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CASE REPORT

Computerised Tomography Appearances of Partial Lipodystrophy Syndrome in Association with Membranoproliferative Glomerulonephritis Type 2

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ABSTRACT

Partial lipodystrophy is a disease in which body fat is lost in an unusual distribution, with patients often having a typically gaunt facial appearance. The disease is sometimes seen in association with mesangiocapillary glomerulonephritis type 2. We describe the computerised tomography features in such a patient and demonstrate the unusual fatty distribution that may occur. To our knowledge these findings have not been previously described in the international literature.

Key words: *Computerised tomography, Mesangiocapillary glomerulonephritis type 2, Partial lipodystrophy*

INTRODUCTION

Partial lipodystrophy is a rare disease in which there is loss of fat, usually from the face and upper part of the body.[1] There is generally sparing of the lower torso and thighs. The disease is frequently associated with mesangiocapillary (membranoproliferative) glomerulonephritis type 2 (MCGN II). This disease complex, although usually seen in the paediatric or young adult age group, should be considered in older patients with characteristic loss of facial fat and signs and symptoms of nephritis. We present a case of a 42-year-old female patient with biopsy proven MCGN II, with characteristic phenotypic findings of partial lipodystrophy disease. The patient underwent a computed tomography (CT) scan, which clearly depicted the abnormal fat distribution that occurs in this rare disease.

CASE REPORT

A 42-year-old female patient was admitted to hospital for investigation of proteinurea and renal impairment. There was no known family history of renal disease. The patient had an unusual facial appearance. Her cheeks appeared sunken with a paucity of facial fat. Her chest also appeared wasted with almost no subcutaneous fat present. There was a large quantity of subcutaneous fat involving the lower abdomen and upper thighs, however, which was clearly out of proportion to the rest of the patient's fat distribution. Clinically the findings were suggestive of partial lipodystrophy in association with MCGN II, a rare but well recognised cause of glomerulonephritis. A renal biopsy was performed which confirmed MCGN II. A CT scan with four single 7 mm thick non-contrast enhanced preselected slices was performed to further depict and document the fat distribution in this condition. The CT slices were taken through the mid-facial, mid-chest, upper abdomen and upper thigh areas (Figures 1 to 4). The scans demonstrated a complete lack of subcutaneous facial and chest fat, along with the presence of additional fat in the abdomen and marked fat deposition within the upper thighs. This very unusual CT fat distribution was compatible with the final diagnosis of partial lipodystrophy in association with MCGN II.

DISCUSSION

MCGN II is commonly seen in patients with the rare disorder, partial lipodystrophy.[2] In the early 1970s, several case descriptions and studies noted that MCGN II and/or partial lipodystrophy was sometimes associated with dysfunction of the complement system. Subsequently, an IgG autoantibody was detected and has been termed the C3 nephritic factor.

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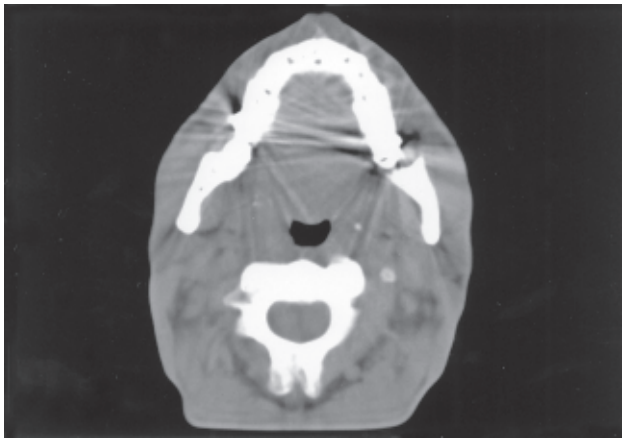

Figure 1. Axial computed tomography scan through the face at the level of the mandible demonstrates minimal facial subcutaneous fat.

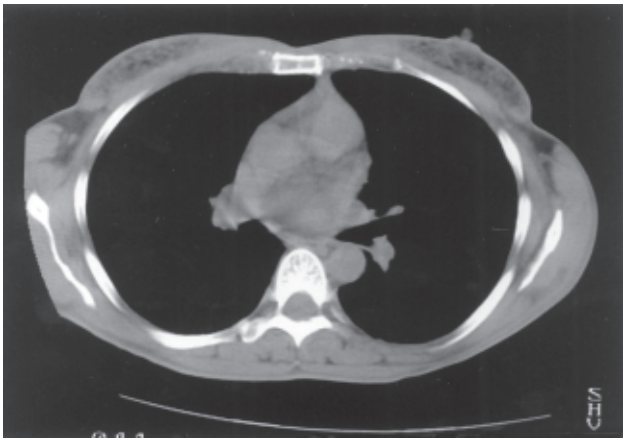

Figure 2. Computed tomography scan through the mid-chest shows minimal subcutaneous fat.

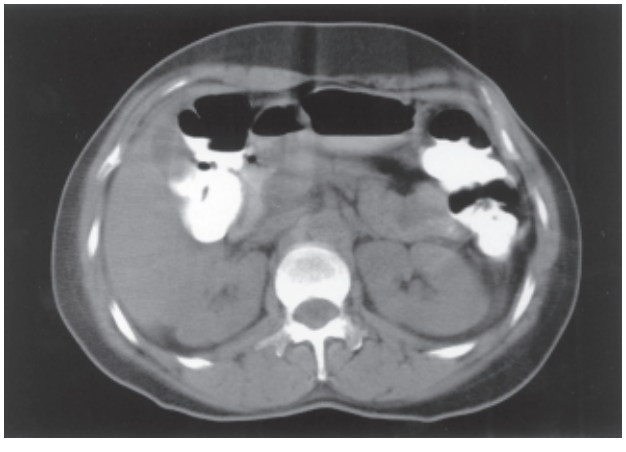

Figure 3. Computed tomography scan at the level of the upper abdomen shows a more normal subcutaneous fat pattern.

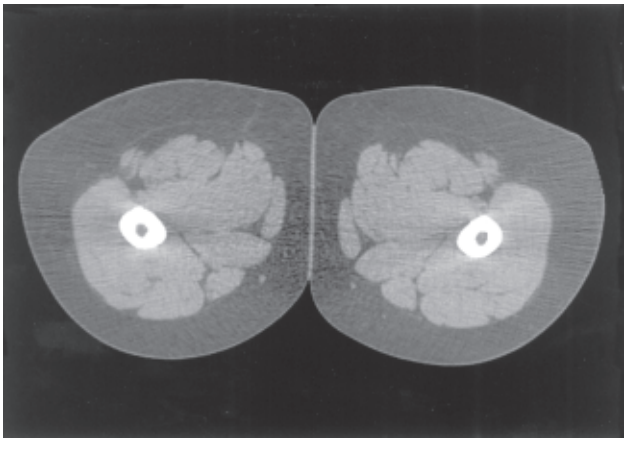

Figure 4. Computed tomography scan through the upper thighs demonstrates excessive fat deposition, which is out of proportion to the fat deposition throughout the rest of the body.

MCGN is a chronic, often progressive renal disease with variable clinical expression. MCGN is divided into three distinct morphological forms, now designated types I, II, and III, on the basis of immunofluorescent (IF) and ultrastructural appearances and complement profiles. Several lines of evidence suggest a genetic basis for at least some cases of MCGN I and III. A significantly higher percentage of those with MCGN I and III have inherited defects of the complement system. MCGN II has been shown to have a definite association with partial lipodystrophy, the typical phenotype showing facial fatty loss. There is typically an upper torso paucity of fat with a more generous distal fat distribution.

Lipodystrophies are rare disorders of the subcutaneous adipose tissue. Two forms are recognised — partial or generalised.[3] Though loss of body fat is a feature common to both types, the two forms have generally been considered mutually exclusive clinical entities, making any unifying pathophysiological concept difficult. Partial lipodystrophy affecting the face has been known to progress and become more generalised on occasions, however.[3]

The cause of the lipodystrophies is uncertain, but myopathy may be a feature, and muscle biopsy studies may help in further defining the syndrome.[4] Facial involvement is the most striking clinical finding of the partial lipodystrophies. While most cases of facial lipodystrophy are bilateral, rarely, unilateral involvement may be seen.[5]

The presence of renal disease in association with an unusual facial fat distribution should alert the clinician to the possible underlying diagnosis of partial lipodystrophy in association with MCGN II. While the radiologist is not routinely involved in the diagnosis of this condition, it is important to be aware of the condition and the characteristic fat distribution which is identifiable on CT.

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Erratum

In the Invited Review of the January-March 2001 issue of the *Journal of the Hong Kong College of Radiologists* (*J HK Coll Radiol* 2001;4(1):16-17), the first sentence of point (1) under the sub-heading 'BREAST CONSERVATION VS MASTECTOMY' on page 16 should read as follows:

"These include:
(1) Patients at higher risk of recurrence with breast conservation treatment (BCT) than mastectomy."
and not
"These include:
(1) Patients at higher risk of recurrence with breast computed tomography (BCT) than mastectomy."

. . . . . .

In the Review Article of the January-March 2001 issue of the *Journal of the Hong Kong College of Radiologists* (*J HK Coll Radiol* 2001;4(1):37-40), under the sub-heading 'CONTROVERSIES IN TECHNICAL DETAIL OF ISOTOPE MAPPING' the first sentence of the second paragraph on page 39 should read as follows:

"Technetium-99m-labelled colloidal rhenium sulphide was used in our series and in the initial three patients, injection and lymphoscintigraphy were done the day before the operation." **and not**

"Technetium-99m-labelled albumin was used in our series and in the initial three patients, injection and lymphoscintigraphy were done the day before the operation."