

Primary thyroid Schwannoma Masquerading as a thyroid nodule: A case report with review of literature

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ABSTRACT

INTRODUCTION: The thyroid gland is a very rare site for head and neck schwannomas⁽¹⁾. Till date there have been only 19 reported cases in English literature⁽²⁾. Only 25 % of Schwannomas occur in the head and neck region⁽²⁾, most of them arising in relation to the peripheral nerves and cervical sympathetic chain^{(3),(4)}.

CASE PRESENTATION: We report a similar case, with clinical and sonological features of a benign thyroid nodule. The diagnosis of schwannoma was established on the final histopathology report and a review of the slides and the imaging available was done to confirm the site of origin. A thorough review of earlier reported cases (only 15 completely reported cases in English literature) showed the entity to mimic a benign thyroid nodule in most cases. **CONCLUSION AND RELEVANCE:** We report this case along with a review of earlier reports to summarise the existing knowledge on the entity, emphasising its radiological appearance, the importance of IHC in pre operative diagnosis on FNAC slides as well the adequacy of a hemithyroidectomy being the treatment standard.

Key Words: Antoni A & B patterns, FNAC, Schwannomas, Thyroid nodule, Verocay Bodies

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INTRODUCTION: Schwannomas, first reported by Verocay in 1908, are the less common form of Peripheral nerve sheath tumours of the thyroid gland⁽⁵⁾. The first reported case of thyroid Schwannoma was by Delaney and Fry in 1964⁽⁶⁾. They typically mimic a benign

thyroid nodule and overlapping features on sonography along with a paucity of cells on FNAC make pre-operative diagnosis very challenging^{(1),(2),(7)}

Such a case, mimicking a thyroid nodule, was diagnosed on histopathology at our institution and is

reported here along with a review of 15 completely described earlier cases .

CASE REPORT

A 47 year old male presented with 6 months history of a progressive swelling in the right lobe of thyroid. He was euthyroid and asymptomatic . Sonography was suggestive of a right hypoechoic thyroid nodule along with a *hyperechoic solid lesion of size 6x3x4 cm adjacent to the posterolateral aspect of the right lobe with cystic degeneration.*

CT scan of the neck (with non-ionic contrast) indicated a heterogeneously enhancing lesion with multiple cystic spaces within along the posterolateral aspect of the right lobe . The lesion was seen to push the adjacent lobe anteriorly and to the left . The lesion was also displacing the right common carotid and the internal jugular vein laterally , however a well-defined fat plane was seen between the lesion and the carotid sheath . No such plane was seen between the lesion and the adjacent thyroid lobe .

Fine Needle Aspiration Cytology (FNAC) showed benign thyroid lesion with Bethesda category II. A right hemi-thyroidectomy was done and intraoperative findings showed a firm and enlarged right lobe with area of

cystic degeneration along its posterior surface , the opposite lobe being normal .

Gross appearance showed a 6 x 4 x4 cm smooth encapsulated right thyroid swelling with posterior cystic area which was yellowish brown in colour. Walls of the cyst were bright yellow with polypoid areas. *Histology showed classical Antoni A (hypercellular areas) with Verocay bodies and Antoni B (hypocellular) pattern , characteristic of Schwannoma , with interspersed thyroid cells along the periphery . The lesion was reported to be arising from within the thyroid gland itself and not extrinsic to it (as had been suggested by the radiological studies .)*

DISCUSSION

Peripheral nerve sheath tumours of the thyroid may be benign or malignant⁽⁶⁾ , the benign variety includes neurofibromas and schwannomas ⁽⁸⁾ . Within the neck , the most common site of Schwannoma is the Vagus , followed by the cervical sympathetic chain⁽⁴⁾ . The origin of Primary thyroid schwannomas -the rarest subset of head and neck schwannomas -has been a matter of speculation and *they are thought to arise from the intrathyroid sensory nerves or from the sympathetic*

and parasympathetic innervation to the thyroid^{(4),(5)}.

The rarity of the diagnosis inspired us to review the FNAC slides, which probably failed to show the presence of spindle cells as a result of the aspirate being drawn from the thyroid tissue surrounding the posteriorly placed lesion. A guided FNAC would have had better chances of providing the correct diagnosis.

The images of the lesion had created a diagnostic dilemma as to its origin, i.e., extrinsic or intrinsic to the thyroid. A review of the sonological and CT images with the radiologist was done to discuss its possible origin. Both the modalities seem to indicate up front that the lesion

was extrinsic to the thyroid. *However a well preserved plane between the lesion and the carotid sheath rules out a cervical sympathetic origin. The possibility of schwannoma of the thyroid bed still remained.*

The Pathologists reporting the specimen were more in favour of a diagnosis of Primary Thyroid Schwannoma saying that *the lesion was arising from within the thyroid with a rim of thyroid tissue around the capsule and a major part of the posterior half of the right lobe had been replaced by the lesion.*

Figure 1: CONTRAST ENHANCED CT NECK (coronal section): Coronal section showing good plane of delineation between the lesion and carotid sheath, along with mass effect on the thyroid gland and the right common carotid.

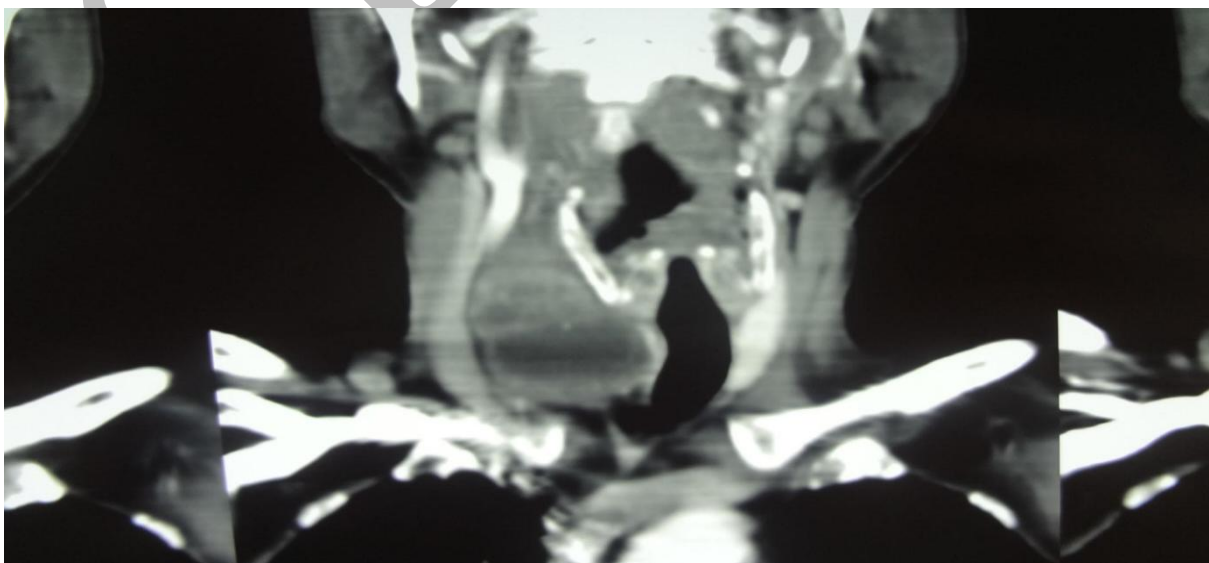


Figure 2 Gross section of hemi-thyroidectomy specimen: A well circumscribed cystic lesion with yellowish colour of the inner cyst wall ;a rim of normal thyroid is seen at the periphery .



Figure 3 Microscopic section - H & E Slide (40x): Well encapsulated lesion with hyper (Antoni A) and hypocellular (Antoni B) . Multiple cystic areas are also seen.

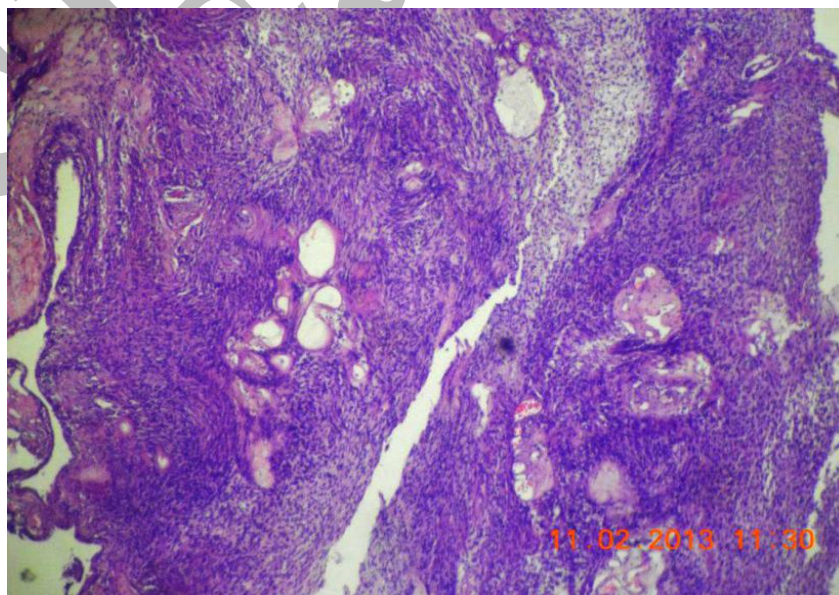


Figure 4: Microscopic section – H & E Slide (40x) Verocay bodies within Antoni A pattern

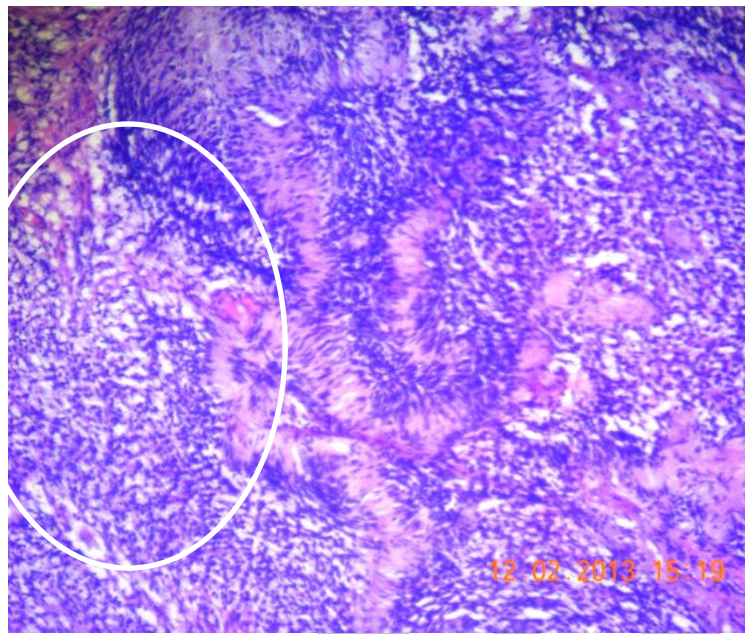


Figure 5: Microscopic section -: H & E Slide (100 xs) magnified view of region in PIC 4 showing Verocay bodies

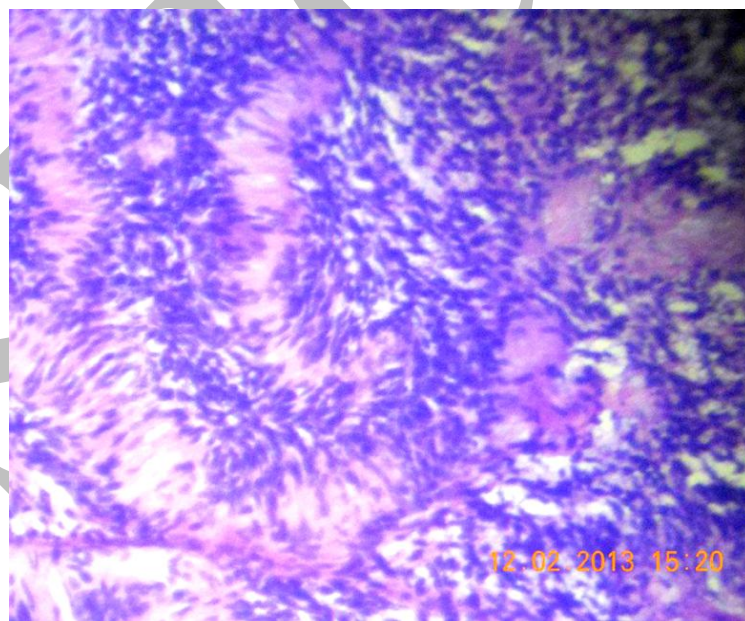


TABLE 1 : SYNOPSIS OF EARLIER PUBLISHED CASES

(Abbreviations: USG – Ultrasonography; FNAC-Fine needle aspiration cytology; k/c/o – known case of ; s/o- suggestive of)

	Presentation	USG	FNAC	CT scan	Surgery done	IHC	Review of FNAC slides
Delaney and Fry et al (first reported case 1953) (8)	Asymptomatic neck swelling	-	-	-	Hemi-thyroidectomy	-	-
De Paoli et al(1)	Enlarging neck mass with foreign body sensation in throat	Hypoechoic nodule with rich vascularity	USG guided aspirate-inconclusive	-	Total thyroidectomy done i/v/o suspected malignancy	S 100 +ve	Aggregate of spindle cells seen on previous slides
Subramaniam et al(2)	Asymptomatic Neck swelling	Thyroid nodule with large cystic degeneration	Colloid goitre	-	Hemi-thyroidectomy	-	-
Jungsuk An et al(6)	Asymptomatic Neck swelling	Hypoechoic nodule with cystic changes	Paucicellular aspirate with few round cells and spindle cells	Well enhancing homogenous mass	Hemi-thyroidectomy	S 100 +ve ; Tgb -ve	-
E Kandil ,Khalek et al(7)	Neck swelling with hoarseness and dysphagia	-	Inconclusive	Enhancing thyroid mass compressing the esophagus	Hemi – thyroidectomy (frozen section done: spindle cell tumour)	S100 & Vimentin +ve	-
Goldstein , Tovi et al(9)	Neck swelling with dyspnoea	Thyroid nodule with cystic changes compressing the trachea	Benign thyroid lesion	-	Total thyroidectomy	-	-
Gustafson et al(10)	Asymptomatic Neck swelling	Hypoechoic thyroid nodule	Inconclusive	-	Hemi-thyroidectomy	-	-
Sujita , Nomura et al(11)	Asymptomatic Neck swelling	Well defined solid thyroid lesion	Inflammatory cells seen-inconclusive	Low density mass on plain CT and moderately enhancing on contrast	Hemi-thyroidectomy	-	-
Jayaram et al(12)	Asymptomatic Neck swelling	Hypoechoic nodule	USG guided aspirate; pallasading pattern of spindle cells s/oschwannoma	-	Hemi-thyroidectomy	S 100 +ve (done on fine needle aspirate)	Both final histology and FNAC slides were indicative of schwannoma
Aron , Kapila et al (2 cases)(13)	Asymptomatic Neck swelling	Hypoechoic nodule with cystic spaces	USG guided aspirates s/o schwannoma	-	Hemi-thyroidectomy	-	Both final histology and FNAC slides were indicative of schwannoma
Mangal ,Agarwal et al(16)	Asymptomatic Neck swelling	Benign thyroid nodule	-	-	Hemi-thyroidectomy	-	-
Chu Gigi et al(15)	Asymptomatic Neck swelling	-	Inconclusive	-	Hemi-thyroidectomy	-	-
Uri , Baron et al(18)	k/c/o Hashimotos with hypothyroid status	Prominent hypoechoic nodule	Suspicious of malignant thyroid neoplasm	-	Total thyroidectomy	-	-
Mikosch, Gallowitsch et al(19)	Asymptomatic Neck swelling	Hypoechoic thyroid nodule	USG guided FNAC: Suspicious of neural tumour	-	Hemothyroidectomy	-	-

REVIEW OF LITERATURE:

A thorough literature search has shown only 15 completely reported cases of Primary Thyroid Schwannoma . (They have been summarised in Table 1)

The most common presentation was that of an asymptomatic neck mass in the 4th or 5th decade without any sex predilection . Most of the lesions appeared as a hypoechoic nodule on USG with variable cystic degeneration. Pre-operative CT scan was done in 4 cases ^{(5),(8),(10),(11)}. The lesion was homogenous and of low density on plain CT, with good post contrast enhancement . *Sujita et al* ⁽¹¹⁾ explained in their article that the well enhancing areas on CT corresponded to the hypercellular Antoni A areas whereas those showing poor enhancement corresponded to Antoni B pattern.

Schwannoma was correctly diagnosed on pre-operative FNAC in only 3 cases ^{(12),(13),(14)}, all 3 being done under sonographic guidance . Most of the remaining cases were reported either as colloid goitre with cystic degeneration or as paucicellular aspirates with scattered spindle cells and labelled inconclusive ^{(1),(2),(13)-(18)} .

On Immunohistochemistry (IHC) schwannomas are positive for S100 and Vimentin and negative for Desmin

and SMA . IHC was used as a confirmatory marker in 4 of the reports cases ^{(1),(6),(18)}.

Intra-operative frozen section was done only in one case ⁽⁵⁾, which showed a spindle cell tumour . This may prove to be a very useful modality to confirm the diagnosis in future cases.

CONCLUSION:

As proved by the paucity of literature available , *Primary Thyroid Schwannoma* is a rare entity which inspired us to report this case along with a literature review . It closely mimics a thyroid nodule in presentation and sonology . This combined with the difficulty of diagnosing it on FNAC, make it a diagnostic challenge. Better dialogue with the pathologist regarding the adequacy of the specimen may increase the sensitivity of FNAC . The use of USG guided FNAC, IHC and intraoperative frozen section are sure to strengthen the chance of diagnosis and prevent over resection of the gland . More reports will improve our understanding and also raise our index of suspicion for *Primary Thyroid Schwannoma* .

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