Benign Calcified Thyroglossal Duct Cyst, - Defying The Hallmark of Papillary Carcinoma, 4th Case Reported In Literature

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ABSTRACT

Thyroglossal duct cyst is the most common upper neck midline lesion in children. Approximately 1% of thyroglossal duct cyst undergoes malignant transformation. Calcification which is a marker of malignancy almost always points out to papillary thyroid carcinoma. Benign case of calcified thyroglossal cyst maybe undiagnosed or under reported. We report a case of a 5 year old girl with a benign calcified thyroglossal duct cyst, a fourth case in world literature. Here the differences between a benign and malignant thyroglossal duct cyst are discussed.

KEYWORDS: Thyroglossal duct cyst, benign, calcified

INTRODUCTION

Thyroglossal duct cyst (TGDC) is a congenital anomaly which rise from the embryogenic remnants of thyroid tissue along an unobliterated thyroglossal duct.[1] Monzen et al, 1991, was the first to report on calcification in a benign thyroglossal duct cyst wall, followed by Kervancioğlu et al in 2000 and Ayala et al in 2003 .[1-3] TGDC makes up 70% and 7% of congenital cystic lesion in children and adults respectively.[4] About 1% of TGDC undergoes malignant transformation. 80% of which is papillary thyroid carcinoma followed by mixed papillary and follicular carcinoma for 8% and Squamous cell carcinoma for 6%.[4] Calcification in a benign lesion is extremely rare. The presence of calcification on imaging is the hallmark of papillary carcinoma thus if detected, a high suspicion of malignancy should be made.

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CASE REPORT

A 5 year old girl presented with an anterior neck swelling for 3 months. The swelling was 1x1 cm slightly to the right of midline at the hyoid region, hard and moved with tongue protrusion. Ultrasound revealed a benign appearing calcified lesion. Fine needle aspiration and cytology showed calcification with fibrin. Due to her parents' request, she was managed conservatively for 3 years until the swelling increased to 4x4 cm in size. Neck CT scan showed a well-defined heavily calcified mass at the right anterior neck just off midline measuring 1.4 x 2.1 x 5.5 cm and in contact with the hyoid bone (Figure 1).

The patient underwent Sistrunk's procedure whereby a 3x2 cm calcified mass at anterior neck arising from hyoid bone was removed (Figure 2). There was no intra or post-operative complication and she was discharge well after a few uneventful follow up visits. On histopathological examination, the mass showed a nodule consisting of numerous islands of calcified bodies separated by fibrous tissue. There was no discernible lining seen around the calcified spaces as well as at the edge of the nodule. No malignant cell was seen. The mass was interpreted as calcified benign cyst.

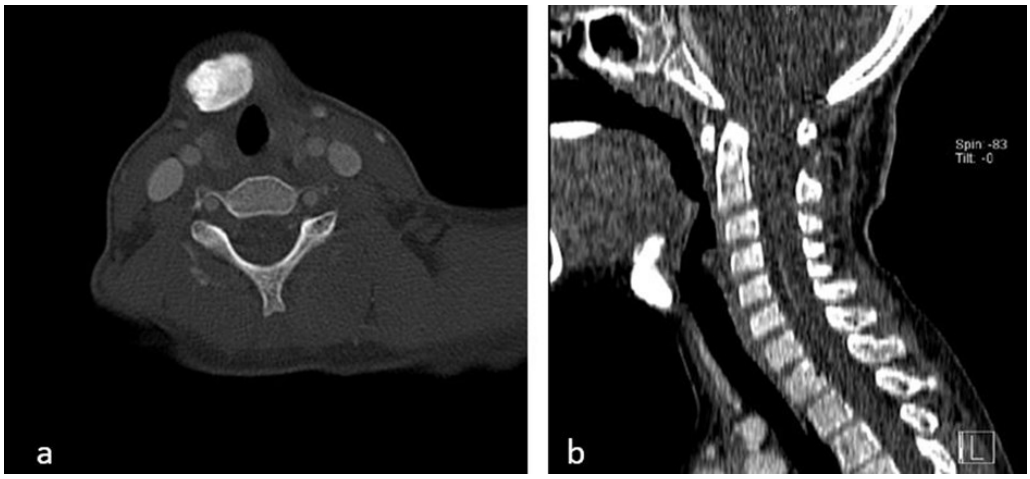

Figure 1 CT Neck: Calcified mass, right of midline at hyoid level (a: axial, b: sagittal section)

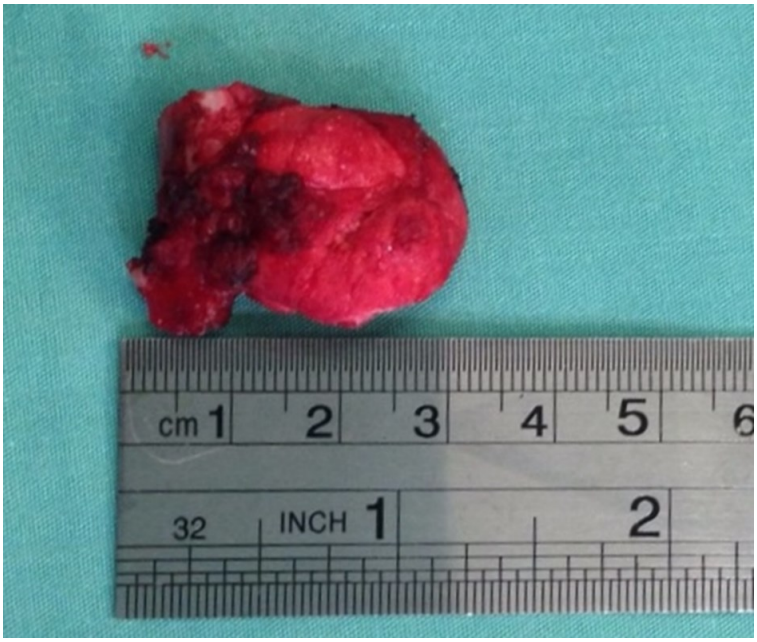

Figure 2 Calcified thyroglossal duct cyst

DISCUSSION

A TGDC classically presents as a midline anterior cervical mass which moves with swallowing and tongue protrusion.[5] Dysphagia, dysphonia, palpable cervical lymph node and a rapidly increasing in size firm mass may indicate carcinoma.[6] However, a rapidly increasing in size firm may also represent a recently infected benign TGDC.[6] The presence of hard mass with fixation to surrounding tissues and old age with a coincident of firm thyroid are more suggestive to underlying carcinoma.[7] There are no specific difference in between presentation of a benign calcified TGDC and a non-calcified TGDC. Here, the differences between a benign and malignant thyroglossal duct cyst are discussed.

Smiti et al suggested for a fine needle aspiration (FNAC) to be done in all TGDC lesion as carcinomas are usually occult until proven by histopathological examination.[8] FNAC is unfortunately limited by the dilution of cells in cystic fluid which may not be representative of the lesion and yields correct results in only 56-62% of cases.[7]

Ultrasound remains the imaging of choice especially in children due to its availability and it evades ionizing radiation or the need for sedation. A benign TGDC may appear as anechoic, homogenously hypoechoic, homogenously hyperechoic (pseudo-solid), or heterogeneous on ultrasound.[8] On CT scan, a benign TGDC appears in a form of midline fluid attenuated mass with smooth wall adjacent to the level of hyoid bone. Infection may increase fluid content and density.[6]

CT scan of TGD carcinoma shows a dense enhancing mural nodule with calcifications within the cyst. 66%

of TGD carcinoma have radiological evidence of calcification and it is a hallmark for papillary cancer.[1] There are only 3 cases of calcifications found in benign TGDC that has been reported so far. In 1991, Monzen et al reported calcification seen in a benign thyroglossal cyst wall.[2] In 2000, Kervancioğlu et al reported calcification seen in both wall and content of a benign thyroglossal duct cyst.[3]

In 2003, Ayala et al presented a mass arising from the hyoid bone containing lobulated calcifications. Histopathological examination revealed psammomatous calcifications. Psammoma bodies are round concentrically lamellated microscopic calcifications and a histological hallmark of papillary carcinoma. It is suggested in the case of carcinoma, the nidus of concentric calcification is caused by the necrosis of the tip of the papilla.[1] Ayala et al reported that no evidence of malignancy was noted in the histopathological examination of their patient's lesion. It is speculated that the calcifications which occurred in the case of benign TGDC is caused by chronic inflammation.

Two theories had been speculated behind TGD carcinoma. The first is that the carcinoma developed de novo from ectopic thyroid which nests within the cyst wall rather than metastases. The second is a multifocal tumour in which a TGD carcinoma and thyroid carcinoma co-exists at the same time.[9] Transformation of benign TGDC into malignancy is unknown.

Sistrunk's procedure is gold standard for treatment of benign TGDC. It involves the removal of the cyst, a section of the hyoid bone and resection of the duct up to foramen caecum.[9] Treatment for TGD carcinoma is Sistrunk's procedure for tumour less than 1cm with no extension of tumour through cystic wall, no lymph node involvement and no abnormality of the thyroid gland. For tumour more than 1 cm or with cystic wall invasion, total or subtotal thyroidectomy is recommended.[10] Thyroid gland is removed if there is any evidence of abnormality since incidence of primary thyroid carcinoma with concurrent thyroglossal duct carcinoma is between 11 and 33%.[10]

In all cases of total thyroidectomy, radioactive iodine (ablation dose) and suppressive T4 therapy is recommended. Neck dissection is performed if cervical node metastases is present.[10] Sistrunk's procedure for the management of TGD carcinoma has the cure rate of 95%.[10] Since benign calcified thyroglossal cyst is hardly documented, we suggest that it be treated with Sistrunk's procedure alone and a longer follow up should ensue to see any possibility of recurrence or malignant transformation.

CONCLUSION

Thyroglossal duct cyst is usually diagnosed clinically. Thyroglossal duct carcinoma maybe highly suspected in the presence of calcification on imaging. Histopathological examination is imperative to distinguish between benign and malignant lesion as to lay out the management plan for either type. Although rare, benign calcified thyroglossal cyst may be kept in mind as a differential for calcified midline lesion of the neck. For a calcified benign thyroglossal cyst we suggest that Sistrunk's procedure should suffice. Total thyroidectomy with radioiodine therapy and suppression T4 therapy with or without neck dissection is recommended in cases of thyroglossal duct carcinoma depending on tumor size, presence of thyroid abnormality, cyst wall invasion and presence of cervical lymph node.

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