Could we use salivary gland ultrasonography as a prognostic marker in Sjögren’s syndrome?

Response to: 'Ultrasonographic damages of major salivary glands are associated with cryoglobulinemic vasculitis and lymphoma in primary Sjögren’s syndrome: are the ultrasonographic features of the salivary glands new prognostic markers in Sjögren’s syndrome?' by Coiffier et al

We thank Dr Coiffier and colleagues for their interesting letter in which they suggest that damages of major salivary glands (SG) might be associated with cryoglobulinemic vasculitis and lymphoma in primary Sjögren’s syndrome. The authors raise the question whether the ultrasonographic (US) features of the salivary glands [our proposed novel ultrasound scoring system] could be used as the new prognostic markers for cryoglobulinemic vasculitis and lymphoma in patients with Sjögren’s syndrome.1

In their recent study of different US scoring systems in 97 sicca syndrome patients, Coiffier et al found three B-cell lymphomas in the primary Sjögren’s syndrome (pSS) group and three, two, and one patient with cryoglobulinemic vasculitis in the pSS, secondary Sjögren’s syndrome (sSS) and control groups, respectively.2 The authors concluded that the detection of B-cell lymphomas or cryoglobulinemic vasculitis was associated with pathological US findings regardless of the scoring used. The reported US pathological features of SG were either numerous cystic lesions without healthy parenchyma or fibrous glands scored as a grade 3 according to the new semi quantitative scoring system described by the SG sub group of the OMERACT US working group.1 Although their findings may suggest specific ultrasound features as a risk factor for developing lymphoma or cryoglobulinemic vasculitis, we think at this point in time it is premature to draw such a conclusion.3 Ultrasound may reveal predisposing factors, but these are not proven to be pathognomonic of lymphoma. Indeed, several predictors of lymphoma in pSS such as epidemiological, clinical (permanent swelling of the SG, palpable purpura, organomegaly), biological (cryoglobulinaemia, or low complement levels) and histopathological findings should also be taken into account.4–9 Large sample and longitudinal studies assessing these clinical and biological predictors of lymphoma with US are currently ongoing and will probably shed more light on this challenging issue. Furthermore, pSS disease activity, for example, assessed by the EULAR SS index, can be used as a clinical predictor of lymphoma development with a dose effect.10

SG enlargement (eg, clinical aspects: unilateral, fixed and hard parotid glands) is regarded as the most dominant clinical symptom for lymphoma in patients with pSS.6 SGUS performed by well-trained ultrasonographers can provide a precise structural assessment of the glands’ surface compared with clinical examination.11 In case of SGUS grade 3, that is, complete destruction of the gland, with numerous hypo-echoic or hyper-echoic bands, the detection of abnormal lymph nodes should raise awareness of possible lymphoma development. Suspicion of abnormal lymph nodes can be confirmed during a long term monitoring of pSS patients especially those with high risk of lymphoma development. In addition, Doppler assessment of gland’s vascularisation in pSS might be of help to detect at risk ultrasound lesions. To this end, the forthcoming results of a longitudinal study for development of consensual Doppler US scoring of gland’s vascularisation in pSS can be quite helpful.

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REFERENCES
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