

## Acquired facial lipoatrophy: A report of 3 cases with imaging features

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### ABSTRACT

Acquired facial lipoatrophy is a rare disease with an unclear etiology and pathological pathway. The distinct causative factors of this disease have been not elucidated, but it is suspected to be associated with immune system-related diseases, most notably AIDS. Although the management of facial lipoatrophy is very important for patients' social life and mental health, no treatment framework has been developed due to the unknown nature of the disease manifestation. The present case report was designed to provide sequential imaging to visualize the disease progression. The clinical backgrounds of the patients are also introduced, helping characterize this disease entity more clearly for maxillofacial specialists. (*Imaging Sci Dent* 2020; 50: 255-60)

**KEY WORDS:** Lipodystrophy; Facial Asymmetry; Facial Hemiatrophy; Maxillofacial Abnormalities; Diagnostic Imaging

The facial contour is affected more strongly by the volume of subcutaneous fat tissue than by the underlying anatomic structures, bone, and musculature. The symmetrical and adequate distribution of subcutaneous fat tissue is considered essential to an aesthetically pleasing face.<sup>1</sup> Lipoatrophy of the face is an uncommon disease that involves a reduction in facial adipose tissue, causing abnormal indentation and asymmetry of the face.<sup>2,3</sup> People with facial lipoatrophy may experience low self-esteem, depression, and social isolation.<sup>2</sup>

Facial lipoatrophy can occur due to natural or pathological processes.<sup>3</sup> Natural or physiological lipoatrophy refers to the loss of facial fat due to senescence.<sup>2</sup> This type of adipose tissue loss generally starts at the age of 20 to 30 years and progresses gradually. Natural lipoatrophy is usually bilateral. In contrast, when pathological adipose tissue loss occurs, patients may lose their facial symmetry to a serious degree within a short time. Common sites of adi-

pose tissue loss are the cheeks, temples, and pre-auricular region.

Currently, the molecular pathways contributing to the manifestation of this disease remain unclear. Several contributing factors are known, and the most commonly established cause is infection with HIV. The treatment for this disease (antiretroviral therapy), as well as HIV infection itself, can trigger inflammation of the subcutaneous adipose tissue, causing the subcutaneous fat to shrink in volume.<sup>3</sup> Lupus erythematosus is another systemic disease that causes the inflammatory loss of subcutaneous adipose tissue, known as panniculitis. Localized scleroderma also involves panniculitis, although it initially is characterized by an inflammatory erythematous patch.<sup>3</sup>

Until recently, due to the ambiguity and rarity of facial lipoatrophy, neither standard clinical criteria for diagnosis nor proper treatment options had been established for this condition.<sup>4,5</sup> However, studies of facial atrophy have been regularly reported in the literature, and the symmetry of the facial contour has become more important for many people in terms of managing their social life and mental health in modern society.<sup>2-5</sup> The present case report aimed to provide information about this rare disease of unclear

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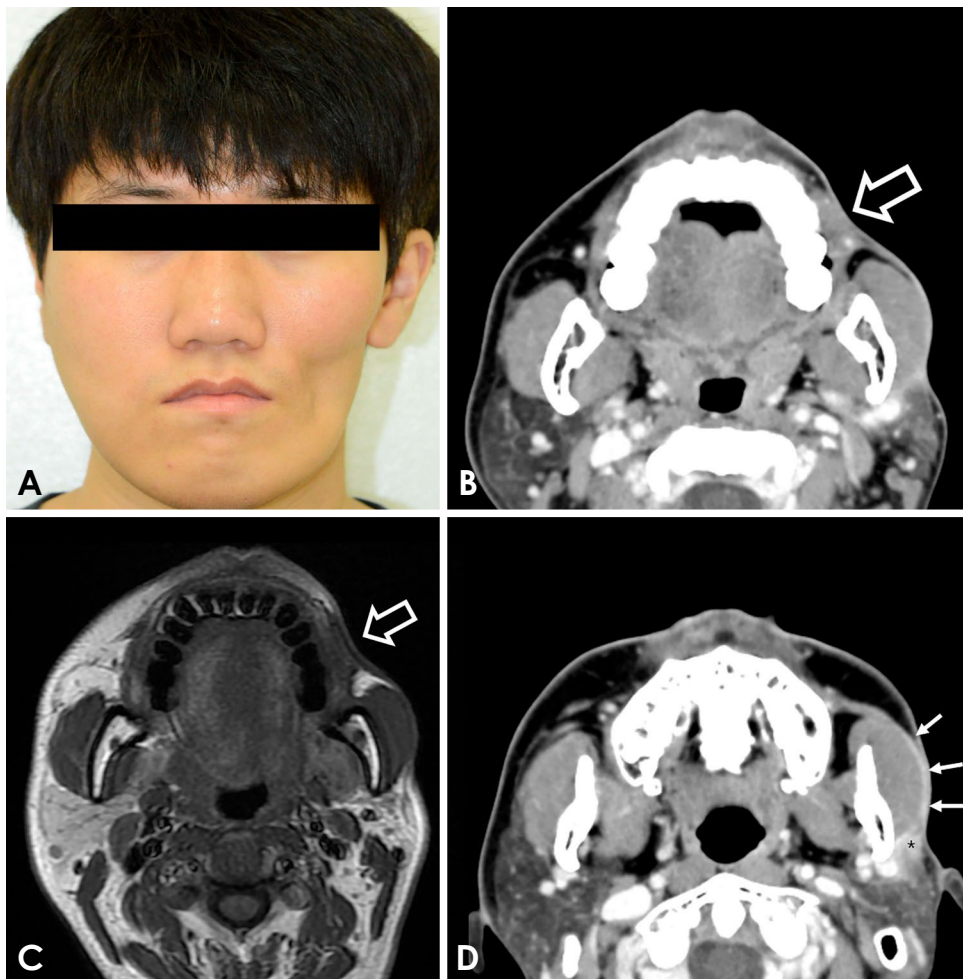
origin to oral and maxillofacial specialists and to help characterize this disease entity more clearly.

## Case Report

### Case 1

A 34-year-old man presented to our hospital with concerns about a left facial depression that he had first noticed a year prior. He was also suffering from muscle spasm and pain in the left face, especially during meals. He reported experiencing frequent headaches, which he thought were caused by the muscle spasms. He had no systemic diseases, and no noteworthy results were obtained from a complete blood count, a blood glucose test, and a test of his serum cholesterol levels. Additionally, the patient was HIV-negative. He underwent computed tomography (CT)

and magnetic resonance (MR) imaging as well as an electromyogram. On the CT and MR images, his left buccal fat pad showed a drastically decreased volume relative to the right side (Figs. 1A-C). The MR images showed no pathologic signal in the nearby soft tissues (Fig. 1B). On the CT images, the left parotid gland showed a decrease in size, with local enhancement in the hilar portion. The left Stensen duct was dilated and showed enhancement as well (Fig. 1D). The patient underwent a nerve block of the masseteric branch of the mandibular nerve, which partially relieved the involuntary masseter muscle spasm. He underwent nerve block therapy twice more separated by a 1-month interval, and the muscle spasm disappeared completely, although he was still worried about the facial deformity. However, 3 months later, muscle spasm reappeared in the left temporal region. Nerve block was per-



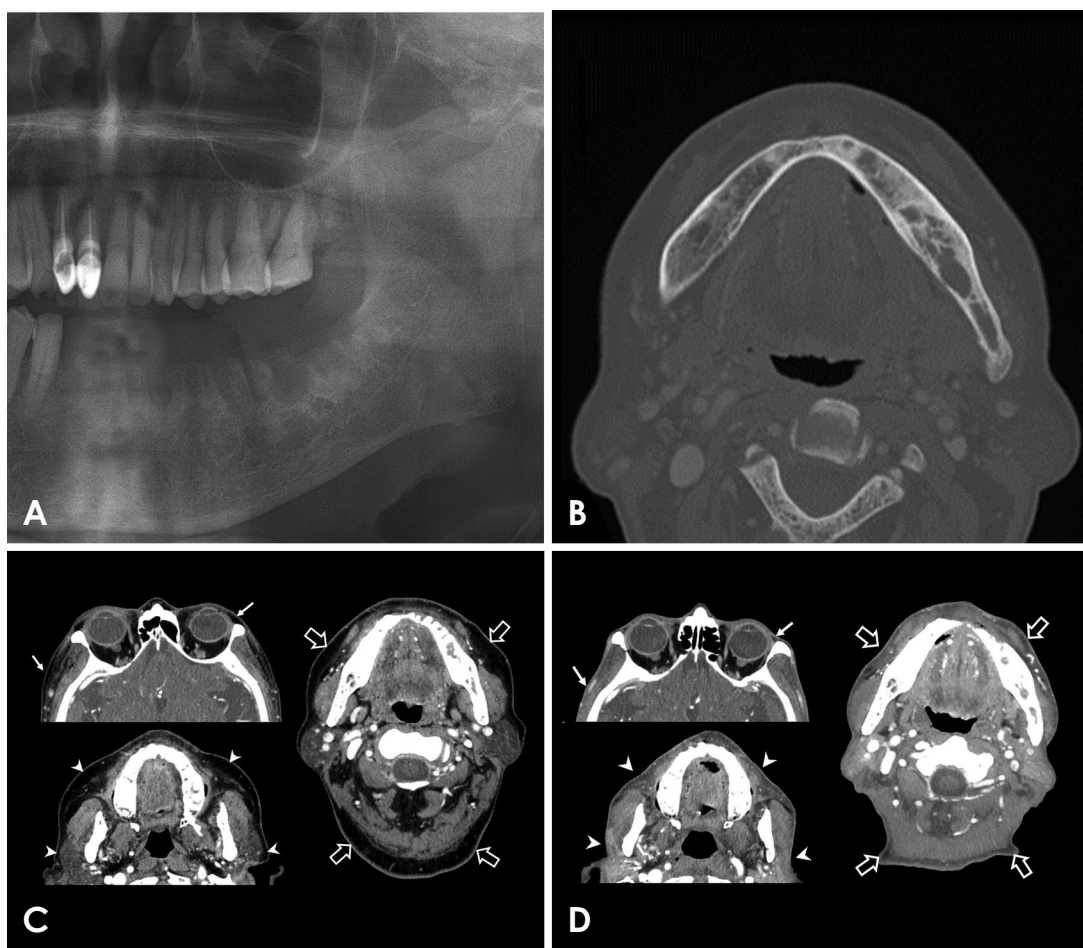
**Fig. 1.** A. A clinical photo of the patient in case 1. Facial asymmetry due to depression of the left buccal cheek is noted. B. A T1-weighted magnetic resonance (MR) image of the patient shows a prominent loss of left buccal fat tissue, while the left masseter muscle shows a normal signal and volume compared to the right side. C. A contrast-enhanced computed tomography (CT) image also reveals a prominent depression on the left buccal cheek (hollow arrow) compared to the right side. D. The left parotid gland shows a decrease in size and local enhancement (asterisk) on a contrast-enhanced CT image. The left Stensen duct demonstrates dilatation with increased enhancement (arrow).

formed on the left masseteric and deep temporal branches of the mandibular nerve. The patient's symptoms were alleviated by the time of the 2-month follow-up visit, and after several additional nerve block treatments, he is now receiving regular follow-up.

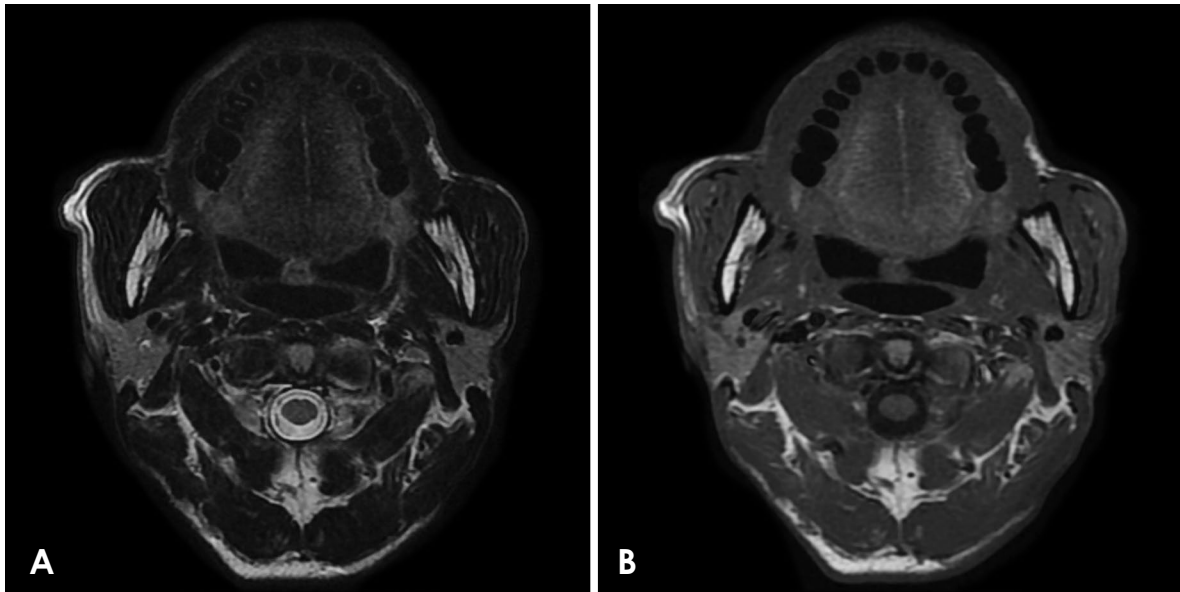
### Case 2

A 65-year-old man presented to our hospital with recurrent pain in the left mandible. The pain had started 4 months before, and tooth extraction with saucerization had been performed at another hospital. The patient had regularly undergone kidney dialysis for 15 years due to end-stage renal disease (ESRD) and also suffered from adrenal deficiency. Panoramic radiography was performed. It showed diffuse sclerosis around the tooth extraction sockets and several osteolytic radiolucencies un-

der the mandibular canal (Fig. 2A). Under suspicion of osteomyelitis, the patient underwent contrast-enhanced CT imaging (LightSpeed VCT; General Electric Medical Systems, Milwaukee, WI, USA). The CT images also revealed features of osteomyelitis on the left mandible (Fig. 2B). After curettage and medication, the patient's symptoms were relieved. However, 8 months later, he again felt pain in the left mandible, which led to a high suspicion of recurrent osteomyelitis. Contrast-enhanced CT images were obtained again and revealed the progression of osteolysis of the mandible. In addition to the osteomyelitis-related findings, the subcutaneous fat tissue of the occipital, periauricular, temporal, and periorbital regions, as well as the neck, exhibited prominently reduced volume (Figs. 2C and D). Additionally, the remaining subcutaneous and intermuscular fat layer of the head and neck and both sides



**Fig. 2.** Panoramic radiography (A) and computed tomography (CT) images (B) in the bone window show diffuse sclerosis around tooth extraction sockets and trabecular bone reduction adjacent to the mandibular canal. C. A contrast-enhanced CT image reveals bone marrow enhancement in the left mandible, while the subcutaneous fat tissue shows no signs of pathology. D. A contrast-enhanced CT image taken 8 months later shows that the subcutaneous fat layer of the head and neck had been obliterated. Both parotid glands show increased enhancement (arrow: periorbital and temporal region, arrow head: preauricular and upper buccal region, open arrow: lower buccal and occipital region, asterisk: parotid gland).



**Fig. 3.** T2-weighted (A) and T1-weighted (B) magnetic resonance images demonstrate severe bilateral loss of the buccal subcutaneous adipose tissue in the patient in case 3. The underlying facial and masticatory muscles and parotid glands show no abnormal signal intensity.

of the parotid gland showed increasing attenuation.

### Case 3

A 61-year-old man presented to our hospital with a chief concern of fat tissue loss in his right buccal cheek area. He had experienced the loss of volume of his left cheek 7 to 8 years prior, and his right cheek had started to lose fat rapidly 2 months before, even as his overall body weight had remained constant. Except for the severe depression on his cheek, he reported no symptoms. Blood testing revealed that he had mild anemia. He had arthritis of the wrist, diagnosed as rheumatoid arthritis. Ultrasonography of the abdomen and endoscopy of the stomach and intestines revealed non-specific findings in the pancreas, liver, spleen, and intestine. MR imaging (Signa HDe; General Electric Medical Systems) at 1.5 T showed a complete loss of the subcutaneous fat tissue of the facial region. The masseter muscle, facial muscles, and neck muscles were very close to the cutaneous layer (Fig. 3). The parotid glands exhibited no pathologic changes but were nearly contacting the cutaneous layer on both sides. The bony structures were also intact, with a normal bone marrow signal and no evidence of severe trauma or infection. Based on the clinical and imaging findings, a diagnosis of facial lipoatrophy was made.

### Discussion

Facial lipoatrophy is not life-threatening; however, pa-

tients' psychosocial health can be considerably affected by this condition. Two of the patients described in the present report visited our hospital with reports of facial depression, and the other patient had incidentally detected facial atrophy. The known causes of lipoatrophy are immune system-related diseases, including HIV infection, lupus erythematosus, and localized scleroderma.<sup>3</sup> Those diseases were not present in the patients discussed in this report. In contrast, the patient in case 2 had adrenal deficiency and ESRD, while the patient in case 3 had mild anemia and was rheumatoid factor-positive. Scant previous research has been presented regarding an association between those diseases and lipoatrophy.<sup>4,5</sup> A case report with a literature review published in 2004 stated that, of 220 cases, only 1 patient had facial lipoatrophy associated with rheumatoid arthritis.<sup>4,5</sup> This report also suggested an association between lipoatrophy and renal disorders, especially membranoproliferative glomerulonephritis (MPGN), as the 220 cases included 45 cases of lipoatrophy (approximately 22%) with histopathologically proven MPGN. MPGN is a type of glomerulonephritis that may progress into renal dysfunction, including ESRD.<sup>6</sup> However, the studies covered in that review were focused on the onset of renal disease due to lipoatrophy, the precise mechanism of which is not yet clear. The patient in case 2 had suffered from renal disease for 15 years, whereas the lipoatrophic changes had taken place more recently. It is thus difficult to conclude that the renal disease developed as a consequence of lipoatrophy; however, the patient's renal condition did require close ex-

amination, since it could have become severe (with a poor prognosis) due to the sudden onset of lipoatrophic changes.

Osteomyelitis was noted on the left mandible of the patient in case 2 after tooth extraction. Misra et al. noted that infection may be a preceding factor of lipoatrophy.<sup>5</sup> The most commonly reported infectious disease preceding lipoatrophy is measles, although 1 report has described lipoatrophy of the face within a few weeks after tooth extraction.<sup>5,7</sup> In case 2, on the follow-up CT images, the subcutaneous and intermuscular fat layers of the head and neck region showed a diffuse increase in attenuation, which is a sign of inflammation. Such general inflammatory changes of the subcutaneous layer were not observed in the other 2 patients, in whom only a severe loss of fat volume was demonstrated. Considering that the patient in case 2 had systemic diseases that were closely related to the immune system, the infection may have spread extensively, making it a possible preceding factor for lipoatrophy.

Unlike the patients in case 2 and 3, the patient in case 1 was relatively young and healthy and had no apparent systemic disease. According to the 2003 study conducted by Misra et al., facial lipoatrophy without any evidence regarding etiology was the most common type reported in the literature.<sup>8</sup> The patient in case 1 was also different from the other patients in that he had discomfort in the affected facial region. Specifically, he complained of muscle spasm and tenderness on the left side of the face during chewing. Lipoatrophy in the extremities presenting with muscle pain has been reported, but those cases were confirmed to be myositis on MR images.<sup>9</sup> However, the electromyogram and MR images of the patient in case 1 showed no specific pathologic findings associated with his facial and masticatory muscles. Although his left cheek had completely lost its subcutaneous fat tissue and his skin was slightly thinned, the ipsilateral masseter muscle maintained its original volume and showed a normal signal compared to that on the right side. The parotid gland on the affected side showed inflammatory and atrophic changes as its volume decreased, and the hilar region was contrast-enhanced on the CT images. Moreover, also on the CT images, the left Stensen duct was thickened and contrast-enhanced, which is a sign of sialodochitis.<sup>10</sup> According to previous research, significant and rapid fat loss may cause increased tightness, with the skin positioned directly on the facial bone.<sup>3</sup> An individual may feel discomfort due to this abnormal tension of the skin.<sup>3</sup> This inflammatory changes of the duct and gland may be due to stenosis of the main duct caused by facial skin tension. The presence of tenderness when chewing may also be explained by these inflammatory changes

in the duct and gland. Still, whether this explanation is relevant to this case is uncertain, because the patient felt that the frequency of involuntary muscle spasms in the masseter muscle decreased after an injection of anesthetic into the masseteric branch of the mandibular nerve. This suggests that his symptoms may have been neurogenic. Progressive facial hemiatrophy, which is known as Parry-Romberg syndrome, is a neurocutaneous syndrome involving the progressive loss of the subcutaneous adipose tissue in the facial region innervated by the fifth cranial nerve.<sup>11,12</sup> The disease varies from mild to severe. In the mild type, the facial depression is barely perceptible, while in the severe type, facial disfigurement is prominent and involves the underlying bone and muscle.<sup>12</sup> The pathogenesis of this disease is not well known. Genetic alterations during embryogenesis, viral or bacterial infections, and traumatic nerve injury have been proposed as etiologies. In a previous study, 45% of patients exhibited migraine and/or facial pain due to trigeminal neuralgia.<sup>12</sup> Some patients have been reported to exhibit lipoatrophic changes involving the salivary gland, as in one of the present cases.<sup>13</sup> Parry-Romberg syndrome is very rare (1 case per 700,000 individuals) and usually occurs in the first or second decade of life, whereas our patient was older.<sup>12</sup> However, the clinical manifestations of our patient were similar to those of this disease, meaning that this syndrome cannot be completely ruled out and further clinical observation is required.

In conclusion, lipoatrophy of the face is an uncommon disease that seems to be associated with various inherited or acquired systemic diseases. A system for the diagnosis and grading of this disease has been established recently, although a framework for treatment is still absent. Thus, oral and maxillofacial specialists should be aware of this entity and perform careful examinations to determine if it is related to an unknown systemic or infectious disease in affected patients. Patients should be carefully followed up with because lipoatrophy may interrupt renal function. Additionally, it is important to correct the disfigurement, since asymmetric facial contour can lead to social stigma and negative psychological effects on patients.

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