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Case Report

CASE REPORT

Thyroglossal duct cyst carcinoma in child†

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Abstract

Papillary thyroid carcinoma occurring in a thyroglossal cyst is a rare condition especially in children, and there is no consensus regarding management. There are only 10 other documented cases in the English literature for children under the age of 12. We discuss one such case. A 10-year-old female child with an 8-month history of a midline neck cyst underwent Sistrunk's procedure as surgical treatment after clinical and ultrasound scan confirmation. An incidental 9-mm papillary carcinoma was seen on histology within the thyroglossal cyst. Following multidisciplinary team (MDT) discussion, the child underwent total thyroidectomy and radioiodine ablation. There is no consensus regarding the ideal management for thyroglossal duct carcinoma in the paediatric literature. We discuss the treatment options and the importance of MDT involvement.

INTRODUCTION

Thyroglossal duct cysts are the second most common neck masses and are the most common congenital cervical abnormality in children. There is an equal male-to-female ratio and thyroglossal remnants are expected to exist in ~7% of the total adult population [1].

Thyroglossal duct cyst carcinoma (TGDCa) is a rare condition which is even rarer in children with only 10 other documented cases of under 12 year olds in the English literature [2]. The incidence rate of papillary carcinoma is ~1% in surgically removed thyroglossal duct cysts [3].

Malignant tumours developing in the thyroglossal duct have two origins: thyrogenic carcinoma arising from thyroembryonic remnants in the duct and squamous cell carcinoma (SCC) arising from metaplastic columnar cells that line the duct and cysts [2]. Approximately 90% of TGDCa are of either papillary or follicular origin, ~5% are SCC and the rest are anaplastic carcinoma, Hürthle cell carcinoma or undifferentiated and unspecified adenocarcinoma [4].

CASE REPORT

A healthy 10-year-old girl presented to the Otolaryngology Department with a 2-month duration of a painless midline neck lump. Clinically, the lump was consistent with a thyroglossal cyst. It was pre-hyoid in position and was not associated with lymphadenopathy. Ultrasonography confirmed a thyroglossal cyst with a mixed echotexture of ~2 cm in size, left of the midline, with no malignant features and a normal thyroid gland. The patient subsequently underwent Sistrunk's procedure.

Histology incidentally revealed a thyroglossal cyst papillary carcinoma. Macroscopically, the pathological specimen showed an irregular cystic mass measuring 2.2 × 2.5 × 1.6 cm. Cross-sectional microscopic analysis of the cyst contained an ill-defined thyroid neoplasia measuring ~9 mm with a predominantly papillary architecture. There was direct infiltration into fibromuscular tissue and focal angiolymphatic invasion with no invasion of the hyoid bone. Immunohistochemistry was positive for CK19, TTF1 and thyroglobulin, which is consistent with a papillary carcinoma.

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Following discussion in both the thyroid and paediatric oncology multidisciplinary teams (MDTs), the decision agreed was for the child to be treated aggressively with a total thyroidectomy and radioiodine ablation. One year following treatment, the patient has been well and under regular follow-up with no evidence of recurrence, no signs of hypocalcaemia, hoarseness and adequate TSH suppression.

DISCUSSION

A thyroglossal cyst is a common cause of head and neck lumps in children. About 16.9% of all congenital anomalies are thyroglossal cysts [5]; however, TGDCa is an extremely rare condition in the paediatric age group [3].

Pfeiffer *et al.* have reported that from the 26 cases of paediatric patients with TGDCa in English literature 21 were papillary carcinoma, 3 mixed papillary-follicular and 1 patient had no pathology information [2]. Lymph node involvement has been found in 50% of paediatric patients with TGDCa, and this is significant when compared with only 11% in the adult population [1, 2, 6]. The duration of neck lump prior to diagnosis is not indicative of malignancy [1]; our patient had the neck lump for 2 months. However, rapid growth, fixed, hard growth, irregular or recent changes may well be indicative of malignancy [1]. Tumour size of <1 cm or less generally has a non-threatening clinical course [1].

There are reports of managing incidental TGDCa by Sistrunk's operation alone for non-metastatic disease with thyroxine suppression. In 31 adult patients who underwent Sistrunk's procedure for TGDCa, there were no disease-related deaths [7]. However, different treatment strategies like thyroidectomy, hormonal manipulation, thyroid ablation and lymphadenectomy are proposed for extensive disease dictated by histology and age [1, 7, 8].

Peretz *et al.* recommend that the management of TGDCa in children should be aggressive, regardless of the extent of the disease and involvement of lymph nodes. All patients should undergo total or near-total thyroidectomy, followed by radioactive ablation and thyroid hormone suppression [1]. This provides therapy for potential tumour remnants and facilitates long-term monitoring of disease recurrence through 131I scintigraphy and measuring thyroglobulin levels [1]. In adults, the incidence rate of TGDCa and coinciding thyroid carcinoma ranges from 25 to 33%. This has not been shown in children; 13 from the total 26 patients who had thyroidectomies showed no evidence of thyroid carcinoma [2]. Consideration should be taken of the potential complications of total thyroidectomy, which include a 3–5% incidence rate of hypocalcaemia, a 1–2% rate of injury to the recurrent nerve as well as the need for life-long thyroid hormone supplements [1]. This is significant when considering there is no difference shown in the prognosis of paediatric patients from either treatment modalities [2] and in adults the 10-year overall Kaplan–Meier survival rate with a well-differentiated TGDCa was 95.6% [7].

There are no evidence-based recommendations for the management of thyroglossal duct carcinomas post Sistrunk's procedure. Our patient had neoplasia completely excised by initial surgery, with the narrowest margin measuring at 0.2 mm. Owing to multifocal nature of papillary carcinoma and for monitoring by thyroglobulin and thyroid stimulating hormone levels, An MDT consensus decision was made for a total thyroidectomy, followed by radioiodine ablation with I131 and thyroxine suppression. Thyroid hormone withdrawal was used instead of recombinant thyroid stimulating hormone, which is not licensed in the paediatric age group, as per radiation oncologist's advice. Thyroxine was started after the completion of radiation treatment. The postoperative histology showed no evidence of malignancy.

In summary, this rare disorder needs a consensus opinion after thorough involvement with all the specialties including paediatric oncologist, thyroid radiation oncologist, head and neck surgeons and other specialist team in the child's care. External opinion may also be required from specialist oncological bodies due to the extreme rarity of such a condition. If the carcinoma is <1 cm in size and there is no neck node involvement, a total thyroidectomy followed by radioiodine treatment with I131 seems a reasonable choice.

CONFLICT OF INTEREST STATEMENT

None declared.

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