

Primary Schwannoma of Thyroid Gland in an Adolescent Male: a Rare Case-report with Review of Literature

Pallavi Agrawal^{1*}, Neha Garg¹, Preeti Jain²

¹Department of Pathology,, Mahavir Cancer Institute & Research Centre, Patna (Bihar), India

²Department of Surgery, Mahavir Cancer Institute & Research Centre, Patna (Bihar), India

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ABSTRACT

Primary neural tumors of thyroid are very rare and are mainly represented by schwannomas and malignant peripheral nerve sheath tumors. This report presents a rare case of schwannoma involving the isthmus of thyroid in an 11-year adolescent male. The patient presented with soft neck mass which moved with deglutition. The histological examination indicated Antoni A type schwannoma. The clinical, radiological and pathological findings of the tumor are discussed emphasizing the difficulty in reaching a correct diagnosis and planning out exact surgical treatment of the patient. Only 20 cases of primary schwannoma of thyroid gland have so far been described in literature and this is the first case report describing this entity in an adolescent age group.

***Corresponding author:**

Dr. Pallavi Agrawal, M.D., DNB, PDCC (Neuropathology), Department of Pathology, Mahavir Cancer Sansthan, Phulwarisharif, Patna – 801505 (Bihar), India
Phone: +91 7739165410
E-mail: dr.pallavimamc@gmail.com



Introduction

Schwannomas are the benign tumors arising from the neuronal sheath of cranial or peripheral nerves.^[1,2] Generally they occur anywhere in the body but 25-45% originates in the head and neck region and most commonly involve the intracranial nerves or their branches. The 8th cranial nerve is the most frequent site involved, followed by the 5th and the 7th cranial nerve. Extra-cranial schwannomas are rare and may develop anywhere from the base of the skull to the thoracic inlet. But they are most commonly found in the middle area of the neck. Thyroid is a rare site for schwannomas.^[3] First case of primary schwannoma of thyroid was reported by Delaney and Fry,^[4] though its description as first non-epithelial tumor of thyroid was made by Frantz.^[5] Only 20 cases of primary schwannoma of thyroid gland have been described in literature. We report the first case of thyroid schwannoma localized in the isthmus of thyroid in an adolescent male.

Case-Report

An 11-year-old male presented with an asymptomatic soft swelling in the mid-line neck region for past 3 months. There was no history of dysphagia, odynophagia and hoarseness of voice. Physical examination revealed a 3×3cm soft, mobile, non-tender swelling that moves with deglutition. Clinically cervical lymph nodes were not palpable. The serum levels of T3, T4, TSH, thyroglobulin and calcitonin were within normal limits. Ultrasound (USG) revealed a round heterogenous midline lesion at the thyroid cartilage level with internal vascularity. Possibility of epidermoid cyst was considered. Fine needle aspiration cytology (FNAC) was performed which showed many benign epithelial cells in a background of eosinophilic material. Non-contrast computerized tomography (NCCT) showed enlarged thyroid isthmus secondary to the presence of well- defined heterogenous nodule measuring 2.6×1.7 cm. Thyroid capsule was intact. Both the lobes were normal. Possibility of colloid nodule was given. To confirm the diagnosis a repeat FNAC from the same lesion was performed which revealed sheets of oval to spindle cells in the background of eosinophilic material. Possibility of spindle cell lesion was considered. To reach on to a definitive diagnosis a tru-cut biopsy was performed which revealed sheets of benign oval to spindle cells. Possibility of fibroma was considered. Following this pre-operative diagnosis the multi-disciplinary team decided to perform wide local excision in the form of isthmectomy. Surgery was indicated because of increase in size of nodule and the intra-nodular vascularization. Intra-operatively a well encapsulated mass adherent to isthmus and part of pyramidal lobe was identified. Both the lobes of thyroid were normal. Post-operatively patient recovered uneventfully. He was discharged within 2 days.

Grossly the excised mass measured 3×2.8×2.4 cm with a solid encapsulated yellowish-white mass measuring 3×1.8×1.5cm. Cut-surface was solid and grey-white. No areas of hemorrhage and necrosis were identified. Microscopic examination revealed an encapsulated nodule composed of mixed patterns of Antoni A and Antoni B areas with a predominant pattern A having Verocay bodies. Immuno-histochemical (IHC) staining showed that the cells were strongly and diffusely positive for S-100, while they were negative for CD34, actin and CD99 (Fig-1).

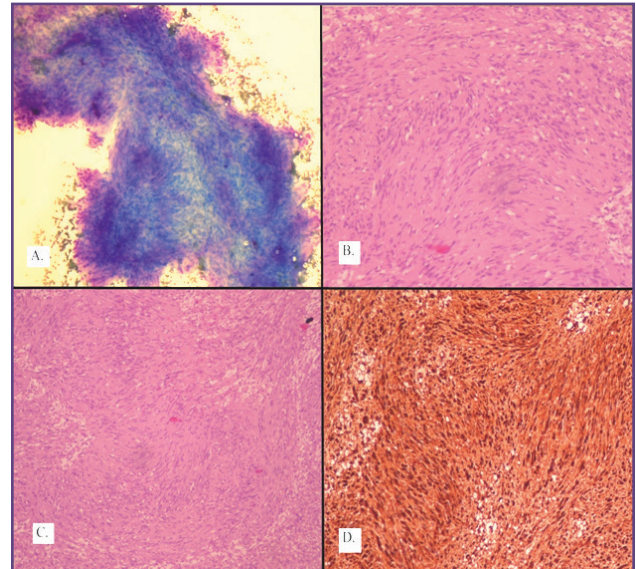


Fig. 1: [A] Cytological smear showing sheets of oval to spindle cells (400X, Giemsa). [B] Tru-cut biopsy showing spindle cell lesion (400X, H&E). [C] Encapsulated lesion showing Antoni A pattern (100X, H&E). [D] Diffuse immune-positivity for S-100 (400x, DAB).

Discussion

Primary non-epithelial neoplasm of thyroid accounts for less than 1% of all thyroid neoplasm.^[6] The lesions included in this category are lymphomas, teratomas, lipomas, hemangiomas and schwannomas. It has been postulated that schwannomas originate from intra-thyroidal sensory nerves or sympathetic and para-sympathetic nerves.^[3] A review of literature revealed only 20 cases of primary schwannoma of thyroid. This is the first case showing involvement of thyroid isthmus by primary schwannoma in an adolescent male. Majority of the cases are reported in the elderly age-group. Details of the cases are shown in Table-1.

It is very difficult to diagnose thyroid schwannomas pre-operatively. Patient generally presents with a mass in the lower neck and rarely with hoarseness, dysphagia or odynophagia. Sometimes due to extension of the mass

	Age/Sex	Clinical Presentation	USG	FNAC	Surgery done	IHC
Delaney and Fry et al,1953 [4]	47/M	Asymptomatic neck swelling			Hemi- thyroidectomy	
Goldstein et al, 1982 [7]	??	Neck swelling with dyspnoea	Thyroid nodule with cystic changes compressing the trachea	Benign thyroid lesion	Total thyroidectomy	
Aoki et al, 1993 [8]	57/M	Asymptomatic Neck swelling	Solid tumor in right lobe	Inconclusive USG guided	Enucleation of thyroid tumor	
Mikosch et al,1997 [9]	31/M	Asymptomatic Neck swelling	Hypoechoic thyroid nodule	FNAC: Suspicious of neural tumour	Hemi- thyroidectomy	
Sujita et al, 1998 [10]	46/F	Asymptomatic Neck swelling	Well defined solid thyroid lesion	Inflammatory cells seen-inconclusive USG guided	Hemi- thyroidectomy	
Jayaram et al, 1999 [11]	41/M	Asymptomatic Neck swelling	Hypoechoic nodule	aspirate; pallsading pattern of spindle cells	Hemi- thyroidectomy	S 100 +ve (done on fine needle aspirate)
Al-Ghamdi et al, 2000 [12]	42/F	Midline neck swelling	Possibility of thyroid malignancy	Inconclusive	Total thyroidectomy	S-100+ve
Gustafson et al, 2001 [13]	20/F	Asymptomatic Neck swelling	Hypoechoic thyroid nodule	Inconclusive	Hemi- thyroidectomy	
Yoshiyuki et al, 2003 [14]	16/F	Neck swelling	Heterogenous mass		Hemi- thyroidectomy	S-100+ve; Vimentin+ve
Baglaj et al, 2004 [15]	12/F	Asymptomatic Neck swelling	Hypoechoic thyroid nodule		Hemi- thyroidectomy	
De Paoli et al, 2005 [16]	63/F	Enlarging neck mass with foreign body sensation in throat	Hypoechoic nodule with rich vascularity	USG guided apirate-inconclusive	Total thyroidectomy done i/v/o suspected malignancy	S 100 +ve
Aron et al (2 cases), 2005 [17]	34/M	Asymptomatic Neck swelling	Hypoechoic nodule with cystic spaces	USG guided aspirates s/o schwannoma	Hemi- thyroidectomy	
Mangro et al, 2008 [18]	52/F	Asymptomatic Neck swelling	3cm solid nodule in right lobe	Inflammatory cells seen-inconclusive	Total thyroidectomy	S-100+ve; Vimentin+ve
Uri , Baron et al, 2009 [19]	57/F	k/c/o Hashimotos with hypothyroid status	Prominent hypoechoic nodule	Suspicious of malignant thyroid neoplasm	Total thyroidectomy	S-100+ve; CAM52+ve; MNF116+ve
Subramaniam et al, 2010 [20]	30/F	Asymptomatic Neck swelling	Thyroid nodule with large cystic degeneration		Hemi- thyroidectomy	
Nidhi Mangal et al, 2010 [21]	25/F	Asymptomatic Neck swelling	Benign thyroid nodule		Hemi- thyroidectomy	
Jungsuk Ann et al,2010 [22]	14/M	Asymptomatic Neck swelling	Hypoechoic nodule with cystic changes	Paucicellular aspirate with few round cells and	Hemi- thyroidectomy	S 100 +ve ; Tgb -ve
Ibrahim et al, 2011 [23]	70/M	Hoarseness of voice	Mass in left lobe of thyroid	Non-diagnostic	Left sided Lobectomy	S-100+ve; Vimentin+ve
Graceffa G et al, 2013 [3]	47/M	Symptomatic cold nodule	Hypoechoic thyroid nodule	Nodular goitre	Total thyroidectomy	S-100+ve; Vimentin+ve
Harsh Dhar et al, 2014 [24]	47/M	Swelling right lobe	Hyperechoic thyroid nodule	Benign thyroid lesion. Bethesda category 2 lesion		S-100+ve; Vimentin+ve

into the retrosternal space patient presents with respiratory distress. The serum levels of thyroid hormones are generally within normal limits in such cases. Radiologically USG can only delineate the extent of the mass and provide the clue whether the lesion is solid or cystic. CT scan does not provide any other specific features. The CT appearance of schwannomas is usually a well circumscribed homogenous mass of soft tissue density. CT findings of inhomogeneity may suggest malignancy in neurogenic tumors. Diagnostic support by thyroid scintigraphy is not always necessary.^[7,10]

About 80% of cold nodules of thyroid are diagnosed as benign epithelial tumor, with the remainder being malignant.^[19] The high risk of malignancy in such cases mandates FNAC of every suspicious nodule. Considering this FNAC was performed in our case which revealed presence of benign epithelial cells. For definitive diagnosis core needle biopsy was performed which revealed presence of benign spindle cells.

The cases which lack typical morphological features of a lesion, many spindle cell lesions enter into the differential diagnosis including mesenchymal lesions like leiomyoma, solitary fibrous tumor and hemangiopericytoma. Leiomyoma shows characteristic whorling pattern and immune-positivity for SMA and vimentin. Solitary fibrous tumor (SFT) represents a single spectrum of mesenchymal tumors, of which hemangiopericytoma is now considered a cellular phenotypic variant. Classically, SFTs are composed of variably pleomorphic spindle cells admixed with collagen and have 'patternless' arrangement or in short fascicles, while tumors within the previously separated hemangiopericytoma spectrum are composed of ovoid, monomorphic cells with thin-walled anastomosing vessels. Histological features of both can be seen in the same tumor and both are CD34 reactive and have dilated staghorn-like vascular network. The typical histopathological features of schwannoma include the presence of cells with slender wavy nuclei, fibrillarystroma, nuclear palisading and presence of verocay bodies. Antoni A areas alternate with hypo-cellular myxoid areas containing blood vessels (Antoni B areas). Mitotic figures are rare. S-100, SMA, CD34 and CD99 positive immuno-staining provide important aid in the diagnosis of neural sheath tumors and may be mandatory for distinguishing schwannomas from other spindle lesions.^[19]

Conclusion

To conclude despite of its rarity, schwannoma like other non-epithelial thyroid tumors should be considered in the pre-operative differential diagnosis of thyroid nodule. Surgical excision is considered to be curative and treatment

of choice. The close routine co-operation is required between the histopathologist, radiologist and surgeon to cut down unnecessary massive surgeries. In addition the surgeon should also ask for frozen section in the absence of pre-operative diagnosis. Thus, the only diagnostic tool for exact typing of the tumor is histopathological examination supported by IHC.

Conflict of Interest

The authors declare that they have no conflict of interest related to the publication of this manuscript.

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