Introduction:
Chronic sclerosing sialadenitis (CSS) is a chronic inflammatory condition of the salivary glands, most commonly affecting the submandibular gland. It causes a firm swelling of the gland, which is clinically indistinguishable from a true neoplasia and is diagnosed after removal of the gland. It was first described by H. Küttner in 1896 (1). Recently it has been suggested that the entity is an autoimmune disease and a part of the IgG4-related diseases characterized by causing mass-forming lesions in various tissues (2,3).

It can affect one or both of the submandibular glands (localized type) and sometimes other organs (systemic type), i.e. the pancreas. Studies have shown a high level of IgG4 in the serum in these patients (4) and immunohistochemical staining of the salivary gland tissue has shown high levels of IgG and IgG4 positive plasma cells (2,3).

To increase awareness of this entity we present the histological and immunohistochemical findings in three Danish patients diagnosed with CSS.

Materials and methods:
Three patients were referred to the Department of ENT, Slagelse Hospital or Odense University Hospital, with a unilateral, firm swelling of the submandibular gland. There were no subjective complaints. The patients went through clinical examination with fine needle aspiration (FNA) and ultrasonography. Based on these examinations malignancy could not be excluded. The patients therefore had either the whole gland surgically removed or an excision made. Histology showed salivary gland tissue with preserved lobular architecture, but with marked fibrosis, acinar atrophy, and a dense lymphoplasmacytic infiltrate. As no aetiology was known, the lesions were diagnosed as CSS with no sign of malignancy. Morphologically, however, CSS can be indistinguishable from sialadenitis NOS (figure 1).

Results:
The salivary gland tissue from the patients diagnosed with CSS showed an increased number of IgG4 and IgG positive plasma cells/hpf (mean 224 and 297 respectively) compared to patients with sialadenitis NOS (mean 4 and 24 respectively) (figure 2).

Discussion and conclusion:
Clinically, CSS can be difficult to distinguish from a true neoplasia. When combining a high level of IgG4 in serum, a benign FNA cytology and ultrasonography, a tentative preoperative diagnosis of CSS can be made. The treatment of choice is still surgery. However, acknowledging this entity of IgG4-related and steroid-sensitive diseases can potentially diminish the need for surgical procedures.

The histomorphological findings are classical with fibrosis, atrophy and a dense inflammatory infiltrate consisting of numerous lymphocytes and plasma cells. Immunohistochemical staining shows abundant IgG4 and IgG positive cells. The IgG4/IgG ratio is high compared to other inflammatory diseases of the salivary glands. The morphological appearance combined with elevated expression of IgG4 is diagnostic for CSS. When diagnosing this entity, it should be considered, if other organs could be involved.

Reference List