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Spontaneous internal jugular vein thrombosis and recurrent laryngeal nerve palsy: a rare simultaneous presentation of an occult malignant neoplasm

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Abstract
Internal jugular vein thrombosis is an uncommon potentially life-threatening disorder caused by various conditions. Non-spontaneous internal jugular vein thrombosis is an uncommon condition associated in the pre-antibiotic area with deep-neck infections. Currently iatrogenic trauma to the internal jugular vein from catheterisation and repeated intravenous injections by drug abusers are the leading causes of thrombosis. Spontaneous internal jugular vein thrombosis may occur when there are no apparent pre-disposing mechanical or inflammatory causes although a few of these patients may harbour an occult malignant neoplasm. Hence, careful investigation and follow-up are vital. Thrombosis in Trousseau's syndrome is usually confined to the vascular system of the extremities and the viscera. However, secondary to the paraneoplastic hypercoagulable state, thrombosis can occur in the large veins of the head and neck region. We understand this to be the first case where spontaneous internal jugular vein thrombosis and ipsilateral recurrent laryngeal nerve paralysis were the only initial manifestations of an occult malignancy.

Key words: Jugular veins; Thrombosis; Trousseau's syndrome; Recurrent laryngeal nerve paralysis

Introduction

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 injury (intimal), alterations of blood flow (stasis), and hypercoagulability of the blood (Robbins *et al.*, 1984). In the pre-antibiotics days internal jugular vein thrombosis was a well known complication associated with acute inflammatory conditions such as tonsillitis, peritonsillar abscess, mastoiditis and dental causes (Cohen *et al.*, 1985). However, since the introduction of antibiotics, the complication of internal jugular vein thrombosis secondary to acute infection has become quite rare although it is still occasionally reported (Yan and Norante, 1980).

Currently the major causes of internal jugular vein thrombosis involve direct iatrogenic trauma to the vein. These include jugular vein catheterisation and repeated injections into the large veins of the neck by intravenous drug abusers (Espritus and Media, 1980). Any associated malignancy, either known or occult, is another uncommon and not well documented aetiology for internal jugular vein thrombosis.

Armond Trousseau in the last century first decribed the syndrome of migratory thrombophlebitis associated with gastric carcinoma (Sack *et al.*, 1977). Studies since then have confirmed and expanded the definition of Trousseau's syndrome to encompass a broad spectrum of coagulation disorders associated with a wide variety of malignant tumours (Rickles and Edwards, 1982). Typically, Trousseau's syndrome is confined to the vascular system of the extremities and occasionally the viscera. Vascular thrombosis secondary to the syndrome presenting in the head and neck has been reported only rarely and is known to occur mainly intracranially. This article reports a rare extracranial presentation of thrombosis in the head and neck occurring simultaneously with unilateral recurrent laryngeal nerve palsy, these being the only initial manifestations of an occult malignancy.

Case report

A previously healthy 65-year-old man was referred with a six-week history of hoarseness of his voice. The symptom had come quite suddenly and he felt increasingly lethargic. He had no significant past medical history and was taking no medication. Physical examination revealed an afebrile, thinly built male with a diffuse, non-tender and doughy left midneck mass with supraclavicular fullness, mostly concealed under the left sternomastoid muscle. There was no obvious prominence of any superficial neck veins with no evidence of facial or retinal oedema.

Direct fibreoptic examination of the larynx and pharynx including postnasal space showed a left vocal fold palsy. Otorhinological examination and general examination was otherwise normal. Following admission to the hospital, routine examination including full blood count, platelet count, coagulation studies and a chest radiograph were all normal. Ultrasound scan suggested the possibility of thrombosis in the internal jugular vein. Fine needle aspiration cytology of the thrombosed jugular vein mass was undertaken which revealed well differentiated squamous cell carcinoma cells. Computed tomography from the skull base down to the pelvic inlet was carried out

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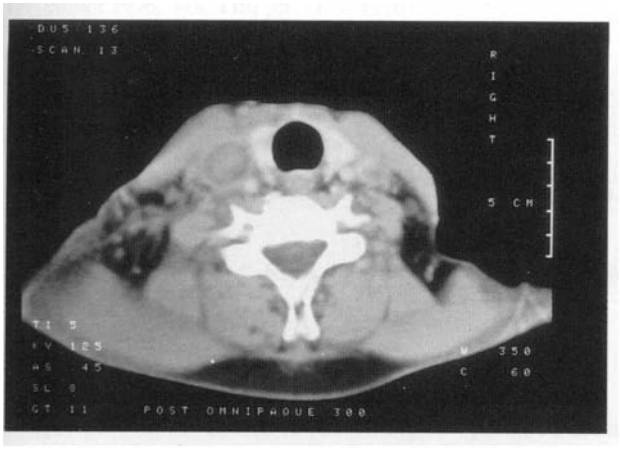

FIG. 1
CT scan showing complete occlusion by thrombosis of left internal jugular vein.

(Figure 1). This confirmed the ultrasound findings of complete thrombosis of the left internal jugular vein with patent subclavian and innominate veins. He was referred to a general physician who found no neurological deficit. Anticoagulation with heparin was commenced. He underwent panendoscopy including bronchoscopy plus washings but no primary lesion was located. A few weeks later he developed cervical lymphadenopathy on the left side distant from the original fine needle aspiration site. This was confirmed as squamous cell carcinoma by repeat fine needle aspiration cytology. His condition deteriorated and though he was maintained on heparin, no further advice management was undertaken except palliative hospice care. He died eight weeks after the initial consultation. His family declined the offer of a post mortem examination.

Discussion

The veins in the head and neck even in the presence of localised disease appear to be less susceptible to thrombosis than those of the extremities and viscera. The structure and location of head and neck veins makes them less pre-disposed to stasis especially as they are mostly valveless. They have an elastic wall, allowing constant collapse and expansion by the action of respiration and pump effect of the heart and gravity aids its central emptying in the upright position (Philip *et al.*, 1985).

In the pre-antibiotic era intimal damage resulting in acute septic thrombophlebitis of the internal jugular vein would result in the dramatic appearance of facial and eyelid swelling, with pitting oedema of the mastoid skin but is highly unlikely to occur today (Goodman, 1917; Cohen *et al.*, 1985).

With catheterisation of the jugular veins and resulting intimal damage becoming a commonplace procedure for different purposes including total parenteral nutrition, the incidence of thrombosis in the internal jugular vein is increasing (Ahmed and Payne, 1976; Chasetre, 1982).

Intravenous drug use is also a well known cause of internal jugular vein thrombosis due to intimal damage. This is as a result of direct injection into the internal jugular vein, the drug abuser having exhausted the peripheral veins (Espritus and Media, 1980).

A high degree of suspicion is necessary to reach the diagnosis of spontaneous internal jugular vein thrombosis when there is no history suggestive of any obvious head and neck infection, any central venous catheterisation or intravenous drug abuse. Hence diagnosis of internal jugular vein thrombosis should be included in differential diagnosis of any painful neck swelling.

The aetiology of spontaneous internal jugular vein thrombosis not associated with stasis or intimal damage brings in the possibility that hypercoagulability is present with a postulated increase in levels of factor 8 and increased production of thromboplastin. Leiberman and his co-workers, (Lieberman *et al.*, 1961) after studying the co-relation between thrombophlebitis and malignancy in a group of patients concluded that it tends to be recurrent or migratory, more resistant to anticoagulation and may quite often precede the diagnosis of neoplasia. They also found that any primary site was possibly associated although it was seen more frequently in neoplasms of the lung, pancreas and female reproductive system. In approximately 50 per cent of the total number of 81 patients in their study, the carcinoma was discovered between two and six months after initial finding of thrombophlebitis while in six patients it was almost a year later.

Isolated spontaneous thrombosis of the larger veins in the neck may thus be the first indication that an occult neoplasm is present. As a result, every patient with spontaneous thrombophlebitis must undergo a careful history, a complete physical examination and a thorough investigation to avoid missing or delaying the diagnosis of a hidden malignancy. This would include full blood count, prothrombin time, partial thromboplastin time and chest X-ray. Radiological investigations with ultrasound, CT scan, MRI scan should be guided by clinical suspicion. The treatment of internal jugular vein thrombosis currently includes appropriate antibiotics and anticoagulation, with exceptional cases subjected to surgery. Anticoagulant therapy largely avoids the risk of pulmonary embolism. Spontaneous internal jugular vein thrombosis especially those associated with malignancies, respond to heparin but are resistant to oral anticoagulants (Liebermann *et al.*, 1961). Patients with spontaneous internal jugular vein thrombosis may thus need to be on long-term heparin therapy.

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