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Case Report

Oral squamous cell carcinoma in a patient with scimitar syndrome

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ABSTRACT

Scimitar syndrome is a rare congenital cardiac condition with a poor prognosis. It is frequently accompanied by concordant pulmonary hypoplasia and aortopulmonary collateral arteries connected to the hypoplastic lung. Here we report a case involving a 58-year-old woman with scimitar syndrome who developed stage II oral cancer. Surgical treatment was deemed high risk because of difficulty in respiratory management through mechanical ventilation during general anesthesia, and hence, she was treated with chemoradiotherapy, which was successfully completed. The findings from this case suggest that chemoradiotherapy is a useful strategy that can contribute to improved clinical outcomes for oral cancer in patients with scimitar syndrome.

1. Introduction

Scimitar syndrome is a rare congenital cardiac condition characterized by total or partial anomalous venous drainage from the right lung into the inferior vena cava [1]. It is frequently accompanied by concordant pulmonary hypoplasia and aortopulmonary collateral (APC) arteries connected to the hypoplastic lung, which result in infection and breathing dysfunction. In newborns and older babies, scimitar syndrome can lead to respiratory disorder and heart failure [2]. Because the radiological appearance of the expanded vein that extends parallel to the right side of the heart up to the diaphragm resembles a Turkish sword, the condition is termed scimitar syndrome [3,4].

On the basis of its clinical presentation, scimitar syndrome is categorized into an infant form, which affects infants less than 1 year of age, and an adult form, which affects older patients [5,6]. Compared with the adult form, the infant form is associated with a worse prognosis and a higher incidence of severe abnormalities, including extracardiac anomalies, APC vessels, coexisting congenital heart disease, and pulmonary hypertension [7]. The adult form causes relatively mild symptoms, including exertional dyspnea and recurrent respiratory infections [8], although patients can occasionally exhibit severe symptoms. Because of recent advances in the diagnosis and treatment of symptoms associated with scimitar syndrome, long-term survival has become common, even among patients with the infant form. However, the risk of developing other diseases is higher in long-term survivors. Here we report a case involving a 58-year-old woman with scimitar syndrome who developed oral squamous cell carcinoma (SCC) that was successfully treated by chemoradiotherapy.

2. Case report

A 58-year-old woman was referred to our department with a chief complaint of ulcer formation on the right side of the floor of her mouth. She experienced slight pain in the involved region and tongue since 8 months. However, her pain gradually worsened and ulceration developed. She first consulted her orthopedic surgeon, who was treating her for rheumatoid arthritis. She was subsequently referred to our department for further evaluation of the oral lesion.

Her medical history included scimitar syndrome with right lung hypoplasia since childhood and rheumatoid arthritis and interstitial pneumonia with respiratory disorder since she was 20 years old. For the treatment of rheumatoid arthritis, she was prescribed several agents, including steroids, methotrexate (MTX), and bisphosphonates, since her initial diagnosis. For the past 2 years, she was taking tacrolimus 1.5 mg/day and alendronate 35 mg/day.

Initial assessments revealed hand deformities, symptoms of cervical spondylosis, and gait disturbance associated with rheumatoid arthritis that necessitated a wheelchair for long-distance movements. A chest radiograph and computed tomography (CT) revealed pulmonary hypoplasia, cystic lung changes, and partial anomalous venous drainage from the right lung into the inferior vena cava

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(Fig. 1A, B). A respiratory function test and arterial blood gas analysis revealed that the partial pressures of oxygen and carbon dioxide were 72.6 and 76.0 mmHg, respectively, at room air. Echocardiography revealed diffuse hypokinesia without any signs of pulmonary hypertension. No enlarged cervical lymph nodes were palpable in her neck.

Intraoral examination revealed an indurated mass measuring 22 mm × 12 mm on the right side of the floor of her mouth. The mass extended to the right side of her tongue (Fig. 1C) and exhibited a partially ulcerated mucosal surface. On palpation, the consistency was found to be elastic hard. T2-weighted magnetic resonance imaging (MRI) revealed a homogeneously enhancing mass with an endophytic appearance on the right side of the floor of the mouth and tongue (Fig. 1D). Increased uptake by the mass was observed on fludeoxyglucose positron emission tomography (standardized uptake value, 25.39). No other specific findings were observed in her neck or thoracic and abdominal regions. The oral mass was clinically diagnosed as a stage II (T2N0M0) malig-

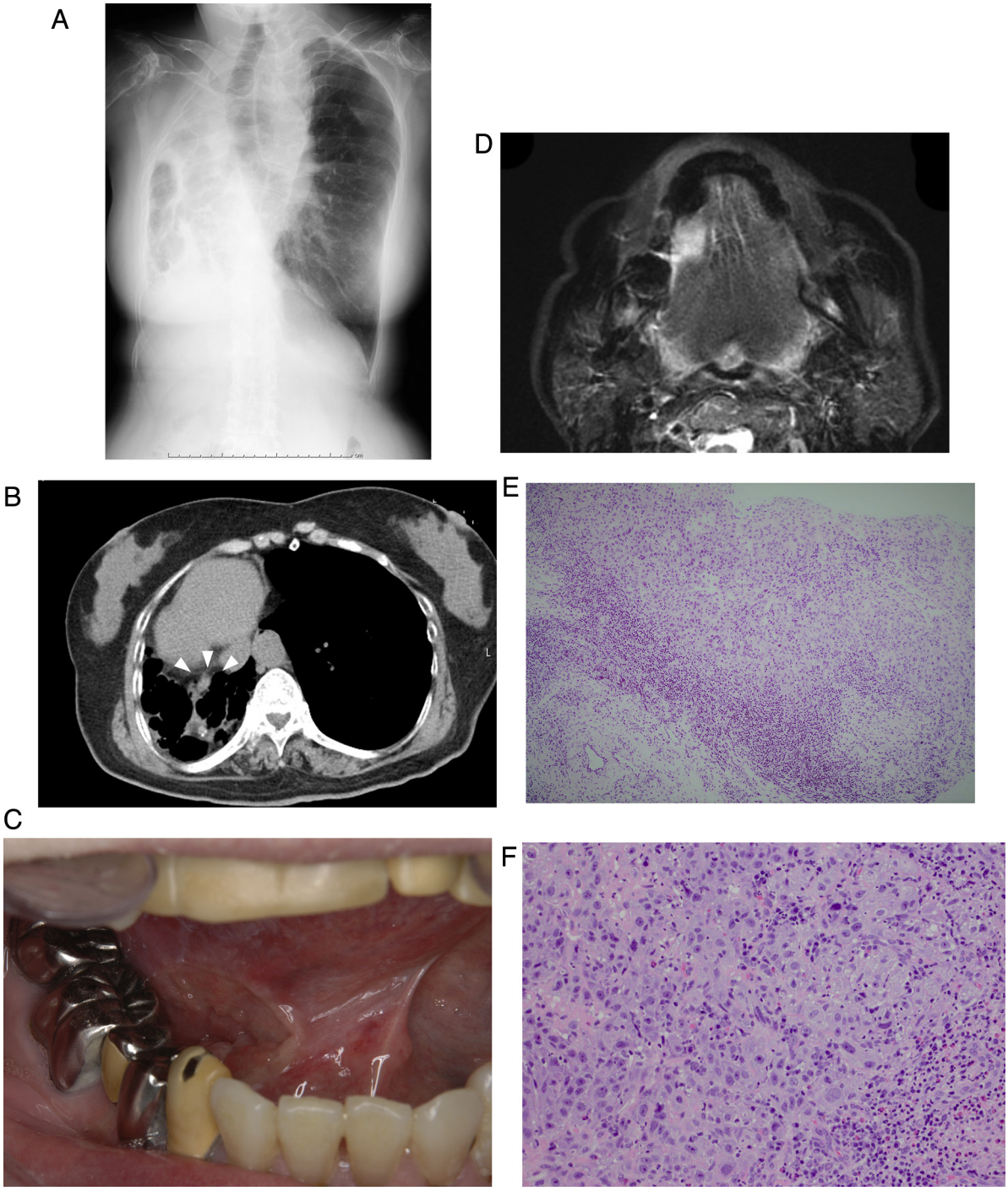

Fig. 1. (A) A chest radiograph showing pulmonary hypoplasia and cystic lung changes. (B) A computed tomography image showing the scimitar vein (arrow). (C) An intraoral photograph showing an indurated mass measuring 22 mm × 12 mm on the right side of the floor of the mouth. The mass is extending to the right side of the tongue. (D) A T2-weighted magnetic resonance image showing a homogeneously enhancing mass with an endophytic appearance on the right side of the floor of the mouth and tongue. (E) Histopathological analysis under a low-power view showing features of poorly differentiated squamous cell carcinoma (hematoxylin-eosin stain, ×100). (F) Histopathological analysis under a high-power view showing poorly differentiated and highly atypical squamous cell carcinoma (hematoxylin-eosin stain, ×400).