Squamous cell carcinoma in a thyroglossal duct cyst and ^{99m}Tc-MIBI findings

Abstract

The incidence of carcinomas in thyroglossal duct cysts is extremely low. The vast majority are papillary carcinomas, with only 14 cases of squamous cell carcinomas reported in the literature. *A 78 years old male* patient presented with an asymptomatic large mass in his anterior neck, considerably immobile, with inflamed overlying skin. Imaging studies confirmed the cystic nature-doughnut shape of the lesion, giving evidence of malignant infiltration of the surrounding tissues, without lymph node enlargement. Fine needle aspiration was positive for a possible squamous cell malignancy. A wide Sistrunk procedure was performed and the mass was excised along with the strap muscles and the infiltrated overlying skin. Histopathology confirmed the diagnosis of a moderately differentiated squamous cell carcinoma. *In conclusion,* scintigraphy with ^{99m}Tc-MIBI showed the characteristic doughnut sign. The rarity of this diagnosis along with differential diagnosis dilemmas are the key points of this presentation.

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Introduction

Thyroglossal duct cyst is the most common anomaly of thyroid gland development [1]. However, the incidence of a carcinoma in a thyroglossal duct cyst is very rare, being about 1% of all these cysts [2]. The vast majority of them are papillary carcinomas [3]. Squamous cell carcinoma of a thyroglossal duct cyst has been sporadically reported with only 14 cases found in the literature. The origin of these carcinomas may be the epithelial lining of the cyst, or normal thyroid tissue present in the wall of the cysts [2]. The clinical presentation of a thyroglossal duct cyst carcinoma is often very similar to that of a benign cyst, therefore, in some cases diagnosis is made during the operation. However diagnostic procedure of this entity requires except clinical examination and different imaging modalities including radionuclide studies [4]. Specifically thyroid scintigraphy with ^{99m}Tc, ^{131}I or ^{123}I play an essential role in the evaluation of neck swelling especially at the midline and has high sensitivity [4].

Carcinoma should be suspected in any cyst that is hard, fixed, irregular or is associated with neck lymphadenopathy and the infiltrations of overlying skin [2]. Thyroglossal duct cysts are usually observed during childhood and in adults after the fourth decade of life. Females are affected slightly more often 3 to 2 times than men [2-5]. We present the 15th case of squamous cell carcinoma arising from a thyroglossal duct cyst reported in the literature. The tumour was removed surgically and the patient is now receiving post-operative radiotherapy. The rarity of this diagnosis along with differential diagnosis dilemmas are the key points of this presentation.

Case description

A 78 years old male patient presented to the Otorhinolaryngology Department of Papageorgiou Hospital in Thessaloniki with a long history of a midline mass of the neck which had recently grown rapidly. The patient was referred from a District General Hospital, where a computed tomography (CT) scan 3 days ago showed a large midline cystic neck mass without evidence of thyroid pathology. Physical examination revealed a large tender mass, considerably immobile, with inflammation of the overlying skin in most of its bulging area with no drainage. There were no palpable lymph nodes of the neck. Laryngoscopic evaluation showed normal findings, without displacement of the laryngeal structures or tongue. Ultrasound examination showed a round cystic mass, without enlarged cervical

lymph nodes or thyroid nodules and fine needle aspiration revealed keratinized epidermoid cells, suspicious for malignancy. Thyroid function blood tests were within normal range. Physical examination and staging with CT scan of head, neck and thorax did not reveal any evidence of a primary squamous cell carcinoma from another origin.

Based on these findings a magnetic resonance imaging (MRI) examination of the neck was performed, which showed a round mass with thick irregular wall, peripheral enhancement and central necrosis. In addition, no pathological enlarged lymph nodes were demonstrated (Fig. 1A, B). Furthermore, a scintigraphic evaluation using the cationic lipophilic complex Technetium-99m methoxy isobutyl isonitrile (^{99m}Tc-MIBI) as an oncotropic radiotracer was performed. The early images obtained 15min after the i.v. injection of 740MBq of the radiotracer revealed increased activity in the periphery of the cystic lesion with a central photopenic area and normal accumulation in the thyroid gland (Fig. 2A, B). However, the delayed image obtained 3h after injection showed faint retention of the tracer in the periphery of the cystic lesion, as a result of low peripheral metabolic activity and the presence of a central cyst.

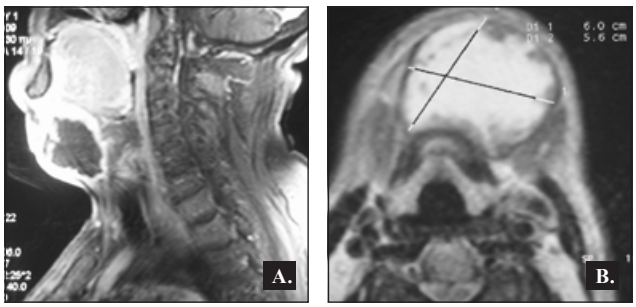

Figure 1. A. Post contrast-enhanced T1-weighted MRI sagittal section of the neck and **B.** T2-weighted MRI axial section of the neck showing an infected thyroglossal cyst with irregular thickened enhancing wall and internal septa.

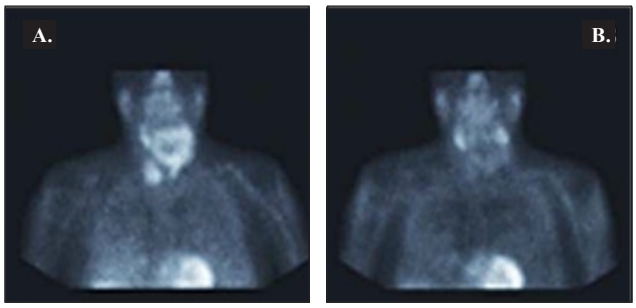

Figure 2. ^{99m}Tc-MIBI scan, 15min (**A**) and 3h (**B**) post the i.v. administration 740MBq. **A.** Above the thyroid gland, a large area of increased radiotracer accumulation is depicted, covering almost all the anterior cervical bed with a photopenic center (doughnut sign). **B.** At the late phase, all the isonitrile has homogenously been washed out from both the thyroid gland and the mass, with no area of abnormal activity left.

A surgical removal of the mass was decided and a wide Sistrunk's procedure was performed. Specifically, Sistrunk's operation includes the removal of the thyroglossal duct cyst through a horizontal neck incision along with the middle portion of the hyoid bone and a small block of muscles around the foramen cecum. Thus, after surgery recurrences originating from tract remnants between the hyoid bone and the base of the tongue are seldom. In our case, a 4x5.5x6cm mass was excised including the anterior third of the hyoid bone and the tissue around the tract up to 1cm in the suprahyoid region. Additionally, the strap muscles and part of the overlying skin were removed as the mass has infiltrated them. Intraoperative frozen sections confirmed the clear resection margins of the excision.

The microscopic examination of the specimen established the diagnosis of inflamed thyroglossal duct cyst, infiltrated by a moderately differentiated squamous cell carcinoma. Specifically, the inner surface of the cyst was thick and infiltrated by numerous inflammatory cells (Fig. 3A), along with foci of squamous cancerous cells (Fig. 3B). There was evidence of invasion of the surrounding strap muscles and the overlying skin. The patient, who had an uneventful recovery, was discharged 7 days after his admission and followed postoperative radiotherapy. At a follow up appointment 6 months later the patient was in good general condition without evidence of recurrence.

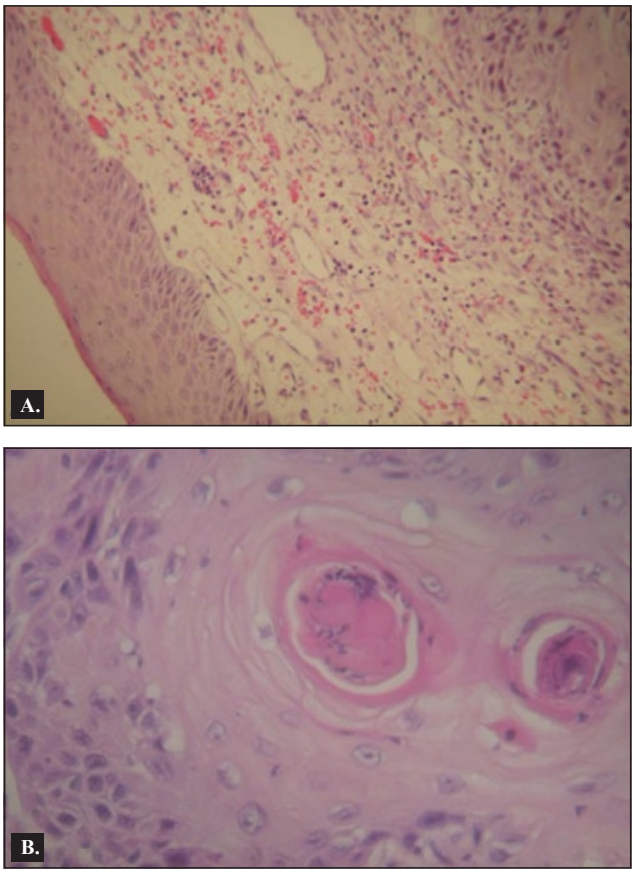

Figure 3. Photomicrographs of the specimen showing **A.** inflammatory infiltration of the cystic wall (H&E x 10) and **B.** deposits of squamous carcinoma cells (H&E x 40).

Discussion

Carcinoma of the thyroglossal duct cyst may arise from the thyroglossal cyst epithelium or from normal ectopic thyroid tissue. More than 90% of the cases originate from this ectopic thyroid tissue [5-6].

Subsequent reports revealed that 80% of the carcinomas in the thyroglossal duct cyst are papillary carcinomas [2-7]. Squamous cell carcinoma in thyroglossal duct cyst is extremely rare, with only 14 cases reported in the literature,

concerning less than 5% of tumours arising in these cysts [7-11]. Clinical criteria for diagnosis are: a cyst at the midline of the neck, exclusion of other carcinomas, presence of normal thyroid tissue, evidence of tumour invasion into surrounding tissues and a transitional zone from normal cystic wall into the squamous cell carcinoma infiltrating area. It should be noted that inflammation could destroy at least partly the epithelium of a thyroglossal cyst, with granulomatous tissue and fibrosis formation causing problems in the diagnostic procedure [12, 13].

In the case presented, the cyst was unnoticed for many years and on the 8th decade of patient's life started growing rapidly and was thus diagnosed. Another interesting point is that, although the tumour tends to invade into the surrounding tissues, does not easily metastasise in the local lymph nodes. From the 15 reported cases (including our case) only two patients had clinically and intraoperatively positive lymph nodes, requiring neck dissection [3, 7].

The diagnosis of a malignant lesion of the thyroglossal duct cyst should best be made preoperatively. However, in one third of the cases it was made after a postoperative histology finding [3]. Elderly patients are less likely to have benign cyst, requiring more detailed preoperative assessment, including fine needle aspiration. Imaging modalities like CT/MRI except their main role in staging of the disease, can detect invasion to the surrounding tissues. Scintigraphy by ^{99m}Tc-MIBI can reveal significant accumulation of tumor-avid radiotracers confined to the tumour, in the cyst and/or in the thyroid gland. Intraoperative frozen sections are recommended for the assessment of cancer free surgical margins.

The number of reported cases is too small for definitive conclusions regarding the optimal treatment of this clinical entity. Patients with limited disease, who had surgery alone, had a better outcome [2, 3, 7, 11], while patients with advanced disease treated with surgery and radiotherapy, had poorer outcome with four of them dying after about one year [2]. This malignancy has a worse prognosis compared with that of a papillary thyroid carcinoma and a high incidence of recurrence, related to incomplete excision or tumour invasion beyond the cystic wall [11].

The role of postoperative radiation treatment has not been well defined in thyroglossal duct cyst carcinoma, partly because of the small number of reported cases. Almost half of the reported patients received postoperative radiotherapy. The indications for radiotherapy include extension of the tumor beyond the cyst wall, residual or recurrent disease, and neck node involvement. Fifty percent of these patients were alive, with no evidence of disease at 1 year follow-up [3, 11].

In cases with extension of the carcinoma beyond the cystic wall, wider excision than Sistrunk's operation is recommended. There are cases reported in the literature with strap muscles excision as in our case [14], of partial or total thyroidectomy [15], of partial glossectomy [16], or even total laryngectomy [17]. There is no report of suppressive treatment in squamous cell carcinoma of the thyroglossal duct cysts. Other authors have stated that these tumours are usually locally invasive and do not easily metastasize to the regional lymph nodes [17]. Neck dissection is recommended only for cases with clinically positive lymph nodes. As these tumours are midline masses with local spread, in a clinically negative neck a prophylactic dissection is generally not advised [2, 9, 10].

In conclusion, ^{99m}Tc-MIBI as an oncotropic radiotracer showed the characteristic doughnut sign of thyroglossal duct carcinoma. The rarity of this diagnosis along with differential diagnosis dilemmas are the key points of this presentation.

All authors declare that they have no conflicts of interest

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