Diffuse Lipomatosis: Imaging Features

KATSUYA TAKAMATSU, MASAHITO HATORI, SHIGERU EHARA and SHOICHI KOKUBUN

Department of Orthopaedic Surgery, Tohoku University School of Medicine, Sendai 980-77, and Department of Radiology, Iwate Medical University School of Medicine, Morioka 020

Diffuse lipomatosis is characterized by overgrowth of mature adipose tissue affecting large portions of an extremity or the trunk associated with osseous hypertrophy (Enzinger and Weiss 1995). The report on the features of computed tomographic scan and magnetic resonance imaging is very scarce (Nishimura et al. 1985; Coode et al. 1991). The purpose of this report is to describe its characteristic imaging features of this rare disorder with emphasis placed on the usefulness of computed tomography and magnetic resonance imaging for its diagnosis.

Case Report

A 54-year-old man was referred to our orthopaedic outpatient clinic, presenting with a large mass in the left gluteal region. When he was one year old, he had a burn at his left toes, resulting in fusion of his great and second toes. His family history was noncontributory. He first noticed the mass when he was 13 years old. He did not seek any medical advice because of no disabilities. However, the mass had slowly increased in size, and he started to have difficulties in laying supine and wearing clothes. On physical examination, the mass in the left gluteal region was elastic hard, non-mobile and smooth. Its size was 40×30 cm (Fig. 1).
local heat or tenderness was noted over the mass. His left lower extremity was diffusely swollen, and the circumference of the left thigh at 10 cm above the upper pole of the patella was 5 cm larger than that of the right thigh. The range of motion of the left hip was restricted to 25 degrees on extension, 10 degrees on flexion and 15 degrees on abduction. Fusion of his left great and second toes due to the old burn was noted. Hematological and biochemical examinations were within normal limits. A radiograph of the pelvis showed significantly prominent soft tissue density over the left hip joint with no localized bony lesions (Fig. 2A). Radiographs of the left foot showed hyperostosis of the phalanges of the third toe and the distal portion of the third metatarsal, and osteoarthritis of the second and third metatarsophalangeal joints (Fig. 2B). Computed tomographic (CT) scans (Sfida; Shimazu Co., Kyoto) of the pelvis, in the prone position, depicted enlarged subcutaneous fat and infiltrating fat into the gluteal muscles: the glutei medius et maximus, and the tensor fascia lata. No localized mass was detected (Fig. 3). Magnetic resonance (MR) imaging (MR Vectro. 0.5 T; GE Yokokawa Medical Co., Tokyo) again demonstrated infiltrating fat signal into the gluteal muscles (Fig. 4). Because of his disabilities, reduction of the volume of the mass was attempted. Approximately two thirds of the gluteus maximus as well as subcutaneous fat was removed with special care to preserve superior and inferior gluteal arteries. Histological examination revealed proliferation of mature lipocytes infiltrating into the muscle fibers. No cellular pleomorphism or lipoblastic activity was
Fig. 2A. Anteroposterior view of the pelvis showing prominent soft tissue density in the left side of the pelvis. No bony lesions were noted.

2B. Anteroposterior view of the left foot showing bony hyperostosis of the third metatarsal and third proximal phalanx, and osteoarthritis of the second and third metatarsophalangeal joints.

noted (Fig. 5). Based on these clinical and histological findings, the diagnosis of diffuse lipomatosis was made. Three years after the procedure, no recurrent growth of the mass was noted.

**DISCUSSION**

Diffuse lipomatosis is a rare disease (Lewis and Geschickter 1925; Oosthuizen and Barnetson 1947; McCarthy et al. 1969; Kindblom and Moller-Nielsen 1975; Nishimura et al. 1985; Klein and Barr 1986; Coode et al. 1991; Takeda et al. 1993), the etiology of which is not known although genetic factors were suggested
Fig. 3. Computed tomograph showing infiltrative fat density into the gluteus medius, the gluteus maximus, and the tensor fascia lata.

Fig. 4. MR imaging showing infiltrative growth of fat signal into the intra and intermuscular regions.
T1-weighted image (spin echo, TR/TE = 300/25 ms)

(Takeda et al. 1993). It has been reported to be associated with various conditions such as psoriasis vulgaris (McCarthy et al. 1969), tuberous sclerosis (Klein and Barr 1986) and poliomyelitis (Kindblom and Moller-Nielsen 1975). Recurrence after removal of the mass is common over a long period of time, although it is not evident in our patient at this time.

For the evaluation of the proliferation of fat, CT scanning and MR imaging
are almost equally accurate, easy to perform, and non-invasive (Dooms et al. 1985). Homogeneous density or signal intensity of fat is a reliable finding of the presence of mature fat, rich in lipids. Such lipid rich fat is seen in typical lipoma, atypical lipoma, well-differentiated liposarcoma, and various forms of lipomatosis. Foci of non-fatty density or signal intensity are often included in the masses where the possibility of coexistent non-fatty tumors should be taken into consideration in the differential diagnosis (Ehara et al. 1995).

Distribution of the proliferation of the fat, as well as clinical history, is important for differential diagnosis. The differential diagnoses of diffuse lipomatosis are macrodystrophy lipomatosa, symmetrical lipomatosis, pelvic lipomatosis and lipoblastomatosis etc. Increased fat and localized gigantism are also seen in macrodystrophy lipomatosa, which occurs in hands and feet, particularly in the distribution of the territory of the nerve, such as the median and plantar nerves (Bansal and Harmit 1989; Greisis and Williams 1991). Hypertrophy of the nerves with proliferation of fat (neurofibrolipomas), often seen on CT scans and MR images, is not seen in diffuse lipomatosis. The lack of neural deficits and involvement of the proximal portions of the extremities or trunk help in differentiating diffuse lipomatosis from macrodystrophy lipomatosa. Benign symmetrical lipomatosis is characterized by symmetrical proliferation of fat in the head, neck and shoulder girdles in middle aged men. Enzi (1984) described
diffuse accumulation of lipomatous tissue under the trapezius muscle and in the peritracheal region on CT scan of a patient with symmetrical lipomatosis. Pelvic lipomatosis occurs chiefly in young to middle-aged black men and consists of an overgrowth of mature adipose tissue in the perirectal and perivesical regions that ultimately may cause hydronephrosis with uremia or gastrointestinal tract obstruction (Klein et al. 1988; Heyns 1991). Lipoblastomatosis often becomes evident in the first 3 years of life and it is classified into the localized and diffuse types. The proliferation of fat in the diffuse type is an infiltrative growth in the subcutaneous tissue and muscles with an appearance similar to that of diffuse lipomatosis. Histological evaluation for the presence of lipoblasts in lipoblastomatosis is needed for differential diagnosis (Chung and Enzinger 1954). Associated bone changes have not been reported in benign symmetric lipomatosis, pelvic lipomatosis or lipoblastomatosis.

The diagnosis of diffuse lipomatosis can be established on clinical and imaging bases. Infiltrative growth of fat and associated osseous hypertrophy are characteristic. Nishimura (1985) stated the usefulness of CT scanning for diagnosing diffuse lipomatosis and determining its distribution. To the best of our knowledge, there has been no report on MR imaging of diffuse lipomatosis. The hallmark of diffuse lipomatosis is an excessive formation of adipose tissue infiltrating the local muscles (Coode et al. 1991). In the present case, MR imaging as well as CT scanning clearly demonstrated an infiltrative overgrowth of fatty tissue in the left buttock and the extremity involving the subcutaneous and gluteal muscular tissue.

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