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Case Report

Nevus lipomatosus cutaneous superficialis on the shoulder: A case report with MRI findings

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ARTICLE INFO

Article history:

Received 21 May 2019

Revised 18 July 2019

Accepted 20 July 2019

Available online 13 August 2019

Keywords:

Nevus lipomatosus cutaneous superficialis

ABSTRACT

Nevus lipomatosus cutaneous superficialis is a rare disease, and its magnetic resonance imaging features have not been reported. A 1-year-old male infant was admitted to our hospital for examination of a mass on his right shoulder. Magnetic resonance imaging revealed hypertrophic skin and a lipomatous subcutaneous mass, consistent with a hamartoma-like lesion or mesenchymal tumor; after surgery, the tumor was pathologically diagnosed as nevus lipomatosus cutaneous superficialis. To the best of our knowledge, this is the first case report focusing on the magnetic resonance imaging features of this disease. Hypertrophy of all skin structures involved (epidermis, dermis, and subcutaneous fat) may be specific to nevus lipomatosus cutaneous superficialis, enabling its differentiation from other tumors.

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Introduction

Nevus lipomatosus cutaneous superficialis (NLCS), a rare hamartoma, is pathologically characterized by hyperplasia of the adipose tissue in the dermis. It is a protruding lesion of the skin, and is often diagnosed dermatologically; thus, radiological examination is not frequently performed, and most radiologists are not familiar with the disease. Herein, we present

a case of NLCS on the right shoulder of a male infant. To the best of our knowledge, this is the first report of the magnetic resonance imaging (MRI) features of NLCS.

Case report

At birth, a male infant had an infantile hemangioma-like lesion on his right shoulder. The length of its major axis was

Declaration of Competing Interest: The authors declare that they have no conflict of interest.

Ethical Approval: This article does not contain any studies with human participants or animals performed by any of the authors.

Informed Consent: Informed consent for publication was obtained from the guardians of the patient in this case report, as potentially identifying information may be included in this article.

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<https://doi.org/10.1016/j.radcr.2019.07.009>

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Fig. 1 – At birth, the patient had a red protruding mass on the right shoulder, which was presumed to be infantile hemangioma. (Color version of figure is available online.)

approximately 3 cm, it was red in color, and its surface was smooth and protruding (Fig. 1). He had undergone multiple laser treatments at another hospital, but the mass had become markedly enlarged. When the infant was 1 year of age, the surface of the mass was slightly red and lobulated, and it was approximately 6 cm in diameter (Fig. 2). This was not typical of an infantile hemangioma, and a malignant tumor could not be ruled out, so he was referred to our hospital.



Fig. 2 – A 1-year-old infant with a large mass on the right shoulder, approximately 3 cm in length. The surface of the mass was pale red, and lobulated. The mass had grown markedly since birth (see Fig. 1). (Color version of figure is available online.)

On MRI, the mass protruded outward from the lateral side of the right shoulder. On T2-weighted images, the epidermis side was thick and exhibited intermediate signal intensity; further, some cystic high signal intensity areas were recognized. The deep side was tumorous and exhibited high signal intensity on both T1- and T2-weighted images; the signal intensity decreased on fat-suppressed T1-weighted images. Fine mesh-like areas with low signal intensity were observed on the deep side on T2-weighted images. On gadolinium-enhanced fat-suppressed T1-weighted images, the epidermis side exhibited strong enhancement, but the lipomatous deep lesion did not exhibit distinct enhancement (Fig. 3). This was our first encounter with such a tumor. In the differential diagnosis, we considered a hamartoma-like lesion, and a mesenchymal tumor such as lipoblastoma or liposarcoma.

Surgical resection was performed: the tumor was located on the deltoid fascia and was completely excised. Pathological examination revealed hyperplasia of adipocytes in the dermis and subcutaneous tissue. The adipocytes formed aggregates around blood vessels, and mature adipocyte proliferation in the form of islets between collagen bundles was apparent. No lipoblasts were observed. The epidermis was thickened and exhibited a nipple-like conformation, and there were no findings suggestive of infantile hemangioma. Hyperplasia of the sebaceous gland and hair follicle was evident in the dermis, which is atypical of NLCS; however, other findings were consistent with NLCS (Fig. 4).

Discussion

NLCS is a rare benign hamartoma that is pathologically characterized by the presence of mature ectopic adipocytes in the dermis [1]. With regard to clinical form, it is classified as either the multiple (or “classical”) form or solitary form.

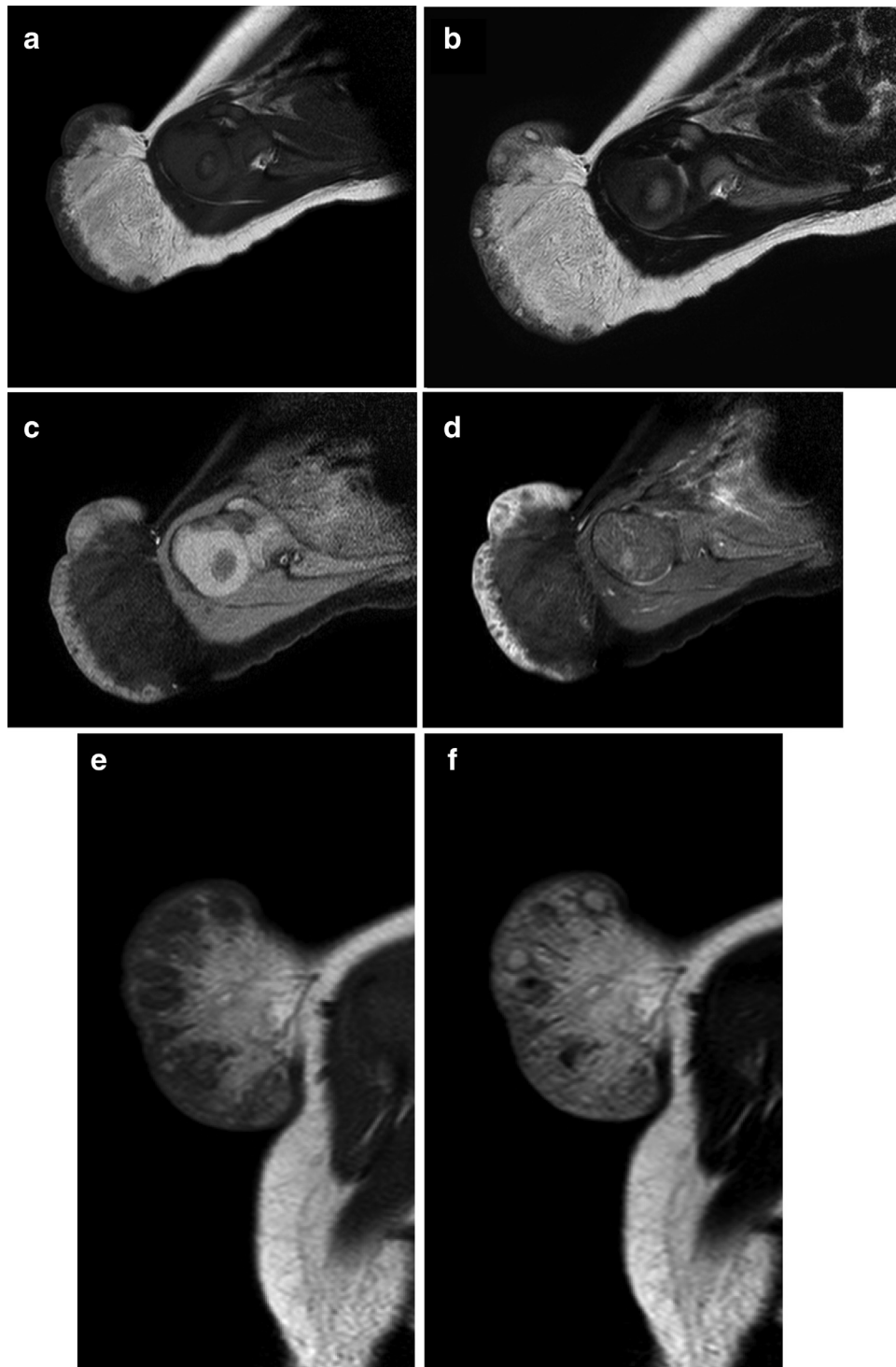


Fig. 3 - A 1-year-old infant with nevus lipomatous cutaneous superficialis, assessed by magnetic resonance imaging. (a) T1-weighted imaging. (b) T2-weighted imaging. (c) Fat-suppressed T1-weighted imaging. (d) Gadolinium-enhanced fat-suppressed T1-weighted imaging. (e) Coronal T1-weighted imaging. (f) Coronal T2-weighted imaging. Both T1- and T2-weighted images depicted a large high signal intensity subcutaneous mass on the patient's right shoulder, and the signal exhibited homogenous reduction on the fat-suppressed image. On the T2-weighted image, the mass exhibited fine mesh-like low signal intensity areas. The thickened skin exhibited strong contrast and contained some well-circumscribed ovoid areas. These ovoid areas exhibited high signal intensity on T2-weighted images and were not enhanced.

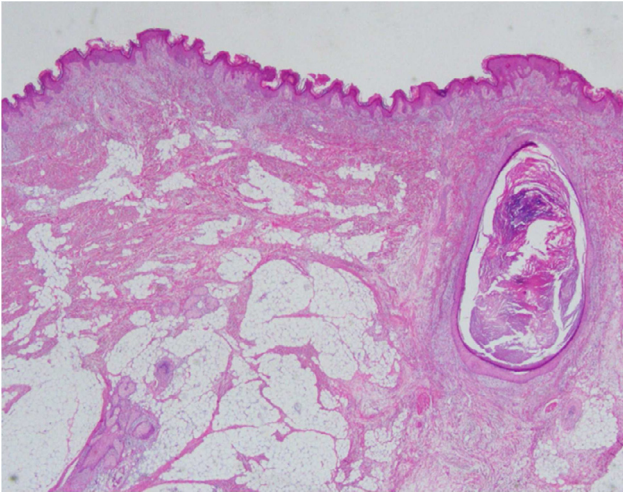


Fig. 4 – A 1-year-old infant with nevus lipomatous cutaneous superficialis, assessed by pathological examination revealed hyperplasia of adipocytes in the dermis and subcutaneous tissue. The adipocytes formed aggregates around blood vessels and exhibited proliferation in the form of islets between collagen bundles. The epidermis was thickened in a manner resembling a nipple-like conformation, and hyperplasia of the sebaceous gland and hair follicle was observed in the dermis.

The multiple form frequently occurs in the lower back and buttocks of young patients, from birth to 20 years of age, and it is characterized by clusters of skin-colored or yellow papules or nodules [2]. There is no observed sexual or familial trend [3]. The solitary form appears more frequently in older patients during the third to sixth decades of life and can occur anywhere on the skin. It exhibits the same dermatologic appearance as the multiple form [4]. Pathological investigation of NLCS typically reveals proliferation of ectopic mature adipocytes in the dermis. The adipocytes commonly form small aggregates around blood vessels or eccrine glands but may also manifest as solitary adipocytes between collagen bundles. The epidermis exhibits acanthosis, basket weave hyperkeratosis, and increased basal pigmentation [2]. NLCS can increase in size. In cases involving the solitary form, there is a possibility of its shape changing over time, broadening from a hemispherical shape to a pedunculated shape [5].

To the best of our knowledge, MRI features of NLCS have not been reported. In the present case, there were several characteristic MRI findings. The NLCS exhibited a lipomatous mass, and T2-weighted imaging revealed fine mesh-like structures inside. There were no nodules or irregular septa, and there was no abnormal contrast enhancement. These MRI findings corresponded with the histopathological observations of evident subcutaneous lipomatous hypertrophy and proliferation of collagen fibers between adipocytes. Therefore, these findings may be characteristic of NLCS. Another potentially characteristic MRI finding in the current case was the homogeneous thickening of the skin. Histopathologically, this indicated hyperplasia of the epidermis and dermis. Fat proliferation in the dermis—which is key to a diagnosis via histopathological examination—was not apparent on MRI.

A finding that was atypical of NLCS was the presence of some sparse, cystic lesions in the dermis. On pathological examination, these were observed as expanded hair follicles and sebaceous ducts, with abundant inflammatory cells. We suspect that these findings represented inflammatory changes due to the multiple laser treatments the patient had previously undergone.

Based on the MRI and histopathological findings in the present case, we believe that a key feature of NLCS may be hypertrophy of all structures involved, including the epidermis, dermis, and subcutaneous fat. This finding may be considered a characteristic of hamartomatous lesions. We considered differential diagnoses of infantile hemangioma and lipomatous tumors including lipoblastomas and liposarcomas, but such hypertrophy is not observed in these diseases.

The most likely differential diagnosis in the present case was infantile hemangioma, which is the most common soft tissue tumor in infancy. Clinical differentiation of infantile hemangioma from other tumors is difficult due to its unusual growth after birth despite its benign nature and the high disease frequency [6–9]. The tumor in the current case exhibited redness and growth after birth, which were indicative of infantile hemangioma. Notably, however, MRI depicted a lipomatous lesion without features suggesting a vascular nature. Therefore, differentiation of such lesions should be easy on MRI.

A lipomatous tumor, such as a lipoblastoma or liposarcoma, could also be considered a differential diagnosis based on both clinical and MRI findings. These tumors can also protrude from the skin and exhibit growth. Lipoblastoma is a rare benign tumor, which typically presents before the age of 3 years and often develops in the extremities [10]. It manifests as a complex fatty tumor with a myxoid component and fibrous septa. On MRI, lipoblastoma only exhibits a small amount of fat tissue, and these lesions are heterogeneous with enhancing soft-tissue components [11]. Enhancement occurs due to a rich capillary network [12]. Lipoblastoma has a myxoid component that exhibits high signal intensity on T2-weighted imaging, and more rough septa than NLCS. As previously mentioned, the most important differentiating feature is that lipoblastoma does not involve thickening of the skin. The MRI features of liposarcoma, particularly myxoid liposarcoma, resemble those of lipoblastoma; however, liposarcoma is extremely rare in infants.

To the best of our knowledge, this is the first case report describing the MRI features of NLCS. Characteristic MRI findings included the depiction of a lipomatous lesion with a fine mesh-like area, and hypertrophy of both the epidermis and the dermis. NLCS should be included in the differential diagnosis of lipomatous cutaneous/subcutaneous mass lesions, and the hypertrophy of all structures involved (epidermis, dermis, and subcutaneous fat) may be specific to NLCS, enabling differentiation from other tumors.

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