

Acquired partial lipodystrophy associated with varicella

Zafer Kurugöl, Zülal Ülger, Ömer Berk, Okan Tuğral

Department of Pediatrics, Ege University Faculty of Medicine, İzmir, Turkey

SUMMARY: Kurugöl Z, Ülger Z, Berk Ö, Tuğral O. Acquired partial lipodystrophy associated with varicella. Turk J Pediatr 2009; 51: 617-620.

Acquired partial lipodystrophy (Barraquer-Simons syndrome) is a rare condition with onset in childhood, and it is characterized by progressive loss of subcutaneous fat in a cephalocaudal fashion. Although it is known that acquired partial lipodystrophy usually follows acute febrile illness, it is very rarely reported to occur in association with varicella. In this case report, we present a seven-year-old girl with progressive loss of fat in her face just after varicella who was diagnosed as acquired partial lipodystrophy.

Key words: acquired partial lipodystrophy, Barraquer-Simons syndrome, varicella, children.

Varicella, caused by varicella-zoster virus (VZV), is a common, highly contagious infectious disease that occurs worldwide. In the absence of a vaccination program, it affects nearly all children before adolescence, especially in temperate climates¹. In this case report, we present a seven-year-old girl having progressive loss of fat in her face after varicella who was diagnosed as acquired partial lipodystrophy (Barraquer-Simons syndrome). Acquired partial lipodystrophy is a rare condition with onset in childhood or adolescence and is characterized by progressive loss of subcutaneous fat of the face, neck, trunk, and upper extremities in a cephalocaudal fashion; it is usually coupled with C₃ hypocomplementemia. Although it is known that acquired partial lipodystrophy usually follows acute febrile illness, it is very rarely reported to occur in association with varicella.

Case Report

A seven-year-old girl presented with a one-year history of thinning of her face. Her parents reported that the loss of fat in her face was progressive and started just after doctor-diagnosed varicella infection one year previously. There was no medical history of a chronic disease or any medication. She was a very healthy child with normal development and normal facial appearance until this varicella infection (Figs. 1, 2). Her parents noticed slow progressive thinning of her face following the VZV infection (Fig. 3). There was no history of



Fig. 1. Physical appearance of the patient one year before varicella.

weight loss, anorexia, chronic diarrhea, polyuria, or polydipsia. Her parents were not related, and there was no history of a similar case in her family. She had two healthy sisters.

On physical examination, she had normal growth with a body weight of 23 kg (50th percentile) and height of 118 cm (50th-75th percentile). The physical examination was unremarkable except for marked symmetrical atrophy of fat over the buccal area and temples. The cheeks were sunken, and there were



Fig. 2. Picture taken just after varicella.



Fig. 3. Picture taken one month after varicella.
Notice slight thinning of cheeks.

bilateral crinkles when she smiled (Fig. 4). The appearance of her trunk, extremities and fat tissue was normal.



Fig. 4. Physical appearance of the patient on admission (one year after varicella).

On laboratory evaluation, complete blood count, erythrocyte sedimentation rate, C-reactive protein, serum electrolytes, renal and liver function tests, creatine kinase levels, fasting blood glucose level, oral glucose tolerance test, and thyroid function tests were all normal. The lipid profile was also normal, with serum cholesterol, high-density lipoprotein-cholesterol, low-density lipoprotein-cholesterol, and triglyceride levels of 148 mg/dl, 50 mg/dl, 72 mg/dl and 84 mg/dl, respectively. Urinalysis and creatinine clearance were normal and there was no proteinuria. Varicella IgG antibody was positive. Antinuclear antibody, anti-double-stranded DNA, rheumatoid factor, and thyroid antibodies were negative, and electromyography was completely normal. Human immunodeficiency virus (HIV) serology was negative. Serum C3 level (27.4 mg/dl) and normal serum C4 level (21.8 mg/dl) were decreased.

The patient was diagnosed as acquired partial lipodystrophy due to painless loss of subcutaneous fat over the face associated with C3 hypocomplementemia.

Discussion

Varicella is generally regarded as a mild, self-limiting viral illness with occasional complications. Even today, varicella is not totally benign. One study suggested that nearly 1:50 varicella cases are associated with complications¹. Although the disease may be mild in some individuals, recent epidemiological studies indicate that there

is significant morbidity and mortality in previously healthy individuals. Complications of varicella include bacterial superinfection of the skin lesions, central nervous system involvement (postinfectious cerebellar ataxia, encephalitis), thrombocytopenia and more rarely hepatitis, glomerulonephritis, arthritis, pericarditis, and orchitis.

In this report, we present a case with acquired partial lipodystrophy following varicella. Acquired partial lipodystrophy in association with varicella is a very rare situation.

Acquired partial lipodystrophy, also known as Barraquer-Simons syndrome or cephalothoracic lipodystrophy, is a very rare condition with onset in childhood or adolescence². It is characterized by progressive loss of subcutaneous fat of the face, neck, trunk, and upper extremities in a cephalocaudal fashion and is usually coupled with C₃ hypocomplementemia. Compared to other types of lipodystrophy, acquired partial lipodystrophy is seldom associated with insulin resistance and its related metabolic derangements³. This may be related to the fact that in this syndrome, patients have limited fat loss. Women are affected approximately three times more often than men. The diagnosis of the disease is mainly clinical. The laboratory workup is needed mainly to investigate for the presence of associated disorders, which are metabolic, autoimmune, and renal diseases. Lipodystrophy progresses slowly and occurs over a period of a few months to two years. Seventy-five percent of patients had significant fat loss when younger than 13 years. The most significant complications are disfigurement in severe facial involvement, renal disease, and rarely, insulin resistance state.

The precise pathophysiology of the fat loss is unclear. Activation of an alternate complement pathway, C₃ hypocomplementemia with C₃-nephritic factor-induced lysis of adipocytes, has been implicated. C₃ hypocomplementemia likely contributes to association of this syndrome with autoimmune diseases and propensity to bacterial infections². Other proposed mechanisms include an autoimmune process and genetic associations⁴. Nephropathy, in the form of membranoproliferative glomerulonephritis, occurs in approximately 20% of the patients². Usually, patients do not have clinically evident renal disease or abnormalities in renal function

until they have had the disease for eight or more years^{3,4}. In our patient, there was no renal involvement, but we will follow the renal function tests of the patient in outpatient clinics visits.

The onset of acquired partial lipodystrophy usually follows an acute febrile viral illness, most commonly measles². The other reported illnesses preceding acquired partial lipodystrophy are pertussis, diphtheria, pneumonia, osteomyelitis, parotitis, infectious mononucleosis, hepatitis B, hepatitis C, and varicella, similar to our patient. Highly active antiretroviral treatments for HIV infection are currently the most frequent cause of acquired secondary lipodystrophic syndromes^{5,6}. Minor surgical procedures and psychological stress have also been reported.

Acquired partial lipodystrophy is also associated with autoimmune disorders like systemic lupus erythematosus (SLE), dermatomyositis, hypothyroidism, pernicious anemia, celiac disease, dermatitis herpetiformis, rheumatoid arthritis, temporal arteritis, and leukocytoclastic vasculitis⁷. There was no sign or laboratory finding of these diseases in our patient.

Patients with acquired partial lipodystrophy have a slowly progressive disease. In the absence of associated renal impairment or insulin resistance, the prognosis is excellent⁷. Educating patients about the disease and associated complications is very important. Parents should be notified about the possible facial changes and the importance of balancing dietary intake to avoid metabolic complications and to ensure healthy development. Several facial reconstruction techniques have been used for cosmetic purpose, with variable success, to restore facial contour. According to guidelines from the American Academy of Dermatology, lipodystrophy is one of the indications for fat transplant. The literature is controversial regarding these procedures. Procedures may include the transposition of facial muscles, adipose tissue transplantation (liposuction), and the insertion of silicone or other implants. The best approach is to individualize the treatment options based on the patient's condition and requirements. However, these procedures are not recommended for prepubertal children.

Acquired partial lipodystrophy is a very rare condition with onset in childhood and can be associated with many disorders like autoimmune

diseases, acute febrile viral infections, hypothyroidism, hepatitis, and antiretroviral treatments for HIV infection. In this report, we presented a case with onset of acquired partial lipodystrophy following varicella. Pediatricians should be aware of this rare complication of varicella.

REFERENCES

1. Choo PW, Donahue JG, Manson JE, Platt R. The epidemiology of varicella and its complications. *J Infect Dis* 1995; 172: 706-712.
2. Misra A, Peethambaram A, Garg A. Clinical features and metabolic and autoimmune derangements in acquired partial lipodystrophy: report of 35 cases and review of the literature. *Medicine* 2004; 83: 18-34.
3. Garg A. Lipodystrophies. *Am J Med* 2000; 108: 143-152.
4. Lenane P, Murphy G. Partial lipodystrophy and renal disease. *Clin Exp Dermatol* 2000; 25: 605-607.
5. Capeau J, Magré J, Lascols O, Caron M, Béréziat V, Vigouroux C. Primary lipodystrophies. *Ann Endocrinol* 2007; 68: 10-20.
6. Waters L, Nelson M. Long-term complications of antiretroviral therapy: lipoatrophy. *Int J Clin Pract* 2007; 61: 999-1014.
7. Pope E, Janson A, Khambalia A, Feldman B. Childhood acquired lipodystrophy: a retrospective study. *J Am Acad Dermatol* 2006; 55: 947-950.