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Case Report

Superior vena cava syndrome caused by mediastinal recurrence of breast cancer with a 13-year disease-free interval: A case report

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Superior vena cava syndrome (SVCS) is an oncological emergency. Lung cancer is the most causative malignancy. In contrast, breast cancer rarely causes SVCS. We report a case in which SVCS was caused by mediastinal lymph node metastasis of breast cancer. The patient was a 60-year-old woman who had undergone breast-conserving therapy at another hospital 13 years previously. Her breast cancer was early stage (T1a(5mm)N0M0; Stage IA), and Luminal type (HER2 negative). She had received adjuvant hormone therapy, but dropped out of treatment two years and six months later. Recently, she had developed cough and face edema, and her extrajugular vein was swollen. CT revealed swollen mediastinal and supraclavicular lymph nodes, lung nodules, pericardial effusion, right pleural fluid, and stenosis of the superior vena cava (SVC). She was diagnosed with recurrent breast cancer with SVCS due to mediastinal LN swelling. A core needle biopsy of a supraclavicular lymph node revealed metastasis; the diagnosis was luminal HER2 positive breast cancer. We initiated treatment with radiotherapy for the mediastinal lymph nodes, and then started hormone therapy and anti-HER2 therapy. These therapies provided relief from her symptoms. She is currently alive and continuing hormone therapy and anti-HER2 therapy. In cases of SVCS due to malignancy, the biopsy findings should be taken into account when possible. An accurate diagnosis is extremely important for the suitable treatment of SVCS, especially in cases caused by malignancy.

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Key words: Superior vena cava syndrome, breast cancer, Luminal HER2 positive, mediastinal lymph node metastasis

Background

Superior vena cava syndrome (SVCS) is an oncologic emergency. SVCS is caused by stenosis or obstruction of the superior vena cava (SVC); thus, symptoms of SVCS include edema the of face, neck arms and upper chest, varicose veins and cough. Although SVCS was most commonly caused by infection approximately 50 years ago[1)], at present SVCS is most frequently caused by malignant disease[2,3)]. Lung cancer is the most common causative malignancy. In contrast, breast cancer rarely causes SVCS. This report describes a rare case of SVCS caused by mediastinal lymph node recurrence from breast cancer.

Case report

The patient was a 60-year-old woman who had undergone breast-conserving surgery and radiotherapy 13 years previously. The final diagnosis was invasive ductal carcinoma, T1a(5mm)N0M0 Stage IA, estrogen receptor(ER) positive, progesterone receptor(PgR) positive and human epidermal

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growth factor receptor type 2 (HER2) negative. She received hormone therapy (tamoxifen citrate), but she dropped out two years and six months later. Recently, she had developed cough and face edema, and her extrajugular vein was swollen. Thoracic X-ray revealed an enlargement of the cardiac shadow and right pleural fluid (Fig 1A). Computed tomography (CT) after drainage and cytology of the pericardial effusion revealed swollen mediastinal and supraclavicular lymph nodes, three lung nodules (14 mm in maximum diameter), right pleural effusion, and stenosis of the SVC caused by the swollen mediastinal lymph nodes (Fig 1B). The differential diagnosis of the lung nodules was either primary lung cancer or metastasis of breast cancer. Cytology of the pericardial effusion revealed breast cancer cells. Core needle biopsy (CNB) of a supraclavicular lymph node and biopsy of a mediastinal lymph node by endobronchial ultrasound-guided transbronchial needle aspiration revealed metastasis of breast cancer. An immunohistological examination revealed the following: ER 95.2%, PgR 20.4%, and HER2 score 3+. She was diagnosed with recurrent breast cancer with SVCS caused by mediastinal lymph node swelling. We considered this to represent an oncological emergency, but it was not considered to be life-threatening because no massive organ metastasis was present.

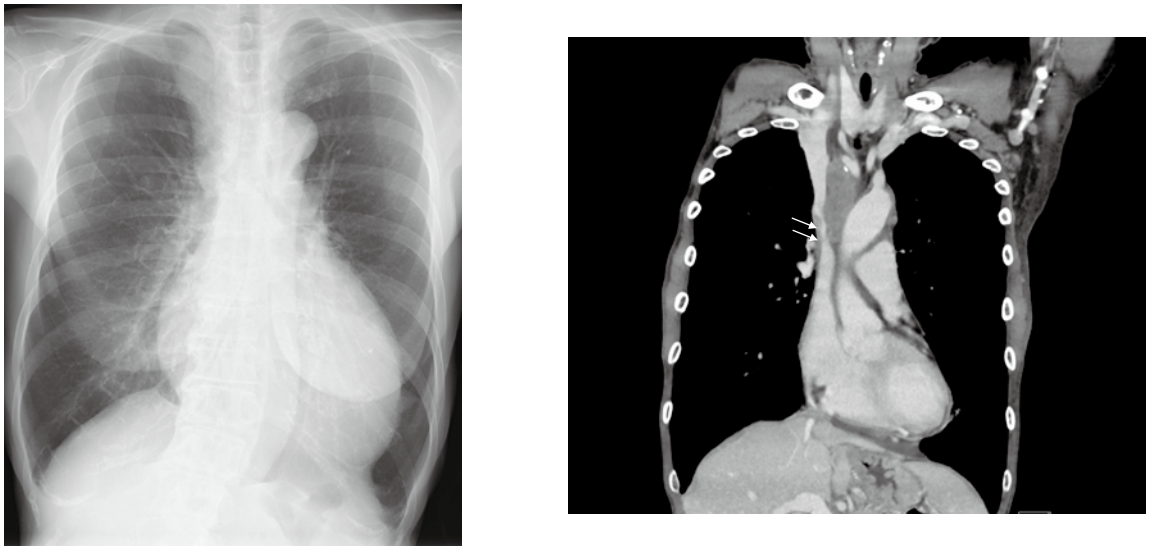

Figure 1.
Pre-treatment state. (A) Chest X-ray revealed an enlargement of the cardiac shadow. (arrows) (B) Computed tomography revealed superior vena cava stenosis caused by swollen mediastinal lymph nodes (arrows).

Thus, radiotherapy (3 Gy/day, total 45 Gy) was first performed targeting her swollen mediastinal lymph nodes. We started hormone therapy (anastrozole) and anti-HER2 therapy (trastuzumab and pertuzumab) after radiotherapy. She felt her symptoms gradually improve. After 12 courses of anti-HER2 therapy, the enlargement of the cardiac shadow was found to have improved on chest X-rays (Fig 2A) and CT showed that the mediastinal and supraclavicular lymph nodes had shrunk, and that the SVC stenosis was relieved (Fig. 2B). Two lung nodules had disappeared while the other one had decreased in size. At thirteen months after the start of treatment, she is continuing treatment with hormone therapy and anti-HER2 therapy, without changes to her daily life.

Discussion

At present, 90-97% cases of SVCS are caused by malignant disease[1,2,3]. The most common causative malignant disease is lung cancer (52-90%), followed by lymphoma (5-25%) and metastatic lesions (5-10%)[1,2,3,4]. Thus, SVCS is a clue to the diagnosis of malignant disease. The signs and symptoms of SVCS include—but are not limited to—facial or arm edema, distended neck or chest veins, facial plethora, dyspnea, cough, and hoarseness[1,5]. More than 50% of patients with SVCS have symptoms before the diagnosis of malignancy[6]. In our case, she had symptoms of SVCS before diagnosis of breast cancer recurrence.

Chest imaging is most important examination for the

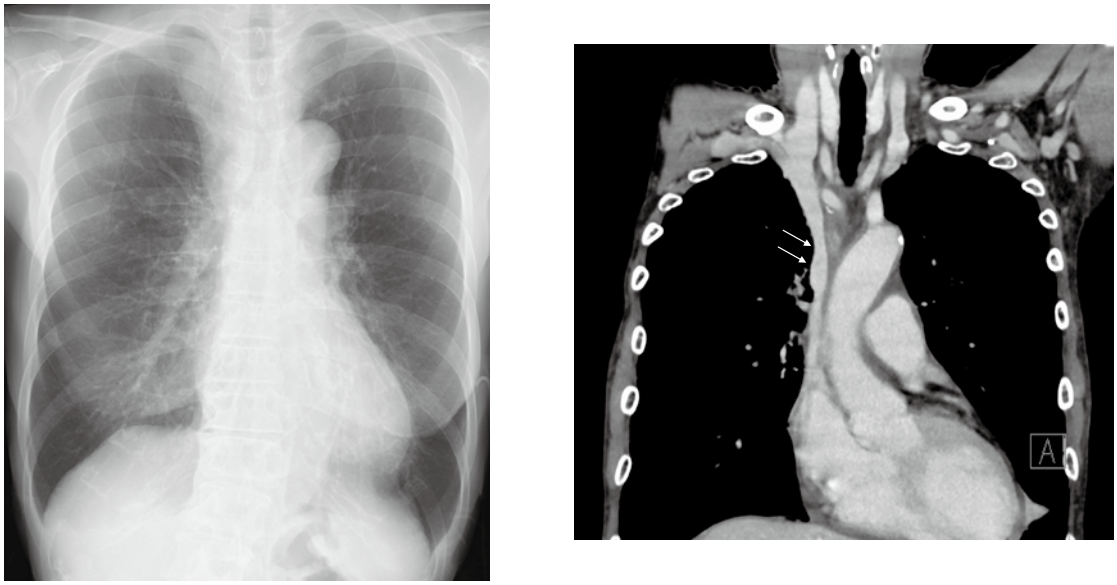

Figure 2.
Post treatment state. (A) An enlargement of the cardiac shadow was found to have improved on chest X-rays. (B) Stenosis of the superior vena cava was relieved on computed tomography.

diagnosis of SVCS. Chest X-rays are reported to show abnormal signs in 84% of SVCS patients[7]. Contrast-enhanced CT is the most useful imaging modality for the diagnosis of SVCS, because CT is able to elucidate not only the cause of SVC stenosis, but also the extent of venous obstruction. Magnetic resonance imaging (MRI) may be useful in cases in which contrast medium cannot be used[1]. Positron emission tomography is sometimes useful, because it may assess the design of the radiotherapy field without contrast enhancement[1]. In our case, chest X-ray and contrast-enhanced CT were particularly useful for making the diagnosis and assessing the therapeutic effect.

Stent placement, radiotherapy and systemic therapy to treat the malignant disease are the most useful treatments for SVCS caused by malignancy. In cases with severe symptoms, stent placement might be chosen as first-line therapy. The time to symptom relief is 0-72 h after stent placement[7] and in 80-95% of the patients, the placement of the stent improved the symptoms more quickly (usually within hours) than after performing radiotherapy[3]. Radiotherapy has long been considered a mainstay of treatment for SVCS. An accurate histological diagnosis before radiotherapy is required for suitable treatment of malignancy[1,7]. Symptoms often improve within 72 h after radiotherapy, with the time to symptom relief ranging from 3 to 30 days[1,7]. Although stent placement and radiotherapy quickly improve symptoms, their effects are only temporary[3]. The most important treatment for SVCS with malignancy is systemic therapy. If SVCS is caused by breast cancer, the subtype of breast cancer is important information for systemic therapy. In our case, we first performed biopsy, and selected radiotherapy as an initial treatment to provide early relief of the symptoms. Based on the findings of CNB before radiotherapy, the patient was diagnosed with metastasis of breast cancer. Thus, we chose hormone therapy and anti-HER2 therapy.

SVCS represents an oncologic emergency; however, most cases are not life-threatening[1,6,7,8]. Although only one case with a fatal outcome was reported in a review of 1986 patients with SVCS[5], the prognosis of SVCS is generally poor, with median survival in all SVCS patients ranging from 1.5 to 10 months[7]. In our case, the SVCS was caused by recurrent breast cancer, however, it was not considered to be life-threatening because there was no liver metastasis, carcinomatous pleurisy or cancerous ascites, which might be associated with a poor prognosis. We are currently continuing to provide treatment with careful follow-up. If such a case is considered to be life-threatening, then we may perform chemotherapy.

In conclusion, local therapy such as stent and radiotherapy for SVCS is important to early relieve the patient from symptoms. Systemic therapy suitable for breast cancer subtype may be important to achieve long symptom-free survival.

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Ethics declarations

Conflict of interest

The authors declare no conflicts of interest in association with the present study.

Ethical approval

This article does not contain any studies with human participants performed by any of the authors.

Informed consent

Informed consent was obtained from the patient for inclusion in this report.

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