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Research Article

LEMIERRE SYNDROME- AN UNCOMMON PHENOMENON

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Abstract: ***Introduction:*** Internal jugular vein thrombosis (IJVT) is a form of venous thromboembolism involving clot formation within the internal jugular vein, resulting in impaired venous drainage from the head and neck. Although less common than lower extremity deep vein thrombosis, the incidence of IJVT has increased with the widespread use of central venous catheters and the rising prevalence of malignant neoplasms and hypercoagulable states. Diagnosis of IJVT is challenging because patients are frequently asymptomatic or present with nonspecific symptoms such as neck pain or swelling. While rare, complications of IJVT can include pulmonary embolism, cerebral venous thrombosis, or septic thrombophlebitis. Early recognition and appropriate treatment are essential to prevent morbidity and mortality.**Case report:** A fifty-seven-year- old female, not a known case of any chronic illness, presented with headache for last four months. It was dull aching, almost continuous type. She developed left neck pain & swelling with cough for last two weeks. She has history of repeated throat infections in last one year, one single episode used to remain for five days and was symptomatically treated by a private- practitioners. There was no history of redness of eyes, blurring of vision, vomiting, difficulty in breathing or weakness of any part of body. She reported first to ENT specialist who on clinical examination found neck swellings but rest all general physical and systemic examination was essentially normal. He advised ultrasonogram of neck which showed bilateral submandibular glands to be small with heterogenous echotexture suggestive of chronic sialadenitis. The left internal jugular vein, left subclavian vein and left brachio-cephalic vein were all thrombosed. The CT neck and chest venography showed a long segment non enhancing intraluminal filling defect involving the left internal jugular vein, extending into terminal portion of left subclavian vein near its venous drainage (involved segment measuring 35 mm) and further into left brachio-cephalic (innominate) vein, causing complete luminal occlusion, suggestive of thrombosis. There was prominence of the external jugular vein and anterior facial veins bilaterally with prominence of multiple smaller superficial venous channels in the neck, representing collateral venous drainage. Mild pericardial effusion was noted. Later on, CTVS opinion was taken who advised for whole body PET CT with CT venography thorax, along with APLA profile, ANA, ENA, C-ANCA, P-ANCA, ACE level, CRP and serum homocysteine levels. She was started on low molecular weight heparin for five days, followed by oral anticoagulants. The whole-body PET-CT showed few faintly FDG avid necrotic left highest mediastinal and left subpectoral lymph nodes-? Infective/inflammatory. The contrast enhanced CT venography neck showed left internal jugular vein, left subclavian vein, brachio-cephalic vein appeared distended and showed no contrast opacification likely thrombosis. Multiple small venous collaterals were noted in neck. The complete biochemical profile including hypercoagulable profile was normal.**Conclusion:** Internal jugular vein thrombosis is a rare condition. All causes should be excluded radiologically and by laboratory tests related to jugular vein thrombosis. IJVT has potentially fatal complications, hence high level of suspicion leading to early diagnosis and treatment under specialist care is mandatory.

Keywords: Lemierre Syndrome, Thrombus, Internal Jugular vein, Subclavian vein, Brachiocephalic vein, Venography, PET Scan

INTRODUCTION

The internal jugular vein (IJV) originates at the jugular foramen. The IJV descends through the lateral neck to join the subclavian vein, forming the brachiocephalic vein, which ultimately drains into the superior vena cava. The IJV lies within the carotid sheath alongside the carotid artery and vagus nerve, typically coursing lateral to the carotid artery and deep to the sternocleidomastoid muscle. Anatomic variation is common, with the IJV most often positioned lateral or anterolateral to the carotid artery, though it may be located medially less commonly.{1} Internal jugular vein thrombosis (IJVT) is a form of venous thromboembolism involving clot formation within the internal jugular vein, resulting in impaired venous drainage from the head and neck. It is less common than lower extremity deep vein thrombosis, the incidence of IJVT has increased with the widespread use of central venous catheters, dialysis catheters, pacemakers, and chemotherapy ports and the rising prevalence of malignant neoplasms and hypercoagulable states.{2-4} Among patients in the intensive care unit, internal jugular vein thrombosis was associated with a platelet count greater than 200,000/μL, platelet transfusion, fresh-frozen plasma transfusion, vasopressor use, mechanical ventilation support, and a central line in place for more than 14 days.{5} Hypercoagulability can also cause IJVT and includes conditions such as factor V Leiden mutation, protein C

deficiency, protein S deficiency, and antithrombin deficiency. {4} Adjacent ear, nose, and throat infections can cause venous inflammation, leading to IJVT. Lemierre syndrome is an oropharyngeal infection that causes septic thrombophlebitis of the internal jugular vein. The most common causative bacterium in Lemierre syndrome is Fusobacterium necrophorum, which translocates to the internal jugular vein, causing thrombus formation. {6} Other less common causes of IJVT include malignant neoplasms, surgical dissection, trauma, local infections, paraneoplastic disease, and ovarian hyperstimulation syndrome. The asymptomatic nature or non-specific presentation with neck pain or swelling poses a diagnostic challenge in IJVT. Many other features like erythema, tenderness and fullness in neck, along with headache may point towards IJVT. Infectious internal jugular vein thrombosis, as seen in Lemierre syndrome, may present with fever, neck swelling, jaw angle pain, and trismus, often following pharyngitis or dental infection. Signs of systemic infection, such as fever and tachycardia, may also be present. {6} The rarer complications of IJVT include pulmonary embolism, cerebral venous thrombosis, or septic thrombophlebitis. A raised D-dimer level can hint at deep vein thrombosis, thus for confirming IJVT specifically, duplex ultrasonography is the first-line imaging modality. A noncompressible vein without any color flow is diagnostic of DVT. Duplex ultrasonography has a sensitivity of 96% and specificity of 93%. {7} The findings of duplex ultrasonography can be confirmed on venography of neck veins. Other investigations like PET CT and tests for ruling out hypercoagulable state should also be done for detailed aetiological workup and future management of the patient. Early recognition and appropriate treatment are essential to prevent morbidity and mortality.

CASE REPORT

A fifty-seven-year- old female, not a known case of any chronic illness, presented with headache for last four months. It was dull aching, almost continuous type. She developed left neck pain & swelling with cough for last two weeks. She has history of repeated throat infections in last one year, one single episode used to remain for five days and was symptomatically treated by a private- practitioners. There was no history of redness of eyes, blurring of vision, vomiting, difficulty in breathing or weakness of any part of body. She reported first to ENT specialist who on clinical examination found neck swellings but rest all general physical and systemic examination was essentially normal. He advised ultrasonogram of neck which showed bilateral submandibular glands to be small with heterogenous echotexture suggestive of chronic sialadenitis. The left internal jugular vein, left subclavian vein and left brachio-cephalic vein were all thrombosed. The CT neck and chest venography showed a long segment non enhancing intraluminal filling defect involving the left internal jugular vein, extending into terminal portion of left subclavian vein near its venous drainage (involved segment measuring 35 mm) and further into left brachio-cephalic (innominate) vein, causing complete luminal occlusion, suggestive of thrombosis. There was prominence of the external jugular vein and anterior facial veins bilaterally with prominence of multiple smaller superficial venous channels in the neck, representing collateral venous drainage. Mild pericardial effusion was noted. Later on, CTVS opinion was taken who advised for whole body PET CT with CT venography thorax, along with APLA profile, ANA, ENA, C-ANCA, P-ANCA, ACE level, CRP and serum homocysteine levels. She was started on low molecular weight heparin for five days, followed by oral anticoagulants. The whole-body PET-CT showed few faintly FDG avid necrotic left highest mediastinal and left subpectoral lymph nodes-? Infective/inflammatory. The contrast enhanced CT venography neck showed left internal jugular vein, left subclavian vein, brachio-cephalic vein appeared distended and showed no contrast opacification likely thrombosis. Multiple small venous collaterals were noted in neck. The complete biochemical profile including hypercoagulable profile was normal.

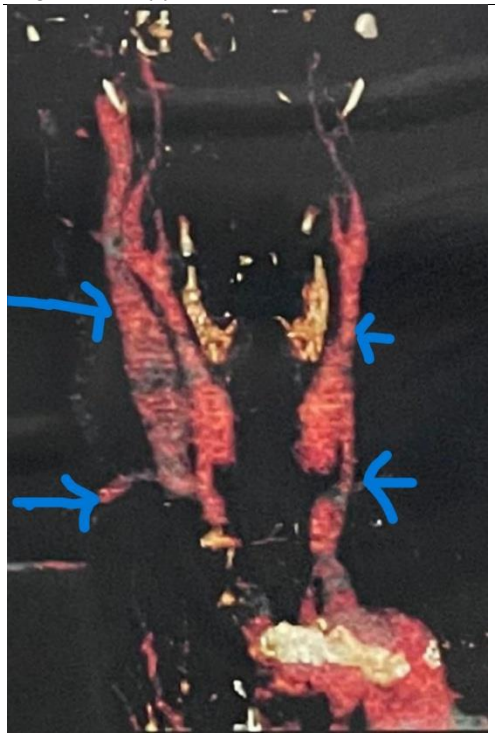

Figure 1- Showing non-visualization Of left IJV (small blue arrows) but Right IJV is seen (large blue arrows)

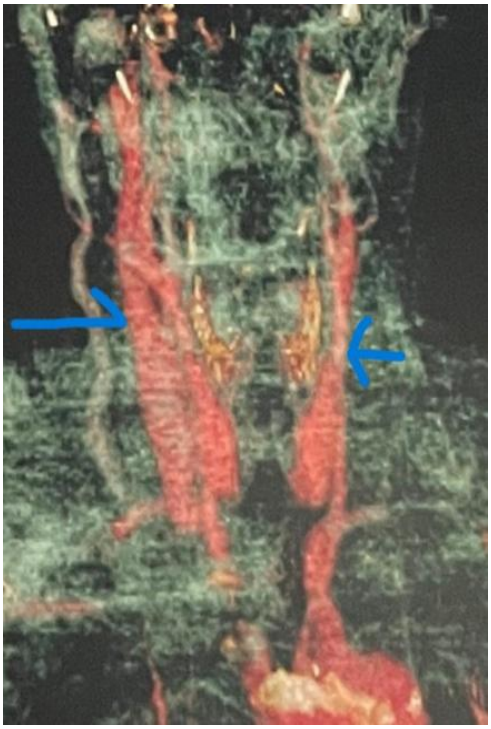

Figure 2- Showing non-visualization of left IJV (small blue arrow) but right IJV is seen (large blue arrow)

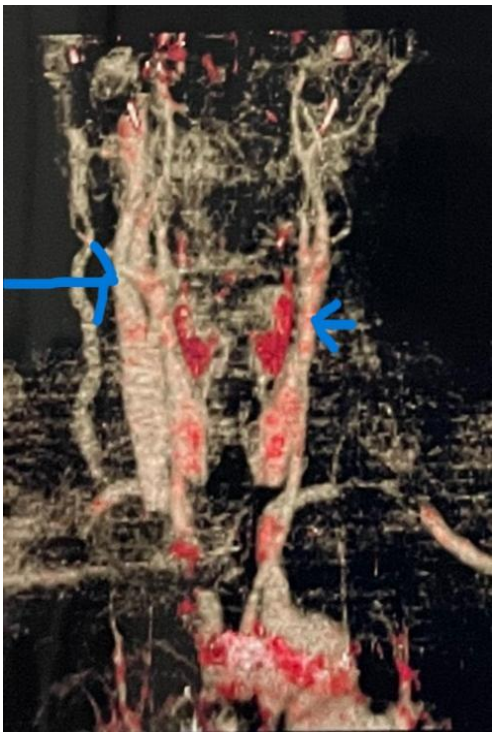

Figure 3- Showing non-opacified Left IJV (small blue arrow) but Right IJV is seen (large blue arrow)

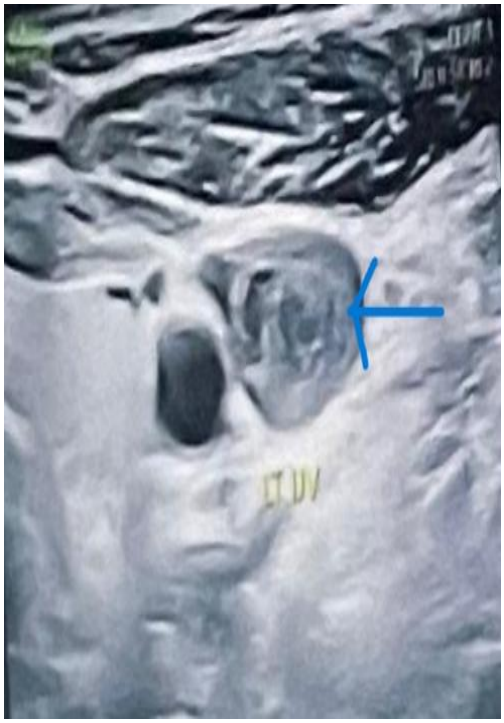

Figure 4- Ultrasonogram neck shows thrombus in left Internal Jugular Vein

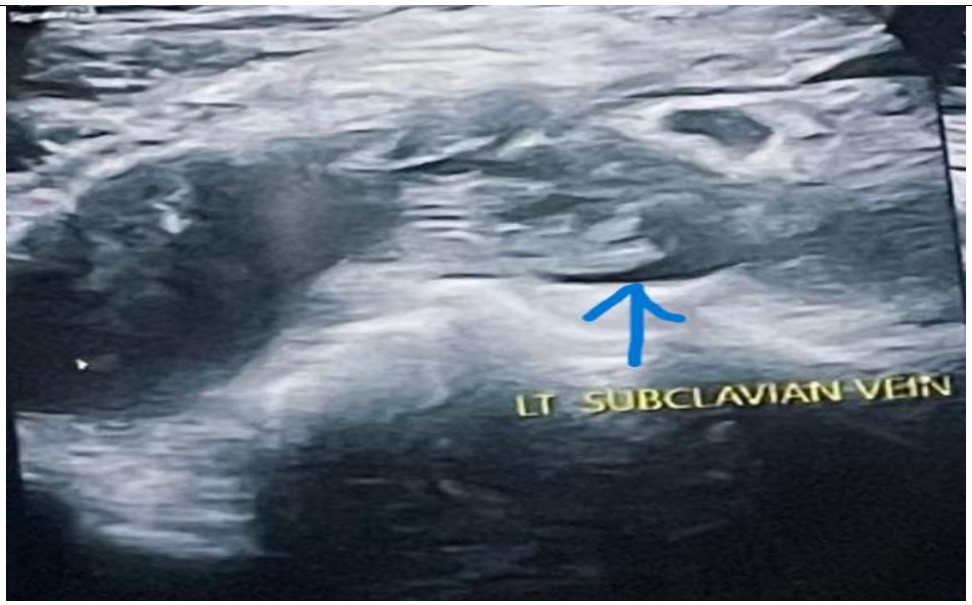

Figure 5- Ultrasonogram neck showing thrombus in Left Subclavian Vein

DISCUSSION

Internal Jugular Vein (IJV) thrombosis is a rare, potentially life-threatening condition defined by a blood clot in the neck's primary vein. It is primarily diagnosed using Doppler ultrasound or CT scans and is treated with a 3 to 6-month course of anticoagulants to prevent serious complications like pulmonary embolism. Thrombosis generally stems from Virchow's Triad i.e. endothelial injury, blood stasis, and hypercoagulability. Internal jugular vein thrombosis (IJVT) is an unusual case of vascular disease of the upper limb veins that could result in multiple complications if left untreated. IJVT can be subdivided into primary and secondary.

Primary IJVT is when thrombosis happens to someone without known risk factors. Most patients present with painful erythematous neck swelling and headache. {8,9} IJVT is also reported to occur spontaneously as a rare case {10} and tends to occur more often in women. In our case, the only aetiology found was infective. She was started on low molecular wight heparin, followed by oral anticoagulants and showed substantial recovery within one month. Her headache and neck swelling has decreased substantially and has been planned for at-least six-month oral anticoagulant and depending on recovery, future continuation of the same will be decided.

CONCLUSION

Internal jugular vein thrombosis is a rare condition. All causes should be excluded radiologically and by laboratory tests related to jugular vein thrombosis. IJVT has potentially fatal complications, hence high level of suspicion leading to early diagnosis and treatment under specialist care is mandatory.

CONFLICT OF INTEREST

The authors declare that there was no conflict of interest and consent was taken from patient before publishing this case report.

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