Surgical Treatment of Enormous Recurrent Dermatofibrosarcoma Protuberans

Shijie Xing[1] Jianjun Wang[1] Wei Zhai[1] Tian Xia[1] Chuanxiao Wang[1]

[1] Department of Thoracic Surgery, Union Hospital, Tongji Medical College, Huazhong University of Science and Technology, Wuhan, China

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Address for correspondence Jianjun Wang, PhD, Department of Thoracic Surgery, Union Hospital, Tongji Medical College, Huazhong University of Science and Technology, Liberation Road, 1277#,Wuhan, Hubei Provience, P.R. China, Wuhan 430022, China (e-mail: xingshijie128@163.com).

Abstract

A patient with enormous recurrent dermatofibrosarcoma protuberans underwent modified three-dimensional histology surgery. Frozen-section examination was used to identify the margins. The patient had a normal postoperative course.

Keywords

- chest wall
- tumor
- surgery/incisions

Introduction

Dermatofibrosarcoma protuberans (DFSP), a primary soft tissue tumor, is relatively uncommon with an estimated incidence rate of 4.2 to 4.5 cases per million persons each year in the United States.[1] It is considered a relatively rare soft tissue tumor with intermediate- to low-grade malignance. The main problem of DFSP is not metastasis, which rarely occurs, but local aggression with a high recurrence rate. Surgery is the preferred treatment of DFSP; three-dimensional histology surgery is a new concept. During the procedure, a narrow lateral strip (1–1.5 cm wide) is excised around the perimeter of the tumor border and then a horizontal slice is excised from the bottom of the tumor. Both the strip and the slice are sent for routine pathologic examination. The procedure will not stop until all surgical margins are tumor-negative. In this article, we described a case of DFSP with modified three-dimensional histology surgery and rush frozen-section examination.

Case Report

A 51-year-old man presented to our department with a 9-month history of progressively growing ulcerated mass in the right anterior chest wall. The patient was a heavy smoker (2 packs/d for at least 30 years). He had been a heroin addict until 2010 and had undergone one operation for chest mass in the compulsory rehabilitation center in 2007. The mass had been around 4 cm in December 2010, without pain, redness, and burning. It gradually enlarged, and the patient experienced chest pain and body weight loss (15 kg at least) for 9 months. On physical examination, an enormous, foul-smelling, easy-bleeding mass in the right anterior chest wall was noted, covered by green mosslike material. The mass was cauliflower-shaped with a stem connected to the deeper layer. Varicose veins could be found on the peripheral skin. Neither enlarged superficial lymph nodes nor metastases were revealed during physical examination. On computed tomography (CT) scan, one thoracic neoplasia, measuring $6 \times 17 \times 20$ cm, was identified outside the thoracic cavity without apparent boundaries to adjacent muscles. The CT value (Ct) of the mass was 13 Hounsfield units (HU) versus the normal surrounding muscle value of 36 to 38 HU. Slight hydrothorax and pneumonia were noted (►**Fig. 1A**). Complete blood count indicated moderate anemia (erythrocytes: 3.17 T (tera, 10^{12})/L, normal range: 4.0 to 5.5 T/L; hemoglobin [Hb] 71 g/L, normal range: 120 to 160 g/L; mean corpuscular volume [MCV] 71.6 fL, normal range: 82 to 92 fL), and increase in platelet counts (PLT; 441 g/L, normal range: 100 to 300 g/L). Coagulation profile, serum biochemistry, and electrolytes were mostly within normal limits except for decreased serum albumin (27.2 g/L; normal range: 40 to 55 g/L). Abnormal results of routine tests, such as respiratory function, electrocardiogram, and ultrasonic echocardiogram, were not reported. The anti-HIV test was negative. Preoperative biopsy results showed dermatofibrosarcoma protuberans. According

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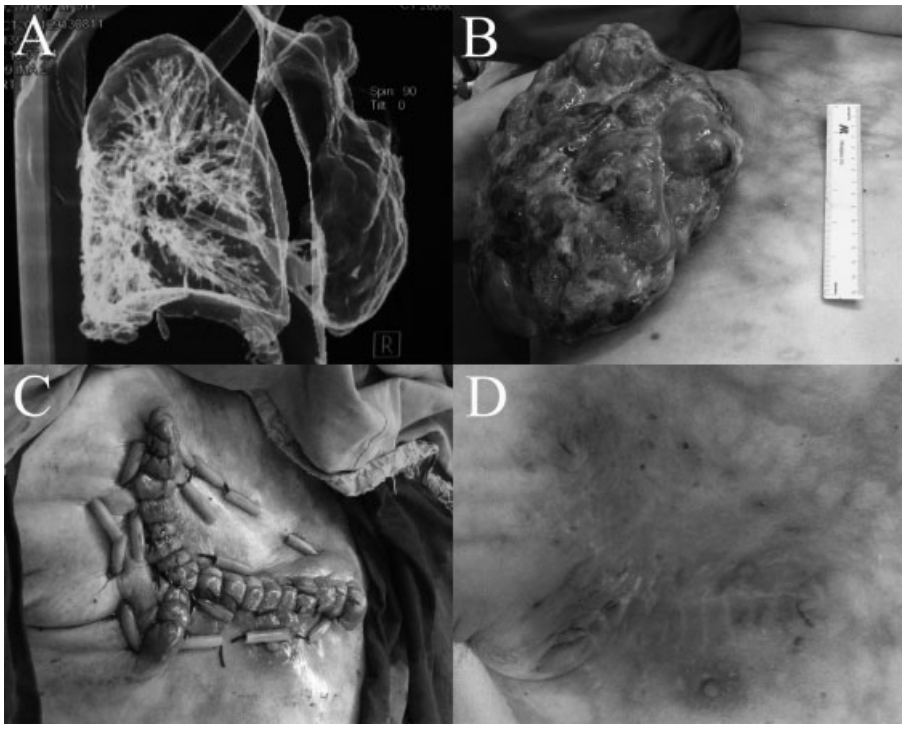

Fig. 1. (A) Preoperative computed tomography scan of the chest showing an expansive mass on the right side. (B) The large, infected, and easy-bleeding mass was identified as a dermatofibrosarcoma protuberans by biopsy. (C) Immediate postoperative view: two methods of suture to relieve wound tension. (D) Postoperative results at 30 days.

to 7th edition of the American Joint Committee on Cancer staging manual,[2] this tumor was stage T2bN0M0 (IIB) and histologic grade was G2.

After administration of general anesthesia and prior to surgery, the patient was placed in a supine position, and single lumen tracheal intubation was used for two-lung ventilation (►**Fig. 1B**). The modified three-dimensional histology surgery was performed. The mass was resected at least 2 cm from the stem (3 cm on average) including the underlying muscles. Then the specimen was sent to the pathology department for frozen-section examination until all incisal margins were negative. The relic ribs and muscles stabilized the chest wall, and the skin defect was closed with adjacent skin by means of tension-relaxing suture (►**Fig. 1C**). A skin flap was not used because of the laxity of the skin. No drainage tube was placed except a latex sheet. Total operative time was ~150 minutes and intraoperative blood loss was less than 500 mL. The postoperative period was uneventful. At the time of discharge (postoperative day 20), the wound had healed satisfactorily (►**Fig. 1D**), and erythrocyte count was 3.08 T/L, Hb was 75 g/L, MCV was 75.8 fL, PLT count was 397 g/L, and serum albumin was 25.4 g/L. The postoperative pathology found dermatofibrosarcoma protuberans, and immunohistochemical staining results were CD34(+), bcl-2(+), anti-smooth muscle antibody (SMA)(−), S-100(−), Des(−), K_i-67(partial +). One year postoperatively, the patient reported no recurrence of the neoplasm.

Discussion

DFSP can arise in any part of the body but occurs mostly on the front of the trunk, head, face, lip, neck, breast, abdomen, and proximal extremities.[3–8] It usually occurs in adults age 20 to 50 years,[9] with the minimum age of 2 years old.[6,10] DFSP rarely metastasizes, despite its local invasiveness. There is no doubt that the initial treatment of DFSP is surgical operation,[1] and every effort should be made to completely resect the mass; thus preoperative biopsy is important at initial therapy.

The tumor, with intermediate- or low-grade malignance, can be removed completely and the patient cured. In 1978, Mohs reported seven patients with DFSP who had micrographic surgery[11]; no local recurrences were reported. Mohs micrographic surgery (MMS) has been increasingly regarded as the standard procedure for DFSP. On the contrary, Johnson-Jahangir and Ratner reported that DFSP located on the head and neck carries a greater risk of morbidity and local recurrence,[12] and some surgeons advocate wide local excision (WLE).[13] The difference between these two techniques is the pathology processing. Under normal circumstances, the specimen from WLE is embedded by conventional paraffin methods; the specimen from MMS is sent to frozen-section examination to check the margins. A successful procedure for DFSP includes resection and wound coverage. WLE can accomplish both in a single-stage operation yet requires more tissue removal. MMS can provide a clear surgical margin but is time-consuming, especially in some cases where reconstruction is needed.[14] However, an enormous tumor can be a challenge for these two procedures. In particular, under some circumstances the stability of chest wall is compromised—for example, for multiple impaired ribs and sternum, chest wall reconstruction is necessary.[15]

The extent of resection is vital. Three-dimensional histology surgery proposed by Moehrle and colleagues not only

avoids the major disadvantages that 3-cm margins are often difficult to obtain, especially when the tumor is close to critical structures, but also preserves normal tissue as much as possible.[16] A retrospective analysis reported that DFSP can be cured by surgery following three-dimensional histology, which emphasizes the importance of negative incisal margins.[17] The first lesion was delimited as a 1- to 1.5-cm margin. Both the horizontal blocks and the vertical blocks were obtained, embedded in paraffin, and sectioned by routine procedures.[18] However, three-dimensional histology with paraffin sections requires that wounds initially remain unsutured, and it is very time-consuming, so it is not usually suitable for an enormous tumor and general anesthesia. Rush frozen-section examination results in considerably shorter waiting time as well as lower anesthesia risk. Frozen section examination of the margins should be routinely performed during these surgeries.

In our case, we modified the technique and first performed a 3-cm-wide resection apart from the stem. The specimen was then sent for the frozen-section examination. The goal was to simplify the process, to potentially ameliorate symptoms of anemia, and to decrease the risk of infection. Skin flap and material planed for chest wall reconstruction preoperatively were not used. No recurrence was noted 1 year postoperatively.

Conclusion

Modified three-dimensional histology surgery and rush frozen-section examination shorten operation time and reduce anesthesia risk. For patients suffering from enormous recurrent DFSP, this surgical treatment is a preferred option.

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The photos were approved by the patient himself.

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