Papillary Carcinoma of Thyroid in A 12 Year Old Child: A Case Report

Ananda Kesavan TM¹, Bincy Varghese², K K Noona³

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Abstract

Background: Pediatric thyroid cancer is an uncommon malignancy. Though rare it is treatable and has an excellent prognosis. The disease characteristics are different among children and adults. In children the disease is aggressive at presentation and often associated with lymph node metastasis, extracapsular extension and a higher incidence of multifocality. This report adds to the few studies detailing thyroid cancers in children.

Clinical Description: 12 year old girl presented with swelling in anterior aspect of neck for the past six months. It was a diffuse swelling with an insidious onset. It gradually increased to the present size. She had no family history of thyroid cancer or any history of exposure to radiation. On examination, child had pallor, lymphadenopathy. The thyroid gland was diffusely enlarged and firm with a hard nodule on the left lateral aspect.

Management: She had normal thyroid function tests and highly elevated anti TPO levels. USG thyroid showed evidence of papillary carcinoma in a background of thyroiditis and bilateral cervical lymphadenopathy. FNAC confirmed the diagnosis. CT evaluation revealed mediastinal lymph nodes. Child underwent total thyroidectomy with modified neck dissection.

Conclusion: Though an aggressive malignancy at presentation with meticulous treatment it is curable, has an excellent prognosis and can avoid complications.

Keywords: Papillary carcinoma; Thyroid malignancy; Pediatric; Antibody.

Author Affiliation: ¹Professor, ²Senior Resident, ³Junior Resident, Department of Pediatrics, Government Medical College, Thrissur 680596, Kerala, India.

Corresponding Author: Ananda Kesavan TM, Professor, Department of Pediatrics, Government Medical College, Thrissur 680596, Kerala, India.

E-mail: dranandiap@gmail.com

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INTRODUCTION

Thyroid cancer in children is a rare entity. Among them papillary carcinoma thyroid accounts for 85-95% of the cases. The usual presentation is that of an asymptomatic neck mass and as a result is often detected late. It usually presents at an advanced stage and has higher recurrence rates compared to its adult counterpart. The condition is treatable and has an excellent prognosis which makes prompt identification and treatment a necessity. Though mortality due to the disease is rare, treatment

complications and sequalae can have long lasting morbidity particularly in children. Here we report a case of papillary carcinoma thyroid in a 12 year old child.

CASE REPORT

12 year old girl presented with swelling in the anterior aspect of neck for the past six months. Swelling was insidious in onset and gradually progressed in size. She had an uncomfortable feeling during swallowing for the past three months. No history of hoarseness of voice or any other symptoms. There is no family history of thyroid cancer or any history of exposure to radiation. On examination, the child had pallor, bilateral cervical lymphadenopathy largest measuring 1.5x1.5 cm. Thyroid gland was diffusely enlarged and firm with a hard nodule over the left lateral aspect (Fig. 1). System examination revealed no positive findings.



 $\textbf{Fig. 1:} \ Preoperative \ photograph \ showing \ the \ thyroid \ swelling.$

On investigation she had normal thyroid function tests (TSH-2.85mIU/L, FT4-1.51ng/dL) but thyroid peroxidase antibody levels were elevated (>1006IU/ml). Ultrasound scan of thyroid showed an enlarged thyroid gland with diffuse microcalcifications, a hypoechoic nodule with microcalcifications in the left lobe. Sonologically, a diagnosis of possible papillary carcinoma in the background of thyroiditis with lymphadenopathy was made. Chest x-ray and USG abdomen showed no significant abnormality.

Fine Needle Aspiration Cytology from the thyroid nodule was performed. Cytological studies

showed a cellular smear with follicular cells in papillary pattern seen in clusters and scattered singly. Cells had round pale nuclei with finely granular chromatin. Nuclear grooves, inclusions and psammoma bodies were noted. Cytological diagnosis was Bethesda Category VI malignant papillary carcinoma thyroid.

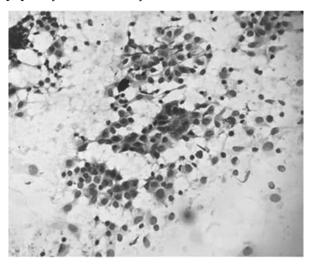


Fig. 2: Smear showing numerous Orphan Annie-eye nuclei.

CT evaluation showed an enlarged left lobe of thyroid with heterogeneously enhancing nodule measuring 1.8x2.4x3.2cm and enlarged mediastinal lymph nodes largest measuring 1.9x1.2cm.

The child underwent total thyroidectomy with central neck dissection and bilateral posterolateral neck dissection. Child is now on thyroid hormone supplementation and on follow up.

DISCUSSION

Papillary thyroid cancer is a differentiated thyroid cancer with a set of distinctive nuclear features. This is the most frequent thyroid neoplasm in adults. The tumor usually appears as an irregular solid mass but can have cystic features too. There can be associated lymph node metastasis. About 10% of patients may present with metastatic disease at initial presentation.2 The prognosis of the disease is good for patients less than 45 years. Pediatric thyroid cancers are rare which accounts for less than 5% of all thyroid cancers of which the major bulk is contributed by papillary carcinoma thyroid. This cancer is aggressive at presentation with frequent lymph nodal metastases and often pulmonary metastases. Though it presents at an advanced stage and has a high recurrence rate compared to its adult counterpart, mortality due to the disease is rare due to its unique biological behaviour.1

The incidence of thyroid cancer in adolescents is 10-fold greater than in younger children. Children below 10 years has an estimated incidence of 1 per million. The incidence is almost same in prepubertal boys and girls.³ After puberty, girls are four to five times more likely to have thyroid cancer as the onset of puberty and related endocrine changes can activate these cancers.³

History of exposure to radiation is an identified risk factor that predisposes to thyroid cancer, eg: childhood cancer survivors who received head and neck irradiation. Francis et al in his study reported that almost 5% of patients with papillary thyroid carcinoma had a family history of the disease.³

The most common presentation is that of a palpable thyroid nodule.³ It can also present as neck nodes with or without a palpable thyroid lesion or occasionally as distant metastasis.⁴ Children have a more extensive disease as compared to adults with regional lymph node involvement and extrathyroidal extension. Devendra et al in his study reported that 56% presented with cervical lymphadenopathy and 19.2% had pulmonary metastasis. In children below 10 years of age the disease is even more aggressive as reported in a study by Kumar and Bal.⁵

The child in our study presented with diffuse thyroid swelling and left sided solitary nodule and bilateral cervical adenopathy. She underwent a detailed clinical examination, thyroid function tests, anti TPO antibody levels, ultrasonography of the neck, fine needle aspiration cytology (FNAC) and CT evaluation. CT neck is essential for surgical planning in children with bulky neck nodes and CT chest when chest x-ray is abnormal.

Total thyroidectomy with neck dissection is the preferred treatment in differentiated thyroid cancer. Lymph node dissection in children with papillary thyroid carcinoma reduces the recurrence risk and provides a progression-free survival.⁶

Demidchik et al found a complication rate of 22%; permanent recurrent nerve damage in 6.2% and permanent hypoparathyroidism in 12.3%.⁷ After thyroidectomy, endocrine specific complication rates are higher in children than in adults.

Thyroxine supplementation for TSH suppression is the next vital step in treatment with a goal of maintaining a serum TSH level of 0.1-0.5µIU/mL in the absence of symptoms of hyperthyroidism. Radioactive iodine therapy is based on the uptake on diagnostic scan and site of the disease. Radioactive Iodine Therapy leads to reduced risk of recurrence

and mortality.² All Indian studies recommend routine post operative radioactive Iodine scanning and therapy due to increased local recurrences and the incidence of pulmonary metastasis not detected on routine imaging.

A recent meta-analysis involving 34,448 patients indicated that positive serum thyroglobulin antibody increases the risk of malignancy twofold in papillary thyroid carcinoma patients compared to the general population. Shan et al in his study observed that thyroid autoantibodies together with an elevated TSH were significantly associated with increased risk of papillary thyroid cancer. Also he observed that preoperative thyroid autoantibodies were associated with better clinicopathological features of papillary thyroid carcinoma.⁸

Lifelong follow up is mandatory as recurrences can occur over a long period of time. Pediatric thyroid cancers though aggressive at presentation has an excellent prognosis. The poor prognostic factors include male gender, non papillary tumour and distant metastasis. Selek et al reported that having autoimmune thyroid disease might be associated with a better prognosis in papillary thyroid carcinoma patients.9 Serum thyroglobulin antibody levels together with serum thyroglobulin levels are valuable markers for early detection of recurrence of papillary thyroid cancer following surgery and radioactive iodine ablation. Children are also followed up with ultrasonogram of neck every 6-12 months during the first 1-2 years of follow-up.4 Hogan et al reported a mean overall survival of 30.5 years. The survival was longer in females.10

CONCLUSION

Thyroid cancer in children is a rare entity. But there is an increased incidence of thyroid cancer in the recent years. Though the disease is aggressive and frequently metastatic, it has an excellent long term survival. The management guidelines are largely adapted from adult guidelines and have evolved from cohort studies. Total thyroidectomy with dissection of involved neck nodes, followed by I131 ablation is recommended. The follow up should be life long with TSH suppression by thyroxin, serial thyroglobulin estimation and radioiodine scanning. Care should be taken to minimize the complications of surgery, which can result in lifelong morbidity. Also, while using radioactive iodine the risk of toxicity and malignancy should be weighed against the potential benefits of the therapy.

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