid thumb amputation and preserve thumb function.⁹

Ethical Approval

This study was approved by our institutional review board.

Statement of Human and Animal Rights

All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008.

Statement of Informed Consent

Informed consent was obtained when necessary.

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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