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Case Report

Extensive tumour thrombosis from a differentiated thyroid carcinoma masquerading as a right atrial mass

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ABSTRACT

Thyroid carcinomas are well recognized for their propensity to cause tumour thrombosis, most confined to the cervical vessels. However, extensive thrombosis involving the mediastinal veins and right atrium is exceedingly rare. Such advanced tumour thrombosis is associated with a high risk of hematogenous dissemination and pulmonary metastasis, adversely affecting overall survival. We report the case of a 74-year-old man with papillary thyroid carcinoma presenting with tumour thrombus extending into the right atrium in the absence of pulmonary metastases. The patient underwent successful single-stage complete surgical resection without significant perioperative morbidity. He remains on regular follow-up and has been disease-free for 2 years following surgery.

Keywords: Right atrial mass, Papillary thyroid carcinoma, Tumour thrombosis

INTRODUCTION

Differentiated thyroid carcinoma (DTC) comprises 90% of all thyroid cancers. Out of all DTCs, follicular carcinoma (10-15% of all cases) is notorious for hematogenous spread and metastasis via microinvasion.[1] Reports of macro angio-invasion are rare. A tumour thrombus involving the internal jugular vein via direct invasion or occult metastasis are often encountered.[2,3] Thrombus extending beyond major cervical vessels into mediastinal vessels is a rarity. We report a case of follicular thyroid carcinoma with extensive cervical and mediastinal venous extension approached with a single-stage surgical procedure.

CASE REPORT

A 74-year-old male with a long-standing anterior cervical mass, presented with signs and symptoms of right heart failure and SVC syndrome. Physical examination showed a large firm, hard, anterior cervical mass with retrosternal extension on the right and a positive Pemberton sign. Upon cardiac evaluation, Echocardiography revealed a 6.4 x 2.9cm right atrial mass extending from the SVC and protruding through the TV into the RV. Further CT scan with angiography showed a highly vascular heterogeneously enhancing thyroid gland (12 × 6.5 × 11.4 cm), right lobe larger than the left with retrosternal extension (4.2 × 2.4 cm). Mass effect on larynx with trachea deviated to the left without luminal compromise. A filling defect s/o thrombus extending from right superior thyroid vein into the right IJV (approx. 6.2cm), inferiorly thrombus extending from bilateral ITV into left innominate, SVC & Right atrium. Ring sign as described by Taib et al was demonstrated on CT confirming no thrombi invasion into the endothelium.[4] Serum thyroglobulin was raised (5876 ng/ml).

Figure 1: Large anterior cervical mass with laterally displaced carotid artery (red arrow) and distended neck veins (blue arrow).

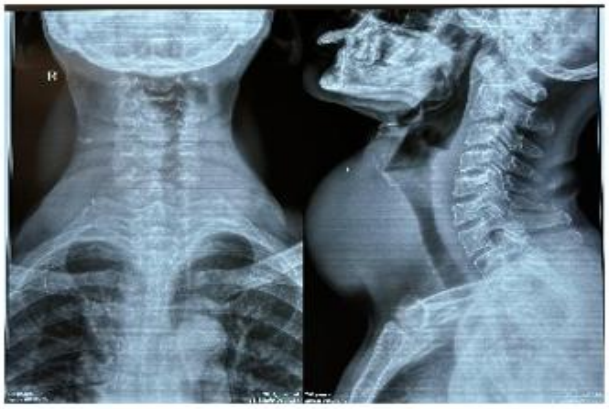

Figure 2: Neck Radiograph showing tracheal deviation towards the left without luminal compromise.

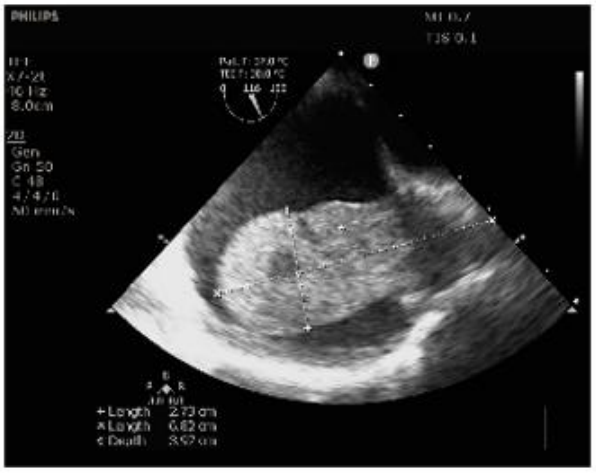

Figure 3: Echocardiography showing a large right atrial mass obstructing the tricuspid valve with extension into right ventricle.

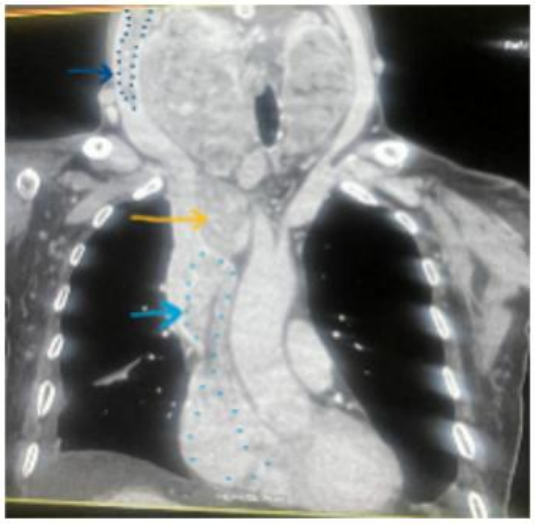

Figure 4: CT scan demonstrating a large heterogeneously enhancing thyroid gland 12 × 6.5 × 11.4 cm with retrosternal extension 4.2 × 2.4 cm (yellow arrow), thrombus extending from right superior thyroid vein into the right IJV of approx. 6.2cm (upper blue arrow), inferiorly thrombus extending from bilateral ITV into left innominate, SVC & Right atrium (lower blue arrow).

Raised serum thyroglobulin and angio-invasion incline towards a malignant aetiology. Whole body PET Scan ruled out metastases elsewhere. Owing to a highly vascular thyroid, FNAC was deferred. With a pressing urgency to prevent pulmonary embolism, patient was optimized for a single-stage surgery with a curative intent. Pre-operative coronary angiography was done which was within normal limits.

Patient was taken up for a total thyroidectomy with extraction of tumour thrombus. As tracheal lumen was not compromised, endotracheal intubation was possible. Since neck veins were involved, central venous and arterial access was secured via femoral vessels. Kocher's position given. Median sternotomy was done first which marginally relieved the venous congestion around the thyroid. An apron incision taken and subplatysmal flaps raised. Enlarged engorged veins with multiple collaterals along with an adherent thyroid gland to the overlying strap muscles noted. Collaterals ligated and strap muscles were cut. Total thyroidectomy done meticulously preserving both the recurrent laryngeal nerves. Retrosternal extension of the thyroid was seen extending along the right inferior thyroid vein and seated over the confluence of the right and left brachiocephalic veins. Excised en-bloc with meticulous dissection over the great veins. Two parathyroid glands were preserved, one reimplanted into the sternocleidomastoid muscle. Right internal jugular venotomy done and tumour thrombus extracted. No adherence to the wall of the vessel noted. A central venous access secured into the right ijv after thrombus extraction and venotomy closed.

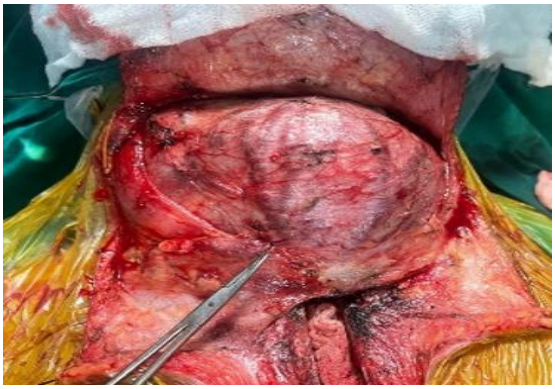

Figure 5: Enlarged thyroid gland with engorged veins with collaterals, as seen after raising subplatysmal flaps.

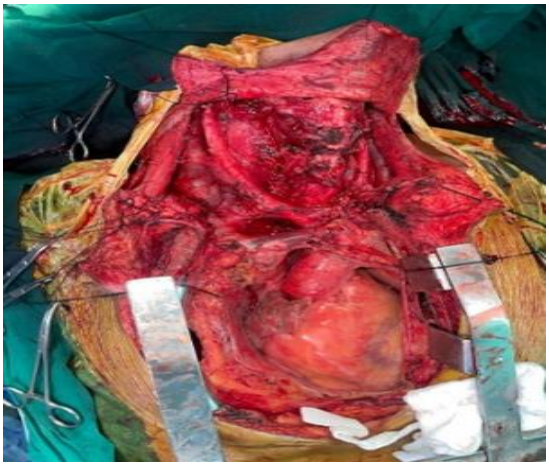

Figure 6: Complete clearance after total thyroidectomy with excision of retrosternal extension.

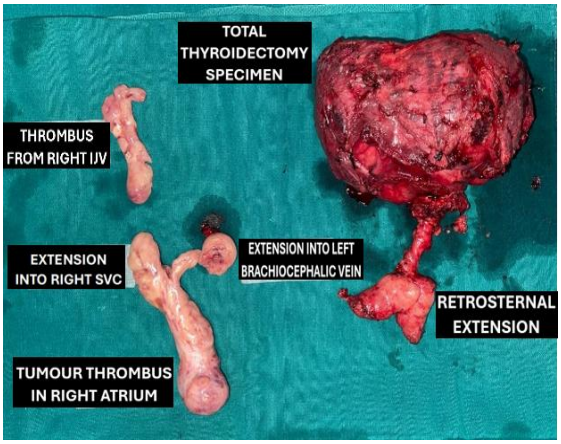

Figure 7: Gross specimen of Total thyroidectomy with retrosternal extension, Tumour thrombus from Right IJV, Inferior tumour thrombus.

Aorta and ivc cannulated and cardiopulmonary bypass established. Ra opened without cross clamping the aorta and tumour thrombus from the right atrium, svc and left innominate extracted. Ra closed. Intraoperative transoesophageal echocardiography and epicardial surface ultrasonography was done, no residual thrombi confirmed. Successfully weaned off bypass. Closure achieved with bilateral pleural, mediastinal drains and a closed suction drain in the neck.

He was extubated on post-operative day 2 with an uneventful post-operative recovery. Full oral feeds tolerated by post-operative day 4 and no change in voice recorded. Histopathology confirmed a follicular neoplasm of the thyroid with tumour thrombosis; pT4a N0 M0. Post-operatively, radioiodine ablation with 120mCi NaI-131 given. On regular follow-up, patient has remained disease free for 2 years.

DISCUSSION

Differentiated thyroid cancers are usually slow-growing and have a long trajectory to recurrence or death. Tumour thrombosis into great veins is a rare, but a well-documented entity in thyroid cancers. It is a strong predictor of malignancy and correlates with poor prognosis by virtue of invasive disease or it's associated complications.[2,5,6]

Tumour thrombosis occurs via direct intravascular invasion, embolization or infiltration into the vessel wall. Follicular carcinomas have been majorly linked with internal jugular vein thrombosis by direct hematogenous spread of malignant cells, further deposition of fibrin and formation of a flourishing thrombus.[7] Extensive vascular invasion is an independent risk factor predicting relapse and distant metastases.[4,5]

Tumour thrombosis is not a common feature of papillary thyroid neoplasm, but if encountered should have the same management approach as a follicular aetiology.[8]

Literature worldwide has documented 12 cases with tumour thrombi extending into the right atrium and ventricle. Out of 12, three were diagnosed on post-mortem.[9] Although rare, when encountered rapid assessment and management are imminent to avoid lethal complications like SVC syndrome, right atrial obstruction, and pulmonary embolism.

Aggressive and comprehensive surgical approach should be offered wherever possible. Offloading the tumour burden with surgery provides a better basis for effective radioiodine therapy. This would provide the best foot forward despite advanced disease.

With advanced pre-operative imaging modalities, if a 'Ring sign' is documented on CT/MRI it implies that the tumour thrombus is free from the endothelial lining of the vessel wall.[4] Such can be approached with thrombectomies. Complex cardiac anaesthesia, cardiopulmonary bypass availabilities enable single-staged surgical interventions. Invasion of the vessel wall

can be tackled with prosthetic restoration using Dacron grafts.[6,10]

CONCLUSION

Tumour thrombosis into major cervical and mediastinal structures is rare in thyroid malignancies, although when diagnosed warrants urgent management. Such extrathyroidal extension correlates with higher recurrence and distant metastases. Management should comprise of total thyroidectomy with extraction of all possible tumour deposits, supplemental vascular reconstructions followed by TSH-suppression therapy and I-131 Radioiodine ablation. Close follow-ups are required.

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