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Clinical Record

Internal jugular vein thrombosis due to distant malignancies: two case reports and literature review

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Abstract

The internal jugular vein is an uncommon site of spontaneous venous thrombosis. Most cases usually result from intravenous drug abuse, jugular vein catheterisation, neck dissection, a hypercoagulable state associated with malignancy (Trousseau's syndrome), neck injury or ovarian overstimulation syndrome. In this paper, we present and discuss two cases of spontaneous jugular vein thrombosis associated with breast and lung malignancies. The possibility of Trousseau's syndrome due to distant malignancy should be considered by otolaryngologists and appropriately investigated.

Key words: Internal Jugular Vein; Venous Thrombosis; Occult Primary Neoplasm

Introduction

Venous thromboembolic disease has an estimated annual incidence in developed countries of one in 1000 people. The disorder commonly affects the legs, but may occur in other veins. The veins in the head and neck, even in the presence of localised disease, appear to be less susceptible to thrombosis, as they are mostly valveless and gravity aids their emptying in the upright position.[1,2]

Although less common than lower extremity deep vein thrombosis (DVT), the incidence of DVT involving the upper extremities may be increasing and accounts for approximately 4 per cent of all DVTs. The most common predisposing factor is the presence of a central venous catheter, which is present in up to 75 per cent of patients with upper extremity DVT.[3] The presence of local or distant malignancy is an important aetiological factor which should be considered.[1] The association of cancer and thrombophlebitis was first observed by Trousseau, and this association still bears his name. The incidence of thrombophlebitis in cancer patients is quite common, and migratory thrombophlebitis is well documented.[3]

Venous thrombosis results from a disturbance of the normal blood flow, with subsequent activation of the coagulation mechanism. The pathophysiology of venous thrombosis is well described in Virchow's triad for vascular thrombosis, which requires the presence of one or more of the following factors: endothelial damage, alteration of blood flow and blood hypercoagulability. Activated clotting factors collect in areas of sluggish or turbulent blood flow, thereby precipitating platelet aggregation. This in turn initiates the thrombotic process.[4]

The internal jugular vein (IJV) is an uncommon site of spontaneous venous thrombosis. Most cases usually result from intravenous drug abuse, jugular vein catheterisation, neck dissection, a hypercoagulable state associated with malignancy, neck injury or ovarian overstimulation syndrome.[4]

In this paper, we present and discuss two cases of spontaneous jugular vein thrombosis, associated with breast and lung malignancies.

Case one

A 58-year-old woman presented with an approximately three-month history of a painful swelling on the left posterior triangle of the neck (Figure 1). Past medical history was unremarkable. The patient was a non-smoker.

On examination, the patient had a diffuse fullness of the left posterior neck, with a deep purple colour of the skin. A palpable mass in the thyroid gland was noted.

Routine laboratory investigations were mostly within normal limits: the haemoglobin level was 12.1 g/dl, white cell count 9800/mm^3 and platelet count 174 000/mm^3. However, the fibrinogen level was 4.0 g/l (normal range 1.8–3.5 g/l).

Magnetic resonance imaging (MRI) of the neck revealed diffuse neck swelling due to thrombosis of the IJV, surrounded by soft tissue oedema. There was a solid mass, about 3 cm in diameter, in the left posterior triangle and multiple nodules in the thyroid gland.

Fine needle aspiration (FNA) of the neck mass showed metastatic adenocarcinoma cells. The primary focus of the adenocarcinoma was a mass in the left breast. Ultrasonography and mammography revealed the radiological malignancy criterion of microcalcification in the breast (Figure 2). A biopsy confirmed an invasive ductal carcinoma.

The patient was heparinised and coumarinised using therapeutic doses for thrombosis resolution. She was then referred to our oncology clinic for treatment of her ductal carcinoma.

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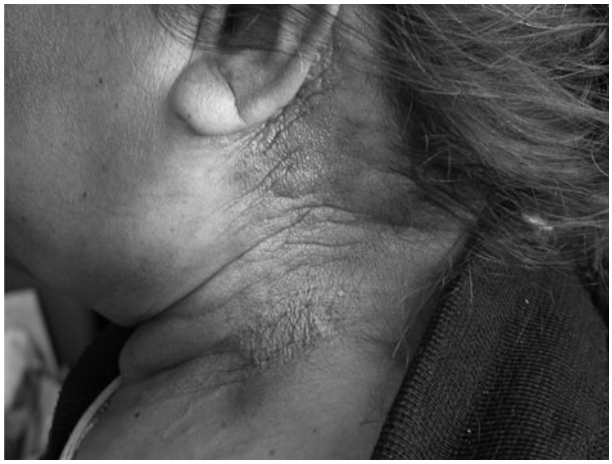

FIG. 1
Swelling and discoloration on the left posterior triangle of the neck.

Case two

A 46-year-old man was admitted with a sudden onset, painless mass on the left neck and a history of 15 days of progressive respiratory distress.

On examination, there was a diffuse, firm, approximately 10×10 cm mass in the left postero-lateral neck. The surface skin was hyperaemic.

A computed tomography (CT) scan of the neck and chest demonstrated a left IJV thrombosis, as well as a soft tissue mass on the left main bronchus and prominent lymphadenopathy in the mediastinum (Figures 3 and 4).

Fine needle aspiration cytology confirmed the diagnosis of large cell lung carcinoma.

The patient died the next day.

Discussion

Spontaneous IJV thrombosis is an extremely rare entity. Although uncommon, reports of venous thrombosis in the neck, arms and upper thoracic veins have increased, mostly in the intensive care setting. Since central venous catheters, haemodialysis and pacemakers are now used frequently, they are common causes of trauma to the IJV, subclavian vein and brachiocephalic vein, with subsequent thrombosis.[1] In our cases, there was no history of central venous catheterisation, haemodialysis or intensive care treatment.

Spontaneous thrombophlebitis may be the first manifestation of an occult malignancy. The possibility of Trousseau's syndrome should be considered and the patient appropriately investigated, taking into account the possibility that the neoplasm might be located in a distant site to the area of thrombosis. Over 100 years ago, Trousseau first reported that cancer patients had an increased incidence of coagulopathies.[1] Since Trousseau published these findings, thromboembolic disorders have been documented in association with a number of tumours, such as lung, pancreas, stomach and colon.[2] These clinical findings have been explained by laboratory studies identifying altered levels of blood clotting factors in the serum of cancer patients. Indeed, the presence of an unexplained deep venous thrombosis can be sufficient clinical indication to prompt screening for occult malignancy.

Mechanistically, it has been proposed that normal host cells, such as platelets, mononuclear phagocytes and smooth muscle cells, are responsible for activating procoagulant and angiogenic pathways.[2] Routine laboratory findings related to coagulopathy, such as prothrombin time and platelet count, were within normal ranges in our patients, except for the fibrinogen level. Thrombosis may have occurred for this reason.

Malignancy-induced thrombosis can be resolved during or after anti-neoplastic treatment. Radiotherapy can be an effective treatment for most malignant causes of superior vena cava (SVC) syndrome, and steroids and diuretics can be used for symptomatic relief. If the patient fails to respond, or the SVC obstruction recurs, a stent can be

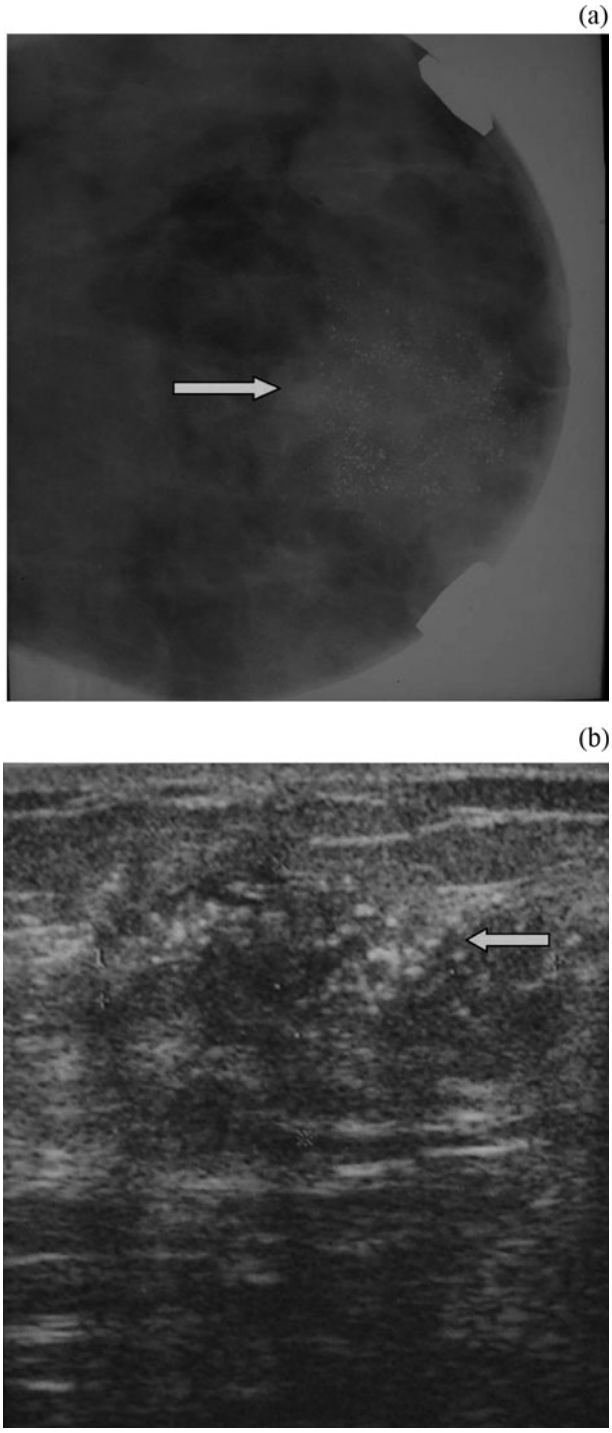

FIG. 2
(a) Mammography and (b) ultrasonography images; arrows indicate microcalcification, a criterion of malignancy.

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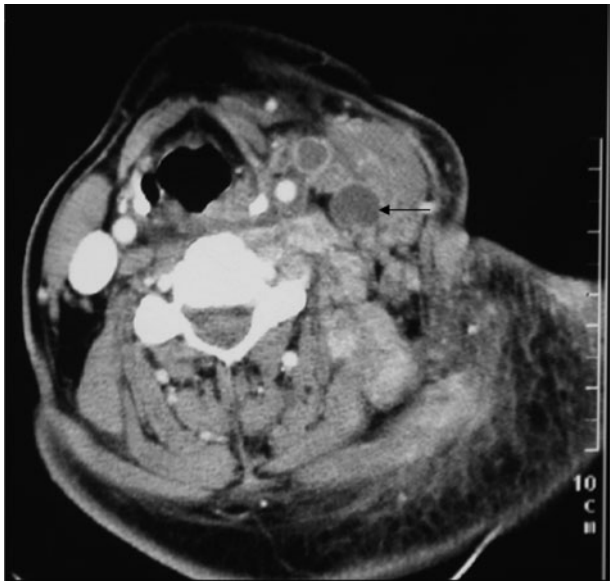

FIG. 3
Axial computed tomography scan; arrow shows the left internal jugular vein thrombosis.

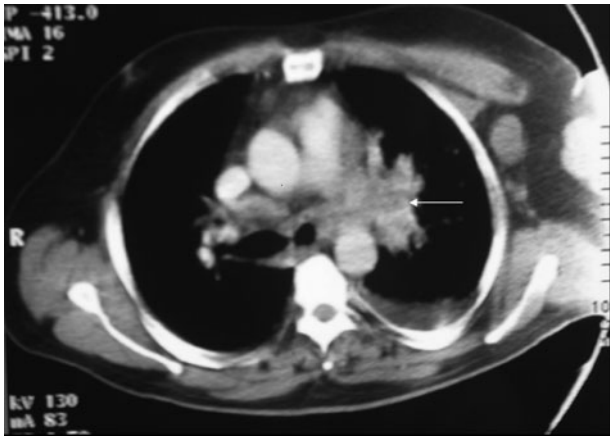

FIG. 4
Axial computed tomography scan; arrow shows the soft tissue mass on the left main bronchus.

used to bypass the obstruction.[1] In our series, the patient with invasive ductal carcinoma was treated with a chemotherapy protocol, and her IVJ thrombosis partially regressed.

The clinical presentations of IJV thrombosis include neck swelling, mass, neck tenderness and fever.[4] Venography is the 'gold standard' for the diagnosis of DVT, but it is an invasive technique with a small risk of allergic reaction or of itself causing venous thrombosis. Nowadays, non-invasive techniques such as ultrasonography are used more frequently. Ultrasonography is considered an excellent diagnostic method and shows an average sensitivity of 97 per cent.[1] However, its assessment of intrathoracic contents is less accurate than CT or MRI. Scanning with CT and MRI gives better definition of soft tissue structures and planes, and is therefore more useful in demonstrating the underlying cause of the thrombosis.[1] We thus investigated our patients with MRI and CT scanning.

- **The internal jugular vein is an uncommon site of spontaneous venous thrombosis**
- **This paper describes two cases of spontaneous jugular vein thrombosis associated with breast and lung malignancies**
- **The association of cancer and thrombophlebitis was first observed by Trousseau, and this association still bears his name**
- **Spontaneous thrombophlebitis may be the first manifestation of an occult malignancy. The possibility of Trousseau's syndrome should be considered in such cases**

The serious complications of IJV thrombosis include pulmonary embolism, septic emboli, generalised septicaemia, facial oedema and pseudotumour cerebri. The differential diagnosis includes cellulitis of the neck, neck space infection (such as Lemierre syndrome) and deep neck abscess.[4,5]

Conclusion

Spontaneous thrombophlebitis may be the first manifestation of an occult malignancy. The possibility of Trousseau's syndrome should be considered in such cases.

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