Background: Carcinoma arising from thyroglossal duct cyst (TGDC) is rare and account for 1 – 2 % of all excised cysts. It is debatable the origin of these tumours whether primarily arising from the TGDC or from metastasis from the thyroid gland. Papillary carcinoma is the most common encountered histological type but the diagnosis usually obtained postoperatively.

Objective: The aim of this study is to highlight this rare condition and to discuss the origin of these tumours. The preoperative diagnostic technique, histological criteria for diagnosis and treatment modalities were highlighted.

Design: Retrospective study.

Setting: Pathology Departments, Military Hospital and Salmaniya Medical Complex, Bahrain.

Method: Patients with TGDC papillary carcinoma who have been diagnosed postoperatively on histological examination after simple Sistrunk’s operation during the study period from January 2002 to December 2008 were followed up for six and two years respectively. Patients’ characteristics, clinical presentations and outcomes were reviewed.

Result: Two patients were encountered during the study period. Both presented with classical features of congenital midline thyroglossal duct cyst. The first patient had a localised tumour and was free of tumour recurrence for 6 years postoperatively. The second patient’s tumor spread to surrounding tissue and lymph nodes, which necessitated aggressive surgery.

Conclusion: Papillary Carcinoma of the TGDC is a rare entity, it is usually a localised disease which has excellent prognosis after simple surgery in most cases We present two cases of carcinoma of TGDC diagnosed postoperatively after simple Sistrunk’s operation; one case had no recurrence and the other had widespread dissemination, which necessitated aggressive surgery. The histological diagnostic criteria of these tumours, their origin from the duct or the main gland were reviewed with treatment modalities.

*Bahrain Med Bull 2008; 30(3):*