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Novel diagnostic imaging features of facial lupus panniculitis: ultrasound, CT, and MR imaging with histopathology correlate

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ABSTRACT

Lupus panniculitis (LP), also referred to as lupus erythematosus profundus (LEP), is a chronic recurrent inflammation condition of the subcutaneous fat. It occurs in 1 to 3% of patients with systemic lupus erythematosus (SLE) and in 10% of patients with discoid lupus erythematosus (DLE), but can also occur as an entity of its own. Patients with lupus panniculitis usually present with persistent, often tender and painful skin lesions, or subcutaneous nodules, that range from 1 to 5 cm in diameter. The overlying skin may appear erythematous; lesions may become ulcerated, and heal with atrophy, skin depression, dimpling and scaring. Lesions tend to resolve spontaneously and may follow a chronic course of remission and exacerbation that persists for months to years.

The imaging features of facial LP are extremely scarce in the literature. We present a case of facial lupus panniculitis and describe the associated characteristic ultrasound, CT, and MR imaging findings along with histopathologic correlation.

1. Case report

A 19-year-old female presented with a chief complaint of left facial swelling and left orbital soft tissue fullness. She recalled falling asleep at her desk while studying prior to noticing her symptoms. Upon awakening, she felt pain and a bump on her left cheek. She described her symptoms as a "swelling and hardening of the face and left cheek over several months." This eventually spread to the right cheek as well and then to her right eyelid. Swelling was noted on the left-greater-than-right side of her face and her face was slightly tender to the touch. She reported no fevers or paresthesias and had no history of facial injections or cosmetic procedures. There was no change in her vision and there were no other lesions. She denied fevers, chills, night sweats, weight loss, weakness/fatigue in her muscles. There was no family history of Lupus or autoimmune diseases. On physical exam, the skin was soft, but mildly puffy appearing. There were no overlying skin color changes, no redness or scaling.

Laboratory findings included positive antinuclear antibody test (which was initially negative) and positive SSA antibodies, negative SSB, normal IgG subsets, no M protein, without other clinical features of systemic lupus or Sjogren's syndrome. Therefore, the diagnosis of lupus panniculitis was made in the setting of cutaneous lupus. Hydroxychloroquine was contraindicated due to her retinal lattice degeneration (which was diagnosed prior to her presentation to our hospital with no clear causative relationship to her facial symptoms). She was started on quinacrine and her symptoms improved. The patient has been followed by the rheumatology department over the past two years and her symptoms remain stable with no features of systemic lupus.

Ultrasound evaluation of her face in an outside institution demonstrated a 2.3 × 1.5 cm mildly hyperechogenic ovoid pseudo mass-like lesion, discrete from the surrounding subcutaneous fat with a thin hypoechoic rim. On Doppler evaluation, there was prominent central increased vascularity within the lesion without any displacement of the vascular structures. There were no cystic components (Fig. 1).

CT of her face demonstrated abnormal fat stranding, with haziness in the deep and superficial fat tissues. There was involvement of the buccal fat pad subjacent to the superficial musculoaponeurotic system (SMAS) bilateraly, left greater than right, which slightly displaced the SMAS laterally. The parotid duct and adjacent facial vessels could be seen within the hazziness and passed through the abnormality without distortion or displacement. There was asymetric prominence of the vascularity in these regions. CT demonstrated similar soft tissue inflammatory changes involving the subcutaneous adipose layer along

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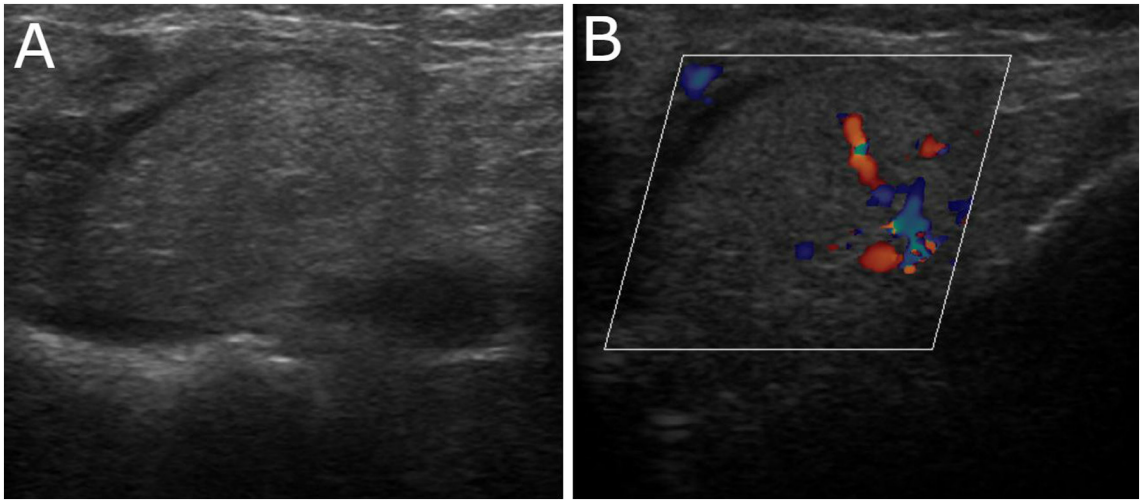

Fig. 1. A) Ultrasound performed over the region of the right face, where the patient was complaining of symptoms, demonstrates a discrete area of intermediate echogenicity without mass effect. There is no cystic component. There are also surrounding inflammatory hyperechoic changes within the subcutaneous adipose layer of the right face. B) Color Doppler ultrasound demonstrates hyperemia, with increased vascular flow. Together, the findings suggest an inflammatory process.

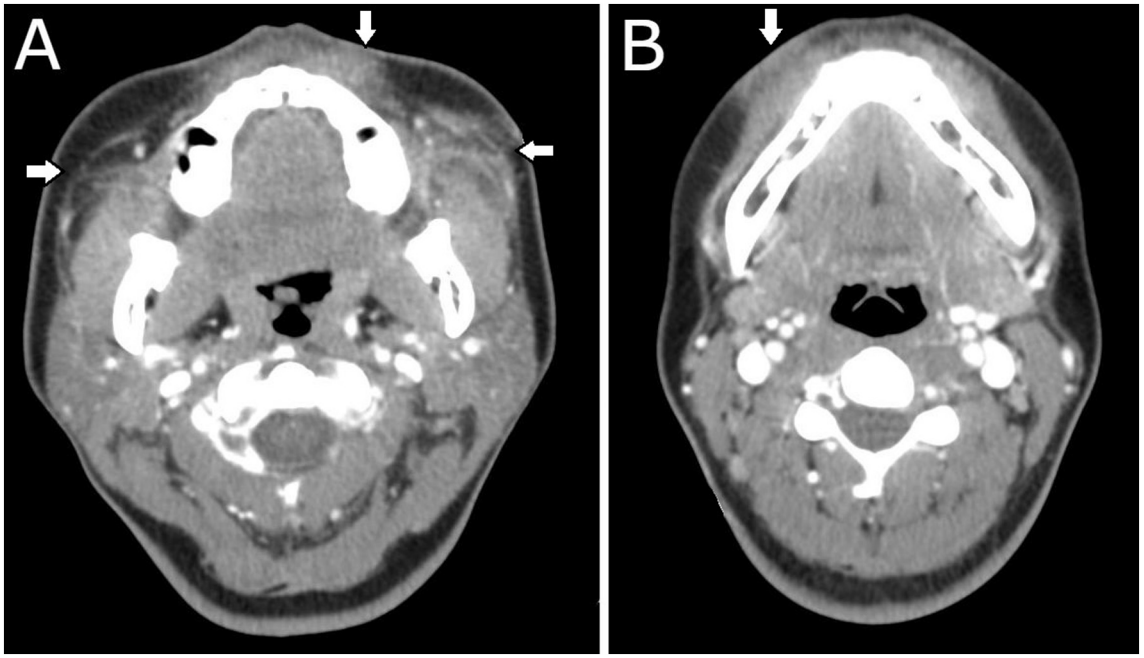

Fig. 2. A) Axial CT at the level of the upper face demonstrates infiltrating soft tissue inflammatory changes without mass effect involving the subcutaneous adipose layer along the left upper lip, with involvement of the left maxillary pre-antral fat. There is no cystic component and no discrete collections. There are also bilateral, left-greater-than-right, inflammatory changes of the superficial musculoaponeurotic system (SMAS) of the face, involving the bilateral buccal spaces, anterior to the master muscles. B) Axial section at the level of the body of the mandible demonstrates inflammatory changes involving the superficial fat and SMAS of the right chin, adjacent to the parasymphysis of the right mandible.

the left upper lip, the right lower lip, with involvement of the left maxillary pre-antral fat space (Fig. 2). There were multiple associated enhancing prominent lymph nodes at level IB and IIA bilaterally, left more than the right. There was no involvement of the parotid glands. There was no sinus or dental disease present. No evidence dental abscess. The overall impression of the findings was that this was likely an inflammatory process, possibly related to lupus panniculitis, rheumatoid disease, or cosmetic injections.

The patient experienced slightly progressive swelling and induration, which gradually worsened over the course of 6–12 months. She underwent contrast enhanced MRI. There was overall progression of soft tissue abnormalities, particularly in the bilateral buccal space, with new abnormalities in the superficial subcutaneous fat. Otherwise, there were no gross changes in the extent of the abnormalities. T1W images revealed reticular infiltration, haziness and ground glass appearance in the superficial and deep fat tissue without mass configuration. Interestingly, the SMAS and vascular structures in the lesion could be clearly identified, with no evidence of displacement or distortion. There was no evidence of a solid mass or extensive edema, which might obscure the underlying fascial and vascular structures. The underlying vascular structures and main parotid ducts could easily be defined within the infiltration. Non-fat saturated T2W images demonstrated linear and reticular hypointensities, haziness and a ground glass appearance in the superficial and deep facial structures, particularly in the buccal spaces. No evidence of significance edema or T2 hyperintensity was appreciated in these regions. There was thickening of SMAS these

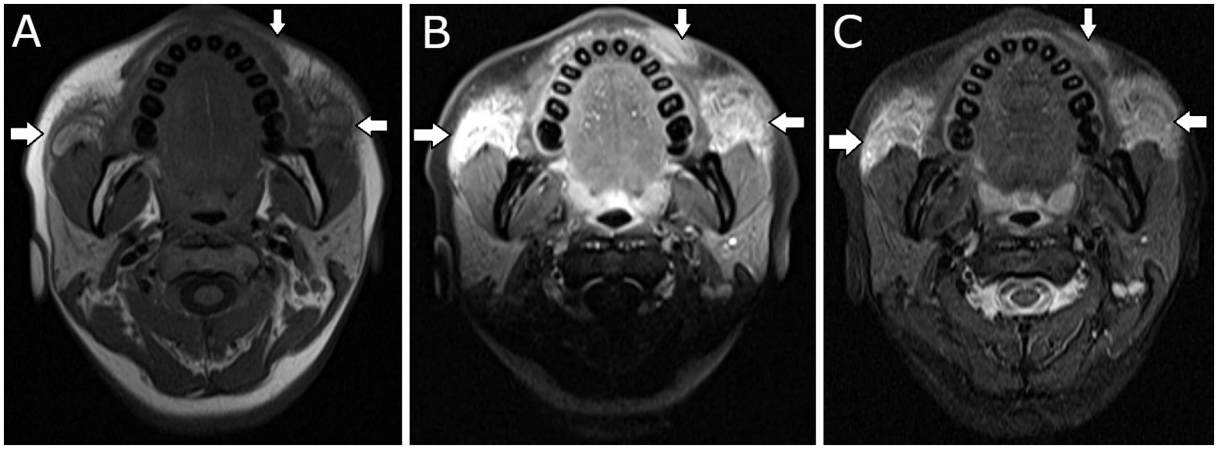

Fig. 3. Axial MR images at the level of the lower maxilla. A) Pre-contrast T1 weighted image demonstrates loss of the normal T1 bright signal within the fat anterior to the bilateral masseters and involving the left upper lip. Instead, there is an infiltrating processes without mass effect, as represented by the dark, reticular T1 signal intensity throughout the fat. The SMAS and vascular structures in the lesion can be seen and there is no evidence of displacement or distortion. B) T1 fat-sat post contrast image clearly demonstrates the inflammatory process, with heterogeneous geographical, reticular enhancement, without mass effect throughout the subcutaneous adipose layer anterior to the bilateral masters and left upper lip. C) Axial STIR at the same level demonstrates abnormal hazy fluid signal intensity, consistent with edema/inflammation.

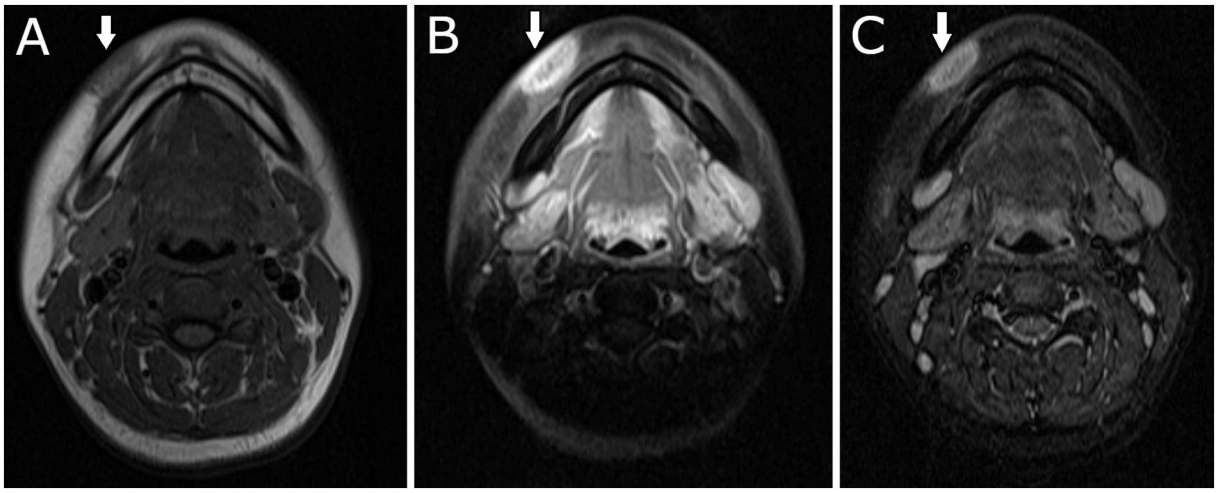

Fig. 4. Axial MR images at the level of the mandible. Pre-contrast T1 (A), Post-contrast T1, fat-sat (B), and STIR (C) images demonstrate loss of the normal T1 bright fat signal, with heterogeneous geographical, reticular enhancement, without mass effect, involving the subcutaneous adipose layer along the right jaw.

regions, left more than the right. On STIR, there was mild, well delineated T2 signal increase in the both buccal spaces, suggestive of edema outlined in the buccal space fat. Underlying vascular structures could be defined in these regions without displacement or distortion. After administration of contrast, there was non-mass-like, moderate, localized enhancement in the fat tissues with preservation of the underlying vascular structures, parotid duct, and fascial structures. Limited diffusion weighted images in the right orbital region revealed mild restricted diffusion, suggesting a hypercellular process (Figs. 3, 4).

The patient's symptoms continued to progress; the initial differential diagnosis included insulin infiltrative diseases such as Melkersson Rosenthal syndrome, lupus, sarcoidosis, amyloidosis, lymphomas, and blepharochalasis. Histopathology from a CT guided biopsy of the patient's right cheek showed lymphoplasmacytic infiltrates and hyaline fat necrosis involving the subcutaneous tissue without involvement of the adjacent skeletal muscle. Higher magnification demonstrated prominent lymphoplasmacytic infiltrates and hyaline fat necrosis (Fig. 5).

The diagnosis of panniculitis was made in the setting of cutaneous lupus. Hydroxychloroquine was contraindicated due to her retinal lattice degeneration. She was started on quinacrine and her symptoms improved.

2. Discussion

The first description of Lupus dates back to the classical period. Rogerious, a thirteenth century physician coined the term. Lupus, which is Latin for “wolf,” was so described because it was thought that the characteristic erosive facial lesions resembled that of a wolf's bite. Systemic manifestations and findings of subcutaneous nodules in patients with lupus were later described by Kaposi in 1883. The first description of patients with lupus panniculitis in the absence of an underlying discoid lupus erythematosus was described in 1956 [1].

Lupus panniculitis affects the deep dermis and subcutaneous adipose layer. It mainly affects females of childbearing age, with a female to male ratio of 4.5:1 [2]. It causes persistent, often tender and painful skin lesions, or subcutaneous nodules, that range from 1 to 5 cm in diameter. The overlying skin may appear erythematous. Lesions may later become ulcerated and can later heal with atrophy, skin depression, dimpling, and scaring [2]. Lesions tend to resolve spontaneously and may follow a chronic course of remission and exacerbation that persists for months to years.

Lupus panniculitis mainly involves the proximal extremities (lateral aspects of the arms and shoulders), thighs, buttocks, trunk, face, and

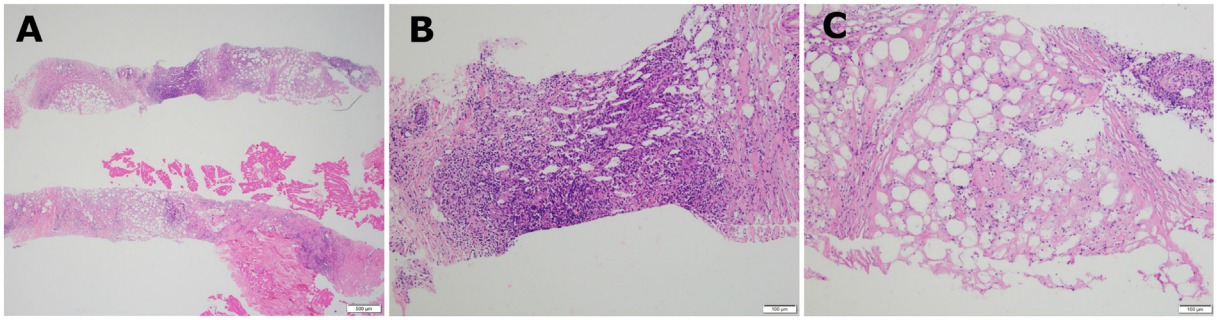

Fig. 5. Histopathology slides from a biopsy of the soft tissue lesion in the patient's right cheek. A. Low magnification photomicrograph demonstrates lymphoplasmacytic infiltrates and hyaline fat necrosis involving the subcutaneous tissue. Note that the adjacent skeletal muscle is uninvolved (Hematoxylin-Eosin stain, 20× magnification). B. Higher magnification clearly demonstrates the lymphoplasmacytic infiltrates (Hematoxylin-Eosin stain, 100× magnification). C. Higher magnification image of the hyaline fat necrosis (Hematoxylin-Eosin stain, 100× magnification).

scalp [3,4]. However, there have also been cases of LP involving the breast, orbit, and even salivary gland involvement [5]. Primarily bilateral buccal fat pad involvement in our case could be explained by the different histologic characteristics of fat in this region, which is mostly composed of brown adipose tissue in early life [6,7]. Based on the histological and ultrastructural data, the buccal fat pad must be considered an example of visceral adipose tissue localized in the face. This is in stark contrast to other adipose tissues in the face, which contain white adipose tissue and a strong collagen network [8].

The exact etiology of panniculitis is unknown. One theory is that an excess of cytokines and circulating immune complexes may lead to an inflammatory response, eventually causing hypodermal necrosis [5]. Patients oftentimes recall a history of trauma in the affected area. In our case, the patient reported noticing her symptoms after falling asleep on her desk, which presumably led to a small amount of micro-trauma, possibly triggering her symptoms.

The diagnosis of LP can be difficult, especially in patients who lack other skin or systemic manifestations of lupus erythematosus. As in our case, the involvement of unusual body sites may be the only presentation and these lesions tend to go undiagnosed or misdiagnosed. The diagnosis is usually based on the characteristic clinical features and confirmed by biopsy and histopathology.

Histopathology examination of a biopsy from a lesion may show two subtypes of panniculitis: septal and lobular. In the septal subtype, the intralobular septa of adipose tissue are affected by necrosis and hyalinization. This contrasts with the lobar type, where the fat lobules are primarily affected [9,10]. In practice, both patterns occur in most patients simultaneously as the inflammatory infiltration is not strictly separated. The two most important histopathologic criteria for diagnosing LP are the presence of lymphocytic infiltrates that involve fat lobules and the hyaline necrosis of those lobule.

Imaging findings can help narrow the diagnosis in facial LP. Ultrasound will likely show inflammatory changes and hyperemia along the involved and surrounding subcutaneous adipose layer. However, the main role of ultrasound is to rule out an underlying abscess or drainable fluid collection or mass. If ultrasound is unrevealing, a CT is the next diagnostic imaging modality of choice. CT will demonstrate infiltrating soft tissue inflammatory changes involving the subcutaneous adipose layer and the superficial musculoaponeurotic system (SMAS) [2]. As opposed to cellulitis, where there is general edema and inflammatory changes primary centered about the skin, the CT imaging characteristics in LP will demonstrate inflammatory changes and stranding primary affecting the subcutaneous adipose layer. CT generally provides excellent spatial resolution and is generally well-available. The down-side to CT is that it exposes the patient to a small amount of ionizing radiation and has worse contrast resolution than MRI. In the authors' opinion, MRI is the imaging modality of choice for diagnosing facial LP. The hallmarks of facial LP on MRI are loss of the normal T1 bright signal within the subcutaneous adipose layer. Post-contrast T1 fat-sat images or STIR images are the most sensitive sequences, which will demonstrate bright, hazy contrast enhancement and edema corresponding to the areas of interest on pre-contrast T1 images.

It is important to distinguish LP from underlying lymphoma. Lymphoma is highly cellular mass with a mass-like configuration which, unlike lupus panniculitis, does not track along the fatty tissue planes. The main way to distinguish LP from lymphoma is by identifying the underlying tissue skeleton. In lymphoma, the underlying tissue skeleton cannot be seen and will not be preserved. Lymphoma will also be hypointense on T2 weighted images due to its increased cellularity, whereas LP will be hyperintense on T2 fat sat images secondary to the edema.

Imaging features of LP are extremely scarce in the literature. This article is the first of its kind to describe in detail the unique MR features of facial LP as well as CT and ultrasound appearances. Vattoth and Cure presented a case of lupus panniculitis, with associated CT imaging features; these authors described an ill-defined inflammatory infiltration of the subcutaneous fat near the masticator space and parotid gland extending to the buccal space fat [2]. In a review article regarding imaging of connective tissue diseases of head and neck region, the CT imaging findings of LP involving the subcutaneous soft tissue of the cheek were described as soft tissue infiltration and thickening [11]. Regarding the MRI findings, Wang et al. described a case of parotid lupus with lupus erythematous panniculitis involving the parotid gland and adjacent subcutaneous fat [12]. Lupus panniculitis rarely involves the breast; Sabaté et al. described a case of LP involving the breast which demonstrated an irregular, heterogeneous mass involving the subcutaneous fat fad with inflammatory skin thickening [13]. We believe that the unique description of ground glass opacities involving the fat on T1 and T2 weighted images (with preservation of the underlying tissue skeleton) and the dark, reticular signal on T1W non-fat-sat images (suggesting cellular infiltration) helps to narrow the differential diagnosis in the appropriate clinical setting.

3. Conclusion

The imaging features of facial LP are not well described and can often be missed. We have clearly defined the characteristic ultrasound, CT, and MRI imaging findings of facial LP. In the authors' opinion, MRI is the imaging modality of choice for diagnosing facial LP, as it can safely be performed and provides excellent contrast resolution of the affected soft tissues. The unique ground glass appearance on T1 and T2 weighted images with preservation of the underlying tissue skeleton and dark reticular signal on T2W non-fat-sat images (suggesting cellular infiltration) is the hallmark of facial lupus panniculitis.

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