Radiology Case Reports

Volume 7, Issue 3, 2012

MRI findings for parotid lupus

Page I. Wang, MD; Erin L. McKean, MD; Jonathan B. McHugh, MD; and Suresh K. Mukherji, MD

This report presents the MRI findings for parotid lupus erythematosus panniculitis. Although the clinical findings of this disorder have been described, very few reports describe the CT findings on this rare disease entity in the parotid region and no reports include its MR appearance. This unusual diagnosis should be considered in the context of proper clinical history.

Case report

Lupus erythematosus panniculitis (LEP), also known as lupus profundus, is a rare but important disease resulting from subcutaneous inflammation of adipose tissue, often associated with discoid and systemic lupus. In the head and neck, LEP has been reported to occur in the submandibular space, periorbital area, cheek, and ear (1-6). To our knowledge, there is no literature on the MR appearance of parotid LEP.

A 41-year-old female with a past medical history of discoid lupus erythematosus (DLE) and two deep-vein thrombi in the postpartum period presented to an otolaryngologist with an asymptomatic "mass on her right cheek." Review of systems was significant for joint pain, fullness in the right preauricular region, and a clicking sensation in her right ear.

On examination, there was a 2 x 3cm firm mass overlying the right parotid with no cutaneous elevation, depression, or erythema. This mass was nontender and diffusely adherent to the underlying soft tissue. There was no associated facial numbness or weakness. Otologic evaluation was unremarkable. There was no palpable cervical lymphadenopathy. Laboratory examination demonstrated negative results for ANA, cardiolipin antibody, C-reactive protein, and rheumatoid factor. Her SED rate was slightly elevated, at 26. Her WBC count was slightly low, at 3,900 mm3.

A contrast-enhanced MRI was performed to further evaluate the lesion. The noncontrast T1 images showed an ill-defined reticulated lesion involving the accessory right parotid gland, with extension into adjacent subcutaneous fat and anterior buccal space (Fig. 1A). The T2 images showed a parotid mass that was isointense to the parotid (Fig. 1B). The postcontrast, fat-suppressed T1W images showed dense focal enhancement of the mass compared to the surrounding parotid gland and periparotid fat (Fig. 1C). There was no evidence of restricted diffusion or perineural spread.

Given the nonspecific MRI findings and concern for neoplasm, a fine-needle aspiration biopsy of the lesion was performed, which was nondiagnostic. The patient developed no additional symptoms and returned for followup six weeks later. The nodular area now appeared mobile and had grown to 4 x 3 cm. A larger needle/punch biopsy histologically demonstrated a panniculitis characterized by a lymphohistiocytic infiltrate including plasma cells, involving fat lobules with associated hyaline fat necrosis (Fig. 2). At higher magnification, nuclear dust (karyorrhectic debris) could be identified within the infiltrate. These histopathologic findings are consistent with LEP.

Discussion

LEP is a rare disease seen more commonly in women, ages 20-45 (7). It presents with persistent, deep, firm erythematosus plaques and nodules and involves the proximal extremities, trunk (including breasts), scalp, face, neck, and buttocks. The rare pediatric cases show a predilection for the face (7). These lesions can be tender and painful; they often heal with lipoatrophy, deep depressions, dystrophic calcifications, and ulceration. LEP is a chronic and recurrent disease. The treatment for LEP includes antimalarial drugs, sunscreen, and systemic glucocorticoids (11).

Citation: Wang PI, Mckean EL, McHugh JB, Mukherji SK. MRI findings of parotid lupus. *Radiology Case Reports*. (Online) 2011;7:667.

Drs. Wang and Mukherji are in the Department of Radiology, Dr. McKean is in the Department of Otolaryngology, and Dr. McHugh is iin the Department of Pathology, all at University of Michigan Hospital, Ann Arbor MI. Contact Dr. Wang at pagew@umich.edu.

Competing Interests: The authors have declared that no competing interests exist.

DOI: 10.2484/rcr.v7i3.667

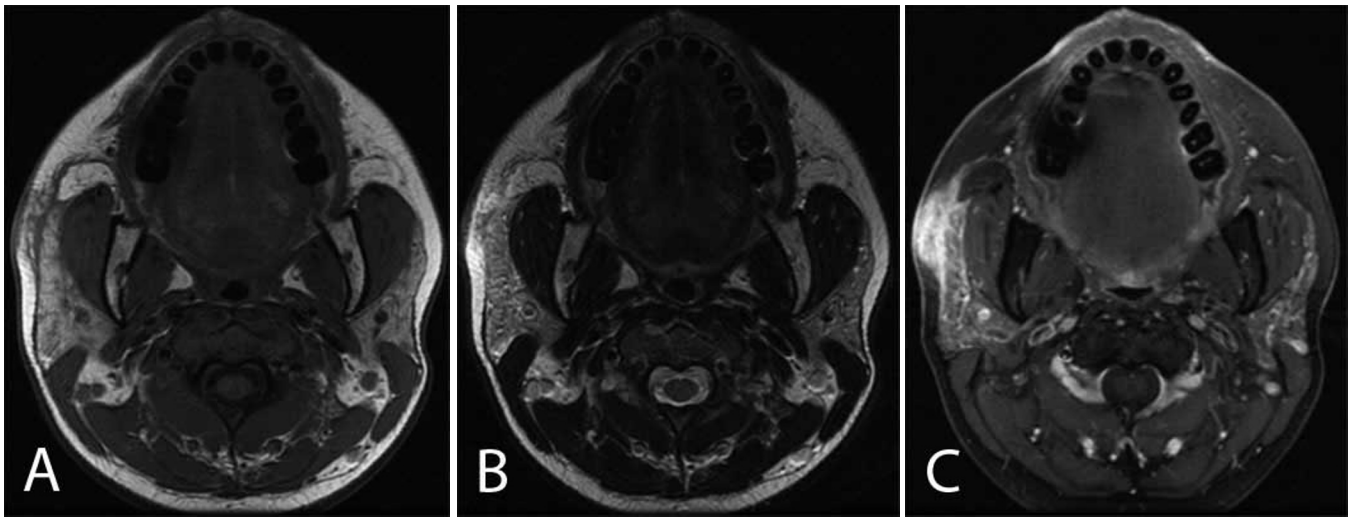

Figure 1. 41-year-old female with lupus erythematosus panniculitis. A. Axial MRI T1 without contrast image at the level of the right accessory parotid shows an ill-defined reticulated lesion involving the accessory right parotid gland, with extension into adjacent subcutaneous fat. B. Axial MRI T2 image shows high signal intensity in the right peripartoid fat. The right parotid itself does not appear hyperintense. C. Axial MRI T1 postcontrast, fat-saturated image shows high signal intensity and marked homogeneous enhancement in the right accessory parotid, with extension into the adjacent subcutaneous fat.

Serologies in patients with LEP are typically normal, but these patients have been documented to have elevated erythrocyte sedimentation rate, leukopenia, anemia, decreased C4 levels, rheumatoid factor, and false-positive

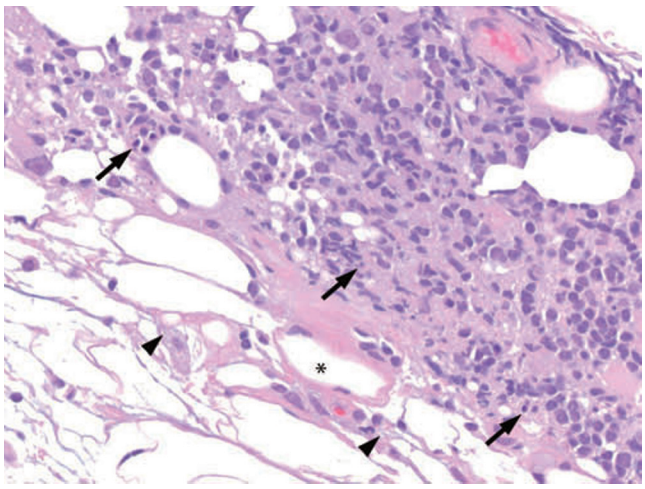

Figure 2. 41-year-old female with lupus erythematosus panniculitis. High-power view demonstrating a lobular panniculitis composed of a lymphohistiocytic infiltrate with occasional plasma cells and with associated hyaline fat necrosis (asterisks), mucin deposition (arrowheads), and nuclear dust (arrows) (400X; hematoxylin and eosin).

syphilis serologies. Patients with concurrent systemic lupus erythematosus (SLE) often have positive ANA titers and positive double-stranded DNA titers (9).

LEP may be an isolated phenomenon, or it may develop in patients with DLE and SLE. Clinical DLE is present in 20-60% of LEP patients, with the histologic diagnosis of DLE present in up to 75% of LEP patients (7). Unfortunately, the diagnosis of DLE may precede or follow the presentation of LEP by several years (7). Clinical SLE is present in 10-42% of patients with LEP, with histologic lupus-band tests positive in 70% of these patients (7). Inflammatory diseases such as alpha 1 antitrypsin deficiency, morphea profunda, sarcoid, pancreatic panniculitis, factitial panniculitis, alopecia areata, and infections (such as mycoplasma and fungal infections) may occasionally mimic the appearance of LEP (9).

The histopathology of LEP consists of septal and lobular panniculitis, often with associated characteristic features of lupus erythematosus in the overlying dermis and epidermis. More specific findings include hyaline fat necrosis, stromal mucin (glycosaminoglycan) deposition, lymphocytic infiltrates forming aggregates (occasionally with germinal centers), and nuclear dust (karyorrhectic debris) associated with the inflammatory infiltrate (10). LEP also contains cytologically atypical mononuclear cells that often rim the periphery of adipocysts. Another characteristic finding is phagocytosis by macrophages of erythrocytes and lymphocytes, resulting in so-called "beanbag" cells.

In clinical practice, LEP needs to be differentiated from other neoplastic, inflammatory, and infectious diseases. The primary neoplasm that presents in a similar fashion is subcutaneous panniculitis-like T cell lymphoma. Subcutaneous panniculitis-like T cell lymphoma can be further differentiated from LEP by polymerase chain analysis of the T-cell receptor gene (both $\alpha\beta$ and $\lambda\delta$) and by immunohistochem-

istry to evaluate for a high proliferation index (M1B1) and, in some cases by identifying CD56 expression (11).

The imaging appearance of periparotid LEP can mimic parotid tumors, sialoadenitis, and parotiditis, and therefore should be listed in the differential of unexplained parotid masses, especially in a patient with a known lupus history. The diagnosis could have been suggested in our case by the ill-defined reticulation identified on the noncontrast T1W images. However, the dense focal enhancement seen on the postcontrast, fat-saturated T1 image was nonspecific and could be seen in a variety of parotid neoplasms. Failure to recognize LEP has led to unnecessary parotidectomy and morbidity (2).

The true incidence of LEP is probably underestimated, as LEP is largely ignored by the major head and neck pathology and surgery textbooks, and this diagnosis can be overlooked by the unsuspecting clinician or pathologist, especially if the patient does not have a lupus history (2). LEP is a treatable and well-established disease that should be included in the differential diagnosis of parotid and periparotid masses, especially if the patient has discoid or systemic lupus erythematosus and no facial nerve involvement.

References

1. Vattoth, S. and J.K. Cure, CT Imaging of Head and Neck Lupus Panniculitis. *AJNR Am J Neuroradiol*, 2008. [PubMed]
2. White, W.L., et al., Periparotid lupus erythematosus panniculitis. Clinicopathologic correlation of two cases presenting as primary parotid disease. *Arch Pathol Lab Med*, 1993. 117(5): p. 535-9. [PubMed]
3. Ogura, N., et al., Lupus erythematosus profundus around the salivary glands: a case resembling submandibular salivary gland disease. *Lupus*, 1997. 6(5): p. 477-9. [PubMed]
4. Magee, K.L., et al., Lupus erythematosus profundus with periorbital swelling and proptosis. *J Am Acad Dermatol*, 1991. 24(2 Pt 1): p. 288-90. [PubMed]
5. Kossard, S., Lupus panniculitis clinically simulating alopecia areata. *Australas J Dermatol*, 2002. 43(3): p. 221-3. [PubMed]
6. Ng, P., S. Tan, and T. Tan, Lupus erythematosus panniculitis: a clinicopathologic study. *International Journal of Dermatology*, 2002(41): p. 488-490. [PubMed]
7. Fraga, J. and A. Garcia-Diez, Lupus erythematosus panniculitis. *Dermatol Clin*, 2008. 26(4): p. 453-63, vi. [PubMed]
8. Wolff K., J.R., and Suurmond D. , *Fitzpatrick's color atlas and synopsis of clinical dermatology*. 5th Ed ed. 2005, NY, NY: McGraw-Hill.
9. Lahita, R.G., *Systemic lupus erythematosus*. 4th Ed. ed. 2004: Academic Press.
10. Peters, M.S. and W.P. Su, Lupus erythematosus panniculitis. *Med Clin North Am*, 1989. 73(5): p. 1113-26. [PubMed]
11. Gonzalez, E.G., et al., Subcutaneous panniculitis-like T-cell lymphoma misdiagnosed as lupus erythematosus panniculitis. *Clin Rheumatol*, 2007. 26(2): p. 244-6. [PubMed]