Papillary carcinoma in a thyroglossal duct cyst

Dear Sir,

The thyroid gland begins to develop on the 24th day of embryogenesis and is identifiable as a segment of mesodermal cells in the floor of the pharynx, between the first and 2nd pouches. The gland subsequently descends on an "S" shaped path anterior to the trachea and reaches the thyroid cartilage by the 7th week. During this decent, the connection between the thyroid gland and the floor of the pharynx (foramen cecum) may persist to form a thyroglossal duct. Secretions by the cells, lining the remnant duct may form thyrogossal duct cysts. Thyroglossal duct cysts are the most common congenital cervical abnormality in childhood, with a frequency of 70% and approximately 7% in adults. Approximately 1% of the thyroglossal duct cysts is histologically malignant but the prognosis is generally good.

We describe a case of papillary carcinoma in thyroglossal duct cyst, to highlight the clinical and pathological features. The patient was a 25-year-old Saudi male with a history of an anterior midline neck mass for several years, but increased in size in the last 5 years. There was no history of compressive symptoms, hoarseness, or symptoms of hypothyroidism or hyperthyroidism. He had no exposure to irradiation. The physical examination revealed a 3.5 cm diameter, smooth, rounded, mobile non-tender cystic mass at the level of the hyoid bone. The mass moved with deglutition and tongue protrusion. The thyroid gland was normal by palpation and no neck lymph nodes were found. The cervical ultrasound showed soft tissue mass, in the midline upper neck, which measured 22 mm x 17 mm x 13 mm. Hyperechoic rim was seen on the inferior aspect of the mass. The mass was identified separate from the thyroid gland. The thyroid scan revealed a thyroid gland normal in shape, size and function. There was no functioning thyroid tissue over the anterior neck mass. Cervical ultrasound and thyroid scan were compatible with the physical findings of a thyroglossal duct cyst.

The patient underwent surgery with this diagnosis, under general anesthesia, and the mass was resected in toto with 0.5 cm of the midportion of the body of the hyoid bone by the usual Sistrunk procedure. The mass was identified separate from the thyroid gland. There were no local signs of invasion of the tissue surrounding the cyst or the duct at surgery. A total thyroidectomy was not carried out as no gross pathological changes were observed in the thyroid during the operation and no enlarged neck lymph nodes were found.

The histopathological examination of the specimen revealed a 3.5 cm x 2.5 cm x 1 cm oval cystic mass. The cyst was partially filled by a solid brownish granular tissue, having an irregular edge indicating capsular invasion. A rim of brownish tissue bordered off the cystic mass.

Histological sections showed the usual features of a papillary carcinoma of the thyroid gland with the characteristic ground-glass nuclei, longitudinal grooving and scattered psammoma bodies (Figure 1). There was evidence of focal capsular invasion, however, the surgical margins were negative. A striking fibrosis was noted within areas of the carcinoma and adjacent to it. Benign thyroid follicles were demonstrated in the tissue near the carcinoma.

Thyroglossal duct cysts are common developmental abnormalities of the thyroid gland. The malignant form of these cysts, however, is rarely encountered with just over 200 cases reported in the literature.[1] The first case was reported in 1915 by Uchermann, then after 12 years with the first English report by Owen.[2] The incidence of thyroglossal duct cyst carcinoma varies from 0.7%-1% of the thyroglossal duct cyst.[3] In most cases the diagnosis is only established after excision of a clinically benign thyroglossal duct cyst. The etiology of such tumour is unclear. There are different theories regarding the origin of this malignancy. In the 60's some authors thought that this carcinoma represented metastatic deposits of thyroid carcinomas. Now, following demonstration of normal thyroid tissue occurrence in the wall of the thyroglossal duct cysts, it was almost universally accepted that a carcinoma may arise from the thyroglossal duct remnants. Previous exposure to radiation has been described as a predisposing factor.

Carcinoma of a thyroglossal duct cyst has been reported in patients from ages 6-81 years (mean, 39.2 years). The thyroglossal duct cyst is a common clinical problem encountered in children. A rare, but significant complication of this common problem is a carcinoma arising in the cyst. There is a slight female predominance of 1.6:1. The common symptom is the presence of an anterior midline neck mass indistinguishable from those of thyroglossal duct cyst. However, neoplasia must be suspected in cases of thyroglossal duct cyst with recent changes in clinical features, particularly a rapid enlargement of the anterior midline neck mass. Imaging techniques (ultrasound, computed tomography) have been used in the preoperative workup of a suspected thyroglossal duct cyst carcinoma. These techniques are certainly not diagnostic. However, the presence of dense or enhancing mural nodules or calcification or both, on computerized tomography (CT)

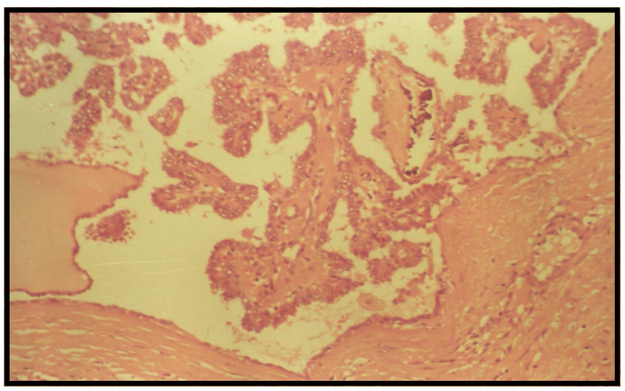

Figure 1 - Photomicrograph featuring papillary carcinoma with the cyst wall (hematoxylin and eosin x 100).

examination of the anterior neck mass, should raise the suspicion of carcinoma of the thyroglossal duct cyst.

Preoperative diagnosis of malignancy by fine needle aspiration biopsy yields a correct result in only 53% of the cases.[4] In positive cases, the aspirated cyst fluid shows cytologic features of papillary carcinoma including psammoma bodies and epithelial cells with papillary clustering, intranuclear cytoplasmic inclusions and positive immunohistochemical reaction to thyroglobulin.

The thyroglossal duct cyst carcinoma causes diagnostic problems for the surgical pathologist and raises therapeutic questions for the surgeon. The histological diagnosis of thyroglossal duct carcinoma requires the presence of malignant cells and also the presence of benign thyroid follicles in the tissue near the carcinoma. A striking fibrosis can occur within the areas of the carcinoma or adjacent to it. Joseph and Komorowski[5] proposed criteria for the unequivocal diagnosis of the thyroglossal duct cyst carcinoma which included, the presence of carcinoma in the duct or cyst, combined with squamous epithelial lining and normal thyroid follicles nests in the duct or cyst wall and the presence of a normal thyroid gland. In addition, the diagnosis must distinguish a cystic metastasis from an occult primary thyroid papillary carcinoma, and primary carcinoma arising from ectopic thyroid tissue located in thyroglossal duct remnants. The demonstration of benign ectopic thyroid follicles in the tissue near the carcinoma is helpful in excluding a metastatic tumour.

The type of neoplasia most frequently described in the literature is that of papillary carcinoma 80%. Other types of carcinomas are also reported in the literature: mixed follicular-papillary carcinoma 8%, squamous cell carcinoma 6%, follicular carcinoma 3%, adenocarcinoma and various unclassified tumours 3%.[1] Medullary carcinoma has not been reported in thyroglossal duct cyst where as anaplastic carcinoma is rare.[3] Unusual cases of Hurthle cell adenoma and carcinoma and mixed papillary and squamous cell carcinoma have also been reported.

The appropriate surgical treatment for thyroglossal duct cyst carcinoma is radical excision of Sistrunk (Sistrunk operation), with a reported cure rate of 95%.[3] However, Kristensen et al[6] mentioned that there should be no extension of the tumour through the cyst wall and no lymph node involvement, in order to increase the safety of the procedure. If abnormalities are detected in the thyroid gland by palpation or ultrasound, a thyroidectomy must be considered. Post operative radiation therapy has been used in some patients with squamous cell carcinoma of a thyroglossal duct cyst.

The prognosis for papillary thyroglossal duct cyst carcinoma is excellent, with occurrence of metastatic lesions in less than 2% of the cases, where as squamous cell carcinoma has a dismal prognosis. In the present case, in accordance with the literature,[6] a follow-up, both clinical and radiological, was recommended with no additional treatment.

In summary, increased suspicion of malignant transformation in a thyroglossal duct cyst should be considered in any patient with a firm anterior midline neck mass who has had rapid enlargement of the mass. Preoperative evaluation should include fine needle aspiration and possible radionucleotide thyroid scanning. Treatment consists of a Sistrunk procedure, with thyroidectomy or neck dissection reserved for patients with known thyroid pathology or nodal metastasis. Patients should have a long-term follow-up, both clinical and radiological, for recurrence or development of metastatic disease.

Vijaya Lakhsmi
Department of Pathology and Laboratory Medicine
Fadel A. Zahra
Department of Otolaryngology
King Fahad Hospital, Al Baha
Kingdom of Saudi Arabia

References

1. Martins AS, Melo GM, Tincani AJ, Lage HT, Matos PS. Papillary carcinoma in a thyroglossal duct cyst. Case report. Sao Paulo Med J 1999; 117: 248-250.
2. Cote DN, Sturgis EM, Peterson T, Miller RH. Thyroglossal duct cyst carcinoma: An unusual case of Hurthle cell carcinoma. Otolaryngol Head Neck Surg (United States) 1995; 113: 153-156.
3. Heshmati HM, Fatourechi V, Heerden JL, Hay ID, Goellner JR. Thyroglossal duct carcinoma: report of 12 cases. Mayo Clin Proc 1997; 72: 315-319.
4. Yang YJ, Haghir S, Wanamaker JR, Powers CN. Diagnosis of papillary carcinoma in a thyroglossal duct cyst by fine needle aspiration biopsy. Arch Pathol Lab Med 2000; 124:139-142.