Case Reports

Journal of INTERNATIONAL MEDICAL RESEARCH

Journal of International Medical Research
50(1) 1–6

DOI: 10.1177/03000605211066002
journals.sagepub.com/home/imr
SAGE

Neuroimaging findings of linear scleroderma of the head and face: a case report

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Abstract

Linear scleroderma of the head and face is a rare connective tissue disorder characterized by linear depressed scarring in the frontoparietal area of the face. Here, we report a patient with linear scleroderma of the head and face with neurological symptoms such as spontaneous epilepsy and numbness of the right limb as well as the presence of white matter lesions. The patient underwent computed tomography and 3.0-T magnetic resonance examinations including diffusion weighted imaging, diffusion tensor imaging, and perfusion imaging. The imaging findings suggested a disrupted fiber tract and decreased relative cerebral blood flow. Our observation may help to improve the diagnosis and treatment of linear scleroderma of the head and face.

Keywords

Linear scleroderma of the head and face, neuroimaging, connective tissue, neurological symptom, scarring, skin abnormality

Date received: 12 July 2021; accepted: 23 November 2021

Introduction

Scleroderma is a connective tissue disease with unknown etiology characterized by hardening of the skin and subcutaneous tissue. It can be divided into systemic and localized types. The former type can affect the internal organs, while the latter type is mainly manifested by skin abnormalities, and internal organs are rarely affected.[1] Localized scleroderma is clinically divided into five subtypes: circumscribed morphea, generalized morphea, linear scleroderma of the limbs or head and face, pansclerotic morphea, and a mixed subtype in which multiple subtypes are present

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simultaneously.[2] The skin lesions resemble a saber strike scar (which is why it is called en coup de sabre) on the frontal parietal scalp and forehead. However, in accordance with the modern nomenclature, it is usually called linear scleroderma of the head and face.[2] The lesions may extend to the nose, cheeks, jaw, and neck, and alopecia is usually present in the affected area of the scalp.[3] Hypoplasia of bone and soft tissue below the lesion may result in lateral facial atrophy. To our knowledge, no previous description using multimodal radiologic technology, including computed tomography (CT), conventional magnetic resonance imaging (MRI), and diffusion tensor and perfusion imaging, applied to linear scleroderma of the head and face has been reported. The aim of this report is to provide additional imaging perspectives for this rare diagnosis.

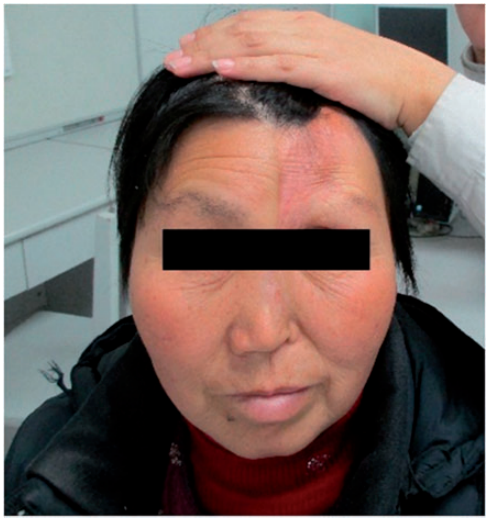

Figure 1. Facial photo of the patient. The skin on the left frontal area is reddish-brown and sunken, resembling a deep saber wound. Mild left facial hemiatrophy was also observed.

Case report

A 62-year-old woman was hospitalized with spontaneous epilepsy and numbness of her right limb for 1 month, and she was then transferred to our hospital. She complained of reddish-brown plaques near the midline of her left forehead and progressive localized pitting of the skin from the age of 5. The patient was asymptomatic at first and did not receive specific treatment. After exhibiting progressive aggravation, a band-like sclerotic skin lesion accompanied by hair loss in the left forehead area and slight left facial hemiatrophy appeared (Figure 1). No members of her family had similar symptoms. Her clinical examination results were normal.

Imaging findings: Brain CT images showed thinning of the left frontal scalp and frontal bone, a hypodense lesion, and speckled calcification in the left frontal lobe (Figure 2). Conventional MRI results showed cerebral morphological changes, including obvious thinning of the scalp

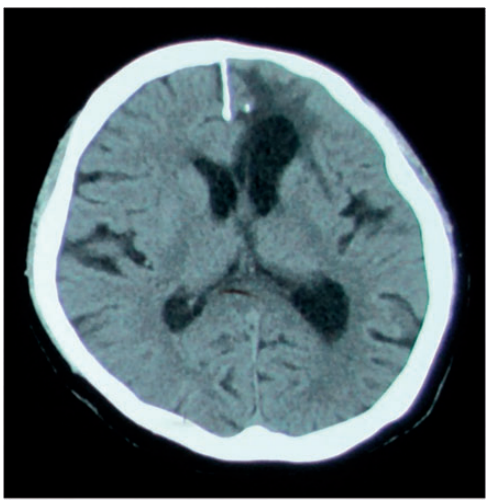

Figure 2. Computed tomography scan revealing thinning of the scalp and frontal bone in the left frontal area, a patchy hypodense area in the white matter around the frontal horn of the left ventricle, and speckled calcification in the peripheral midline of the left frontal lobe.

and subcutaneous soft tissue in the left frontal area, slight thinning of the local skull, a thickened cortex, and a shallow sulcus in the left frontal lobe. Local white matter atrophy was observed, and the boundary between the gray and white matter was blurred. The left lateral ventricle was enlarged, with the frontal horn significantly enlarged. The MRI results showed abnormal signals including patchy long T1 and T2 signals in the white matter area around the frontal horn of the left ventricle. A T2 fluid attenuated inversion recovery sequence showed hyperintensity, and diffusion weighted imaging showed hypointensity. Another cystic area in front of the frontal horn of the left frontal lobe is indicated by arrows in Figure 3. The average diffusion coefficients calculated by diffusion tensor imaging showed increased values in the white matter lesion shown on T2-weighted images (Figure 4a). The fractional anisotropy map showed decreased anisotropy compared with that in the contralateral area (Figure 4b). Diffusion tensor tractography images showed a disrupted fiber tract in the left frontal lobe, which was sparse and irregular compared with that in the contralateral area (Figure 4c). Perfusion

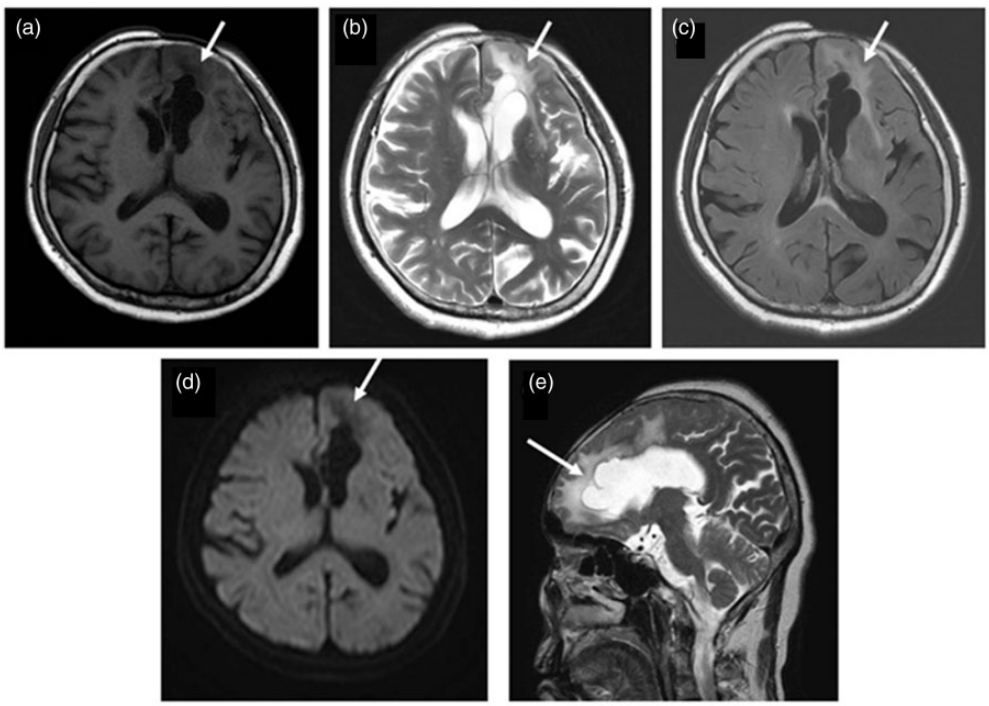

Figure 3. Magnetic resonance imaging, demonstrating thinning of the left frontal scalp and skull, thickening of the left cerebral cortex, and narrowing of the sulcus. The white matter in the left frontal lobe was atrophied, and the boundary between the gray and white matter was blurred. The left lateral ventricle was enlarged. Patchy long T1 and T2 signals were observed in the white matter area around the frontal horn of the left ventricle. The white arrows indicate abnormal signals in the images. Another cystic lesion was seen in the left frontal ventricle anterior to the frontal horn (axial view. a; T1-weighted image, b; T2-weighted image, c; fluid attenuated inversion recovery image, d; diffusion weighted image. sagittal view. e; T2-weighted image).

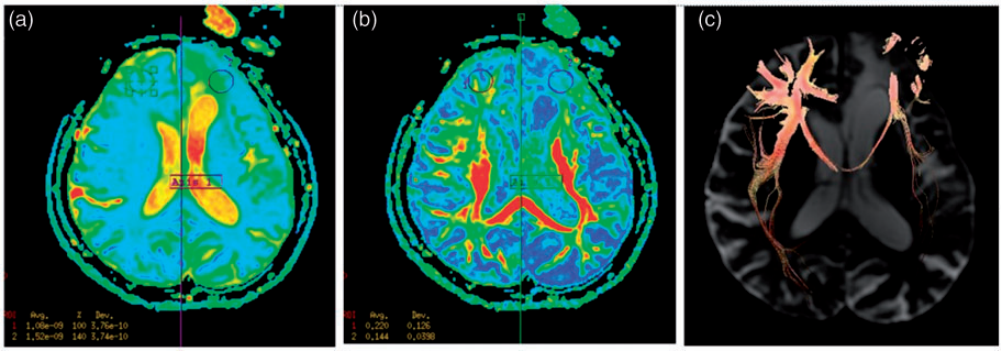

Figure 4. Diffusion tensor imaging, showing that the average apparent diffusion coefficient value in the lesion area of the left frontal lobe was higher than that in the contralateral area (a), and the fractional anisotropy value was lower in the corresponding area (b). Diffusion tensor tractography images showed that fibrous tracts were disrupted in the left frontal and parietal lobes and were sparser and more irregular than those on the contralateral side, especially in the frontal lobe (c).

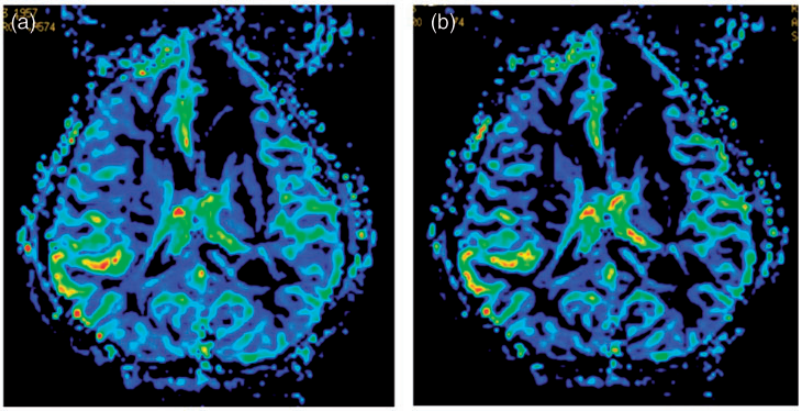

Figure 5. Perfusion weighted imaging. The relative cerebral blood flow (a) and relative cerebral blood volume (b) values were decreased in the left frontal lobe lesions compared with those in the contralateral area.

weighted imaging showed that the relative cerebral blood flow and relative cerebral blood volume in the lesion area were significantly reduced (Figure 5).

The patient was treated in the dermatology department of our hospital. After excluding other infectious etiologies, the patient was subsequently treated with prednisone and methotrexate, in addition to her longstanding carbamazepine treatment, and showed some improvement of her symptoms.

The combination of dermatologic findings with neurologic symptoms and ipsilateral brain radiological findings supported the diagnosis of linear scleroderma of the head and face. The patient provided consent for treatment and written consent for publication. The study protocol was approved by the ethics review committee of Qilu Hospital of Shandong University. The reporting of this study conforms to the CARE guidelines.[4]

Discussion

There are several hypotheses regarding the mechanism of nervous system involvement in linear scleroderma. First, destruction of endothelial cells leads to fibroblast activation, followed by collagen contraction and cell cavity narrowing, which result in local ischemia.[5] Second, the nervous system is affected by inflammatory lesions, especially angioinflammatory lesions. Brain biopsies of children have shown infiltration of perivascular lymphocytes in angioinflammatory lesions. Abnormal and dilated blood vessels were observed on biopsies and by angiography, supporting the hypothesis that angioinflammatory lesions are the basis of brain tissue involvement.[6] Third, abnormalities in the nervous system were present in cases where sclerosis, fibrosis, and glial cell hyperplasia (astrocyte hyperplasia, which usually leads to scarring) involved the brain parenchyma, meninges, and vascular system.[7]

Epilepsy and headache are the most common symptoms of linear scleroderma of the head and face involving the nervous system, and sometimes these symptoms can precede the onset of skin changes by months or years.[8] Hypoesthesia, cognitive impairment, poor discrimination, and unclear pronunciation may also occur.[9] CT and MRI images can reveal skull and intracranial abnormalities,[1] and imaging changes may occur even in asymptomatic patients.[10] Imaging abnormalities include brain atrophy, white matter lesions, intracerebral calcification, meningeal changes, and skull atrophy,[11] which are mainly located on the same side as the skin lesions but can also occur on the opposite side.[7] In this case, patchy long T1 and T2 signal shadows were observed in the white matter area around the frontal horn of the left ventricle. Perfusion MRI revealed relative decreases in cerebral blood flow and cerebral blood volume, which were consistent with previously reported results.[12] The cerebral blood flow may have been reduced because of inflammation in the lesion area as previously reported.[6] According to diffusion tensor tractography imaging, the fiber bundle in the left frontal lobe was interrupted and sparse, suggesting demyelination of nerve fibers or possible damage or atrophy of nerve axons. CT is sensitive to calcification in the brain and blood vessels, and MRI can accurately show meningeal changes and white matter lesions.

Linear scleroderma of the head and face can be diagnosed by skin changes. It rarely presents with central nervous system involvement. Imaging findings of lesions in the central nervous system are not specific and are not easily diagnosed. The combined application of CT and multimodal MRI can improve the accuracy of diagnosis not only by detecting cutaneous changes but also by indicating central nervous involvement in this disease. For asymptomatic patients with linear scleroderma of the head and face, neuroimaging examinations can be used for early diagnosis and to reduce the occurrence of sequelae.

Author contributions

Linlin Meng drafted the manuscript. Qing Wang critically revised the manuscript for important intellectual content. All authors agreed to participate and approved the submitted version of the manuscript.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

Ethics statement

The authors obtained written informed consent from the patient for submission of this manuscript for publication.

Funding

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

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