Follicular Thyroid Carcinoma with Direct Tumour Extension into the Great Cervical Veins and Right Atrium: Is Transcervical Thrombectomy a Safe Option?

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Direct tumour extension into the internal jugular veins (IJV) and right atrium in thyroid cancer is extremely uncommon. We report three cases of advanced thyroid cancer invading into the IJV and right atrium. All three patients had well-differentiated thyroid cancer and presented with typical clinical picture of the superior cava syndrome coupled with significant compressive symptoms. Two patients had obvious tumour thrombus extending to the superior vena cava (SVC) and right atrium, whilst in one patient, the thrombus extended to the SVC close to the edge of the right atrium. In all three patients, the intraluminal tumour thrombus was clinically palpable. Involvement of the IJV, SVC and right atrium was confirmed with computed tomography (CT) scan and echocardiography. The decision to extract the thrombus transcervically was made on the basis of the positive “ring sign”, which is a thin rim of contrast surrounding the tumour thrombus documented on CT. All three patients underwent total thyroidectomy with segmental resection and ligation of the IJV. The tumour thrombus was successfully extracted transcervically. The histopathology report confirmed follicular carcinoma in all three patients. Two patients had radioiodine ablation therapy postoperatively, and were well 18 months after operation. One patient who had lung metastases documented on chest CT succumbed to the disease due to massive haemoptysis 3 weeks after operation. [Asian J Surg 2007;30(3):216–9]

Key Words: direct tumour infiltration, internal jugular veins, right atrium, thyroid cancer

Introduction

Direct tumour extension into the internal jugular veins (IJV) and the right atrium in thyroid cancer is extremely uncommon.1–10 In the literature, about 20 cases have been reported,1–10 with eight cases requiring surgical resection. Seven required major vascular intervention involving resection of the great veins and graft bypass. Most of the cases involved only one side. Those with superior vena cava (SVC) obstruction required SVC thrombectomy, three cases had thrombectomy from the subclavian vein5 and one report had resection of bilateral innominate veins with graft bypass.5 Radical surgery is reported to be superior in achieving local control,5–6,11 and the longest survival was 5 years.5 In this case report, we would like to suggest that the transcervical approach is a safe option. In all three cases in this report, the “ring sign” was present in the IJV, where there was a rim of contrast demonstrable around the thrombus. This is an important sign which indicates that the thrombus is not adherent or invading...
into the endothelium lining of the veins, thus allowing the thrombus to be removed transcervically.

Case reports

Case 1
L.Y., a 66-year-old Malay lady, was referred for critical airway obstruction with a tracheostomy. She had a past history of total thyroidectomy for follicular thyroid cancer, but had defaulted treatment. Computed tomography (CT) scan showed an inhomogeneous enhancing recurrent thyroid mass extending from the right cervical region to the thoracic inlet. The mass was encasing and displacing the right carotid artery and IJV. There was obvious extension of the tumour thrombus into the IJV, SVC and right atrium. Figure 1 shows the positive ring sign in the SVC. Echocardiogram revealed tumour thrombus in the right atrium. Lung function test showed mild restrictive lung function. Figure 2 shows the gross specimen of the recurrent follicular thyroid carcinoma with tumour thrombus.

Case 2
F.A., a 62-year-old Malay lady, was admitted for right-sided thyroid swelling of 6 months’ duration, which had progressively enlarged and was associated with dyspnoea and dysphagia. There was a large lobulated mass seen on the right anterior neck with prominent dilated veins in the upper chest. There was another palpable fusiform mass extending from the right jaw to the right clavicle. Fine needle aspiration cytology (FNAC) of the thyroid mass confirmed a follicular neoplasm. Chest radiograph showed a soft tissue mass extending to the upper border of the sternum, but there was no obvious lung metastasis. CT showed an inhomogeneous mass extending from the right cervical region into the thoracic inlet and retrosternal area. This was an intraluminal tumour extension into the IJV, SVC and the right atrium (Figure 3). Echocardiography showed that the tumour thrombus was in the right atrium. Figure 4 illustrates transcervical thrombectomy after ligating the IJV.

Case 3
H., a 45-year-old Malay lady, presented with anterior neck swelling of 2 years’ duration and dysphagia of 5 months’ duration. On clinical examination, there was a right-sided thyroid swelling measuring 6 × 4 cm associated with a firm
fusiform swelling over the submandibular region extending to the clavicle, and FNAC revealed a follicular neoplasm. Neck CT showed the right lobe of the thyroid encasing the thyroid cartilage and infiltrating into the posterior part of the right submandibular gland. Tumour thrombus was seen in the right IJV from the angle of the jaw to the lower end of the right brachiocephalic vein. The ring sign was clearly seen. There were multiple lung metastases seen on CT. Preoperative echocardiography did not reveal any tumour thrombus in the right atrium. She underwent total thyroidectomy and transcervical thrombectomy. She was discharged well with a tracheostomy but unfortunately, she succumbed to the disease 3 weeks after operation with massive haemoptysis.

Discussion

Intraluminal invasion of vascular structures have been reported in other types of cancer, namely renal cell carcinoma where intraluminal propagation into the inferior vena cava has been reported in about 5% of all cases. Follicular thyroid carcinoma known for haematogenous spread may behave similarly. Follicular thyroid carcinoma is frequently reported in the literature with regard to intraluminal vascular involvement; however, papillary carcinoma, Hurthle cell, insular and anaplastic carcinoma have also been reported.1–11

In all three patients, the final diagnosis was follicular thyroid carcinoma. Follicular carcinoma is known to spread by the haematogenous route as compared to papillary carcinoma, which mainly spreads via the lymphatic system. The tumour invades into the IJV through the thyroid veins and propagates, but the most distal portion would be freely mobile and can present as a leaf-like structure, much like the vein thrombosis progression in deep vein thrombosis. The possible reason for why the endothelium of the great vessels is not invaded could be due to the high velocity of flow in these large vessels, thus not allowing the tumour to lateralize and invade into the wall.

CT should be done in patients with SVC obstruction to differentiate intraluminal extension or extrinsic compression in thyroid cancers.8 The operability and surgical approach of intravascular extension can be assessed with CT scan.8 In the first case where the carotid artery was encased, we managed to remove all of the tumour tissue and skeletonized the carotid artery. There was a plane between the IJV and the carotid artery. Encasement was actually due to the IJV wrapping itself longitudinally over a firm structure like the artery. In all three cases in this report, the ring sign was present in the great cervical veins, where there was a rim of contrast demonstrable around the thrombus. This sign indicates that the thrombus is not adherent or invading into the endothelium lining of the great veins, thus allowing the thrombus to be removed transcervically. With the improvement of medical imaging, i.e. spiral CT and three-dimensional reconstruction, further evaluation can be done to confirm this phenomenon.

The transcervical approach is justified, as the tumour thrombus does not usually involve the endothelial lining of the great veins.3 The ring sign seen in the SVC in cases 1 and 2 and brachiocephalic in case 3 demonstrates that the thrombi can be removed by a venotomy in the neck. However, segmental resection and ligation of the IJV upon removal of the thrombus were indicated in all patients as the segment of the vein was involved with tumour. Furthermore, in our cases, the tumour involved the vessels unilaterally, and resection of the IJV on one side is a safe option. The transcervical approach offers the patient a lesser procedure and good postoperative recovery without requiring a sternotomy.

During operation, the patient must be in Trendelenburg position to prevent air embolism. Upon removal of the lower end of the tumour thrombus, it was essential that there was back flow of blood before the IJV was ligated. Postoperative echocardiogram showed no residual tumour thrombus in the right atrium in the first two cases.

Radioactive iodine-131 ablation was given in two courses (200 mCi) to ablate the remaining functioning tissue in the
neck and mediastinal region in the first two cases. In the first two cases, the radioiodine scan revealed residual uptake in the neck and lungs. $^{131}$I total body scan (TBS) is essential to ensure that metastatic disease is radioiodine-sensitive, especially as the three patients were older than 40 years. Indiscriminate use of radioiodine for ablation is futile in cases with negative $^{131}$I TBS as there are risks of salivary and lacrimal complications. In patients with extensive distant metastases, e.g. in the lungs and bones, total thyroidectomy and tumour debulking should be performed in most cases, as they play a major role in the efficacy of postoperative radioablative treatment.

Thyroglobulin levels were not routinely measured in our hospitals, as it is not readily available. However, this test can be done to ensure that thyroid and tumour tissue have been adequately debulked and surveillance can be done in the post-treatment period. Our three patients did not have the test done preoperatively. The short follow-up time for the first two cases did not allow us to make conclusions on survival. In terms of local control, surgery offers the best hope as for our case 1, it allowed the patient to be without a tracheostomy. The unfavourable outcome in case 3 may be explained by erosion of the metastatic lesion in the lungs into a blood vessel, resulting in massive haemoptysis and fatal injury.

Transcervical tumour thrombectomy of intraluminal extension of thyroid carcinoma into the great cervical veins and right atrium with segmental excision of the IJV is a safe surgical option in patients with a positive ring sign on CT. Aggressive surgery is acceptable and justified in advanced well-differentiated thyroid cancer to decrease the probability of local recurrence and prevent tumour embolism. It also provides a better basis for effective radioiodine therapy by reducing the tumour mass and hence offers the best hope for prolonged survival.

References