CASE REPORT

Unilateral parotid mass as an unusual presentation of lupus erythematosus profundus

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SUMMARY

Unilateral parotid swelling or mass in the lupus erythematosus profundus person is a rare or under reported clinical scenario in existing literature. This is a case of a 49-year-old man with such presentation for whom medical management of the underlying condition led to the resolution of the swelling. Lupus erythematosus profundus must be considered as a differential diagnosis of unexplained parotid swelling to prevent incorrect diagnosis, unnecessary surgical intervention (eg, parotidectomy) and the morbidity that comes with it (eg, facial nerve weakness, Frey's syndrome etc).

BACKGROUND

Lupus erythematosus profundus (LEP), also referred to as lupus panniculitis, is a rare but important disease. It is considered to be a type of discoid or cutaneous lupus erythematosus (DLE) but can occur in isolation or in association with DLE and the systemic form of lupus that is, systemic lupus erythematosus (SLE). Salivary gland involvement, though acknowledged, has sparsely been reported.^{1–4}

Intermittent and long-standing swellings of the major salivary glands are frequently referred to the head and neck specialists for investigation and management. Their close association with important neurovascular structures means that surgical intervention may result in relatively high morbidity. Management must be based on an accurate definitive diagnosis in the context of the given clinicopathological scenario. This case report demonstrates the importance of multidisciplinary input, correct diagnosis and subsequent management of a unilateral parotid swelling presenting as a part of a systemic disease entity. This report adds to the currently sparse literature on the salivary gland involvement of this disease and adds LEP to the list of differentials to the causes of unilateral parotid swelling.

CASE PRESENTATION

A 49-year-old surgeon developed a significant asymptomatic, firm, mobile swelling of the left parotid gland of relative sudden onset. There was no report of oral discomfort, dryness of the mouth or eyes. No other swellings, general malaise, weight loss or night sweats were reported either.

Medical history was unremarkable apart from recent diagnosis of DLE of the scalp in the occipital region.

An examination revealed a 2.5×3 cm well-defined localised, firm, mobile swelling in the preauricular region of the left parotid gland. Overlying skin was normal. Facial nerve function was noted

to be grossly intact with no regional lymphadenopathy. Otological and intraoral examinations were unremarkable. Oral mucous membrane was healthy and adequately moist with no obvious salivary ductal obstruction. The contralateral parotid and other major salivary glands appeared normal in structure and function.

INVESTIGATIONS

Blood investigations revealed a normal full blood count, slightly raised erythrocyte sedimentation rate (19 mm/h), a negative autoimmune screen, normal renal and liver functions and normal serum ACE and complement levels. The ultrasound scan demonstrated a localised area of glandular enlargement within the anterior aspect of the gland close to the main parotid duct. No discrete masses or ductal dilation were seen. An incisional biopsy of the scalp lesion showed dense lymphoid infiltrate with positive IgM and complement C3. The predominance of dermal and epidermal changes coupled with focal involvement of subcutaneous tissue made LEP a more likely diagnosis than the other possibility of subcutaneous panniculitis like T-cell lymphoma. Subsequent fine needle aspirate (FNA) of the parotid lesion showed a dense lymphocytic infiltrate (mainly T-cell population). Repeat FNA confirmed a dense, mainly lymphocytic, cell infiltrate with no loss of acinar tissue, granulomatous changes or malignant cells. Immunocytochemistry performed on the sample demonstrated a mainly T-lymphocyte population again with normal CD4:CD8 ratio. Further MRI scan with contrast confirmed a single focal homogenous abnormality 2.2×1.3 cm within the superficial lobe of the gland (figures 1 and 2) with no areas of necrosis or cystic change. These findings were consistent with localised sialadenitis. Several prominent lymph nodes adjacent to the gland were also detected. The plain chest radiograph demonstrated normal appearances and failed to show any significant abnormalities or features suggestive of sarcoidosis. The conclusions of this concurred with sialadenitis of a short history and further reinforcement of the 'observe only' course of management.

In light of the patient's recent diagnosis of DLE and evident inflammatory process affecting the parotid unilaterally, the clinicopathological scenario prompted a second opinion from a lupus specialist and rheumatologist. Further history taking failed to indicate the presence of other connective tissue disorders. His scalp lesion now appeared as a deep area of subcutaneous fat atrophy with alopecia and some surrounding nodules. A new autoantibody screen revealed his

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Figure 1 Post-contrast MRI coronal image showing 2.2×1.3 cm focal homogeneous abnormality within the superficial lobe of the left parotid with no areas of either necrosis or cystic change. This has poorly defined interface with the surrounding soft tissues.

antinuclear antibody (ANA)—an antibody with no well-defined clinical association—to be weakly positive at 1 in 80 with a speckled pattern on immunofluorescence. A weak perinuclear antineutrophil cytoplasmic antibodies pattern was also demonstrated—which was deemed to be most likely due to the presence of ANAs or a non-specific finding secondary to recent infection or inflammation. The patient's anti-SS-A and anti-SS-B, anti-Sm and anti-RNP were all negative. The immunoglobulins—IgG, IgA and IgM and C3, C4 and rheumatoid factor levels were all within normal range. A conclusion was reached that the same process was affecting both the scalp and parotid—lupus erythematosus panniculitis.

DIFFERENTIAL DIAGNOSIS

Unilateral parotid involvement can be due to primary salivary gland neoplasms, secondary deposits, acute bacterial sialadenitis

Figure 2 Post-contrast MRI axial image showing further abnormal signal change along the line of the parotid duct.

or abscess and chronic sialadenitis. Salivary gland exocrinopathy in association with other autoimmune disease such as SLE is termed secondary Sjogren's syndrome. The unilateral focal parotid swelling, lack of sicca symptoms and the immune profile ruled out secondary Sjogren's syndrome in this case. The cutaneous lupus erythematosus covers a spectrum of lesions and includes both discoid lupus and lupus profundus. Histological changes in lupus profundus are best appreciated with deep biopsies, including subcutaneous fat which might not be easy from the scalp and the parotid area without risking deeper vital structures.

TREATMENT

Treatment was initiated with 20 mg prednisolone daily for 20 days with 200 mg hydroxychloroquine daily but subsequently as maintenance therapy which resulted in almost complete resolution of the parotid swelling. The scalp lesion remained asymptomatic following three intralesional corticosteroid injections with no adverse clinical changes.

OUTCOME AND FOLLOW-UP

At 5 years, the patient has remained systemically well. The scalp lesion has been occasionally itchy but remained clinically stable and the patient remains on a maintenance dose of hydroxychloroquine. All that remains of the parotid mass is a small non-tender subcutaneous induration with no residual swelling. It has been recommended that should the parotid swelling recur and enlarge, it should be excised for definitive histology.

DISCUSSION

LEP was first described in 1883 by Kaposi⁵ but later given these titles in 1940 by Irgang.⁶ Typically, LEP presents as (often chronic) painless, mobile, *subcutaneous* nodules or plaques with normal or abnormal overlying skin. The face, proximal limbs and buttocks are most commonly affected. Young and middle-aged females are most commonly affected. Diagnosis is based on clinical and histological findings. Response to antimetabolites such as hydroxychloroquine is well documented, especially in LEP associated with DLE. Systemic corticosteroid therapy and intralesional steroid injections also have their place.⁷ Lesions typically heal with atrophy and/or scars. Surgery should only be attempted when other less invasive approaches have been unsuccessful and debilitation is appreciable.² In this case LEP of the parotid appeared to resemble sialadenitis histologically. This suggests that such conditions may have been under reported due to underdiagnosis or misdiagnosis.⁸

Learning points

- ▶ Lupus erythematosus profundus also referred to as lupus panniculitis is a rare but important disease.
- ▶ This is considered to be a type of discoid or cutaneous lupus but can present in isolation or in association with discoid or systemic lupus.
- ▶ Salivary gland involvement though acknowledged, has been sparsely reported.
- ▶ This report adds lupus erythematosus to the list of differential diagnosis of unilateral parotid swellings, especially in the context of concomitant presence of discoid or cutaneous lupus lesions.

LEP has already been acknowledged as a differential diagnosis of an unexplained submandibular mass in a patient with SLE.⁸ This case report adds to the currently sparse literature that identifies LEP as a cause of unilateral parotid swelling. This patient was managed effectively non-surgically via a multidisciplinary approach to diagnosis and treatment.

Contributors All the authors have been involved in writing and reviewing the paper.

Competing interests None.

Patient consent Obtained.

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