Laryngeal Gout: An Extremely Rare Cause of Life-Threatening Upper Airway Obstruction Required Emergency Tracheostomy

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Abstract

Gout is a chronic and systemic disease due to purine metabolism disorder and reduction of uric acid excretion that may involves multiple parts of the body. However, laryngeal involvement is extremely rare with only 18 cases reported worldwide. The patient may present with life-threatening upper airway obstruction and need surgical airway management. The history of chronic gouty arthritis with multiple joints involvement, together with laryngeal symptoms should raise the suspicious of laryngeal gout. Diagnosis is based on clinicopathological features, while radiology findings are non-specific. The treatment of laryngeal gout involved combination of surgery and long-term urate lowering therapy. It important to treat patient as a whole which required multidisciplinary team approach to manage the local symptoms, systemic derangement, rehabilitation and psychosocial well-being. We present a case of chronic gouty arthritis with multiple joints involvement presented with acute respiratory distress which required emergency tracheostomy under local anaesthesia.

Keywords: Laryngeal gout, gouty tophi, tracheostomy, urate lowering therapy

Introduction

Gout is a chronic and systemic disease caused by purine metabolism disorder and reduction of uric acid excretion, which result in hyperuricemia. Long term hyperuricemia leads to deposition of monosodium urate crystals in and around the joints, most commonly at first metatarsophalangeal joint.[1] Without treatment, gouty tophi may form when the serum monosodium urate levels exceed the saturation point (>7.0 mg/dL).[2] It affected 1-4% of worldwide population, more prevalence in male and increases with age.[3]

Involvement of the larynx is extremely rare with only 18 cases reported worldwide up to date.[2,4,5] None of these cases required emergency tracheostomy under local anaesthesia. The management of laryngeal gout is challenging due to its rarity and involvement of the airway. The patients may have significant morbidity secondary to disturbance in respiration, speech, and swallowing and risk of aspiration. The treatment usually needs multidisciplinary approach, surgical removal of gouty tophi, long-term urate lowering therapy, maintain patency of the airway and speech and swallowing rehabilitation.

Case Report

A 41-year-old male with underlying chronic gouty arthritis presented with six months history of progressive worsening of shortness of breath, hoarseness, dysphagia, and anterior neck swelling. He had noisy breathing and odynophagia for the past one week. There were no aspiration symptoms, neck trauma or recent upper respiratory tract infection. He had chronic multiple joints pain, swelling and deformities and was diagnosed with gouty arthritis 7 years ago. He was started on allopurinol (a purine base analogue xanthine oxidase inhibitor) however developed allergic reaction to the medication and subsequently defaulted follow up. He became partially

dependent, and wheelchair bound for the past 3 years due to the severe deformities of the bilateral upper and lower limb joints secondary to the chronic gouty arthritis. Patient denied history of smoking and alcohol consumption.

On examination, patient was tachypnoeic with respiratory rate of 23 breaths per minute and present of hoarseness, inspiratory stridor and chest recession. The oxygen saturation was 90% to 93% on room air. Other vital sign parameters were normal. Neck examination revealed bony hard swelling over the thyroid cartilage with loss of the normal laryngeal framework configuration (Figure 1A). There was no cervical lymph node palpable. Flexible nasopharyngolaryngoscopy examination showed present of smooth surface mass at the left laryngeal surface of epiglottis extending to left aryepiglottic fold and pyriform sinus caused significant narrowing of the airway (Figure 1B). Bilateral vocal cords and subglottic region were not visualised as obscured by the mass. Examination of the nasal cavity, nasopharynx, oral cavity, oropharynx, and ear were unremarkable. Upper and lower limbs examinations revealed swelling and deformities of multiple joints namely bilateral metacarpophalangeal, interphalangeal, knee and metatarsophalangeal joints (Figure 2). His blood investigations were normal, except serum uric acid of 9.5mg/dL.

Lateral soft tissue neck x-ray showed radiopaque and scattered calcification of thyroid cartilage and loss of cervical lordosis (Figure 3). The airway below the cricoid cartilage was patent. Emergency tracheostomy under local anaesthesia was performed as anaesthetist team unable to intubate the patient. Subsequently, patient underwent contrasted computerised tomography (CT) scan of neck which showed heterogenous enhancing lobulated mass in the hypopharynx with its epicentre in the visceral space of left supraglottic region, approximately measuring 3.6cm x 3.9cm x 4.9cm causing significant airway narrowing (Figure 4A and 4B). This mass extends outward to involve the aryepiglottic region, ipsilateral hyoid muscles without extracutaneous extension and cause mass effect to the adjacent thyroid cartilage. The bilateral vocal cords and the airway below the cricoid cartilage appeared normal. The findings suggestive of laryngeal neoplasm or vascular malformation. Laryngeal gout was less likely due to atypical location.

Further imaging was performed with magnetic resonance imaging (MRI) of the neck to rule out vascular malformation as one of differential diagnosis. The extension of the mass is similar to the CT scan findings with low signal intensity on T1 weighted, mixed signal intensity on T2 weighted with heterogeneous enhancement, predominantly at the peripheral aspect post contrast (Figure 4C – 4E). In addition, there was peripheral calcification and central necrotic/ cystic component. No flow void within the mass. Generally, the MRI findings more suggestive of laryngeal malignancy and less likely laryngeal gout.

The patient underwent direct laryngoscopy, tracheoscopy and biopsy. Intraoperative findings revealed hard mass with normal overlying mucosa at the left laryngeal surface of epiglottis, extending laterally to aryepiglottic fold and pyriform fossa, and inferiorly to false cord. Tracheoscopy was performed with 2.9mm zero-degree telescope showed normal bilateral vocal cords and trachea. Mucosal incision was made at the junction between the epiglottis and the mass to raise mucosal flap (Figure 5A). Multiple gouty tophi and inflamed tissue were visualised after the mucosal flap was raised (Figure 5B). Biopsy was taken and gouty tophi were removed. Histopathology examination revealed tissue fragments composed of fibrofatty and lesional tissues admixed with blood and fibrin clots. The lesional tissue comprising of islands of amorphous crystalline material surrounded by macrophages and multinucleated giant cells (Figure 6). The overall findings suggestive of gouty tophi with no feature of malignancy.

The patient was referred to rheumatologist and started on non-purine analogue xanthine oxidase inhibitor (Febuxostat 40mg once daily). The airway much improved (Figure 7) after eight months on the medication and subsequently able to decannulate from tracheostomy. The tracheostomy site was well healed, and the patient is still on our regular follow up. After one year of decannulation, the airway remained patent, dysphagia was resolved and voice back to normal. The serum uric acid was lowered to 4.9mg/dL.

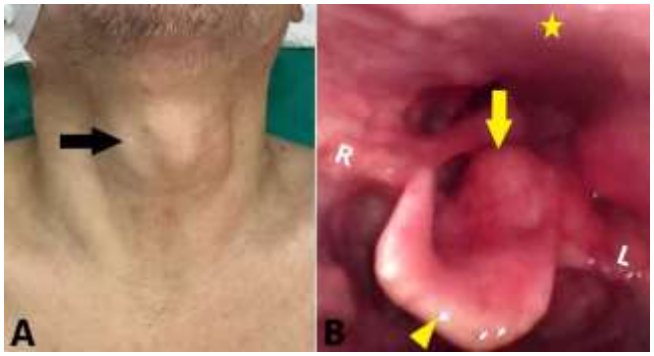

Figure 1: A. Bony hard swelling over the thyroid cartilage with loss of the normal laryngeal framework configuration (back arrow). B. Indirect laryngoscopy view of larynx shows present of smooth surface mass (yellow arrow) at the left laryngeal surface of epiglottis (yellow arrowhead) extending to left aryepiglottic fold and pyriform sinus caused significant narrowing of the airway. Yellow star – posterior pharyngeal wall; R – right; L – left.

Figure 2: Swelling and deformities of multiple joints of bilateral upper and lower limbs. A. Right hand (metacarpophalangeal joints and interphalangeal joints). B. Left hand (metacarpophalangeal joint and interphalangeal joints). C. Left knee joint. D. Right foot (metatarsophalangeal joints and interphalangeal joints). E. Left foot (metatarsophalangeal joints).

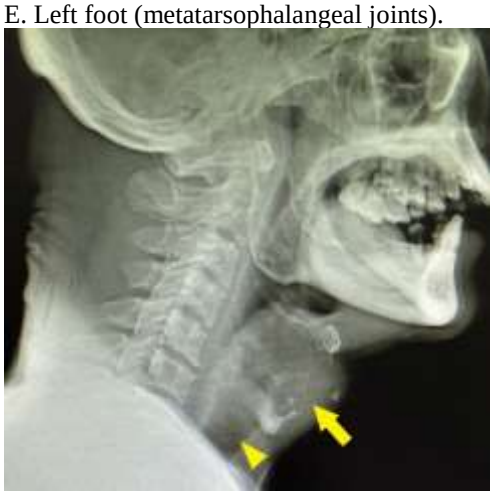

Figure 3: Lateral soft tissue neck x-ray shows radiopaque and scattered calcification of thyroid cartilage (arrow) and loss of cervical lordosis. The airway below the cricoid cartilage was patent (arrowhead).

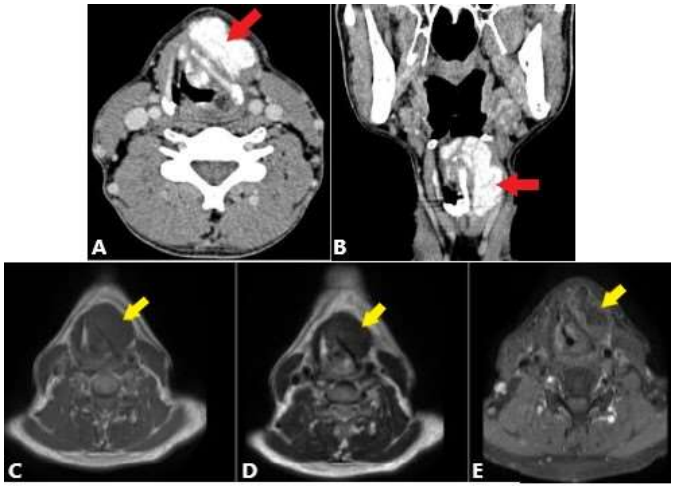

Figure 4: Contrasted computerised tomography scan of neck in axial (Figure A) and coronal (Figure B) views show heterogenous enhancing lobulated mass in the hypopharynx with its epicentre in the visceral space of left supraglottic region, causing significant airway narrowing. Magnetic resonance imaging of the neck shows the mass is low signal intensity on T1 weighted (Figure C), mixed signal intensity on T2 weighted (Figure D) with heterogeneous enhancement, predominantly at the peripheral aspect post contrast (Figure E).

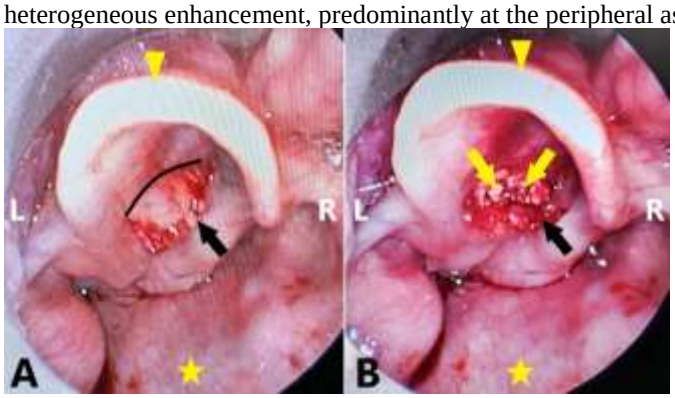

Figure 5: A. Direct laryngoscopy view of larynx shows mucosal incision (black curved line) was made at the junction between the epiglottis (yellow arrowhead) and the mass to raise mucosal flap (black arrow). Multiple gouty tophi and inflamed tissue (yellow arrow) visualised after the mucosal flap raised (black arrow). Yellow star – posterior pharyngeal wall; R – right; L – left.

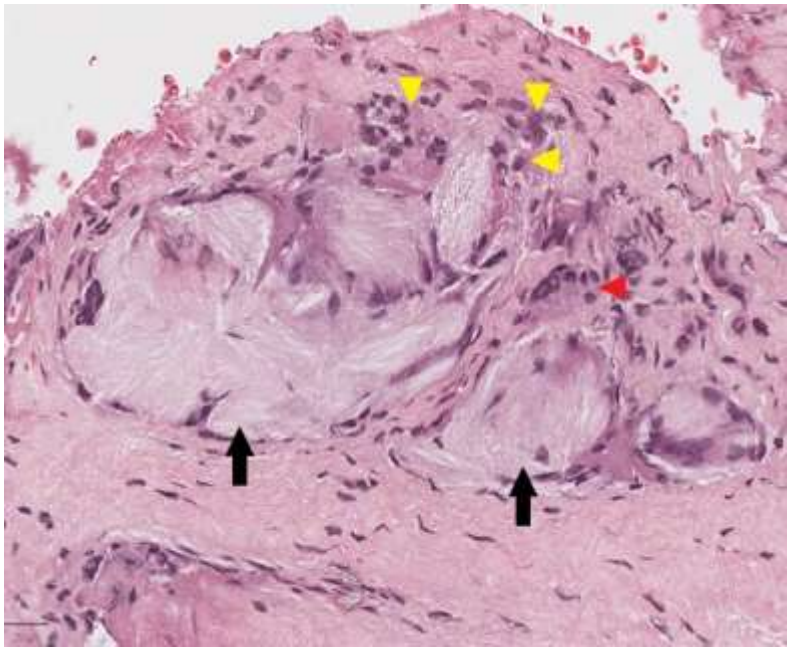

Figure 6: The lesional tissue comprising of amorphous crystalline material (arrow) surrounded by macrophages (yellow arrowhead) and multinucleated giant cells (red yellowhead).

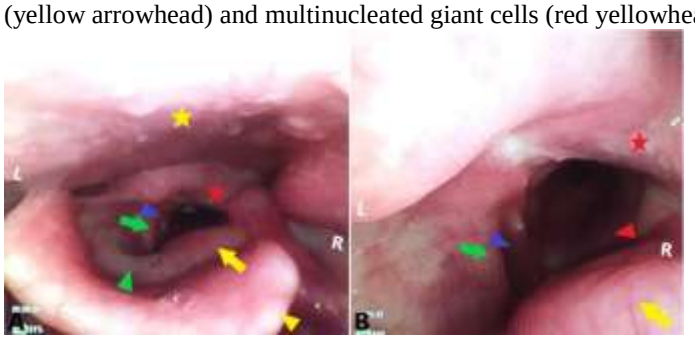

Figure 7: A. Direct laryngoscopy view of larynx shows airway much improved after eight months on the medication. B. Closed view of the laryngeal inlet. Yellow star – posterior pharyngeal wall; Red star – posterior commissure; Green arrow – left false cord; Yellow arrow – gouty mass smaller in size; Blue arrowhead - right vocal cord; Red arrowhead – left vocal cord; Green arrowhead – secretion; R – right; L – left.

Discussion

Larynx is atypical site for gouty arthritis and its involvement almost always preceded by the involvement of upper and lower limbs. The most common site of involvement is first metatarsophalangeal joint, followed by the wrist, ankle, and knee joints.[2] Although is rare, involvement of larynx causes significant morbidity in term of speech, swallowing, respiration and aspiration. Laryngeal gout was first described since 1863 by Garrod, while the proper case report with pathologic confirmation was first described by Goodman *et al.* in 1976.[5] Apart from larynx, gouty tophi also may occur at other part of head and neck like pinna, middle ear, nose, throat, temporomandibular joint, sternoclavicular joint and eye structures.[2,6,7]

The clinical course of gout typically starts with asymptomatic hyperuricemia, followed by recurrent attack of acute arthritis, accumulation of gouty tophi, chronic gouty arthritis, and deformity of the joint.[1] Deposition of urate crystals lead to inflammation of the affected joint and explains the acute flare of the symptoms. The symptoms generally include pain, swelling, redness, and heat. The presenting case presented with acute odynophagia and worsening respiratory distress could be due to the acute flare in the larynx.

The chronic symptoms of laryngeal gout are dependent on the site of involvement and severity, ranging from asymptomatic to neck swelling, hoarseness, dysphagia, aspiration, and shortness of breath. The neck swelling

usually due to the deposition of gouty tophi on the thyroid cartilage lamina or other laryngeal framework. Hoarseness could be due to the direct deposition of gouty tophi on vocal cord or secondary to the deformity of cricoarytenoid joint. In addition, involvement of cricoarytenoid joint also may result in aspiration and upper airway obstruction secondary to vocal cord immobility. Dysphagia is developed when the gouty tophi on the larynx especially at the cricoid cartilage is large enough to cause external compression on upper oesophagus. Other than that, large gouty tophi also may disturb laryngeal excursion which may further contribute to dysphagia. Apart from vocal cord immobility, deposition of gouty tophi on internal site of larynx will obstruct the upper airway either at supraglottic, glottic or subglottic regions. Although extremely rare, patient may be presented with life threatening upper airway obstruction which may need emergency tracheostomy as seen in the presenting case.

Although laryngeal involvement is rare, high clinical suspicious of gout should be made in cases of chronic gouty arthritis with multiple joints involvement and presenting with laryngeal symptoms. The goal standard of diagnosis of laryngeal gout remains histopathological examination.[2,5,7] Biopsy should be performed whenever possible to confirm diagnosis and exclude other pathology especially malignancy prior to any definitive treatment or surgical intervention. Recently a study had shown association of head and neck cancer particularly laryngeal cancer with gout.[8] The typical histopathological finding of gout is core of urate crystal and basophil surrounded by macrophages and epithelioid cells.[7] Needle-shaped uric acid crystals are water soluble and could not be seen in our specimen as they were dissolved by the solvents used during tissue processing.

Soft tissue neck x-ray is an initial investigation that can be done bedside at emergency room in unstable cases. It may not provide anatomical detail of larynx for diagnosis and extension of the disease but provide adequate information on the patency of trachea in case emergency tracheostomy is needed especially under local anaesthesia as seen in presenting case. Contrasted CT scan is an important imaging tool for laryngeal gout because it provides anatomical detail especially for bony structures, shows extension of disease and may exclude other possible differential diagnosis. However, because of the rarity of the disease, laryngeal gout usually in low priority of the differential diagnosis. The characteristic feature of tophi on CT scan is reported as denser than soft tissue but less dense than calcification with Hounsfield units of 160–170.[9] In our presenting case, there was heterogenous enhancing lesion with calcification, which share similar feature with malignant or vascular lesions. Dual energy protocol CT scan is a promising emergent tool in detecting gout with good sensitivity and specificity,[10] but unfortunately not available in our centre. MRI findings usually show homogeneous low signal intensity on T1-weighted image while T2-weighted image has variable signal intensity.[9] It is rarely necessary in gout cases, but we had proceeded with this imaging in this presenting case mainly to rule out vascular lesion. It is important to exclude this lesion first as biopsy may result in catastrophic bleeding in the airway. The imaging modalities either CT scan or MRI cannot confirm the diagnosis of laryngeal gout on its own but provide valuable information especially for targeted biopsy and evaluate extension of the disease.

The treatment of laryngeal gout is different from those that only affected upper and lower limbs. While medication in usually the only treatment for gout involves upper and lower limbs, laryngeal gout usually required combination of surgery and medication. Secure the airway is paramount important and usually the first step in cases with upper airway obstruction as seen in presenting case. Emergency tracheostomy under local anaesthesia was required in our case as patient presented with life threatening respiratory distress and anaesthetist unable to intubate through oral or nasal due to the present of hard mass at supraglottic region. There is high risk to put patient under general anaesthesia as the patient will loss his breathing effort. In addition, attempting intubation in a small airway patient without visualising glottic structures has high possibility of fail intubation and injury to the surrounding structures. Intubating through the mass without knowing the nature of the lesion also may lead to severe bleeding and obstructing the lower airway. After consideration of these factors, tracheostomy under local anaesthesia is safer as patient still able to control his breathing effort while inserting tracheostomy tube and avoiding injury to the supraglottic structures and mass. Ideally CT scan should be performed prior to the tracheostomy or other surgical intervention. However, our patient was unstable and unable to lie flat for CT scan, thus tracheostomy was performed first.

The goal of definitive surgery is to remove the gouty tophi as much as possible to provide adequate airway while preserving the normal speech and swallowing structures and functions. The surgical approaches either endoscopic or transcervical are dependent on the location and severity of the disease.

The key strategy of medical treatment is long term urate lowering therapy with the target value of serum urate of <5mg/dL which usually result in monosodium urate crystals dissolution.[5] Purine analogue xanthine oxidase inhibitor (allopurinol) is most commonly used as the first line medical therapy but unfortunately our patient allergic to this medication. Therefore, he was changed to non-purine analogue xanthine oxidase inhibitor

(febuxostat) and responding well to this medication. Other medications that can be used as alternative for chronic gouty arthritis are uricosuric like probenecid and sulfinpyrazole, and uricolytic like rasburicase and pegloticase. In acute flare of gouty arthritis, combination of analgesic and anti-inflammatory medications like steroid, non-steroidal anti-inflammatory drugs and colchicine are the core treatment.

Patient with laryngeal gout need long term follow up with otorhinolaryngologist and rheumatologist to make sure the airway is adequately patent, and serum uric acid level is controlled with medication respectively. Speech and language therapy team is also important for rehabilitation as some of the patients may have residual hoarseness and dysphagia. Other than that, patients with multiple joints deformity should have follow up with orthopaedic, rehabilitation and physiotherapy teams. Some of the patients may need psychiatrist and counselling consultation to alleviate anxiety or depression of become dependent to family and loss of the job and income. The patient also needs to be motivated with lifestyle changes like avoiding high purine foods and alcohol and do more exercise for weight reduction.[11]

Conclusion

Laryngeal gout is extremely rare, challenging in management and need long term follow up. It may present with life-threatening upper airway obstruction that warrant emergency tracheostomy under local anaesthesia. Securing the airway with subsequent definitive surgery and long-term urate lowering therapy are the central strategy of treatment. Multidisciplinary team approach is usually required as majority of patients also have other anatomical area involvement.

Disclosure

The authors declared no conflicts of interest. Informed consent was obtained from the patient.

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