Rare disease

Uncommon presentation of a common cancer papillary thyroid cancer in paediatric population

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Summary

Carcinoma of thyroid gland is rare in the paediatric age group before the age of 15 years. A 12-year-old girl presented with a swelling in the midline of neck, with no exposure of radiation in the past. On ultrasonography and fine needle aspiration cytology, the findings were suggestive of papillary carcinoma of thyroid. The case is being reported on account of its rarity.

BACKGROUND

Thyroid cancer is a rare pathology in childhood and adolescence being responsible for 1.5–3% of all carcinomas in this age group, with differentiated thyroid carcinoma most commonly found, especially papillary carcinoma. In the case being reported the rarity is due to the age of presentation, no prior radiation exposure and no family history; hence a case of sporadic papillary carcinoma in the paediatric age group.

EPIDEMIOLOGY

The statistics pertaining to the Gujarat Cancer Research Hospital Ahmedabad, reported in the past 18 months, 12 cases of papillary carcinoma with the youngest being of 19 years at the time of detection¹ (the present case was subsequently referred there, hence is included).

CASE PRESENTATION

A 12-year-old female child to consanguineously married muslim parents (first cousins) was brought to this hospital with swelling in the midline of the neck since 3 months. The swelling was painless and almost pea size initially but had rapidly increased in size in the past 2 months with another subsequent swelling development in the left upper cervical region of 1-month duration. The midline swelling was 2–3 cm in size in its greatest axis in

the midline of the neck, mobile, firm in consistency, smooth surfaced and it moved with deglutition. The girl had complaints of increased perspiration, on and off diarrhoea, fatigue and irritability. The girl being referred for ultrasonography (USG) of thyroid from the ear, nose and throat department as the preliminary investigation and subsequent clinical and pathological correlation helped us to diagnose this sporadic case of papillary carcinoma with added features of hyperthyroidism (increased T3 levels).

INVESTIGATIONS

On USG:

- ▶ Both lobes of the thyroid showed an altered echopattern, predominantly hypoechoic.
- ▶ The left lobe was further enlarged with irregular margins and few specks of calcification.
- ▶ Increased chaotic vascularity on Doppler.
- ▶ Bilaterally enlarged cervical lymphnodes more on the left, the largest about 2×1.2 cm in the left upper jugular chain.

The findings were characteristic of papillary thyroid carcinoma (figures 1–6).

Chest x-ray: was normal and showed no evidence of metastasis (figure 7).

Figure 1 Sex versus age disposition graph.

Figure 2 Left lobe of thyroid, heterogenous echopattern.

Figure 3 Right lobe of thyroid, homogenous echopattern.

OUTCOME AND FOLLOW-UP

Subsequently, USG-guided fine needle aspiration cytology (FNAC) from the thyroid nodule showed papillary carcinoma of the thyroid gland. Haematological profile and liver/renal function tests were within normal limits. The child had been posted for total thyroidectomy.

The biopsy specimen showed a papillary pattern having grooved nuclei and microcalcification, thus proving our USG diagnosis correct.

TREATMENT

Surgery is the treatment of choice. Total or subtotal thyroidectomy is recommended. No clinical trials have established whether total thyroidectomy with lymph node dissection is better than subtotal thyroidectomy. ^{131}I whole body scan should be performed approximately 6 weeks after the initial thyroid resection, followed by a therapeutic dose of radionuclide (^{131}I) to abate residual tissue and treat residual metastatic disease. Radioiodine ablation has been shown to decrease the risk of local recurrence.

Total thyroidectomy and bilateral posterolateral neck dissection were undertaken in the present case.

DISCUSSION

Palpable thyroid nodules can be diagnosed in 4–7% of the adult population. The high-resolution ultrasounds are able to detect nodules around 19% of the adult population, reaching up to 67% in populations at higher risk such as women and elderly individuals.

Thyroid cancer is a rare pathology in childhood and adolescence being responsible for 1.5–3% of all carcinomas in this age group in the USA and Europe. Specifically in adults, the differentiated thyroid carcinoma is the most commonly found, especially papillary carcinoma. In this population, age, family history of thyroid disease and radiation exposure are very important factors as already shown in various series. Thyroid carcinoma in children and adolescents tends to behave in a more biologically aggressive manner when compared with the disease in adults. Despite this, the prognosis has been reported to be very good in several series. Papillary carcinoma is the commonest type encountered.

The most common presentation is cervical lymphadenopathy or firm palpable thyroid nodule with or without cervical lymphadenopathy. Six per cent of papillary carcinoma presents with metastasis to lung or upper

Figure 4 Chaotic vascularity on doppler in left lobe of thyroid.

Figure 5 Degenerative changes of lower jugular chain lymphnodes on left.

mediastinal lymphadenopathy. But it almost never occurs in the absence of cervical lymphadenopathy.

Studies of comparative analysis of radiation induced and sporadic thyroid carcinomas in children demonstrate clear differences in some epidemiological characteristics, morphology and types of the ret oncogene alteration between these two groups of paediatric neoplasms. There is a significant difference in sex distribution between both groups, with a dramatic female predominance among sporadic cases.²

Thyroid scan with ^{123}I or $^{99\text{m}}\text{Tc}$ usually shows parenchyma with normal uptake and one or more hypofunctioning nodules.

USG PROVIDES CHARACTERISTIC INFORMATION ABOUT THYROIDS WHICH ARE DISTINCTIVE FOR PAPILLARY CARCINOMA

- ▶ Hypoechogenicity (90% of cases), resulting from closely packed cell content, with minimal colloid substance.
- ▶ Microcalcifications, appearing as tiny, punctuate hyper-echoic foci, either with or without acoustic shadowing.
- ▶ Hypervascularity (90% of cases), with disorganised vascularity, mostly in well encapsulated forms.
- ▶ Cervical lymph node metastasis, which may contain tiny, punctuate echogenic foci caused by microcalcifications.³

Figure 6 Enlarged upper jugular chain lymphnodes on left.

The USG diagnosis can be corroborated by FNAC if unsatisfactory core or excision biopsy is recommended.

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Figure 7 Normal chest x-ray.

therapeutic dose of radionuclide (^{131}I) to abate residual tissue and treat residual metastatic disease. Radioiodine ablation has been shown to decrease the risk of local recurrence.⁵

Learning points

- ▶ Ultrasonography (USG) features for papillary carcinoma are extremely characteristic.
- ▶ Hypoechogenicity, resulting from closely packed cell content, with minimal colloid substance.
- ▶ Microcalcifications, as tiny, punctate hyperechoic foci, either with or without acoustic shadowing.
- ▶ Disorganised hypervascularity in well-encapsulated form.
- ▶ Cervical lymphnodal metastases, occasionally with cystic degeneration or containing tiny, punctate echogenic foci.
- ▶ Presentation of the USG features in adult or paediatric population more or less remains similar.
- ▶ Rare sporadic cases can be diagnosed quite accurately with non-invasive modalities as USG and diagnosis can be confirmed with FNAC.

Competing interests None.

Patient consent Obtained.

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