



Primary Cavernous Hemangioma of the Thyroid- “An Infrequent Case with An Unusual Presentation”

Authors

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Abstract

Primary cavernous hemangioma of the thyroid is an extremely rare case with only 17 cases reported in literature. We are reporting an unusual case of a 75 year old female with a firm cystic swelling on the right lobe of thyroid. Ultrasonography shows well defined large predominantly cystic heterogenous lesion with septations and solid areas in the right lobe. Fine needle aspiration cytology was unsatisfactory due to bloody nature of the smear. A preoperative diagnosis of solitary nodule of thyroid was made. Right hemithyroidectomy was done and a diagnosis of primary cavernous hemangioma of thyroid was made based on the histopathological findings.

Case Report

A 75 year old female, non-smoker and non-alcoholic presented with swelling on the right side of neck for 20 years with difficulty in swallowing for 1 year. The swelling was insidious in onset and gradually progressive. It was not associated with neck pain, breathing difficulty or hoarseness of voice. No complaints suggestive of hyper/hypothyroidism was seen. No history of trauma or surgeries in the neck and prior investigations like FNAC was done for the patient. No history any irradiation to the neck. No history of hypertension, bronchial asthma, COPD or diabetes mellitus. Local examination reveals a globular firm swelling of

approximately 6 x 4cm in the right side of neck. Surface of the lesion was smooth, pinchable and moving with deglutition. Margins were well defined and lower border of thyroid was clearly visible. Trachea was mildly displaced to the left and berry sign was negative. No visible or palpable pulsations were seen over the swelling. Skin over the swelling appears normal with no dilated veins over chest wall. No palpable cervical lymph nodes were noted. Routine investigations done showed-Haemoglobin: 12.4 g/dl, hematocrit-36%, serum urea and creatinine- 34mg/dl and 1.2mg/dl respectively. Thyroid function tests were within normal limits.

Ultrasonography of the thyroid shows a well defined large predominantly cystic heterogeneous lesion with septations and solid areas in right lobe of thyroid, wider than tall shape with mobile internal echoes seen within the cystic area. No calcifications were seen. (Figure 1) On colour Doppler, vascularity was noted within the septations and in the periphery of the lesion.

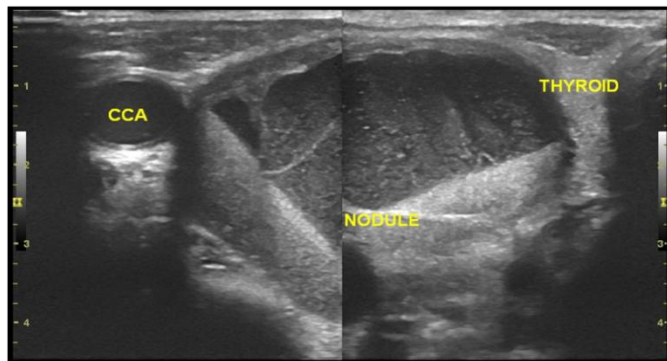


Figure 1: Ultrasonography- well defined large heterogeneous lesion with septations and solid areas in right lobe of thyroid

Fine needle aspiration cytology was done for the patient and nine smears from two passes were prepared, all of them displaying only haemorrhagic material and thus reported the case as an unsatisfactory smear- Bethesda system of reporting thyroid cytopathology (BSRTC)- Category I and a repeat FNAC was advised which was denied by the patient on her subsequent visit. (Figure 2)

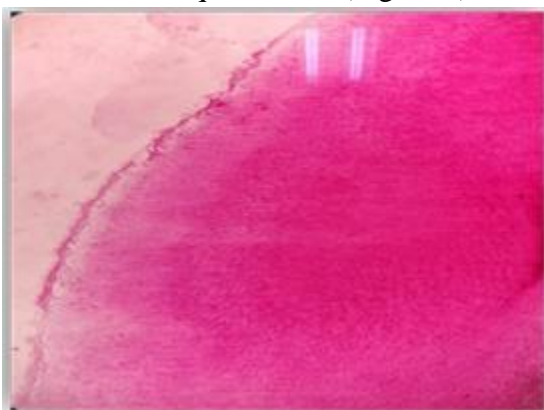


Figure 2: Cytology of thyroid depicting bloody smear

A right hemi-thyroidectomy was performed and specimen was sent for histopathological evaluation. The specimen measuring 8 x5 x3 cm, grossly was completely replaced by a well encapsulated cystic

tissue, divided into multiple lobules by thin fibrous septae and the cystic lumina filled with blood clot. There was focal areas of calcification, myxoid change, haemorrhage and fibrosis noted. No solid/papillary growth or extra-thyroidal extension of the tumor was seen.

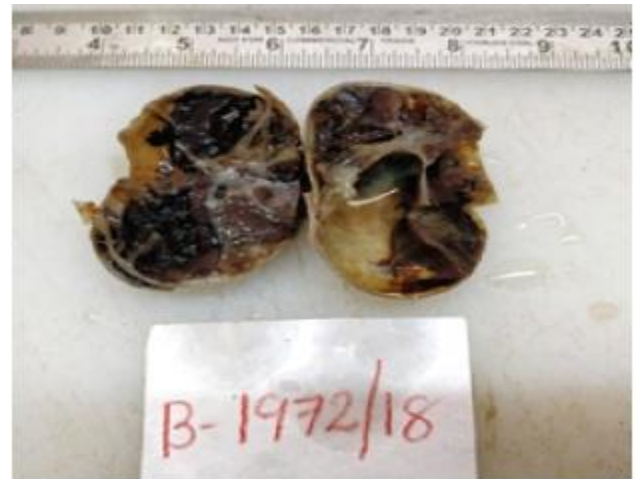


Figure 3: Gross details of the specimen displaying a cystic tissue divided into multiple lobules separated by thin fibrous septae

Microscopic findings was a replica of gross details exhibiting a capsulated lesion composed of lobules of thin walled discrete and branching intercommunicating predominantly large and many small vessels lined by flattened endothelium, filled with blood and fibrin with intervening hypocellular fibromyxoidstroma. There were secondary changes observed like fibrosis, hyalinization, calcification, fibrin exudation, sheets of foamy histiocytes, cystic macrophages, haemorrhage and cholesterol clefts. Remnants of thyroid follicles were observed at the periphery. However no phleboliths, necrosis, mitosis or atypical cells were seen. No extra thyroidal extension noted. Based on the gross and microscopic findings a diagnosis of primary cavernous hemangioma of the thyroid was made. (Figure 4)

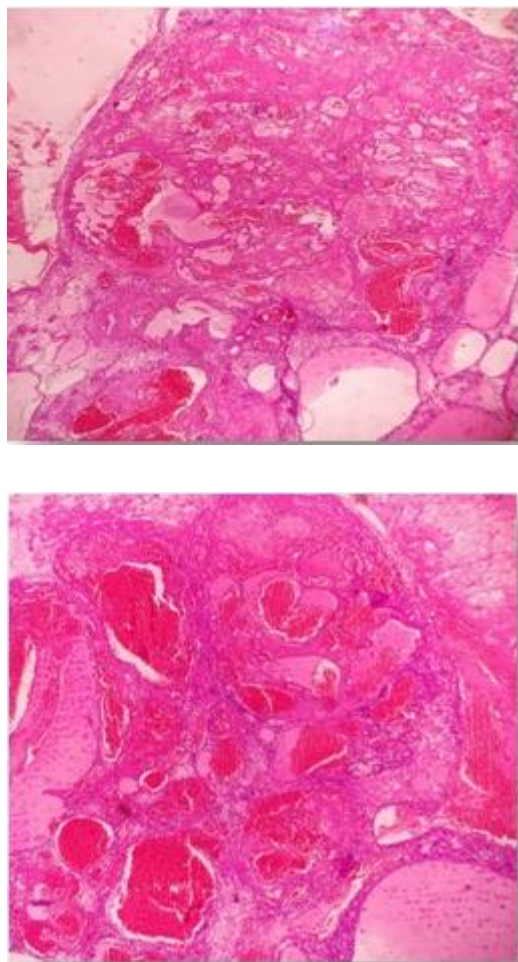


Figure 4: Microscopy showing lobulated vascular lesion

Table 1: Tabular column showing the total number of cases reported in literature

Patient	Age/sex	Size (cm) and laterality	Authors, year published
1	56/M	7.5x3, Left	Pickleman, 1999, USA
2	53/M	6x3.5, Right	Pendse, 2006, India
3	53/M	4x4cm, Right	Kumar, 2006, India
4	48/F	5x4, Left	Rios, 2008, Spain
5	63/F	5x4, Left	Graham, 2008, Spain
6	21/M	5.5x3, Right	Kano, 2009, Japan
7	66/M	17x16, Left	Lee, 2010, Korea
8	64/M	7x6, Right	Ciralik, 2004, Turkey
9	25/M	4.9x4.4, Left	Datta, 2001, India
10	71/F	5.2x4.8, Left	Akhiro, 2002, Japan
11	78/M	4x4, Right	Michalopoulos, 2011, Greece
12	80/F	22x21, Left	Maciel, 2003, Brazil
13	84/F	6x4, Left	Gutzeit, 2012, Switzerland
14	38/M	6x5, Left	Dasgupta, 2014, India
15	48/M	4.5x4, Right	Jie maio, 2017, China
16	78/M	5x5, Right	Emmanuel, 2010, UK
17	72/M	4.2x3.2, Left	Nikolas, 2009, France
18 (Our case)	75/F	8x5, Right	Libin et al, 2018, India

Discussion

Primary cavernous hemangioma of the thyroid is an extremely rare thyroid tumour with only 17 cases reported in literature⁽²⁾. It commonly affects childhood but any age group can be affected. The condition shows a male preponderance with left lobe more commonly involved than the right lobe. Median size of the lesion is 4-5cm, but lesions as large as 20cm have been reported in literature⁽³⁾. The exact etiology of the lesion is not well known but it has been postulated to be an abnormality of angioblastic mesenchyme to form canals^(1,3). Another theory postulated is a previous thyroid injury or past history of FNAC resulting in hematoma organization and vascular formation. More substantial evidences are required to confirm the association of genes of cerebral cavernous hemangioma namely KRIT1 gene: K- rev interaction trapped 1 gene (CCM-1), Malcavernin (CCM-2) and Programmed death cell protein 10 (CCM-3) with thyroid hemangioma.

The condition clinically presents as a firm to hard cystic pulsatile tumor of the thyroid. Intra-lesional haemorrhage could result in rapid growth and compression manifestations. The hard consistency of the lesion is due to presence of phleboliths and coarse calcifications within the tumour⁽⁴⁾. The clinician can wrongly diagnose as a thyroid malignancy considering the firm to hard consistency of the lesion.

Ultrasonography of the thyroid shows a non-specific finding of hypoechoic/isoechoic lesion with coarse calcification⁽³⁾. Fine needle aspiration cytology yields only blood on multiple attempts resulting in inconclusive unsatisfactory reports. Hence the routine preoperative diagnostic tests are not very helpful in preoperative diagnosis of this rare condition⁽⁷⁾. Some radiological diagnostic modalities like magnetic resonance angiography (MRA), single photon emission computed tomography (SPECT), digital subtraction angiography (DSA) and technicium-99 RBCs scan are helpful in preoperative diagnosis of thyroid hemangioma but unavailability and cost, limits its use⁽¹³⁾. Magnetic resonance angiography shows the

classical serpentine/unwinding pattern but this diagnostic modality is not routinely done for any thyroid lesion.

Thyroidectomy is the treatment of choice and conservative management has no role in management^(5,6). Tumour can rarely infiltrate the skeletal muscles so careful resection of the lesion is required to avoid avulsion of the nearby vital structures. A lesion with larger size and extensive haemorrhage during operation should be overlooked as case reports with more than 2L of blood loss has been reported during surgery. Hence preoperative blood and fluid requirements for proper surgical management is necessary^(8,9). The condition has a fairly good prognosis if sufficient preoperative preparation is done.

Conclusion

Primary cavernous hemangioma is a rare benign thyroid tumor. Classical pre-operative suspicion which should alert the clinician includes: 1. Male patient. 2. Hard to firm pulsatile cystic thyroid nodule on clinical examination. 3. Hypoechoic cystic nodule with coarse calcification on ultrasonography of thyroid. 4. Inconclusive unsatisfactory cytology on atleast 2-3 attempts. MRA (if possible) showing a serpentine pattern.

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