Unusual association of diseases/symptoms

An uncommon shape of epiglottis misleading as acute epiglottitis

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Summary

We report a case of a 31-year-old Japanese woman with an uncommon shape of epiglottis misleading as acute epiglottitis. She presented with high fever, arthralgia, general fatigue and strong throat pain. Laryngeal fiberoscopy showed a markedly swollen epiglottis suggestive of acute epiglottitis, though she had no signs of asphyxia. A careful observation and detailed medical history ruled out acute epiglottitis. However, without clinical discretion, it might have been misinterpreted as acute epiglottitis. She was diagnosed later with Behçet's disease, which is reported to present pharyngeal stenosis. Two years later, a repeat laryngeal fiberoscopy showed exactly the same appearance of the epiglottis, suggesting its shape to be permanent. The cause of this misleading shape of the epiglottis in this patient is yet unknown. For similar cases, a possibility of this kind of epiglottis should be considered before diagnosing as acute epiglottitis.

BACKGROUND

Acute epiglottitis is common especially in the emergency room. Progressive throat pain and shortness of breath followed by asphyxia is a typical course. When overlooked or underdiagnosed, it will become life threatening by complete airway obstruction. Therefore, intubation or tracheostomy is indicated for sudden or progressive asphyxia. Although diagnosis is established based on clinical presentation, a typical fibroscope image of markedly swollen epiglottis is key to the diagnosis.¹ We report a case with an uncommon shape of epiglottis misleading as acute epiglottitis on laryngeal fiberoscopy in a 31-year-old Japanese woman. Although she did not present any sign of asphyxia, other clinical manifestations such as high fever, severe throat pain and blood tests raised the suspicion of severe acute epiglottitis. Our case might have been misdiagnosed as acute epiglottitis without clinical discretion.

CASE PRESENTATION

A 31-year-old Japanese woman consulted our hospital for fever of 38.2 °C, severe arthralgia of the left ankle joint, right first metatarso-phalangeal joint, both knee joints and both shoulder joints and general fatigue. Laboratory tests showed leucocyte count of 12 000/μl (neutrophils 70%) and C-reactive protein (CRP) of 6.0 μg/dl. Palpation of the neck showed bilateral lymphadenopathy in the posterior cervical region. She was admitted to the Department of Internal Medicine on the same day under unconfirmed diagnosis. The history included tonsillectomy at the age of nine. Skin eruptions associated with erythema nodosum were noted for several years. She had been followed up at a university hospital but treatment was discontinued recently because of stable symptoms. She was not a smoker or alcohol drinker. Body weight was 52.4 kg and height was 154 cm. She was treated with

oral loxoprofen sodium and intravenous injection of penicillin G, but showed only a temporary response. On day 5 of admission, she complained of severe sore throat. A few mucosal aphthous ulcers were noted on the posterior pharyngeal wall, tip of the tongue and mucosa of the lower lip. Body temperature was still as high as 39.0°C. The pre-existing skin eruptions increased in both size and number accompanied by pain. She was referred to the Department of Otolaryngology for assessment of oral aphthous ulcers and severe throat pain. She underwent laryngeal fiberoscopy, which showed a markedly swollen epiglottis (figure 1A). Although she did not have any difficulty in breathing, she was advised about possible acute epiglottitis. At that stage, she indicated that she had been suspected of exactly the same diagnosis and potential need for tracheostomy a few years ago at another hospital where she underwent laryngeal fiberoscopy, various blood tests and a CT scan. All tests at that time were unremarkable except for the swollen epiglottis, and the patient was told that the swollen epiglottis was probably congenital. This history was convincing as the patient did not present dyspnoea or stridor since admission. Acute epiglottitis was more unlikely because she had no dysphagia or drooling. Nevertheless, the patient was advised about possible asphyxia. Continuous monitoring of breathing and SpO₂ with a pulse oximeter for a few days showed normal SpO₂. Laryngeal fiberoscopy was repeated on day 7 and also on day 14 but showed similar morphology of the epiglottis. In the meantime, the final diagnosis was established as Behçet's disease based on oral aphthous ulcers, external genital ulcers and skin lesions. No ophthalmic symptoms/signs were noted.

DIFFERENTIAL DIAGNOSIS

The differential diagnoses include Behçet's disease and congenital anomaly as the cause of this bizarre shape of

Figure 1 (A) The image of initial laryngeal fiberoscopy at our facility. The epiglottis appears swollen suggestive of acute epiglottitis. (B) Laryngeal fiberoscopic image taken 2 years later. Note the similar appearance to (A) 2 years earlier.

epiglottitis. Sarcoidosis^{2 3} and tuberculosis^{4 5} may also be included, which are known to cause upper airway obstruction, but are less likely because roentgenography of the chest was unremarkable.

TREATMENT

She was treated with colchicine on day 9 of admission and showed marked improvement including arthralgia, sore throat and fever. On day 16, she was referred to a tertiary care centre for treatment of Behçet's disease under the care of collagen disease specialists.

OUTCOME AND FOLLOW-UP

Two years later, when the symptoms of Behçet's disease were stable, she visited our department for a throat check-up. Laryngeal fiberoscopy showed exactly the same findings noted 2 years earlier of swollen epiglottitis (figure 1B). Although we recommended a biopsy to confirm the diagnosis, the patient's consent was not obtained.

DISCUSSION

Acute epiglottitis occasionally requires immediate treatment including intubation or tracheostomy for sudden asphyxia. The fiberoscope image of the epiglottis is usually most important for the diagnosis of acute epiglottitis. Other manifestations such as sore throat, fever, odynophagia, muffled voice, dyspnoea, stridor, dysphagia drooling and laboratory data such as leucocytosis or high CRP may also help the diagnosis.¹ Some of the clinical features of our patient, including high fever, throat pain and leucocytosis, were supportive of acute epiglottitis together with a misleading fiberoscope image. If she complained of other symptoms supportive of acute epiglottitis such as stridor, dysphagia or drooling, even unrelated to the deformed epiglottis, she might have been misdiagnosed as acute epiglottitis. An unnecessary treatment such as intubation or tracheostomy might have been performed. When looking back retrospectively, the fiberoscope image of the swollen mucosa gives an impression of chronic hypertrophy rather than acute oedema and

careful observation could rule out acute epiglottitis as unlikely. The airway obstruction was mild enough to exclude the need for immediate airway management. However, such judgement would remain in doubt under the diagnosis process with possible deterioration of the compromised airway. Fortunately, we could discretely rule out acute epiglottitis by a detailed history taking and careful observation of the larynx. The lack of typical symptoms such as dysphagia, drooling or stridor was also helpful.

To our knowledge, no similar case has been reported previously. What is the cause of the swollen epiglottis noted in our patient? We made two assumptions: The first is a 'congenital' anomaly. Chen and Holinger⁶ found anomalies in 33 of 115 laryngeal specimens examined, though none resembled ours. The second is a yet unreported manifestation of Behçet's disease. Although Behçet's disease is well known for the classical triad affecting the mouth, eyes and genitals, it may also present with various ear, nose and throat manifestations including hearing loss and disequilibrium.⁷ Although pharyngeal stenosis has been reported in some cases of Behçet's disease,⁸⁻¹¹ laryngeal involvement is scarcely reported. The case described by Nonomura *et al*¹² showed pharyngolaryngeal stenosis with deformed epiglottis, requiring surgical treatment including tracheostomy. This epiglottis was oedematous, folded and deformed into an omega shape and is partially similar to ours. Our case may represent the early stage of pharyngolaryngeal stenosis of such a case. The laryngeal hypertrophy may progress very slowly leading to future stenosis necessitating surgery. However, this still remains a speculation and further follow-up is needed for confirmation. The latest review⁷ describing the relation between Behçet's disease and laryngeal manifestations refers only to the above-cited case.¹²

On seeing a similar case of epiglottitis, the examiner should take into account the possibility of pre-existing deformity before making an erroneous diagnosis of acute epiglottitis.

Learning points

- ▶ A complete airway obstruction caused by acute epiglottitis is an imminent risk and intubation or tracheostomy is indicated for sudden or progressive asphyxia.
- ▶ The epiglottis of our case was markedly swollen and might have been misinterpreted as acute epiglottitis.
- ▶ Acute epiglottitis was discretely ruled out by a detailed history taking and careful observation of the larynx. The lack of typical symptoms such as dysphagea, drooling or stridor was also helpful for ruling out.
- ▶ The swollen epiglottis was possibly caused either by a congenital anomaly or a yet unreported manifestation of Behçet's disease.
- ▶ Before diagnosing as acute epiglottitis, the examiner should take into account the possibility of a similar epiglottitis as in our case.

Competing interests None.

Patient consent Obtained.

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