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Thyroglossal Duct Cyst Carcinoma in Child and Adult. Two Case Reports

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Rezumat

Cancerul chistului de canal tireoglos la copil și adult. Două cazuri clinice

Carcinoamele chistului de canal tireoglos sunt rar menționate în literatură însumând 0,7-1,6% din cazurile care prezintă aceste incluzii embrionare restante. Sunt prezentate două observații de carcinom papilar al chistului tireoglos: o fetiță de 14 ani și un bărbat de 44 ani operate în serviciul nostru, ambele diagnosticate postoperator. În primul caz prin operația Sistrunk standard a fost extirpată o formațiune chistică asimptomatică de 3 cm Ø, examenul la parafină descoperind incidental un carcinom tiroidian papilar milimetric în peretele chistului. Al doilea caz prezenta un chist de 4 cm Ø cu caractere clinice și ecografice benigne asociat cu un nodul de 8 mm Ø situat in lobul tiroidian drept. Puncția biopsie cu ac subțire a fost neconcludentă astfel că exereza tip Sistrunk a fost completată de o lobectomie tiroidiană dreaptă. Histologia a relevat prezența unui focar canceros limitat în peretele chistului dar și a unui microcarcinom papilar tiroidian. Evoluție postoperatorie simplă verificată timp de trei ani. Prevalența cazurilor proprii a fost superioară altor serii. Conduita conservatoare adoptată In cele două observații datorită aspectului clinic "inocent", dimensiunilor subcentrimetrice ale leziunilor și absenței unor factori de risc a realizat vindecări stabile, verificate in timp confirmând prognosticul favorabil al acestei rare patologii.

Cuvinte cheie: chist de canal tireoglos, carcinom, operația Sistrunk

Abstract

Thyroglossal duct cyst carcinoma is rarely mentioned in literature representing only 0,7-1,6% of cases with these embryonic remnants. Two patients with thyroid duct cyst carcinoma, a 14-year-old girl and a 44-year-old man operated on in our department, both diagnosed postoperatively are described. In the first one a classical Sistrunk operation was performed removing a 3 cm Ø asymptomatic mass, a milimetric papillary carcinoma being incidentally discovered at paraffin section pathology. The second case presented a 4 cm Ø cyst with benign clinical and ultrasonic features excepting a 8 mm Ø nodule in the right thyroid lobe. Cytology was inconclusive such as a Sistrunk procedure together with a right thyroid lobectomy were done. Pathology revealed a limited carcinomatous focus in the cyst wall but also a papillary thyroid microcarcinoma. Both cases was thereby diagnosed only after microscopic examination. Conservative approach adopted due to "innocent" clinical appearance, subcentimetric size and absence of any risk factor in the two cases achieved stable, verified over time healing confirming the favorable prognosis of this rare pathology.

Key words: thyroglossal duct cyst, carcinoma, Sistrunk operation

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Introduction

Thyroglossal duct remnants are the most common developmental anomalies turning up in the midline cervical lesion, the majority of them presenting in as an enlarging mass in the first decades of life. On the contrary thyroid duct cyst carcinomas (TDC Ca) are significantly less common, only about 274 cases being mentioned in literature after the first descriptions of Brentano (1911) and Ucherman (1915).(1,2)

Two theories explain the origin of TDC Ca, i.e. the de novo theory sustaining the tumor development from the ectopic thyroid tissue of the wall duct or cyst and the metastatic theory in which the carcinoma originate from a more or less occult primary thyroid lesion. Both these conceptions recommended also – together with size, spread and patient risk factors – the surgical attitude with only genuine Sistrunk operation in cases originating from the own cyst and respectively cystectomy extended to thyroid exeresis followed by I 131 ablation and TSH supression in the so called metastatic lesions. Despite the large spectrum of TDC Ca, the general prognosis of these lesions is favorable. (3,4,5,6,)

Case reports

First patient a 14-year-old girl was addressed to our clinic with a gradually enlarged painless mass in her neck which she had noted one year earlier. Local consultation revealed a 3 cm Ø nontender mass in the midline below the hyoid bone moving with swallowing. (Fig. 1) The thyroid gland was normal on palpation and no cervical lymph nodes were perceived. She did not complain offshore throat, hoarseness or dysphagia and had no history of neck irradiation. Indirect laringoscopy examination was unremarkable.

Ultrasound (US) showed a subcutaneous cystic lesion with irregular content. (Fig. 2) Appearance, shape, and size of thyroid gland were normal. Laboratory investigations are in regular limits. Fine needle aspiration biopsy (FNAB) was carried revealing only features of simple TDC.

A standard Sistrunk procedure was performed completely excising the lesion. Postoperative gross examination of the piece showed a patchy cystic structure which was adherent to the central zone of the hyoid bone and strap muscles. Paraffine section showed the cyst wall tapered by cubic or cilindric carcinomatous cells with marginal papillae with fibrovascular cores, lined by typical ground grass nuclei appearance. (Fig. 3) Postoperative evolution and long term follow up were uneventful without any complications or reccurrences certifying the cure of this case.

Second case: A 44-year-old male presented with an asymptomatic midline neck swelling which had been progressively increased in size for two years. On physical examination a round-shaped mass located in the upper part of the anterior neck was found.The mass moved with swallowing as well as at tongue protrusion were found. Palpation of the thyroid gland was unremarkable and no lymph node enlargement was identified in the neck. US of the neck revealed an "innocent" cystic mass under the hyoid measuring 4 cm in diameter, associated also with a thyroid hypoechoic nodule of 8 mm Ø situated in the center of the right unmodified thyroid lobe. Thyroid function tests and other baseline blood investigations was within normal range. Repeated US guided FNAB of the cyst was inconclusive A clinical diagnostic of TDC was made.

The patient underwent a Sistrunk operation (Fig. 4) but the frozen section of the thyroid nodule revealed a doubtful aspects therefore a total right lobectomy was peformed. Final pathologic analysis identified a papillary minute carcinoma confined in the wall cyst showing ductal structures with papillary proliferations and psammoma bodies but also a papillary microcarcinoma in the thyroid specimen. (Fig. 5)

The limited character of both lesions did no required any surgical correction, radioactive iodine therapy or endocrine suppression, the patient being in regular follow-up without any relapse after 3 years.

Discussions

Thyroglossal duct cysts represents the most common congenital anomaly of the neck, present in 2-4% of all masses of this region, comprising more than 75% of midline growths in children and founded also in 7% of such cases in adults. They derive from the persistent epithelial tract, the

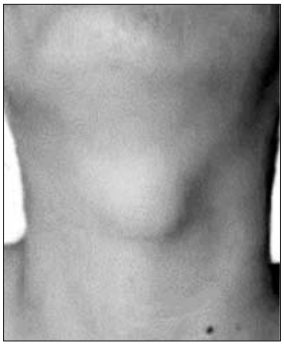

Figure 1. *Case 1. Cervical thyroglossal lump with banal clinical appearance*

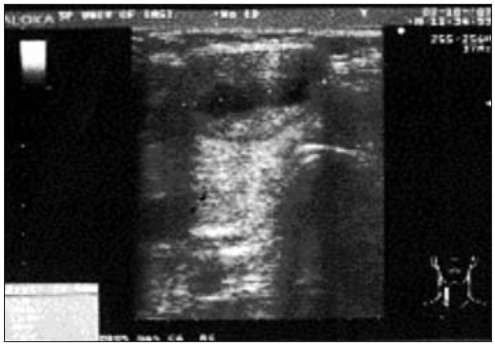

Figure 2. *Case 1. US aspect*

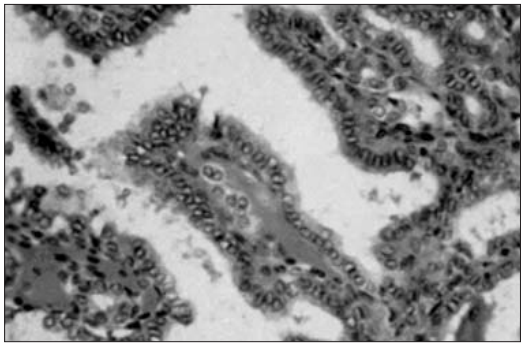

Figure 3. *Case 1. Microscopic showing papillary fonds with ground glass appearance of nucleus HE x 400*

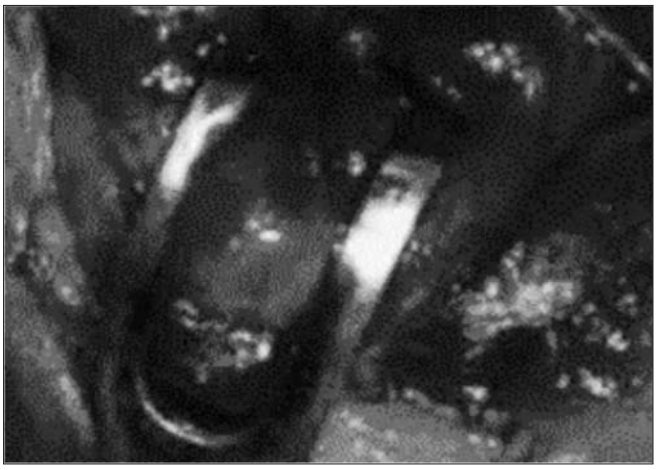

Figure 4. *Case 2. Intraoperative vision of the tumour*

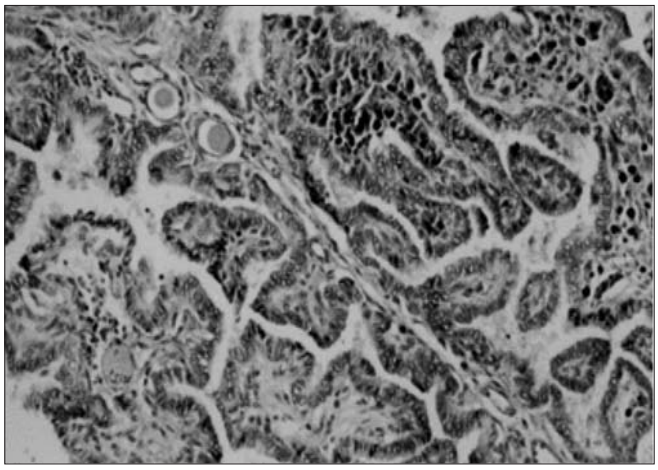

Figure 5. *Case 2. Microscopic papillary thyoid carcinoma HE X 400*

duct forming with the descent of the thyroid from the foramen caecum to its final position in the front of the neck. Normally this duct obliterates early in the fetal life. On the contrary TDC Ca are rare lesions only 1% arising in TDC. (2,7,8)

Patients with TDC carcinoma have an average age of 40 years (range 6-81). The females are affected more often than males in a 3:2 ratio.

The reported TDC Ca are distributed as papillary thyroid carcinoma in 81,7% of the cases, mixed papillary-folliculary in 6,9%, squamous-cell carcinoma in 5,2%, follicular and adenocarcinoma in 1,7% each and epidermoid and anaplastic carcinoma in 0,9%. (9,10,11,12)

The clinical appearance of ordinary TDC Ca may be indistinguishable from that of benign TDC presenting as small round, (2-5 cm Ø), soft, painless and mobile mass moving with deglutition and tongue protrusion. Features that should arouse the suspicion of malignancy include large or increasing, size, firm or irregular shape, pseudoinflammatory signs and fixity, to which may be added previous exposure to ionizing radiation. However 4% of these neoplasia were found to be locally invasive, while 11% were found to include metastasis to cervical lymph nodes. (3,4,11,12)

Although the great majority of TDC Ca have an banal clinical presentation as complete as possible diagnostic algorithm must be watched avoiding the surprise of a malignant diagnosis. Diagnostic tandem composed of US and FNAB should routinely be required but is not alway revealing for diagnosis.

Cervical US is the initial investigation of choice objectifying in significant cases the presence, size, extent, cystic or solid structure, coexisting thyroid lesions or lymph nodes of TDC Ca but often the findings of cancer are unevident. Sometimes millimetric focus of TDC Ca can be identified on US while the presence of specific markers for malignancy as solid components, wall thickening and especially scattered microcalcifications are present only in advanced cases. (13,14)

As for preoperative FNAB unfortunately few cases of of TDC Ca were therefore investigated. Cytologic examinations can show papillary groups of atypical epithelial cells exhibing enlarged hyperchromatic nuclei with increased nuclear/cytoplasmic ratio and conspicuous nucleoli. Nevertheless the diagnostic pitfalls are common FNAB yielding correct results in only 55-66% of cases of TGD Ca. However Miccoli that underwent repeated FNAB under US guidance obtained a100% sensitivity and specificity. Frozen-section intraoperative histopathology may be useful in assessing malignancy mainly in thyroid lesions. (15,16,17)

In confirmed cases CT features of malignant TDC can show a cystic component containing a fluid level and a high intensity of the cyst wall together with a small, eccentric, irregular shaped, dense or enhanced mural nodule or/and irregular small foci of calcification within either in the primary carcinoma or in lymph metastatic node. Eventual spread of adjacent structure is added. MR showed more or less regular solide (micro)nodule within the cystic wall with higher signal intensity than that of surrounding tissue in both the T1 and T2-weighed images. (18,19)

Concerning the treatment of TDCCa some practical realities must be accepted. Therefore in a number of cases only standard Sistrunk procedure (removal of complete cyst together with a portion of hyoid bone and persitent duct up to the foramen caecum) alone is performed establishing the diagnosis of malignancy and however providing definitive healing of the condition in notable proportions. Actually the knowledge of the embryogenesis of the TDC and the pathology of carcinoma arising in these structures is mandatory for an appropriate management of TDC Ca but the rarity of confirmed cases impede a consensus regarding the optimal management for these lesions.

So the most practiced initial approach for patients with suspected or confirmed so called "de novo" TDC Ca having under 45 years of age, presenting tumors less than 1,5 cm Ø confined to the lesion and a normal US and intraoperative thyroid gland, is a standard Sistrunk procedure Conversely in patients from high risk groups i.e. older than 45 years with

larger tumors, fixed or spreaded to adjacent soft structures but mainly pre or intraoperative presence of thyroid foci or nodules and local/regional lymph nodes a subtotal or total associated thyroidectomy (with compartment radical or modified neck dissection if applicable), radioactive iodine ablation and thyroxine suppressive therapy is the most frequently recommended treatment. Many authors suggest that papillary cancer arising from a TDC be managed in the same way as if it started within the thyroid gland. (11,20,21,22,23,24,25,26)

Generally, the prognosis of TDC Ca is extremely good with rare mentions of recurrences or metastases.

Conclusions

Thyroglossal duct cyst carcinoma is a rare clinicopathological finding encountered in practice. In most of cases the malignant growth is not suspected preoperatively but during surgery or after paraffine section. Because their rarity and difficulties encountered in the diagnosis there are many controversies regarding the optimal surgical strategy of this condition. Surgeons should be aware of possibility of malignancy in these lesions and adapt the optimal operative strategy for cystic neoformation but also for thyroid and lymph node lesions.

Conflict of interest

The authors declares that there is no conflict of interest regarding the publication of this article.

Contribution of authors

MRD conceived the study and performed surgery, IC, MG and RT members of operative team, contributing to the conception and design of the study, SD diagnosed and monitorised the pediatric patient.

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