Internal Jugular Vein Thrombosis: A Case Study And Review Of The Literature
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Citation

Abstract
Internal jugular vein (IJV) thrombosis, associated with suppurative infection of the upper aero-digestive tract was first described a century ago. Although the true incidence of IJV thrombosis is unknown it appears to be increasing. The complications associated with IJV thrombosis are potentially lethal and it is therefore important to recognise and treat this condition early. This article describes a case of infective IJV thrombosis secondary to chronic otitis media in a nine year old girl and reviews the literature.

CASE STUDY
An eight-year-old girl presented to our department with a longstanding history of left sided otorrhoea and a five-day history of pyrexia and otalgia. On examination her temperature was 39.8°C and her left ear was filled with mucopus. Examination of her left ear revealed a subtotal perforation of the tympanic membrane with no evidence of cholesteatoma. There was slight tenderness over her mastoid bone with no evidence of a mastoid abscess. Her facial nerve function was intact. An audiogram revealed a 60 dB conductive hearing loss in the left ear and normal hearing in the right ear.

She was admitted to hospital and treated with intravenous Ampicillin and Metronidazole. After three days she still had persistent high temperatures and developed a post-auricular mastoid abscess. A CT scan was performed which showed an opacified mastoid bone and a possible sigmoid sinus thrombosis. There was no evidence of an intra-cranial collection.

A modified radical mastoidectomy was performed which revealed a large amount of granulation tissue in the middle ear and mastoid. The sigmoid sinus was exposed and revealed an infected thrombus which was evacuated. Specimens were sent for histology and culture.

Over the following five days she continued to spike high temperatures and developed neck pain. Examination revealed a tender, cord-like mass on the left side of her neck. An ultrasound scan of her neck revealed a thrombosed left internal jugular vein. The specimens sent previously during mastoid surgery cultured Pseudomonas sensitive to Gentamicin and Amikacin. Because of the positive culture result and the jugular vein thrombus we added intravenous Amikacin to her antibiotic therapy.

Over the next two days the patient continued to spike high temperatures and developed a productive cough. Chest X-ray revealed an abscess and infiltrate in the right lung. (Figs 1, 2) CT scan of her chest demonstrated a right middle lobe lung abscess, a loculated empyema in the posterior base of the right lung and multiple calcified hilar lymph nodes suggestive of previous tuberculosis. (Fig 3)

Figure 1
The thoracic surgeons performed a thoracotomy to evacuate the loculated empyema and drain the lung abscess. Biopsies and microbiology specimens from the empyema revealed Pseudomonas. After long discussion we decided to start the patient on low molecular weight heparin. Over the next six days her temperature settled and she made a full recovery.

**DISCUSSION**

Internal jugular vein (IJV) thrombosis refers to an intraluminal thrombus occurring anywhere from the origin of the IJV in the cranium, down to where it joins the subclavian vein to form the brachiocephalic vein. IJV thrombosis associated with suppurative infection of the upper aerodigestive tract was first described at the beginning of the twentieth century by Courmont, Goodman and Mosher [1,2,3].

In 1936 Lemierre defined it further. He described Lemierre syndrome as an infected IJV thrombus, caused by extension of oropharyngeal infection [4]. Today it is also known as Necrobacillosis or post-anginal septicaemia [5].

The incidence of IJV thrombosis is still unknown. What we do know is that 66% of patients with IJV catheters have proof of thrombus formation on ultrasound or at autopsy, and that up to a third of patients following a neck dissection will have a thrombus in the IJV [6,7]. Harada et al showed that the most significant narrowing of the IJV following neck dissection occurs in the first week after surgery and that patency is gradually restored within three months [8]. The incidence appears to be increasing secondary to the increased use of central venous catheters placed in the IJV and subclavian vein, and because of the increased use of the IJV by intravenous drug abusers [9].

Etiological factors can best be described in relation to Virchow’s triad. Any factor causing endothelial damage, alteration in blood flow or hypercoagulable state can lead to IJV thrombosis. IJV catheters can potentiate thrombus formation by causing endothelial damage during insertion, or the catheter itself can act as a nidus for clot formation [6]. The mechanism of thrombus formation in oropharyngeal infections is likely to be the result of systemic hypercoagulability (caused or exacerbated by infection), venous stasis (from vessel occlusion by the infectious process of inflammation) and endothelial damage (via direct endovascular invasion by microbes or through perivascular inflammation) [10]. IJV thrombus with otitis media or mastoiditis is as a result of progression of sigmoid sinus thrombosis. Endothelial damage and introduction of infection are the main causative factors of thrombosis in intravenous drug abusers. Malignancy potentiates thrombus formation by causing direct compression from tumour or nodes and by causing a hypercoagulable state.

Organisms involved in septic IJV thrombosis are determined
by the etiology. In intravenous catheter-related IJV, the most likely organisms are gram positives with Staphylococcus Aureus the most common. If oral cavity sepsis is the causative factor, Anaerobes are the most common. Gram negative organisms, especially Proteus and Pseudomonas are the most common organisms in otologic sepsis.

Clinical manifestations of IJV thrombosis depend on whether it is infected (complicated) or not infected (uncomplicated). Uncomplicated cases present with pain and swelling in the neck, and a cord can be palpated beneath the sternocleidomastoid muscle. Tovi et al described the following clinical manifestation in a large series of patients with septic IJV thrombosis: fever (83%), leucocytosis (78%), cervical pain (66%), neck swelling (72%), cord sign (39%), sepsis syndrome (39%), pleuro-pulmonary complications (28%), superior vena cava syndrome (11%), chylothorax (5%) and jugular foramen syndrome (6%).[11]

Laboratory investigations required in patients with IJV thrombosis depend on whether the thrombus is infected (complicated) or not infected (uncomplicated). Patients with complicated IJV thrombosis require culture from the infection source (oropharynx, ears, catheter tip etc.) and blood cultures. Uncomplicated IJV thrombosis requires more in-depth investigations, for example protein C, S, antithrombin-3-deficiency tests and DIC screen which includes prothrombin time (PT), activated partial thromboplastin time (APTT), fibrin split products and fibrinogen.

Imaging studies useful in IJV thrombosis include contrasted computed tomography (CT) scan, magnetic resonance imaging (MRI), nuclear medicine scan, ultrasound and contrast venogram.

Contrast venogram used to be the gold standard, but because it is invasive and may dislodge thrombus and cause an embolus, it has lost favour. Ultrasound scan is safe, non-invasive, and cost-effective and a doppler can detect flow rate. Unfortunately it is sub-optimal for detecting thrombosis deep to the mandible and clavicle.

CT scan with intravenous contrast is considered by many to be the study of choice. CT scan findings include identification of a low-density intraluminal thrombus, a sharply defined bright vessel wall (because of contrast uptake by vasa vasorum), soft tissue swelling surrounding the IJV and a distended IJV proximal to the thrombus.

MRI provides better soft tissue definition and sensitivity to blood flow rates compared to CT scanning and does not require exposure to contrast material or radiation.[12].

Nuclear medicine scanning has high false positive rates and prolonged study time. Furthermore the studies have to be performed in a nuclear medicine area which means that a patient, who is often critically ill, has to be transported to undergo the investigation.

Medical treatment of patients with IJV thrombosis depends on whether the thrombus is infected or not. In non-infected thrombosis the causative factor, for example an IJV catheter, should be removed first. The role of anti-coagulation therapy is controversial. There are insufficient studies to guide physicians and the fact that it is a fairly under-diagnosed condition with very few serious complications raises the question whether anti-coagulation therapy is necessary. As regards thrombolytic therapy, only a few isolated case series exist and the safety has not yet been established.[13].

In infected IJV thrombosis the primary site of infection should be treated first, for example neck abscesses should be drained and mastoidectomy done for mastoiditis. Most patients with infected IJV thrombosis will do well on antibiotics alone. The choice of antibiotic will depend on the causative factor. If the thrombus was caused by an indwelling catheter the most likely organism will be gram positive and Vancomycin should be used until the organism is isolated and its sensitivity established.[14]. In case of a primary ear infection the patient should be treated with Amikacin to cover gram negative organisms until the organism and its sensitivity has been established.[15]. In case of an oropharyngeal infection the patient should be started on a broad spectrum antibiotic with anaerobic cover until the organism and its sensitivity has been determined. It is important that all patients with infected thrombosis receive a total of four to six weeks of antibiotic therapy.[16].

The role of systemic anti-coagulation in an infected thrombus has not been established. It should only be considered if there is clot propagation or septic emboli. It is important to keep in mind that there is a risk of bleeding and expansile haematoma with potential airway compromise.

Surgery should be reserved for complicated cases only. The indications for surgery include associated deep space neck infections, carotid sheath involvement (to prevent extension into the carotid artery), intra-luminal abscess and failed medical treatment. It must be remembered that response to
medical treatment might be slow. A number of procedures have been described in the literature and include drainage of collections, debridement of necrotic tissue and even ligation or excision of the IJV.

IJV thrombosis may be associated with complications like pulmonary embolism, subclavian vein thrombosis, superior sagittal sinus thrombosis, superior vena cava syndrome, pseudotumor cerebri and laryngeal and lower airway oedema. Infected thrombophlebitis can cause complications like systemic sepsis syndrome, septic emboli to lungs, liver, spleen, brain, skin, muscle, and bone marrow, empyema, septic arthritis, renal failure, hepatic dysfunction and cerebral oedema [17].

Lemierre syndrome had a mortality exceeding fifty percent in the pre-antibiotic era. The mortality today is difficult to determine because most patients have multi-system involvement and a concurrent critical illness. This makes the contribution of the thrombosis itself to mortality difficult to determine.

CONCLUSION

When recognized early and treated with appropriate aggressive medical and surgical therapy, death from jugular vein thrombosis is uncommon today. By being aware of jugular vein thrombosis, the physician can be more vigilant for potential complications and diagnose and treat them earlier.

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