CASE REPORT

Isolated non-hereditary angioneurotic oedema of uvula (Quincke's disease) in an adolescent

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SUMMARY

A 19-year-old primipara woman was admitted in labour with positive vaginal swab for group B *Streptococcus* and given benzylpenicillin and ranitidine. She used Entonox for pain relief. She received bupivacaine in the epidural catheter and shortly after developed sensation of foreign body in her throat with muffled voice and isolated angioneurotic oedema of the uvula. She responded well to H1 and H2 histamine blockers and steroids and epinephrine were kept as standby. She recovered well soon after delivery. Penicillin, Entonox or bupivacaine seemed possible cause of the Quincke's disease. Hereditary form was ruled out by normal creatine kinase and reaction to bupivacaine by C1 esterase inhibitor assay. It can be associated with spectrum of anaphylactic reactions and the resuscitation team should be alerted. Airway maintenance should be the primary management strategy. Most cases respond to observation, oxygen therapy and antihistamines with or without steroids. Epinephrine may be required occasionally.

BACKGROUND

Isolated non-hereditary angioneurotic oedema of uvula (Quincke's disease) in a pregnant woman during labour is uncommon and has not been reported.^{1–14} It is a rare clinical entity and in majority of the patients, the exact aetiology is unknown.^{1–2} Quincke³ first described angio-oedema in the medical literature in 1882 and since that time, isolated angio-oedema of uvula has been known as Quincke's disease. We encountered a case recently in a primigravida patient in labour and our experience has been presented.

CASE PRESENTATION

A 19-year-old primigravida patient at 39 weeks gestation was admitted to birth centre with labour pain. Her antenatal period was uneventful except she was group B *Streptococcus* positive found on vaginal swabs. There are no known drug allergies on her history.

On admission, temperature was 37.7°C, pulse 104 bpm, blood pressure 131/87 mm Hg and respiration was 18/min. Her body mass index was 24. Vaginal examination showed cervical dilation about 3–4 cm with intact membrane. Membrane ruptured spontaneously at 21:05 and clear liquid drained. She reported feeling sick and vomited four times. Epidural catheter was sited around midnight. She was still having labour pain and epidural top up was given at 00:45 with levobupivacaine without fentanyl.

About 5 min later, she felt something stuck in the back of her throat. She denied shortness of breath, difficulty in breathing and itchiness. On examination, temperature was 36.7°C, pulse 101 bpm, blood pressure 152/91 and there were no clinical signs of respiratory distress or airway obstruction.

Local examination revealed grossly swollen isolated uvula with normal tonsils and normal epiglottis. Chest examination was normal with bilateral air entry with no audible wheeze. The impression was allergic reaction to penicillin, nitrous oxide or bupivacaine. On detailed enquiry, she disclosed having skin rash with penicillin in her childhood. There is also significant family history of penicillin allergy in her mother. She did not recall any history of a similar episode and no family history of angio-oedema.

INVESTIGATIONS

Cardiotocograph showed baseline pulse rate of 140 bpm, variability >5 bpm, accelerations present, with no decelerations. It was classified as normal. Urinalysis was normal and full blood count showed haemoglobin 134 g/L, white cell count 16×10⁹/L, neutrophils 13.1×10⁹/L and platelets 184×10⁹/L. Her blood group was B negative. High vaginal swab following postcoital bleeding taken 3 days ago isolated group-B streptococci, which were sensitive to erythromycin and penicillin. Creatine kinase level was 86 U/L (<165), aspartate aminotransferase 40 U/L (<40) and C1 esterase inhibitor level was 0.21 g/L (0.15–0.35), functional C1 inhibitor activity 85% (40–150), complement C3 1.26 g/L (0.75–1.65), complement C4 0.30 g/L (0.14–0.54).

DIFFERENTIAL DIAGNOSIS

A differential diagnosis of isolated uvular angioneurotic oedema secondary to adverse effect of one of the three drugs, namely, penicillin, nitrous oxide or bupivacaine used in that order was considered.

TREATMENT

She received penicillin G 3 g intravenously at 21:30. Entonox was given at 21:45 and she was given ranitidine 150 mg and paracetamol 1 g. Piriton 10 mg intravenously given at 01:08. Dexamethasone, epinephrine and resuscitation trolley were kept on standby in the room.

OUTCOME AND FOLLOW-UP

Her labour progressed well and she was fully dilated at 1:35 and delivered a live male infant in good condition at 2:11. Entonox and epidural catheter was discontinued. She was observed closely in the postnatal period.

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The uvula swelling was found to be settling down within a few hours after delivery and her symptoms improved. She was reviewed by anaesthetic team in the morning and decision for steroids was deferred as she made a significant recovery. Uvula oedema had resolved almost completely and she was discharged from the hospital the same day in the evening.

DISCUSSION

Isolated oedema of uvula is a rare occurrence and it usually manifests as fullness of the oropharynx and difficulty in speaking. It can be associated with difficulty in breathing as it affects the vocal cords.³

The aetiology of isolated angio-oedema of the uvula is usually due to type 1 hypersensitivity reaction and as its name indicates isolated angio-oedema of uvula, it is not associated with other hypersensitivity reactions such as pruritus, skin rash, hypotension or tachycardia. It is usually localised, recurrent, non-pruritic subcutaneous swelling that usually appears rapidly and resolves spontaneously.^{4,5}

Following are the possible causes of recurrent oedema of the uvula^{6,7}: idiopathic (commonest cause), hereditary (C1 esterase inhibitor deficiency), trauma (eg, oropharyngeal intubation), infections (group B streptococci, *Haemophilus influenzae* type B, *Candida* and anaerobic aetiological agents), allergy (airborne, food and drugs), medications and drugs (non-steroidal anti-inflammatory drugs (NSAIDs), ACE inhibitors, cannabis, glucosamine sulfate, ipratropium bromide), snoring and obstructive sleep apnoea.

In our case, there are three possible agents responsible for isolated oedema of the uvula: benzylpenicillin, Entonox or bupivacaine. Penicillin was given about 5 h before the occurrence and, therefore, penicillin allergy is unlikely for isolated uvula oedema.

The patient was using Entonox continuously for 4 h for pain relief. There is a possibility of oxidation of nitrous oxide to nitric oxide by hydrochloric acid due to vomiting which she had and forceful aspiration of Entonox. Nitric acid is a potent vasodilator and might cause angio-oedema of uvula. There is no case report about isolated uvula oedema due to Entonox or penicillin. Local anaesthetics are known to be the cause of uvula angio-oedema in patients known to have C1 esterase enzyme deficiency.^{8,9}

In our case, bupivacaine may be responsible as symptoms develop about 5–10 min after epidural catheter was sited and top-up given. However, the laboratory tests have shown C1 esterase and other related enzymes and complements to be completely normal negating the possibility of bupivacaine as the agent responsible. Therefore, either penicillin or Entonox seems to be responsible for this effect.

Infective cause was unlikely in our case as tonsils and epiglottitis were normal on examination and bloods shows normal inflammatory markers.

If type I hypersensitivity was thought to be a cause, it would be usual practice to check mast cell tryptase immediately (as a baseline), then at 1–2 h and 24 h.¹⁵ It is worth mentioning that as this had not been done initially in our case, it did not seem instructive to check levels several hours after the reaction.

In management of uvula angio-oedema, airway maintenance remains the primary management strategy. In emergency situations, intravenous H1 and H2 histamine blockers, corticosteroids

and, infrequently, epinephrine may be required. The on-call anaesthetist team and resuscitation trolley should be kept handy at all times. Sometimes, very rarely invasive techniques of securing airways in the form of needle decompression and uvulectomy should be performed by specialist head and neck surgeons experienced in the procedure. Overall prognosis is good and recurrence rate is low.^{12–14}

Learning points

- For all medical professionals to be aware of this rare occurrence of isolated angio-oedema of uvula.
- Isolated angio-oedema of uvula can be associated with a spectrum of anaphylactic reactions and resuscitation team should be alerted.
- Airway maintenance should be the primary management strategy at all times.
- Most cases respond to continuous clinical observation, oxygen therapy and H1 and H2 blockers antihistamines, with or without steroids.
- Epinephrine may be required occasionally and surgical airway management is rarely required.

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