

Anterior chest wall reconstruction for cutaneous involvement of Hodgkin's lymphoma

Hodgkin lenfomasının deri tutulumu için anterior göğüs duvarı rekonstrüksiyonu

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Abstract

Hodgkin lymphoma with initial thoracic involvement is common and it is more frequently localized in the mediastinum, followed by lung parenchyma and the pleura. However, skin involvement of Hodgkin's lymphoma is extremely rare. We report this case to illustrate a rare and unique presentation of a potentially curable disease, with resistance to standard treatment. Free tissue transfer was used as an adjunct to provide a better quality of life for the patient with Hodgkin lymphoma complicated with extensive anterior chest wall ulceration exposing lung and pericardium.

Keywords: Hodgkin lymphoma, Cutaneous, Anterior chest wall, Reconstruction

Öz

Toraksik tutulum gösteren Hodgkin lenfoma yaygındır ve mediastende daha sık lokalize olur, bunu akciğer parankimi ve plevra izler. Ancak Hodgkin lenfomanın cilt tutulumu oldukça nadirdir. Bu vaka sunumunda, tedavi edilebilir bir hastalığın nadir ve tedaviye dirençli prezentasyonunu sunmaktayız. Akciğer ve perikardiyumun teşhir olduğu yaygın göğüs ön duvarı ülserasyonu komplikasyonu ile başvuran Hodgkin lenfomalı hastanın yaşam kalitesini arttırmak amacıyla serbest doku transferi yapıldı.

Anahtar kelimeler: Hodgkin lenfoma, Kütanöz, Anterior göğüs duvarı, Rekonstrüksiyon

Introduction

Hodgkin's lymphoma (HL) encompasses 20 to 30% of all lymphomas [1]. The cutaneous involvement in Hodgkin's disease is almost always secondary to visceral or nodal involvement and is a rare condition, occurring in only 0.5 to 3.4% of the cases that typically occurs late in the progression. This unusual condition is believed to have declined in incidence in recent decades, likely due to improved treatment of patients with Hodgkin's disease who received modern combined chemotherapy, radiation therapy, and the role of peripheral blood stem cell transplantation [2].

Hodgkin's lymphomas are usually treated medically, with surgery being utilized for local control. Commonly, the cytotoxic chemotherapy is preferred for the treatment of malignant lymphoma. In well-chosen cases, surgical treatment can significantly contribute to a good outcome and better quality of life [3].

The patient described in this case report is unique whereby she presented with huge anterior chest wall ulceration which was successfully treated by using a combination of chemotherapy and free tissue transfer.

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Case presentation

A 40-year-old woman was diagnosed with stage IV (liver) mixed cellularity subtype Hodgkin lymphoma with bulky mediastinal mass in January 2006. She completed ABVD (Adriamycin, Bleomycin, Vinblastine, Dacarbazine) regimen and achieved partial remission with a significant residual anterior mediastinal disease. There was a delay in the commencement of salvage chemotherapy as she eventually was lost to follow-up for a year and presented back with anterior chest wall ulceration (Figure 1). Two months prior to presentation, she had skin ulceration over the anterior chest wall preceded by persistent discharging sinus. The infected wound secreted copious amount of foul smelling discharge. There was no constitutional symptom.

Clinically, the wound measured 15x12 cm with 4 cm depth with sloughy wound bed, exposed and eroded sternal cartilage with foul smelling serous discharge. Multiple cervical nodes were palpable. She had no sign of upper airway obstruction. Chest CT scan demonstrated multiple enlarged calcified nodules seen within the mediastinum, extensive sternal bone destruction, and extension to the anterior chest wall, exposing right pleura and pericardium, from the level of T2 to T11 costal cartilage. No lung involvement and no osteomyelitic changes were seen. Biopsies of cutaneous lesions revealed histological subtypes of Hodgkin disease consistent with those observed in the lymph nodes.

She underwent wound debridement and excision of the inflamed margin with preservation of thoracic cage. The wound bed was covered with pedicled omental and supercharged pedicled transverse rectus abdominis myocutaneous (TRAM) flap and concurrent chemoport insertion on right upper chest (Figure 2A, 2B, and 2C). Postoperatively, it was complicated with wound infection and dehiscence resulting in partial flap loss (Figure 3). The wound was then re-debrided prior to secondary coverage with free latissimus dorsi myocutaneous flap using perforators from internal thoracic as recipient vessel (Figure 4). During the recovery period, multiple sinuses developed on the lateral aspect of the flaps with continuous serous discharge. It took almost six months of regular dressing with concurrent treatment with monoclonal antibody targeting CD30 antigen on Hodgkin cells, brentuximab vedotin monotherapy for the wound to finally heal.

Upon completion of four cycles of brentuximab, her mediastinal disease slightly increased in size with presence of new lung nodules and hence her treatment has now switched to GDC (Gemcitabine, Carboplatin, Dexamethasone) protocol palliative chemotherapy. There is no evidence of cutaneous disease recurrence at one year of follow-up and she is able to resume most of her daily house chores (Figure 5).



Figure 1: Huge ulceration over the anterior chest wall



Figure 2A: Wound bed preparation



Figure 2B: Pedicled omental flap and pedicled TRAM flap dehiscence



Figure 2C: Flap inset



Figure 3: Wound breakdown after the first operation



Figure 4: Free latissimus dorsi myocutaneous flap to cover the wound

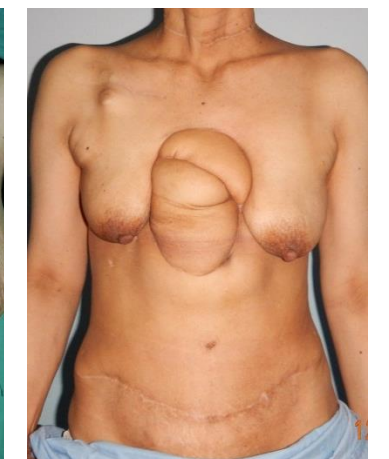


Figure 5: Final result after 1 year post-operatively

Discussion

Most patients with HL present with asymptomatic superficial lymphadenopathy. The usual locations of the disease include cervical (more than 80%), supraclavicular and mediastinal lymph nodes (more than 50%) while sub-diaphragmatic presentations, bone marrow, and hepatic involvement are rare.

Secondary cutaneous HL occurs in 0.5 to 3.4% of cases and is almost always secondary to visceral or nodal involvement. Histologically, it resembles nodal disease [1]. The mechanisms for the skin involvement in HL are unknown but suggested to be retrograde lymphatic spread derived from tumour-involved lymph nodes, direct extension into the skin by tumour cells in underlying lymph nodes, or haematogenous spread of the tumour [6]. The commonest clinical presentation usually involves a single or multiple dermal or subcutaneous nodules. Cutaneous involvement usually denotes stage IV disease, and it carries an ominous prognosis [1].

Our patient demonstrated an exceptional manifestation of mediastinal Hodgkin's lymphoma, an initial appearance with an anterior chest wall ulceration without symptoms of upper airway compression. The invasion to the sternum may have been directly from the anterior mediastinal lymph nodes.

The advent of more effective treatment options has improved the 5-year survival rates (81%) that are unmatched in any other cancers over the past four decades. Every patient with newly diagnosed HL has a possibility of being cured with the proper treatment [5]. However, 20–30% with advanced disease will be refractory to initial treatment. In the management of refractory disease, salvage chemotherapy is used, followed by autologous stem-cell transplantation and high dose chemotherapy [7].

There are several case reports and case series depicting the behaviour and treatment of the cutaneous involvement of HL but none so far describing the incorporation of free tissue transfer in the management of HL [2,4,8,9].

Prior to commencement of surgical management of this patient, histological diagnosis is warranted to rule out different malignancies in patients presenting with a mediastinal mass invading the anterior chest wall. Possible tumours include carcinoma and sarcoma of primary sternal tumour or the breast itself.

The ultimate goal in chest reconstruction is to eliminate dead space, preserve adequate chest wall solidity and deliver coverage while preserving form and function [10].

The chest wall defect can be categorized as superficial and deep whereby the superficial defects that involve only the soft tissues of the chest wall, without exposure of vital structures are comparatively easier to manage and can be treated effectively with skin grafts or local flaps. Full thickness defects exposing bone and cartilage, or even vital structures, for example lungs, heart, or a major vessel, necessitates different reconstructive options [11].

In this patient, the defect was wide, exposing the eroded sternum and adjacent ribs. The wound bed was sloughy with a copious amount of serous discharge. Thus, local infection was treated with regular dressing and all devitalized soft tissues debrided prior to coverage with vascularized soft tissue flaps.

The patient was underweight and cachexic, and not enough tissue bulk was available to fill the defect of a large area within the deep cavity, hence the need for an additional procedure. This was achieved by usage of vascularized omental flap to cover the bottom half of the defects [12].

The supercharged pedicled transverse rectus abdominis muscle flap provides an additional bulk for resurfacing the chest wall and cutaneous coverage to completely seal the defect. Therefore, both flaps were able to be raised through a single incision, involving an adjacent surgical field to the defective area.

The wound dehiscence caused her to lose a quarter of the coverage of the wound subsequently causing the upper part of the initial defect to be exposed due to gravitational effect and wound contraction. The free latissimus dorsi myocutaneous flap was used to salvage the remaining defect. The development of the sinuses on the lateral part of the flaps were treated with regular flushing and frequent dressing changes until it was fully healed.

Post-operatively, the patient was started on brentuximab. It is indicated in relapsed Hodgkin lymphoma and relapsed systemic anaplastic large-cell lymphoma after FDA accelerated approval, based on the result of two single-arm trials [13]. Unfortunately, her disease progressed after completion of four cycles. She was then started on GDC (Gemcitabine, carboplatin, dexamethasone) therapy and the treatment is still ongoing. Despite that, she regained part of her quality of life without the troublesome wound on her chest.

Conclusion

Although the cutaneous involvement of Hodgkin lymphoma is rare and the prognosis is thought to be extremely poor, our case among others demonstrates that a good response to current treatment and surgical management does offer an aide in the management of extensive cutaneous involvement in Hodgkin's disease.

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