CASE REPORT

Spectrum on Non-Surgical Causes of Internal Jugular Vein Thrombosis In Otorhinolaryngology Practice

Mohd Hazmi Mohamed[1], Azlan Iskandar Ishak[1], Wan Aliaa Wan Sulaiman[2]

[1] Department of Otorhinolaryngology, Faculty of Medicine and Health Sciences, Universiti Putra Malaysia
[2] Department of Neurology, Faculty of Medicine and Health Sciences, Universiti Putra Malaysia

ABSTRACT

Internal jugular vein thrombosis is an uncommon entity secondary to various aetiologies. In the pre-antibiotic era, it was commonly associated with Lemierre syndrome, a septic thrombophlebitis of the internal jugular vein secondary to oropharyngeal infection. With the advent of antibiotics, Lemierre syndrome has become a rare entity and is often overlooked. Internal jugular vein thrombosis may also occur in the setting of malignancy, a combination referred to as Trousseau's syndrome, which is not well documented.
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Corresponding Author:
Mohd Hazmi Mohamed, MS (ORL-HNS)
Email:mdhazmi@upm.edu.my
Tel: +603-97692360

INTRODUCTION

Venous thromboembolic disease, primarily deep venous thrombosis is a common occurrence in medical practice. However internal jugular vein thrombosis, an upper extremities vein thrombosis remains uncommon. While deep venous thromboses typically occur in the lower extremities, they constitute only 4-10% of all venous thrombosis in the upper extremities, with the incidence of internal jugular vein thrombosis being lesser (1).

Internal jugular vein thrombosis is potentially life-threatening, as it can cause airway oedema, systemic sepsis and pulmonary embolism. Patients typically present with neck swelling and neck pain, although the initial symptoms may be subtle. The condition often follows infection in the head and neck region, neck surgery, trauma, complications of central venous catheterization, malignancy, or a hypercoagulable state. Thrombosis can occur anywhere from the intracranial segment of the internal jugular vein to the junction of the internal jugular vein and subclavian vein.

CASE REPORTS

Case 1

A 33-year-old obese lady with a history of diabetes mellitus presented with a sudden onset of painful right neck swelling and fever lasting for 3 days. She had a preceding upper respiratory tract infection. Examination revealed a firm, warm, erythematous swelling in the right neck (level II to level III) measuring 5x6 cm. No trismus or medialization of the lateral pharyngeal wall was observed.

Initial blood investigation showed elevated C-reactive protein (71.6 mg/l) and high D-dimer (1529 ng/ml). Computed Tomography (CT) revealed a heterogenous hypodense mass in the right parapharyngeal space measuring 5.5x5.3x9.5 cm, with a predominant solid component, and concomitant focal right internal jugular vein thrombosis. The provisional diagnosis was parapahryngeal space inflammation.

She was started on a broad-spectrum antibiotic with intravenous ceftriaxone. Despite clinical improvement of the swelling and neck pain, D-dimer progressively increased. A repeat CT scan showed the previously heterogenous mass already liquefied forming an abscess collection. The internal jugular vein thrombosis had extended up to the right brachiocephalic vein (Figure 1). Given the septic thrombophlebitic of the internal jugular vein, she was diagnosed with Lemierre syndrome.

She underwent neck exploration and abscess drainage under general anaesthesia. She recovered well post-operatively and was started on an anticoagulant with warfarin as advised by the infectious disease. Her neck swab culture later showed a growth of klebsiella species, sensitive to ceftriaxone. She was discharged home with a six-week oral antibiotic and a continuation of warfarin.

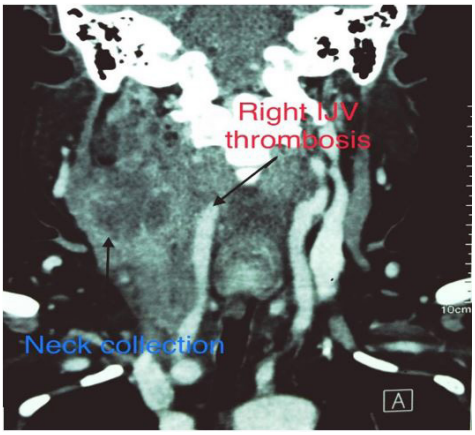

Figure 1: Contrast Computed Tomography Slice Showing the Right IJV Thrombosis with Adjacent Heterogenous Right Neck Collection

Case 2

A 77-year-old female presented with left-sided neck swelling for 6 months. The swelling, initially small, gradually increased in size. She had similar episodes 2 years earlier but completely resolved after a course of antibiotics. She reported weight loss but denied fever, night sweats and direct contact with tuberculosis patients.

Fine needle aspiration cytology (FNAC) of the swelling was inconclusive initially, with the second test showing reactive lymphadenitis. Concerned about her condition, she sought a second opinion in our centre.

Computed tomography (CT) of the neck revealed an enlarged left submandibular gland and multiple cervical lymphadenopathies, mostly in the left neck, with some of the nodes showing a hypodense centre suggestive of a necrotic component. A long segment of the left internal jugular vein was thrombosed from the level of C4 down to the left brachiocephalic vein (Figure 2).

Since her previous FNACs were inconclusive and the CT scan finding was suggestive of lymphoma, she underwent submandibulectomy and excision biopsy of left cervical lymph nodes. Both specimens showed Diffuse Large B Cell lymphoma (DLBCL). Given the long segment left internal jugular vein thrombosis, she was started on warfarin and was referred to the haematology unit for DLBC treatment.

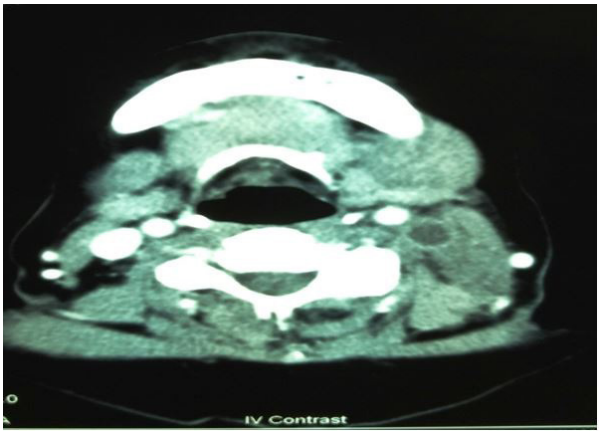

Figure 2: Contrast Computed Tomography Slice Showing Lymph Nodes Hypodense Centre, Suggestive of a Necrotic Component

DISCUSSION

Lemierre disease is a septic thrombophlebitis of the internal jugular vein following an oropharyngeal infection. It was in the pre-antibiotic era with a high mortality rate. With the widespread use of antibiotics in treating throat infections, the number of cases was drastically reduced. The syndrome was initially called as postanginal septicaemia and eventually named Lemierre syndrome in honour of Andre Lemierre, who in 1936 reported 20 cases of internal jugular vein thrombosis with septic emboli; of whom 18 died (2).

Fusobacterium necrophorum; a gram-negative obligate anaerobe, is overwhelmingly the common organism isolated from patients with Lemierre syndrome. Other organisms isolated include *Bacteroides melaninogenicus, Eikenella corrodens*, and non-group *A Streptococcus*. Singaporewella et al in 2006 reported a case of Lemierre syndrome with a pus culture positive for *Klebsiella pneumoniae* in a patient with uncontrolled diabetes (3). In our first case, we described a similar species of organism, klebsiella species, in a diabetic patient. Klebsiella is a common pathogen isolated in neck abscesses and may over over-represented in diabetic patients. While Fusobacterium necrophorum remains the most commonly identified pathogen, it is crucial to acknowledge that our case, along with others in the literature, highlights the diversity of bacteria associated with Lemierre syndrome (4).

In Case 1, initial computed tomography (CT) showed signs of neck inflammation. Despite of broad spectrum of antibiotics, the disease progressed into an abscess and extension of internal jugular vein thrombosis. Even if antibiotics used are effective against causative organisms in vitro, they may not always be effective in deep neck infections. Surgical drainage may, therefore, be necessary for Lemierre syndrome with purulent collection.

D-dimer, a fibrin degradation product, may help in the diagnosis of thrombosis and be used as a prognostic, although its clinical value in Lemierre syndrome has been poorly described so far. In our first case, despite broad-spectrum antibiotics and clinical response, the D-dimer value was elevated, prompting an early repeat CT scan which showed worsening IJV thrombosis. Additionally, our case contributes to the growing body of evidence indicating that thrombosis associated with Lemierre syndrome can extend in the first days of hospitalization. This observation aligns with findings from recent meta-analyses, such as the study by Valerio et al (4).

Accurate diagnosis of the internal jugular vein and its causes is difficult, with imaging playing a crucial role. While contrast venography is still considered the gold standard for diagnosis of venous occlusion; it carries a significant risk of clots dislodgement and dissemination of septic emboli. Non-invasive imaging such as contrast-enhanced CT or Magnetic Resonance (MR) are usually better options in the diagnosis of internal jugular vein thrombosis since the extent of the thrombus, anatomy of surrounding structure and any localised infection can be identified. These investigations may give additional information regarding the causes of internal jugular vein thrombosis such as direct invasion of thyroid carcinoma or other neck masses.

Trousseau in the 1860s delineated an association between cancer, venous thrombosis and hypercoagulability. He asserted that if the diagnosis of a suspected carcinoma of an internal organ could not be verified, the sudden and spontaneous appearance of thrombophlebitis in a large vein afforded necessary proof for diagnosis; an occurrence referred to as Trousseau syndrome.

Head and neck region malignancy with thrombosis is uncommon. In a series of 541 cases with Trousseau's syndrome, Sack et al reported that the majority of underlying malignancies were identified in the lung (25.6%), pancreas (17.4%), stomach (16.8%), colon (15.2%), prostate,(6.5%) and head and neck (2%) (5). In our second case, we present a case suggesting a potential direct cause of internal jugular vein thrombosis arising from neck lymphoma.

Upon a diagnosis of internal jugular vein thrombosis, anticoagulant therapy should be considered in view risk of pulmonary embolism. However, there is no clear guideline regarding anticoagulants in internal jugular vein thrombosis. Treatment of the underlying cause or malignancy is the most definitive therapy. In cases of infection, patients may respond well with antibiotics alone without anticoagulant therapy. Nevertheless, in the presence of septic emboli or propagation of emboli, anti-coagulant usage should at least be considered.

Nevertheless, more causes of thrombosis are known in medicine outside the scope of Otorhinolaryngology practice; for instance, in family medicine, thrombosis is seen in intravenous drug users and critical care central line-associated thrombosis of the jugular vein.

CONCLUSION

The presented cases highlight the diverse aetiologies of internal jugular vein thrombosis, emphasising the importance of a thorough diagnostic approach. Additionally, imaging, such as contrasted CT scan, is pivotal in accurate diagnosis and identification of contributing factors.

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