CASE REPORT

Radiotherapy and collagenosis: is it safe? - A case report of reirradiation in a patient with scleroderma

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Abstract

Historically, scleroderma and other collagenous diseases have been considered a relative contraindication to radiation. The literature has few studies describing poor outcomes and cosmesis in this situation and there are almost no data concerning about reirradiation and colagenosis. The authors describe a case of a patient with a soft tissue sarcoma in the arm submitted to conservative surgery. They describe the outcome, cosmesis and function of this rare twice-irradiated scleroderma patient.

Keywords: brachytherapy, case report, collagenase, localized, radiotherapy, scleroderma.

INTRODUCTION

The relatively rare incidence of soft tissue sarcoma (STS) associated with a large quantity of anatomic sites affected and histopathologic diversity contributes to the treatment becoming a challenge; when it occurs in distal parts, the association of conservative surgery and external radiotherapy (ERT) is an effective alternative to radical surgery[1].

However, the described case presents a peculiarity that historically has proved to be a relative contraindication to teletherapy: scleroderma[2]. The authors describe the results of primary treatment using an intraoperative high dose rate of brachytherapy (IO-HDR) and reirradiation using ERT.

CASE DESCRIPTION

A 54-year-old female, Caucasian, was admitted to Hospital A.C.Camargo on November 24, 2008, complaining about the appearance of a nodule on her left elbow, appearing earlier in May 2008.

Physical examination revealed a nodular lesion with cystic content involving the lateral and proximal faces of the left forearm (near the elbow), apparently superficial. There were no neurological symptoms.

The patient claimed to have undergone an ultrasound in her city of origin, which showed a cystic lesion, subsequently drained (content despised). As there was a remaining nodule and progressive growth, the lesion was resected (April 11, 2008). Histopathology analysis revealed a high-grade pleomorphic sarcoma with overlapping edges.

Additional tests were made as follows:

- Chest CT scan: no evidence of metastatic lesion;
- Echocardiogram: mild pericardial effusion;
- Chest radiograph: fibrosis scars;
- Abdominal ultrasound: no changes.

After a multidisciplinary discussion, observing the patient already had cardiopulmonary complications due to an underlying disease, the therapeutic proposal was conservative surgery associated with intraoperative radiotherapy and plastic repair with surgical flap.

On May 12, 2008, the patient underwent procedure: the tumor measured approximately 8.0 x 3.0cm and was involving superficial and deep layers, in contact with the joint and in proximity to the medium nerve. Three-dimensional resection was performed *en bloc* with the joint capsule and the deep border in the cranial part of the ulna´s periosteum. Margins were free (T2bNx pathological stage).

For intraoperative brachytherapy a silicone mold was used with five drive catheters and an after-loading automatic system using Iridium-192 source. 18.5 Gy was prescribed in a single dose to 1.0cm from applicator's surface.

The surgery was completed using triceps flap for reconstruction of the joint. The definitive histopathology examination revealed a high-grade spindle cell sarcoma. Immunohistochemical analysis was compatible with high--grade pleomorphic sarcoma.

After 9 months of follow up, despite having no complications related to the IO-HDR, there was a growing exophytic lesion in the surgical bed measuring about 0.5 cm. Biopsy confirmed local recurrence (same histological features).

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On March 6, 2009, the patient was re-operated: a pathological exam revealed free margins of a high-grade spindle cell sarcoma measuring 0.8 cm in diameter and infiltrating derm and hypoderm.

Less than two months later, ERT was performed using a three-dimensional technique and conventional fractionation. The treatment plan was concluded in two phases: 40 Gy to operative bed and scar followed by 20 Gy boost to operative bed.

The initial tolerance to teletherapy was good, with patient presenting grade 1 fatigue and grade 1 radiodermatitis (RTOG). In the long term, however, the patient developed scar dehiscence which evolved to a chronic ulcer measuring about 2.0 cm in diameter, unresponsive to conservative medical treatment.

On March 30, 2010, a new surgical procedure was performed for the ulcer treatment, requiring skin grafting.

After almost three years from the teletherapy (last follow up on March 1, 2012), the patient is being followed up without evidence of local or systemic recurrence. Functionally, what is observed is a small limitation of 10 degrees in the elbow extension, as well as its full flexion (minimum angle of about 30 degrees).

DISCUSSION

The treatment of STS in distal parts has evolved from the limb amputations in the 70s to conservative surgery associated with radiotherapy in the 90s, without loss in the local control rates and disease-free survival.[1]

Eventually, when anatomical implications of the tumor implantation makes the application of conventional radiotherapy difficult, there is the possibility to use intraoperative radiotherapy as a plausible complementary modality instead of teletherapy[3].

In 2007, Gold published a study with radiotherapeutic management of 20 patients of the Mayo Clinic (*Rochester, MN, USA)* suffering from scleroderma. Of all patients, 15 (75%) had acute adverse effects, three of them classified as serious. In relation to chronic adverse effects, 13 (65%) were diagnosed, and also 3 (15%) were classified as serious[4].

Due to the peculiarity of the case (high probability of occurrence of adverse effects of treatment) and based on literary evidence, we chose to perform an intraoperative high dose rate of brachytherapy associated with conservative surgery. The objective of minimizing the side effects of radiotherapy has been reached, since after 10 months from the procedure (May 5, 2008 to March 6, 2009) the patient remained asymptomatic. This period is greater than the average time reported for the onset of late toxicity observed by Gold et al.[4].

In fact, after recurrence and the second course of radiotherapy, a chronic ulcer could be noticed, but was successfully treated surgically. No other serious toxicities could be noticed which is considered a positive result - in agreement with other publications[5] and remembering the baseline diagnosis of scleroderma and its potential detriment.

Despite the diagnosis of scleroderma, reirradiation using IO-HDR and ERT was feasible for this patient. After more than 2 years, it seems to have reasonable results in terms of tumor control and functionally.

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