A 17-year-old male presented with a painless swelling in the right parotid region of 2 years duration. The swelling was progressively increasing in size. On examination there was a round, firm, smooth-surfaced and non-tender swelling in the right infra-auricular region. CT scan revealed a smooth well-defined mass with a lobulated surface and homogeneous soft tissue density. Clinical differential diagnosis included benign salivary gland tumours.

Fine needle aspiration was done from the swelling. Smears showed reactive population of lymphoid cells in various stages of maturation along with a few clusters of benign ductal epithelial cells, suggesting the diagnosis of reactive lymphoid hyperplasia of intra-parotid lymph node. Subsequently, the patient underwent superficial parotidectomy and the specimen was sent for histopathological examination. It was a single globular soft tissue mass measuring 4.5 x 4 x 3 cm. Cut section showed a well-defined homogeneous greyish brown mass surrounded by a thin rim of normal salivary gland tissue. Microscopically, the lesion was composed of lymphoid follicles of varying sizes with atrophic germinal centres traversed by hyalinised vessels (Figure 1). Mantle zone lymphocytes showed characteristic concentric onion skinning around the atrophic germinal centres (Figure 2). There was prominent vascular proliferation in the interfollicular area. A few ductal epithelial cells in the form of salivary inclusions representing embryologic remnants were also seen. Normal compressed salivary tissue was identified at the periphery (Figure 3). On the basis of these findings, a diagnosis of Castleman’s disease (hyaline vascular type) of intra-parotid lymph node was made.

Discussion

Castleman’s disease is a peculiar type of lymph node hyper-
plasia and is known by different names including lymphoid hamartoma, giant lymph node hyperplasia and angiofollicular lymph node hyperplasia. It affects young adults between 15-35 years of age and has no sex predilection.

The most common sites of this lesion are the mediastinum, head and neck, cervical lymph nodes, axilla and the abdomen. Though the head and neck is the second most common site for this lesion with 61 cases reported, the involvement of the parotid gland is extremely rare with only 20 case reports in the English literature. The majority of the cases of Castleman’s disease of the parotid arise within the para-glandular or intra-glandular lymph nodes. In the present case the lymphoid lesion was localised within the salivary gland with epithelial inclusions within it and was surrounded by normal salivary tissue, thus indicating that the disease process was taking place in the intra-parotid lymph node.

The origin of this entity is still unknown. Some authors favour a theory of lympho-proliferation due to chronic antigenic stimulation by a virus or chronic inflammation while others consider it to be a lymphoid hamartoma. Two histological types are recognised: classic hyaline vascular type which is usually unicentric, and plasma cell type which can be multifocal and associated with systemic symptoms. It is hypothesised that the plasma cell lesion is an early active process and the hyaline vascular lesion represents the final outcome.

Castleman’s disease often presents a diagnostic challenge because of the paucity of signs and symptoms, lack of diagnostic tools and its tendency to mimic neoplasms. Patients present with an asymptomatic mass lesion and rarely with vague systemic symptoms like fatigue, fever and sweats. Diagnostic tests are inconclusive; however, they help to rule out other differential lesions like infections, tuberculous lymphadenopathy, salivary gland neoplasms and lymphoma. Diagnostic techniques include routine laboratory tests, complete blood count, blood biochemistry, Mantoux test, chest X-ray and fine needle aspiration. Definitive diagnosis is made only on histopathological examination.

Morphological differential diagnoses include lymphoepithelial cyst of the parotid and follicular lymphoma of mucosa associated lymphoid tissue (MALT) type. In lymphoepithelial cyst, in addition to follicular lymphoid hyperplasia, there are cysts lined by squamous epithelium containing keratin. Follicles in lymphoma are uniform in size, densely packed, with very little interfollicular area, and are composed of a homogeneous population of small, cleaved cells.

The treatment of choice for this lesion is excision followed by histopathological examination. Thus excision is both diagnostic and therapeutic. Recurrences are rare in the hyaline vascular type; however, follow-up is required in plasma cell type lesion.

References