CASE REPORT

Variant of Lemierre's syndrome with internal jugular vein aneurysm

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Accepted 11 December 2017

SUMMARY

Internal jugular vein (IJV) aneurysm is a rare entity, and a thrombosed aneurysm poses diagnostic and management challenges. We came across a 53-year-old woman who presented with fever, vomiting and right neck swelling for a week. Laboratory investigations showed neutrophilic leucocytosis, raised acute phase reactant and blood culture yielded *Klebsiella pneumoniae*. Ultrasound and contrast-enhanced CT neck revealed a large fusiform aneurysm of the right IJV with filling defect extending from the aneurysm into the right transverse sinus. There was a cavity at the right lower third molar tooth. MRI confirmed the findings with additional enhancing focus at right lower periodontal region. The swelling reduced after 2 weeks of medical therapy, and follow-up imaging 4 months later showed complete resolution of the aneurysm with residual thrombosis. After extensive workup, dental infection remains the only identifiable primary source leading to thrombophlebitis of the right IJV and subsequent sequelae.

BACKGROUND

Neck abscess is often considered when patients with septicaemia present with progressive neck swelling. A thrombosed internal jugular vein (IJV) aneurysm is not commonly encountered and the diagnosis can only be confidently established on cross-sectional imaging. We intend to highlight not just the rare nature of the disease but also the thought processes and clinical decisions that led to the diagnosis and subsequent management.

CASE PRESENTATION

A previously healthy 53-year-old Indian woman presented with a painful right neck swelling for 1 week, associated with intermittent fever and chills. She also complained of dizziness and right ear tinnitus, along with a few episodes of vomiting. Toothache was reported 2 weeks prior to the onset of neck swelling but resolved spontaneously. Otherwise, she denied sore throat, productive cough, shortness of breath, chest pain, headache or other focal neurological symptoms.

On examination, she was afebrile, normotensive with no tachycardia. Random plasma glucose was 10.8 mmol/L and oxygen saturation was 97% on room air. She was alert but appeared lethargic. There was a 5×4 cm firm, non-fluctuant tender swelling over the right submandibular region which was warm without overlying erythema.

There was no palpable lymphadenopathy, no sign of tonsillitis and uvula was not deviated. Fiberoptic nasopharyngolaryngoscope revealed normal vocal cords. Otoloscopic examination showed normal tympanic membranes. Oral hygiene was generally poor and chronic generalised periodontitis was noted on dental assessment. Cardiovascular, respiratory and cranial nerves examination were grossly normal.

INVESTIGATIONS

Initial laboratory investigations showed leucocytosis (white cell count $23.4 \times 10^9/L$ with 92% neutrophil predominance), hypochromic microcytic anaemia (haemoglobin 84 g/dL, mean cell volume 76.9 fL, mean cell haemoglobin 24.4 pg, mean cell haemoglobin concentration 318 g/dL) and raised acute phase reactants (C-reactive protein 155.6 mg/L, erythrocyte sedimentation rate 120 mm/1 hour). Blood culture and sensitivity yielded *Klebsiella pneumoniae* which was sensitive to amoxicillin/clavulanic acid, cotrimoxazole, cefuroxime and ampicillin/sulbactam.

She was diagnosed with type II diabetes mellitus with a fasting plasma glucose of 15 mmol/L and HbA1c of 10.8%. There was no proteinuria.

Ultrasound neck showed a collection with mobile debris with epicentre at the right angle of mandible. The right IJV inferior to collection is patent but there were no demonstrable Doppler signals at the level of the collection. Contrast-enhanced CT neck revealed a large fusiform aneurysm (size 4.2 cm (W) × 4 cm (AP) × 5.6 cm (CC)) of the right IJV with filling defect extending from the aneurysm into the right transverse sinus (figure 1). There was a cavity at the right lower third molar tooth. Contrast-enhanced MRI confirmed the filling defect and aneurysm. However, the filling defects were T1 hypointense and T2 hyperintense with no blooming artefact on gradient echo sequences. There was an additional enhancing focus at right lower molar periodontal region (figure 2). Both CT and MRI showed peripheral enhancement surrounding the aneurysm and thrombosed vein. Overall features were consistent with infected thrombus.

Chest radiograph and abdominal ultrasound scan did not reveal any focal infection or evidence of malignancy.

Echocardiogram revealed normal chamber size, mild mitral and aortic regurgitation with ejection fraction of 65%. There was no intracardiac thrombus or pericardial effusion.

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To cite: Chua SH, Ong SCL, Liew YH. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-223371

Figure 1 Contrast-enhanced CT neck at sagittal (A), coronal (B) and axial (C) sections showed a fusiform aneurysm at the right internal jugular vein (asterisk) with filling defects extending into the right transverse sinus (arrow). There is peripheral enhancement surrounding the aneurysm and thrombosed vein.

DIFFERENTIAL DIAGNOSIS

1. Neck abscess—difficult to differentiate from IJV aneurysm and thrombosis on clinical grounds.
2. *K. pneumoniae*-associated Lemierre's syndrome—thrombophlebitis, thrombosis and resultant mycotic aneurysm of the IJV.
3. Primary IJV aneurysm complicated with infection and thrombosis—due to fusiform appearance of the aneurysm.

TREATMENT

With the initial impression of right neck abscess, patient was commenced on intravenous ceftriaxone and intravenous metronidazole. In view of extensive IJV thrombosis and aneurysm as evidenced on the CT scan, subcutaneous enoxaparin (1 mg/kg twice daily) was commenced on day 2 of admission. Patient also had dental extraction done on the right third upper and lower third molar teeth by the dental team.

Antibiotic was later switched to amoxicillin/clavulanic acid. The neck swelling started to improve after 2 weeks of antibiotic treatment and dizziness subsided completely. Inflammatory markers were reducing in trend.

In total, patient received 4 weeks of antibiotic. She was discharged on day 29 with subcutaneous enoxaparin, planned for 6 months. Enoxaparin was the choice of anticoagulant as patient was resistant to warfarin with persistent subtherapeutic international normalised ratio despite 70 mg of warfarin per week.

Figure 2 MRI of the internal jugular vein aneurysm (asterisks) at axial sections in gradient echo sequence (A) showed no blooming artefact. Contrast-enhanced MRI (B) showed peripheral enhancement of the aneurysm with central non-enhancing component. There is also enhancement at the right lower molar periodontal region (arrow).

Figure 3 Contrast-enhanced CT neck 4 months later at sagittal (A), coronal (B) and axial (C) sections showed completely resolved internal jugular vein (IJV) aneurysm with residual thrombosis of the IJV. There is still filling defect within the right transverse sinus (arrow).

OUTCOME AND FOLLOW-UP

Patient remained asymptomatic and the neck swelling has completely resolved 4 months later. Repeated inflammatory markers were normal. Follow-up CT scan and MRI showed complete resolution of the aneurysm with residual thrombosis in the right IJV, sigmoid sinus and transverse sinus (figure 3). Signal characteristics of the thrombosis on MRI were identical to the initial scan. Previously seen enhancement at right lower periodontal region has also resolved.

Subsequent laboratory tests to rule out secondary causes of IJV thrombosis showed subclinical hypothyroidism (thyroid stimulating hormone 7.29 mIU/L, free T4 13 pmol/L), negative anti-nuclear antibodies, normal tumour markers (Ca-125, CA19-9, alpha-fetoprotein and carcinoembryonic antigen) and negative anticardiolipin antibody, anti- β 2-glycoprotein 1 antibodies and lupus anticoagulant.

We initially planned for a total of 6 months anticoagulation in view of persistent thrombosis with intracranial extension but patient defaulted treatment after 4 months of subcutaneous enoxaparin injection. Follow-up CT scan at 6 months showed no significant interval change and anticoagulation therapy was stopped.

DISCUSSION

Venous aneurysms are not often discussed in the list of differential diagnoses of neck swelling. IJV aneurysms are rare congenital being mostly reported in the paediatric literature. This exhibit intends to highlight the challenges in distinguishing a thrombosed IJV aneurysm from neck abscess on clinical grounds and in formulating an optimal management plan.

Venous aneurysm can be regarded as a focal venous dilatation that communicates with a main venous structure by a single channel without arteriovenous malformation or a pseudoaneurysm.¹ Macroscopically, true aneurysm is described as fusiform and pseudoaneurysm as saccular.² The former involving IJV is thought to be congenital and has fewer predilections for thrombosis.^{2,3} According to Calligaro *et al*,⁴ the natural history of IJV aneurysms are benign. In our case, however, there was little evidence to suggest that the IJV aneurysm was congenital, in spite of its fusiform appearance. Sudden progressive neck swelling with septicaemia and complete resolution of aneurysm following treatment, in addition to atypical MRI signals of the filling defects suggest that it was most likely an acquired condition.

After extensive workup, the seemingly trivial dental infection remains the only identifiable primary source leading to thrombophlebitis of the right IJV and subsequent sequelae. We notice a great similarity with nine other cases of IJV thrombosis (though none with aneurysm), diagnosed as *K. pneumoniae*-associated Lemierre's

syndrome.⁵ Interestingly, eight out of nine patients were diabetic. We can infer that there is a possible correlation between IJV thrombosis, *K. pneumoniae* infection and diabetes mellitus. There is impaired phagocytosis of neutrophils due to hyperglycaemia in diabetic patients leading to increased risk of infections caused by *K. pneumoniae*, especially the capsular serotype K1 and K2 strain.⁶

According to Gbaguidi *et al*,⁷ median duration of anticoagulant therapy in IJV thrombosis from any cause was 6 months. Seventy-seven per cent of patients had echographic persistent thrombosis at 3–6 months' follow-up. For IJV thrombosis due to Lemierre's syndrome, however, many had reported successful treatment with or without anticoagulant in conjunction with antimicrobial therapy. IJV thrombosis may also persist long after resolution of infection, ranging from 4 weeks to 6 months.⁸ Anticoagulation for at least 3 months is recommended in the American College of Chest Physicians 2012 guidelines to reduce recurrent thromboembolism in patients with proximal deep vein thrombosis. Prolonged anticoagulation therapy beyond 3 months is not recommended in patient with upper extremity deep vein thrombosis (DVT) in view of substantial lower risk of

recurrence thromboembolism compared with those with proximal leg DVT.⁹

Contributors The authors SHC, SCLO and YHL have contributed sufficiently to the project and have met all criteria to be included as authors.

Disclaimer All authors have no relevant financial interest in this manuscript and no activities, affiliations or relationships to disclose. All authors have not published or submitted any related papers from the same study.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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Learning points

- To be aware of internal jugular vein (IJV) thrombosis and aneurysm as a differential diagnosis in neck swelling.
- There is a strong possible association between Lemierre's syndrome, *Klebsiella pneumoniae* and diabetes mellitus.
- Management of IJV thrombosis should focus on treating the underlying cause.
- Duration of anticoagulant therapy should be guided by the risk of thromboembolism rather than resolution of thrombosis.

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