Thyroglossal duct carcinoma

A case report and review of the literature

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Summary

A case of papillary thyroid carcinoma arising in a thyroglossal duct cyst is reported. The tumour is rare and is usually not suspected pre-operatively, and it is almost invariably only on histological examination that it is diagnosed as malignant. The clinical and pathological features are reviewed and management is discussed.

Case report

In July 1978 a 30-year-old White man incidentally discovered an asymptomatic lump in the midline of the front of his neck. A few weeks later he reported his finding to his general practitioner. There were no symptoms of thyrotoxicosis. The only other feature of interest was that he had had an attack of rheumatic fever in childhood. On examination the pulse rate and blood pressure were normal and there were no signs of thyrotoxicosis. The cardiovascular, respiratory and gastro-intestinal systems were normal. A midline well-circumscribed firm nodule, 2.5 cm in longest diameter, was present in front of the thyroid cartilage. On swallowing it moved with the pretracheal fascia. The main thyroid gland did not appear to be well defined. A diagnosis was made of a thyroid nodule, probably in the pyramidal lobe. Laboratory evaluation of thyroid function failed to detect abnormality. A thyroid scan showed slightly irregular activity in the right lobe, possibly in keeping with a multinodular gland, but no cold nodule was present; the nodule palpated in the pyramidal lobe region did not show up clearly. At operation the nodule was found to be attached to the isthmus by means of a thin fibrous cord. The thyroid gland itself appeared normal and there was no evidence of lymphadenopathy. The nodule, which was cystic and fluid-filled, was excised.

Microscopic examination showed a cystic structure with a thin fibrous wall lined with ciliated columnar, cuboidal and squamous epithelium. In the cyst wall occasional lymphoid aggregates were present. Arborescent papillary structures and trabecular fibrous tissue extended from the wall into the cyst lumen (Fig. 1). Their epithelial covering consisted of a cuboidal or columnar epithelium, and the nuclei of a few of these cells exhibited ground-glass features of varying size. Focal multilayering of the epithelium, atypism and focal squamous metaplasia were noted; no calciospheres were present. Fresh haemorrhage as well as haemosiderin pigment were present in the cyst. Small regional lymph nodes showed no evidence of tumour spread, but contained haemosiderin pigment. No blood vessel invasion was noted. A diagnosis of papillary carcinoma arising within a thyroglossal duct cyst was made.

The patient’s postoperative course was uneventful and he was given thyroxine (Eltroxin). He was found to be free of symptoms on regular follow-up examination.

Discussion

The median thyroid anlage originates as an evagination of the endodermal epithelium in the floor of the pharynx, the foramen caecum of the tongue representing the point of origin. The proliferating epithelium of this tubular structure becomes invested in part by tongue and hyoid bone, which develop later than the tract. Below the hyoid bone, this midline tract passes anterior to the thyroid cartilage and from this most caudal portion the thyroid gland develops. A persistent duct ends in the pyramidal lobe of the thyroid gland. Normally the tract undergoes fibrosis with resultant obliteration of the lumen. Should the embryonic cells of the tract develop and mature, a thyroglossal duct or thyroid follicle may result. The intimate but variable relationship of the duct with the hyoid bone is an important consideration when surgical removal of the persistent duct and cyst is performed. Ducts and cysts may be found between the foramen caecum of the tongue and the normally situated thyroid gland. For positive identification of thyroid follicles should be present near the duct or cyst. When thyroid follicles are not identified, the pathological diagnosis depends on the location and is necessarily only ‘consistent with thyroglossal tract structures’. Malignant tumours of the thyroglossal tract are rare. Several reviews exist in the literature.

The most recent review covers 52 cases of primary carcinoma of the thyroglossal tract. These authors state that thyroid tissue

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may be found in 7% of thyroglossal duct cyst walls and in one-third of thyroglossal tracts tested by scan. Logically, primary carcinoma may originate in these areas. Some cases may present problems, however. Firstly, thyroid gland carcinoma may spread along the thyroglossal duct, which may act as a natural conduit, but in Joseph and Komorowski's series only 2 of 11 thyroid glands examined histologically were found to contain carcinoma. In our patient the thyroid gland was unremarkable on scan and at subsequent operation. Secondly, cystic lymph node metastases from a papillary carcinoma of the thyroid gland may be associated with total effacement of lymph node architecture. However, if the mass has been present for a long time, if normal thyroid tissue is found within the cyst wall, or, as in our case, if the cyst wall was epithelium-lined, it is most likely a thyroglossal cyst. Joseph and Komorowski also state that one should not assume that one is dealing with a cystic metastasis to a lymph node on the grounds that there is a dense lymphatic infiltrate within the cyst wall. Thyroglossal duct cysts showing no evidence of a rapid change often contain a lymphocytic infiltrate, especially if the cyst has been inflamed previously. Many tumours also elicit a lymphocytic response. Thirdly, multicentric carcinoma arising in thyroid tissue may account for a tumour arising in a thyroglossal cyst or duct, as well as in the thyroid gland. An attempt to exclude the latter growth must be made clinically, on scan and at operation, and on histological examination if thyroidectomy is decided upon. Fourthly, carcinoma of thyroglossal duct origin need not necessarily be of squamous epithelial origin, despite the fact that a duct or cyst is lined by squamous epithelium. Bhagavan et al. point out that the stromad of epithelium lining the thyroglossal duct with its embryological potential for thyroidogenesis might possibly produce a carcinoma resembling that of thyroid gland origin. Females are affected more frequently, in a ratio of 2:1, and the incidence is highest in the 30-60-year-old group. This is comparable to the pattern for papillary carcinoma of the thyroid gland proper. The midline lesion is usually asymptomatic. However, the most common symptom, as in patients with benign cysts, is a non-tender, slowly growing palpable mass; the size averages 2-4 cm, as in benign cysts. The lesions rise and fall with deglutition and protrusion of the tongue. More than half of the patients gave a history of more than 1 year's duration. However, the duration of presence of a palpable mass has ranged from 10 days to 40 years. The lesions are firm, non-tender and slightly cystic. Patients have sometimes been aware of a recent rapid growth, and an accelerated growth rate has been observed in pregnancy. Usually the pre-operative diagnosis is that of a benign thyroglossal tract cyst. A Sistrunk procedure is performed, thus removing the lesion and the tract, including the central part of the hyoid bone. The pathologist usually finds a fluid-filled cyst with a warty growth extending from one part of the cyst wall, which proves to be papillary carcinoma.

The most common microscopic pattern is that of papillary adenocarcinoma, which represented 78.8% of Joseph and Komorowski's cases. The lesion was found to be a mixed follicular and papillary adenocarcinoma in 5.8% of their cases, a pure follicular carcinoma in 1.9% and a squamous cell carcinoma in 5.8%; the remainder were solid basal cell epitheliomas, malignant strumas and adenocarcinomas not subclassified. The histological types therefore parallel those of carcinoma of the thyroid gland, papillary carcinoma being the most common and generally being associated with a favourable prognosis. Pre-operative indications that the lesion is malignant may include cervical lymph nodes which appear suspiciously enlarged on clinical examination or a thyroglossal cyst larger than the mean for the series. Aspiration biopsy is likely to be useful in the diagnosis of papillary carcinoma arising in a thyroglossal cyst.

The Sistrunk operation for benign lesions also controls the rare case of papillary carcinoma of the thyroglossal tract. In the larger group of patients in whom the carcinoma is limited to the thyroglossal duct, the cyst should be opened after removal and inspected for a warty mural plaque or a solid mass in the lumen. A frozen section should be obtained irrespective of the presence of such a mass. If no lymph node enlargement is present on clinical examination, no masses are felt in the thyroid gland and the chest radiograph is negative, the open wound should give the surgeon the opportunity to palpate the thyroid and to open the carotid sheath so as to evaluate the jugular chain more accurately. If the latter findings are negative and if the thyroglossal neoplasm is merely a small plaque, the incision can be closed. The patient should be followed up at regular intervals to check for the development of thyroid nodules or enlarged lymph nodes. When there is no evidence of extension of the lesion beyond the confines of the cyst most patients can be managed by local resection with the Sistrunk procedure and suppressive doses of thyroid hormone.

After examination of permanent sections, a determination of spread outside the cyst wall means that a wider excision would be required to remove possible residual papillary carcinoma invading surrounding soft tissues. If there is a mass in the thyroid gland microscopic examination is required and if papillary carcinoma is found total thyroidectomy is recommended. If lymph nodes are palpably enlarged a modified radical neck dissection, preserving the sternocleidomastoid muscle, the jugular vein and the spinal accessory nerve, is recommended. The presence and extent of metastatic disease should be determined at the initial operation. The patient with metastatic carcinoma should be treated by local resection, hormonal manipulation, thyroid ablation and lymphadenectomy as indicated by age and sex, the cell type of the tumour and the extent of local and metastatic disease. Metastasis to the lungs and liver has been recorded, but this is rare. Very few of these patients have metastatic disease.

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REFERENCES