A Rare Cause of Upper Limb Lymphoedema

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This is the first case report of upper extremity lymphoedema caused by a thyroid tumour. An 87 year old female patient with grade 3 lymphoedema of the upper right extremity was admitted. The lymphoedema had developed over the course of two years. Investigation identified a thyroid tumour, which compressed the mediastinal structures at the right superior thoracic outlet, causing venous congestion, oedema, and lymphoedema. The patient underwent thyroidectomy which markedly improved the lymphatic oedema. Resolution of the compression mechanism was an effective treatment, despite the severity and chronicity of the initial presentation.

INTRODUCTION

Oedema of an extremity may be caused by obstructive venous diseases or reflux based venous diseases, phleboedema, or by a lymphatic drainage deficit caused by an alteration of the lymphatic system, lymphoedema. It is possible to distinguish them clinically because lymphoedema usually manifests as a swelling in the dorsal surface of the hand with a characteristic blunt “squared off” appearance of the fingers. Lymphoedema usually causes swelling of the distal extremity and then progresses proximally, and is typically characterised by a pitted or dimpled texture of the skin (peau d’orange). In advanced stages the skin becomes hyperkeratotic and develops verrucous cobblestone like papules, plaques, and nodules with underlying woody fibrosis. Lymphoedema may be classified as primary or secondary according to the underlying cause. The causes of primary lymphoedema are unknown and relate to organic or functional deficits of the lymphatic system. Secondary lymphoedema is related to injuries of the lymphatic vessels and/or of the lymph nodes. The majority of upper extremity lymphoedema is secondary to axillary lymph node surgery or radiotherapy for the treatment of breast cancer.

This case report focuses on an unusual cause of upper extremity lymphoedema.

REPORT

An 87 year old female patient presented with lymphoedema (grade 3) of the upper right extremity that progressed over the course of two years (Fig. 1A). She complained of hoarseness and dysphagia that had settled in the last three months. There was no history of surgery, radiotherapy, trauma, or chronic infection. On inspection, right breast lymphoedema (Fig. 1B) and a cervical mass were apparent. Mammography, breast ultrasound, and biopsy ruled out inflammatory breast cancer. Ultrasound also showed no axillary lymphadenopathy or axillary venous thrombosis. The chest computed tomography scan (CT) revealed a solid left thyroid mass measuring 63 x 40 x 96 mm with mediastinal structure compression and deviation to the right (Fig. 2A and B). This scan also showed breast fat stranding and skin thickening. A thyroid biopsy showed papillary carcinoma. The upper limb lymphoedema was initially treated with zinc paste bandages to reduce the oedema and then with an elastic arm sleeve. Minor improvement of the lymphoedema was observed after compression therapy. The patient was also treated with antibiotics and non-steroidal anti-inflammatory drugs (NSAIDs) in the first week, together with lymphatic active drugs (diosmine) continuously. There was also an initial one week course of antibiotics because of a slight redness and warmth of the forearm and because infection could not be...
Figure 1. (A) Right upper extremity lymphoedema at presentation. Swelling of the dorsal surface of the hand. The skin is hyperkeratotic and presents verrucous cobblestone like papules (grade 3 lymphoedema). (B) Right upper extremity and breast lymphoedema. (C) Right upper extremity lymphoedema five weeks after thyroidectomy (grade 2).
Figure 2. (A) and (B) Axial and coronal CT images show a solid left thyroid mass (asterisk) compressing the mediastinal structures, in particular the lymphatic vessels, right lymphatic duct, and subclavian vein at the right superior thoracic outlet (arrow). (C) CT axial image showing right breast enlargement, fat stranding (asterisk), and skin thickening (arrow) reflecting the lymphoedema.
completely ruled out. The patient was later treated by total thyroidectomy and the upper extremity lymphoedema improved significantly, beyond the changes observed after compression therapy.

CONCLUSION

To the authors’ knowledge, this is the first case of upper extremity and breast lymphoedema caused by a thyroid tumour. It appeared that the thyroid mass compressed the mediastinal structures, in particular the lymphatic vessels, right lymphatic duct, and subclavian vein at the right superior thoracic outlet, causing venous congestion, lymphoedema, hoarseness, and dysphagia, underlining the mass effect. The oedema was not caused by metastatic lymphadenopathy, despite the diagnosis of papillary thyroid carcinoma (adenopathies were not clinically evident, nor identified after CT examination). The right upper extremity lymph is drained by superficial and deep channels, which converge in the apical group of axillary lymph nodes. These are received by the right lymphatic duct, which drains into the right subclavian vein at its junction with the internal jugular vein, or with various combinations at the right subclavian vein and jugular trunks. In this case the lymphatic outflow was compromised with the resulting development of upper extremity and breast lymphoedema. Despite initial elastic compression therapy, the lymphoedema only subsided significantly after the central obstruction was alleviated through surgery, reinforcing the clinical diagnostic hypothesis.

CONFLICTS OF INTEREST

None.

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REFERENCES