Differentiated Thyroid Carcinoma During Childhood and Adolescence: A Report of 4 Cases

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Abstract

Background: Thyroid carcinoma is uncommon in children, but there has been a rising incidence in recent time. This study was undertaken to determine the pattern in Kano.

Patients and Methods: Four cases of differentiated thyroid carcinoma (DTC) in children and adolescents seen between October 1997 and September 2005 were retrospectively studied. The parameters studied included the demographic, and clinicopathologic characteristics, and treatment outcome.

Results: There were 3 females and 1 male (M: F=1:3), with a mean age of 13.2 years. All the patients presented with asymptomatic thyroid nodule, with cytological diagnosis of malignancy in 3. There were no serious complications after total thyroidectomy. Histological examination confirmed the diagnosis of papillary thyroid carcinoma in 3 patients, and one case of follicular thyroid carcinoma. Two out of the 3 papillary thyroid cancers were multicentric. Follow up was poor, the longest period being one year.

Conclusion: DTC in children and adolescence present most commonly as a solitary thyroid nodule. Fine needle aspiration cytology (FNAC) is an important means in preoperative investigations. Total thyroidectomy by experienced thyroid surgeons, has reduced morbidity and mortality.

Key words: Solitary thyroid nodule, FNAC, papillary thyroid carcinoma, total thyroidectomy, childhood

Introduction

Although thyroid cancer is the most frequent endocrine malignancy with variable geographic and ethnic incidence around the world, thyroid carcinoma is uncommon in children. It represents about 1% of pediatric malignancies , and 90% of these are of the papillary type[1]. The incidence of differentiated thyroid carcinoma (DTC) has increased in recent times as a result of radiation exposure[2]. Other etiological factors include endemic goiter, thyroiditis and Gardner's syndrome[1].

The clinical characteristics and biologic behavior of DTC in children and adolescence are different from those in adults[1-3].

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Fine needle aspiration cytology (FNAC) has become an important means of investigation of goiters with high specificity and sensitivity[4]. It has increased early detection of malignancy and timely treatment. This article is a report of our experience with management of thyroid carcinoma in children in Kano, Nigeria.

Patients and Methods

From October 1997 to September 2005, 4 children aged 11 to 17 years were managed for differentiated thyroid carcinoma at Aminu Kano Teaching Hospital Kano, Nigeria. Data which formed the basis of this study were collected from clinical and pathological records (FNAC). Preoperative investigations included Fine needle aspiration cytology, chest x-rays, thyroid function test, indirect laryngoscopy, full blood count, urea and electrolytes. Total thyroidectomy was

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performed in cytologically proven cases of malignancy.

Case Reports

Case 1

An 11-year old girl presented with swelling of the right lobe of the thyroid of 9 month-duration. The swelling was firm, and measured 4 by 3 cm. There was no cervical lymphadenopathy (T2N0M0). Ultrsonography of the neck showed a well defined isoechoic mass in the right lobe. FNAC of the mass revealed clusters of papillary epithelial cells showing Orphan-Annie nuclei. The biochemical, haematological, and thyroid function profiles were within normal ranges. Total thyroidectomy was performed. Diagnosis of multicentric papillary carcinoma was confirmed on histological examination. The patient developed wound infection which was managed by daily dressings. She had suppressive and replacement therapy with L-thyroxine, but was lost to follow up after about four months after surgery.

Case 2

A 12-year old girl presented with a 6-month history of a painless anterior neck swelling, with no features of toxicity. Examination revealed a hard nodule measuring 3.5 by 2 cm. in the isthmus of the thyroid gland. There was no cervical lymphadenopathy (T2N0M0). The thyroid hormones were within normal limits. FNAC of the nodule showed papillary carcinoma. Total thyroidectomy was done, and she has L-thyroxine therapy postoperatively. The postoperative course was uneventful. She was lost to follow up after nine months after surgery.

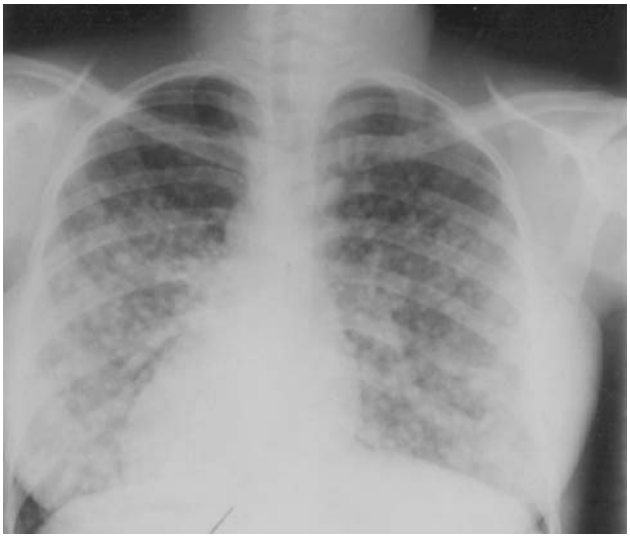


Fig. 1: Pulmonary metastasis of thyroid cancer in a young boy.

Case 3

A 17-year old girl developed a right-sided neck swelling which was excised in a peripheral hospital, but the tissue was not subjected to histological examination. There was no cough, compression features or weight loss. Examination showed a hard mass measuring 4 by 2.5 cm in the right lobe of the thyroid gland, with multiple ipsilateral cervical lymphadenopathy (T2N1M1). FNAC of the mass confirmed papillary carcinoma with lymph node metastases. Chest X-ray demonstrated multiple pulmonary metastases (Fig.1). The vocal cords were free at laryngoscopy, and the thyroid The biochemical, haematological, and thyroid function profiles were within normal ranges. At surgery the nodes along the internal jugular vein were enlarged. Total thyroidectomy was performed. Post-operatively, the patient had suppressive and replacement therapy with L-thyroxine. She is still being followed up in the clinic a year after surgery.

Case 4

A 14-year old boy presented with a nodular enlargement of the left lobe of the thyroid gland, measured 10 by 8.5 cm in its longest diameter. There was no lymphadenopathy (T3N0M0). Ultra-sonography showed multiple isoechoic and hypoechoic nodules. FNAC of the mass revealed features of a benign lesion. Subtotal thyroidectomy was done, but the tissue histology examination showed follicular carcinoma performed. Post-operatively, he had suppressive and replacement therapy with L-thyroxine, but he was lost to follow up 7 months after surgery.

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Discussion

In this report, as in others[1,3], three out of four differentiated thyroid cancers (DTC) were of papillary type, occurring more in females than in males.

Genetic factors and radiation exposure are strong aetiological factors in childhood thyroid cancer[2]. The thyroid gland in children exhibits enhanced sensitivity to radiation-induced genetic change and neoplasia[5]. Rearrangements of the RET/PTC proto-oncogens are found in 3-33% of papillary thyroid carcinoma unassociated with irradiation, and 60-80% of those occurring after irradiation[5]. About 3% are familial and inherited in an autosomal dominant pattern[5]. No known aetiological factors could be confirmed in any of the patients in this study.

DTC in children and adolescents have peculiar clinical chareacteristics. These include high frequency of nodal spread in and pulmonary metastasis at the time of diagnosis[3]. Occasionally, the first sign of the disease could be lymph node metastases or rarely lung or bone metastases. In a study, cervical lymph node involvement was 52%[3]. the third patient in the present report had cervical lymphadenopathy and pulmonary metastases. Multicentric lesions are more common in children, and recurrence rates are higher[3,6,7].

Fine needle aspiration cytology (FNAC) has become a useful diagnostic means in the preoperative evaluation of goiters[4,8]. It is cost effective, accurate and reliable. The diagnostic accuracy is more impressive when compared to ultrasonography, CT-scan, MRI, and radio-isotope scan8. In the current study, FNAC confirmed the diagnosis of thyroid carcinoma in 3 patients preoperatively. This is similar to what is described in literature[4,8]. One limitation of FNAC is its difficulty to differentiate follicular carcinoma from adenoma, as was experienced in one of our patients. In such a case histology is preferable. The diagnosis of a well differentiated follicular carcinoma of the thyroid is based on its vascular or capsular invasion, which cannot be determined on cytology.

The treatment of DTC in children is controversial, regarding the extent of primary surgical excision, the need for extensive node dissection, and the role of iodine-131 therapy. However, total thyroidectomy with modified lymphadenectomy is the preferred surgical approach[1,3,9,10]. Total thyroidectomy is favoured because it reduces the incidence of tumour recurrence and increases disease-free survival, when compared with limited thyroidectomy[3,6,7,11]. Additionally, total thyroidectomy facilitates the efficacy of iodine-131 therapy and monitoring, and it allows the use of serum thyroglobulin (Tg) as a sensitive marker in case there is a recurrence[3,5,9,10].

The only substantial argument against total thyroidectomy is its potentially higher complication rate, especially permanent hypoparathyroidism and recurrent laryngeal nerve damage[3,9]. However with meticulous surgical technique, treatment-related morbidity can be minimized[3,10]. None of the patients in this study, all the patients who had preoperative diagnosis of malignancy had total thyroidectomy. One patient, who had hemi-thyroidectomy for a supposedly benign tumor that proved to be follicular carcinoma, declined a redo (total) thyroidectomy. The reason for this decline was not clear, probably it was on financial grounds or because of fear of complications that could attend to a repeat surgery.

Surgery should be followed by I^{131} therapy, to eradicate any remaining normal and malignant thyroid tissues. This reduces the long term risk of recurrence[1,3,5]. Radio-iodine therapy also increases the sensitivity of subsequent iodine-131 total body scanning, and the specificity of serum Tg for the detection of persistent or recurrent disease[5]. The ablative dose and indication of are controversial. However, its use is recommended in patients at high risk of recurrence and death, which includes large tumour mass with capsular and vascular involvement and distant metastasis, incomplete tuomour excision, microscopic features of

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aggressive disease, and certain histological types[5]. Post operative radioiodine therapy. Post operative radioiodine therapy was not administered to any of our patients for logistic reasons.

The long-term prognosis of DTC in children is excellent, even in those with pulmonary metastases[3,11]. The mortality rates are low[3,6,11]. Vassilopoulou et al[12], noted a 100% survival at 10 years, and in some other series[1], the 10-year survival rate was 93%. Overall, the mortality rate is less than 10%6. Death is related to progressive disease or radiation induced pulmonary fibrosis[7].

After treatment, majority of the patients will require regular follow up over several years, because recurrence or metastasis can occur any time. Unfortunately, follow up is poor in our environment, the longest being one year in this study. Follow up should be directed at detecting thyroid cancer tissue, and this consists of clinical assessment, regular thyroglobulin (Tg) assays, and chest x-rays. Detectable serum levels of Tg after suppressive doses of levothyroxine sodium, is indicative of recurrence or metastastic disease, as 80% of such patients will show positive iodine-131 uptake[7]. Whole-body positron emission tomography with 18F- fluorodeoxyglucose can also be used to detect metastasis, when whole-body iodine-131 scan is negative.

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