Primary malignant lymphomas of salivary glands are relatively rare and usually affect the parotid gland. They account for 0.3% of all tumors and 2-5% of salivary gland neoplasms. Their incidence increases with age and reaches its peak about the 55th year of life. Male to female ratio is 2:1. NHL may be extranodal, originating from MALT and nodal, - from a lymph node inside the gland. In contrast with other locations extranodal parotid NHL are with lower malignancy and better prognosis.

Etiology
NHL pathophysiology is unknown, but it is considered that acquired or drug induced immunodeficiency increases the risk 50 to 100 times. Some autoimmune diseases are recognized as risk factors. In a recent metaanalysis of 12 studies over 29423 patients with Sjogren’s syndrome (SS) was found 100-fold increase of the risk for NHL. Patients with SS are five times more likely to develop NHL. Some autoimmune diseases are associated with a high frequency of NHL. Autoimmune disorders are observed in up to 44% of NHL patients.

Diagnosis and treatment
The clinical findings are unspecific and the diagnosis can often be avoided. Parotidectomy is considered, unnecessarily extensive surgery as consequence of unnecessary radicalism with all accompanying risks. NHL is treated by chemo- and radiotherapy. The role of surgery is purely diagnostic.

Aim
To stress the importance of clinical thinking towards malignant lymphomas of the parotid and review the data from history, presentation and radiological investigations, which may direct us to the diagnosis.

Clinical case
We present a 54 years old female with indolent formation in the left parotid region, dating from 3 months, with hard-elastic consistence, adherent to the skin but mobile in account of the underlying tissues, measuring 4/3/3cm. CT-scan showed infiltration of the overlying skin and the left lobulus and leftsided submandibular and jugular lymph nodes ranging from 9 to 15mm (fig.1).

On surgery was found a formation, originating from the parotid gland and infiltrating the skin and an enlarged adjacent lymph node. A fresh frozen section showed poorly differentiated carcinoma of the gland with skin infiltration and metastasis in the lymph node. Radical parotidectomy with facial nerve preservation was undertaken with partial resection of the left lobulus, excision of the affected skin, and selective neck dissection in regions Ib, II and III. The skin defect was covered by local plasty. Paraffin sections showed salivary gland, muscular fibers, 1 lymphatic node, subdermal tissues and skin with deep infiltration of small, monomorphic lymphocyte like cells, some with bigger hyperchromatic nuclei, which might correspond to small cells undifferentiated carcinoma, neuroendocrine tumor or NHL. The lymph nodes from the neck dissection were with hyperplastic lymphadenitis. IHH suggested follicular lymphoma G1-2 or atypical follicular hyperplasia. There were no postoperative complications. No dissemination was found. The patient underwent radio- and chemotherapy and was disease free 1 year after the operation.

CT symptoms:
• poorly defined border (fig.2a);
• infiltration (fig.2b);
• multifocality (fig.2c).

Options for histological diagnosis
• The existence of locoregional lymphadenopathy may give the option for biopsy without parotidectomy.
• In cases with infiltration of skin or superficial infiltration incisional biopsy can be done without breaking the oncological principles.
• FNAB may be diagnostic, but its reliability depends on competence of the pathologist and the possibility for cytopathologic investigations, including flowcytometry.

Prognosis
5-years survival rate in parotid lymphoma varies between 50% and 75%. No statistically significant correlation between survival and histological variation is observed. There are data that in lesions sized less than 6cm, 5-year survival reaches 87%, and in bigger is about 61%. Surgical reduction of the tumor does not improve prognosis.

Conclusions
Primary parotid NHL is a disease with good prognosis and high local tumor control. If it is considered, unnecessarily extensive surgery can often be avoided. Parotidectomy is indicated only in cases when fine needle aspiration biopsy is impossible, no other tumor tissue is accessible for sampling and the results from FNA or frozen sections are unreliable. The role of surgery is purely diagnostic. The proved NHL is treated exclusively conservatively.

Reference