Spontaneous internal jugular vein thrombosis and metastatic adenocarcinoma of unknown primary

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Internal jugular vein thrombosis is difficult to diagnose clinically. Real time ultrasound should be the investigation of choice. For spontaneous jugular vein thrombosis associated with adenocarcinoma of unknown primary, further imaging to search for the primary tumour is probably not justified because of the poor prognosis of the patient.

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Introduction

Spontaneous internal jugular vein thrombosis (IJVT) is rarely the first clinical diagnosis given when patients present with neck swelling and fever or a neck mass. Ultrasound, computerised tomography (CT) and magnetic resonance imaging (MRI) can all give the correct diagnosis. Because of the practicality and non-invasiveness of ultrasound, we recommend this as the investigation of choice. Patients with adenocarcinoma of unknown primary have a poor prognosis and their response to chemotherapy is not affected by the location of the primary site of malignancy. An enthusiastic search for the primary tumour is therefore not recommended.

Case report

A 56-year-old Chinese woman presented with anorexia and localised swelling of the left side of the neck of three weeks' duration. She was afebrile and clinically fit. There was no history of trauma or coagulation disorder. The mass was tender. A high-resolution ultrasound of the neck was performed to exclude lymphadenopathy. A 2 cm segment of distal left internal jugular vein was distended

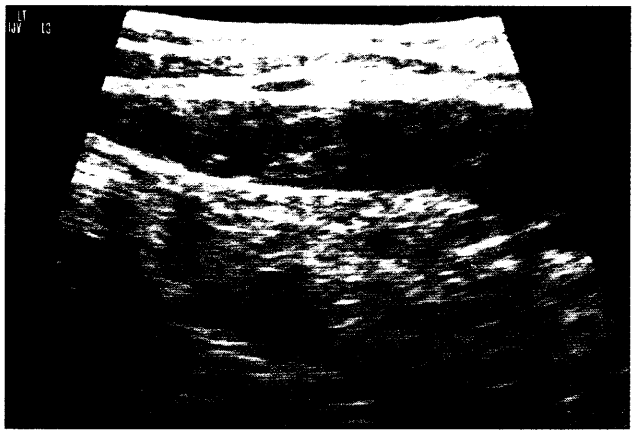

Fig 1a. Longitudinal section of left internal jugular vein showing echogenicity inside the lumen(*)

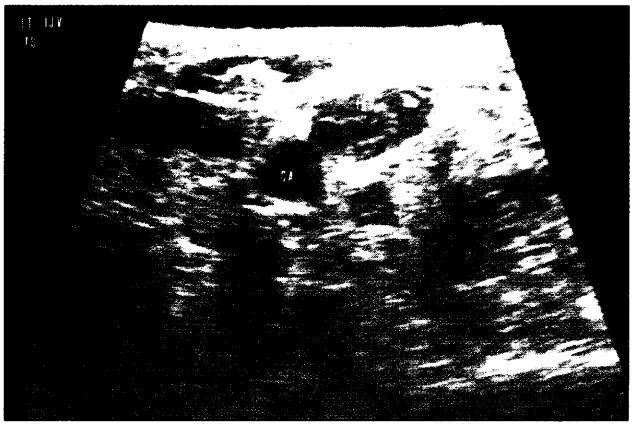

Fig 1b. Tranverse section of left internal jugular vein showing the thrombus and distension of the vein IJV = internal jugular vein, CA = carotid artery(*)

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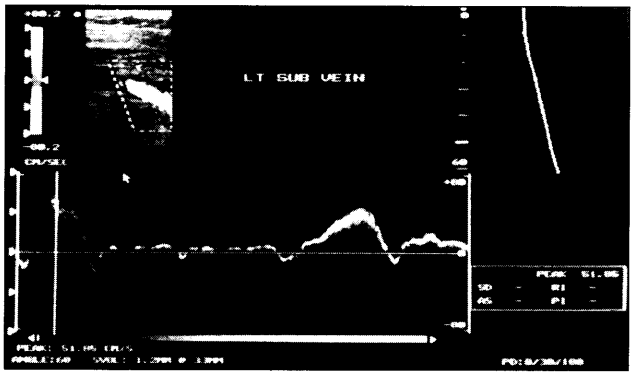

Fig 2. Doppler ultrasound showing patency and pulsatile waveform indicating good flow and no obstruction in the left subclavian vein

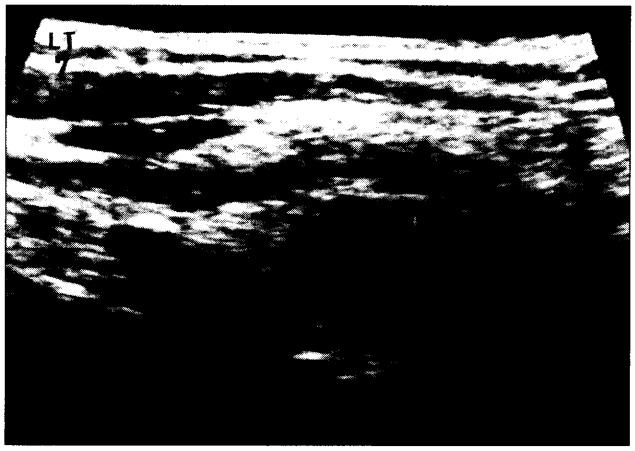

Fig 3. The largest lymph node in left supraclavicular fossa, measuring 1 cm in diameter. Note the hypoechoic centre and irregular outline suggestive of malignancy

and filled with an echogenic substance (Figs 1a and 1b). Colour Doppler ultrasound confirmed that there was no flow in the thrombosed segment. The left external jugular vein was distended, while the left subclavian and axillary veins were patent (Fig 2). There was no associated lymphadenopathy. Anticoagulation for spontaneous IJVT was given to reduce the risk of pulmonary embolism.

A repeat ultrasound was performed three weeks later to assess the patient's response to therapy. The extent of thrombus remained unchanged, however, there were multiple lymph nodes in the left supraclavicular fossa. These were not compressing on the major vessels. The largest lymph node measured 1 cm in diameter and had a hypoechoic centre and irregular outline (Fig 3). The ultrasound appearance of the lymph node was suggestive of malignant involvement. Excision biopsy of the lymph node revealed adenocarcinoma. Chest radiography, barium studies, and an abdominal ultrasound to search for the primary site of malignancy were unremarkable. The patient progressively deteriorated and died six months after her initial presentation.

Discussion

There are multiple causes of IJVT. The most common cause is an indwelling central catheter. Other recognised causes include localised infection, surgery, intravenous drug abuse, and compression by intrinsic or extrinsic tumours.[1] It has also been reported to be associated with distant primary neoplasm[1,2] and adenocarcinoma of unknown primary.[3] The most common presentation of IJVT includes neck swelling, fever, neck tenderness, or the presence of a mass. Making a clinical diagnosis of IJVT is often difficult. Frequently, clinicians make an initial diagnosis of infection or tumour and imaging such as ultrasound or CT subsequently reveals the correct diagnosis.

Complications of IJVT include pulmonary embolism, which has an incidence of 5%. Other recognised complications include septic emboli, generalised septicaemia, facial oedema, and pseudotumour cerebri.[4] Because of the risk of pulmonary embolism, anticoagulant therapy is usually recommended. Oral warfarin should be continued for three months except in the case of intravenous drug abusers who have a high incidence of haemorrhagic complications.

This condition can be diagnosed using venography, scintigraphy, ultrasound, CT, and MRI. The last three imaging modalities have the advantage of being non-invasive and may give additional information about the cause of IJVT, such as extrinsic compression or direct invasion by thyroid carcinomas or other neck masses. Ultrasound is readily available, involves no radiation, and is rapid and convenient in terms of post-treatment follow-up. The only drawback is when investigating a thrombosis extending into the mandible or beneath the clavicle, as ultrasound is unable to delineate the full extent of these thrombi. However, the full extent of the thrombosis is usually not a critical factor in determining treatment.[5] The diagnostic criteria in real time ultrasound include enlarged jugular vein containing solid material consisting of thrombus, lack of compressibility of the vein, lack of venous pulsations, lack of dilatation on Valsalva manoeuvre, and dilatation of the contralateral jugular vein.[6] Colour Doppler ultrasound can detect turbulance

or the absence of flow, and is very helpful in detecting an acute thrombus which has a low level of echogenicity.

Internal jugular vein thrombosis is diagnosed in CT by the increase in size of the vein, and the presence of the thrombus as a low-attenuation non-enhancing filling defect. The vessel wall may be enhanced (possibly due to blood flow through the vasa vasorum) and secondary reactive soft tissue swelling may be present. Computerised tomography has two disadvantages—the need to introduce contrast media and the use of radiation. Magnetic resonance imaging can also be used to detect thrombosis, however its use is limited because of its expense and restricted availability.

Patients with metastatic adenocarcinoma of unknown site have a poor prognosis. The median survival time is only 15 weeks.[5] Despite an enthusiastic search for the primary tumour and even an autospy study, the primary tumour is often not found. Although different chemotherapy regimens have been under trial to improve the survival time, the response is not very promising.[4-6] Our patient rapidly deteriorated and died after six months. We were unable to detect the primary neoplasm despite the various imaging performed. In this clinical setting, ultrasonography alone will suffice. Chest radiography is the only investigation to be added, if any. Unnecessary investigations should be avoided as this will not alter the management or prognosis of the patient.[4]

We believe that real time ultrasound should be the recommended imaging modality for the diagnosis of IJVT. The use of Doppler ultrasound further increases the sensitivity. In patients with IJVT associated with adenocarcinoma of unknown primary, further imaging to search for the primary tumour is probably not indicated because of the low yield and poor prognosis of the patient.

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