Significant laryngeal destruction in a northern European cohort of Behçet’s disease patients

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ABSTRACT
Objective. Behçet’s disease (BD) is a multisystem autoimmune disease of unknown origin typically affecting the triad of oral and genital mucosa and the eye. Limited data are available in the literature regarding the otolaryngology-related manifestations of BD, particularly in northern Europeans. This is a novel study detailing surprising and significant laryngeal structural changes in a northern European cohort of BD.

Methods. Patients meeting the International Study Group for Behçet’s Disease (ISGBD) and the International Criteria for Behçet’s Disease (ICBD) criteria for diagnosis were identified from an institutional database. Patients underwent examination with an otolaryngologist, including flexible laryngoscopy. Intra-oral, pharyngeal and laryngeal manifestations of BD were documented and characterised. Patients underwent hearing assessment with pure-tone audiometry.

Results. Fifteen patients with BD were identified (4 male, 11 female; median age 36 years). 60% (n=9) showed evidence of disease on examination and flexible laryngoscopy. 33% (n=5) showed laryngeal changes related to BD. 13% (n=2) demonstrated bilateral sensorineural hearing loss. The 5 cases demonstrating laryngeal manifestations of disease are described in detail with photographic records.

Conclusion. Limited data has been published regarding the laryngeal manifestations of BD, particularly in a northern European population. Our cohort of BD patients demonstrate significant laryngeal structural changes. It would appear that these clinically relevant changes may be more common than was previously thought. Raised awareness of the risk of laryngeal pathology in BD patients, often in the absence of overt clinical symptomatology, may result in earlier diagnosis and treatment.

Rheumatologists and otolaryngologists should consider closer multi-disciplinary co-operation in the management and follow up of patients with BD.

Introduction
Behçet’s disease (BD) is a complex, inflammatory, systemic disease of unknown aetiology. Although Hippocrates first described a patient with a triad of mouth ulcers and symptoms affecting the genitals and eyes 2,500 years ago, Professor Hulusi Behçet, a Turkish dermatologist, in 1937 provided the first characterisation of the trisymptom ‘occulo-buccal-genital’ complex (1, 2). The clinical manifestations of BD are believed to be attributable to vasculitis, which could involve blood vessels of any size from arterial or venous circulation. The prevalence of BD shows significant geographical variation with a reported prevalence ranging from 80–400/100,000 in Turkey and Middle-Eastern countries, to less than 1/100,000 in western Europe (3, 4). Pathogenesis of the disease is not well understood, however, environmental and genetic predisposition may play a role in the development of the disease (3, 4).

No pathognomonic laboratory test exists to diagnosis BD. As such, the diagnosis of BD is guided by validated criteria, most commonly used are the International Study Group for Behçet’s Disease (ISGBD) or the International Criteria for Behçet’s Disease (ICBD). In the ISGBD criteria, which were published in 1990, recurrent oral aphthosis three or more times in a year is mandatory, with the presence of any two of the following: genital ulceration, ocular or cutaneous manifestation, or skin pathergy.

While having excellent specificity, the ISGBD criteria lacked sensitivity, and for this reason the ICBD criteria were developed and first presented in 2006 (5, 6). For this classification, which is
based on points, oral aphthosis is not mandatory, and vascular manifestations were added to the existing five items. While genital ulceration and eye manifestations were given two points, other remaining items were given one point each; getting three or more points confirms the diagnosis.

The principle morbidity of BD relates to its vascular and ophthalmic complications, which, left untreated can lead to blindness or death (7). There are many other well-described manifestations of BD, which were not included in the ISGBD or ICBD criteria, in particular cardiac, gastrointestinal, pulmonary, arthritic and neurological involvement (7). The otolaryngology-related manifestations of BD, though previously described, have been less thoroughly explored in the literature. These manifestations are said to be relatively rare and include hearing loss, pharyngeal and laryngeal ulceration and nasal mucosal involvement (8).

A patient with extensive pharyngeal and laryngeal ulceration (Case 1 below) presented to our institution following a period of non-compliance with medical therapy. The extent of findings on laryngoscopy in this patient and the relative paucity of information relating to laryngeal changes in BD led us to undertake a screening using flexible laryngoscopy and audiometric assessment of all other patients with BD treated at our institution.

### Methods

Patients with BD were identified from a prospectively-maintained database kept by the Rheumatology service at our institution. Patients whose diagnosis of BD conformed to ISGBD and ICBD criteria for diagnosis were included. All patients had a negative autoantibodies profile which include anti-nuclear antibody (ANA), anti-phospholipid antibody, anti-proteinase 3 and anti-myeloperoxidase anti-neutrophil cytoplasmic antibodies (anti-PR3 and anti-MPO ANCA). Relevant microbiologic studies were completed and did not identify any causative organisms. Following informed consent, patients underwent examination of the oral cavity, flexible laryngoscopy and pure-tone audiometry assessment at a multi-disciplinary outpatient clinic attended by Rheumatology and Otolaryngology services. Intra-oral, pharyngeal and laryngeal manifestations of BD were documented and characterised. Photographic and video records were obtained in all cases with patient consent. Basic demographic data and relevant clinical data were also compiled.

### Results

Fifteen patients with BD were identified from a prospectively-maintained institutional database (Table II). 27% (n=4) were male, 73% (n=11) were female. Median age was 36 years. 60% of patients (n=9) showed evidence of disease on oral examination and flexible laryngoscopy. Of these, 55% (n=5) showed laryngeal changes related to BD (33% of the total cohort). Of the remaining patients with findings 45% (n=4) demonstrated evidence of active oral ulceration (26% of the total cohort). 13% of patients (n=2) demonstrated reduced hearing on pure-tone audiometry.

The 5 cases demonstrating laryngeal manifestations of disease are described below with photographic records and summarised in Table I.

#### Case 1

A 23-year-old female diagnosed with BD at the age of 16 following presentation with recurrent oral aphthosis, genital ulceration, papulopustular rash of the lower limbs, fatigue and arthralgia, fulfilling the ISGBD and ICBD criteria. Overall clinical improvement was achieved with commencement of azathioprine and colchicine, however, following a lengthy period of non-compliance with her medical therapy, symptoms of odynophagia and dysphagia developed at the age of 23.

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**Table I. Five patients with positive findings at flexible laryngoscopy (n=15).**

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Sex</th>
<th>Diagnosis</th>
<th>Laryngoscopy findings</th>
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<tbody>
<tr>
<td>1</td>
<td>23</td>
<td>F</td>
<td>7 years</td>
<td>Shortened, thickened epiglottis. Posterior wall stricture obscuring the hypopharynx. Fistula to oesophagus.</td>
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<tr>
<td>2</td>
<td>69</td>
<td>M</td>
<td>5 years</td>
<td>Epiglottic thickening, irregular surface. Areyepiglottic fold irregular surface</td>
</tr>
<tr>
<td>3</td>
<td>33</td>
<td>F</td>
<td>6 months</td>
<td>Shortened, partially obliterated epiglottis</td>
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<tr>
<td>4</td>
<td>51</td>
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<td>16 years</td>
<td>Shortened, thickened, tethered epiglottis. Unilateral thickened aryepiglottic fold</td>
</tr>
<tr>
<td>5</td>
<td>48</td>
<td>F</td>
<td>6 years</td>
<td>Tracheal mucous casting. Atrophy of nasal structures</td>
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**Table II. Clinical manifestations of patients with BD in the study group (n=15).**

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<tr>
<th>Patient</th>
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<th>GU</th>
<th>OL</th>
<th>CL</th>
<th>PP</th>
<th>VI</th>
<th>GI</th>
<th>JI</th>
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<tr>
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<td>F</td>
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</table>

AOU: Recurrent aphthous oral ulcers; GU: Genital ulcers; OL: Ocular lesions; CL: Cutaneous lesions; PP: Positive pathergy test; VI: vascular involvement; GI: GI involvement; JI: Joint involvement; ENT: ENT manifestations.
Following referral to the Otolaryngology service, flexible nasendoscopy was performed revealing giant confluent mucosal ulcers throughout the tongue base and lateral pharyngeal wall, with oedema of the epiglottis. Pharyngeal ulceration extended from the glossopiglottic ligament to the aryepiglottic fold. Pharyngeal biopsy revealed nonspecific deep ulceration with marked numbers of neutrophils and eosinophils, however, no evidence of vasculitis was seen.

Following commencement of systemic corticosteroids and concomitant infliximab infusion (a TNF-α blocker), the patient’s symptoms resolved. At interval flexible laryngoscopy, ulceration was resolved, however, a shortened, thickened epiglottis and extensive change of the hypopharyngeal architecture was observed (Fig. 1a–b). An area of fistulation through a pharyngolaryngeal stricture of the posterior wall represented the sole inlet of the oesophagus (Fig. 1b).

**Case 2**

A 69-year-old male diagnosed with BD at the age of 64 following presentation with recurrent oral aphthosis with odynophagia to solids and fluids, hoarseness, progressive hearing loss, papulopustular rash and arthritis. Ocular examination revealed bilateral anterior uveitis, with overall presentation fulfilling the ISGBD and ICBD criteria. The patient underwent first Otolaryngology assessment at the time of initial presentation, which showed supraglottitis and thickening of his left vocal cord.

Clinical remission was achieved with combination therapy of adalimumab and methotrexate. Flexible laryngoscopy at otolaryngology assessment as part of this study identified thickening and mucosal irregularity of the epiglottis and aryepiglottic folds, despite the patient being entirely asymptomatic (Fig. 2a). A band of thickened soft-palatal tissue in the oropharynx from the posterior tonsillar pillars to the uvula was also noted (Fig. 2b).

**Case 3**

A 33-year-old female diagnosed with BD at the age of 33 following presenta- tion with a five-month history of recurrent oral aphthosis, genital ulceration, and generalised arthralgia. Subsequently sterile inflammatory neutrophilic dermatosis (Fig. 3a) developed on the right lower limb with concurrent elevation of acute phase reactants (erythrocyte sedimentation rate (ESR) 36 mm/h; C-reactive protein (CRP) 13 mg/l). A diagnosis of BD was made, with symptoms and clinical findings fulfilling the ISGBD and ICBD criteria. Resolution of clinical symptoms and normalisation of her acute phase reactants was seen with oral prednisolone, in combination with infliximab infusion and predsol mouthwash. Despite having no previous ear, nose or throat symptoms and being asymptomatic at the time of otolaryngology assessment, flexible laryngoscopy identified a shortened, partially obliterated epiglottis (Fig. 3b).

**Case 4**

A 51-year-old male diagnosed with BD at the age of 35 following presentation with recurrent oral aphthosis, genital ulceration, bilateral anterior uveitis, small, sterile neutrophilic dermatosis on his right shin and a positive pathergy test, associated with fatigue and arthralgia, fulfilling the ISGBD and ICBD criteria. He was commenced on infliximab, colchicine and low dose prednisolone.

The patient remained in clinical remission for seven years, when he developed abdominal discomfort and cramping. Colonoscopy revealed aphthous ulceration of the caecum and a thickened terminal ileum, consistent with intestinal BD. An alternative TNF-α
blocker, adalimumab, was commenced without further exacerbations since that time.
Flexible laryngoscopy at otolaryngology assessment identified a thickened, tethered epiglottis with reduced movement and unilateral aryepiglottic fold thickening associated with an aberrant movement pattern (Fig. 4a-b). Again, this patient was asymptomatic at the time of review. Pure-tone audiogram highlighted bilateral, symmetrical sensorineural hearing loss, requiring referral for hearing aid assessment.

Case 5
A 48-year-old female diagnosed with BD at the age of 42 following presentation with recurrent oral aphthosis with associated arthralgia, fever, fatigue, papulopustular rash and subsequent genital ulceration, fulfilling the ISGBD and ICBD criteria.
The patient concurrently developed significant dysphonia with associated chronic productive cough. Respiratory system examination and investigations were suggestive of upper respiratory tract findings. Initiation of infliximab infusion and oral prednisolone gave immediate dramatic response, with complete resolution of orogenital ulcers and associated BD symptoms. Intermittent dysphonia and upper respiratory tract symptoms persisted, however. Infliximab infusion was discontinued for an 18-month period following the patient’s wish to conceive, which was associated with a recurrence of orogenital symptoms. After recommencement of infliximab infusion, significant improvement was again noted, with complete resolution of the orogenital ulcers. Dysphonia and upper airway symptoms continued, however, and referral was made for respiratory medicine assessment.
At bronchoscopy, widespread pharyngolaryngeal aphthous ulceration with thickened mucous castings covering the larynx and upper trachea were noted. Otolaryngology review was sought in light of potential risk to the airway. Intravenous corticosteroids were commenced in combination with inhalation antimicrobials, resulting in gradual symptom resolution.

Follow-up flexible laryngoscopy at the time of this showed residual mucous secretions in the trachea (Fig. 5b). Nasendoscopy showed atrophy of nasal structures and mucosal scarring in keeping with the appearances of an ‘empty nose’ syndrome, with the same pattern seen on computed tomography (CT) (Fig. 5a).

Discussion
We describe the first series of patients with laryngeal manifestations of BD identified following a screening assessment with flexible laryngoscopy at a multidisciplinary clinic. Limited descriptions of manifestations of BD related to the speciality of otolaryngology exist in the literature. A
review from Webb et al. in 2008 identified 27 key papers on the topic (9). Otolaryngeal features of BD are described in a number of series, with 22–59% of patients found to have sensorineural hearing loss (10–12). Two patients in our series, 13.3%, demonstrated bilateral, symmetrical sensorineural hearing loss. While this figure is lower than those reported in previous literature, our northern European patient cohort differ to most other series, as discussed below.

Vestibular and inner ear disease has also been described (13, 14). Nasal mucosal involvement was seen in 7.8% of 400 BD patients in one series, with associated dysosmia in 3.8% (15). Two single-patient case reports describe sinus disease in the setting of BD (16, 17). The pattern of ‘sinusopathy’ described by Martins et al. is similar to that of Case 5 in our study, however, complete radiographic imaging are not available in this case (16). Further isolated case reports describing pharyngeal and laryngeal manifestations of BD have been described (Table III). The earliest case, published in 1951 by Kenet et al., describes widespread pharyngeal and laryngeal aphthous ulceration which was refractory to medical therapy and required tracheotomy to preserve the airway (18). Similarities with Case 1 in our series can be drawn here, as the severity of our patient’s findings on flexible laryngoscopy at presentation following a hiatus in medical therapy warranted consideration for tracheotomy. Other reports describe a broad variety of pharyngeal scarring and stenosis patterns noted in symptomatic patients (19–21). More recent single case reports have described abnormalities of the epiglottis which could be misdiagnosed as epiglottitis, as well as pose potential airway challenges (22).

Few previous cases in the literature have described surgery in the setting of laryngeal BD (23, 24). In one, Nonomura et al. report initial resection of supraglottic stenosis resulting in over-scarring necessitating further open surgery and grafting two-years after the index procedure. Follow-up data in this case was not available beyond 6 months post-operative following the second surgery. This tendency to mucosal over-scarring makes operative intervention in patients with BD challenging, in particular with pharyngolaryngeal disease where failure may result in permanent tracheotomy.

A number of large series exist which describe the commonest presenting features and affected systems in BD across a number of geographic regions (25–27). None appear to include complete otolaryngological examination however, or clarify whether examination included flexible laryngoscopy. Morales-Angulo et al. in 2014 retrospectively reviewed the records of patients with BD for otolaryngology-related manifestations (28). In total, 33 patients were identified from a 22-year period at their institution. Following review of medical records, 12% (n=4) demonstrated odyophagia secondary to oropharyngeal ulceration. This data was collected from paper records only, however, was retrospective, did not describe whether flexible laryngoscopy was used in all cases and was not video or photo-documented. A variety of structural changes caused by BD activity were identified at flexible laryngoscopy in our study. These findings affected a number of anatomic subsites within the oropharynx, pharynx, larynx and trachea and posed potential dangers from an airway perspective. While many of these patients were asymptomatic, undertaking flexible laryngoscopy documenting these changes in order to allow for reassessment of disease progression appears common sense. Further, the changes identified could allow for planning in the event of a requirement for later anaesthetic intubation for surgery. Misdiagnosis of these changes, if not recognised as a part of BD pathology, could lead to unnecessary investigations or treatment.

These laryngeal manifestations do not occur in a normal population. They are intrinsically destructive in origin and can only conceivably occur in destructive/erosive conditions such as post radiotherapy, post caustic ingestion and possibly post severe aggressive bullous pemphigoid. None of our patients had received radiotherapy, ingested caustic substances or had bullous pemphigoid. Destructive lesions as reported in this cohort are not for instance a feature in conditions such as laryngo-pharyngeal reflux disease, aphthous ulceration or sleep apnoea syndrome.

In 1985, Firestein et al. proposed the term MAGIC syndrome as an acronym for Mouth and Genital ulcers with Inflammation and Gastrointestinal, Arthritis and Carditis. However, this syndrome is not now considered to be a disease entity. In our patient, MAGIC syndrome was not identified as a part of BD pathology, could lead to unnecessary investigations or treatment.

The profile of our cohort of BD patients is atypical. Despite a reported prevalence as low as <1/100,000 in northern Europe we have a relatively large identified patient cohort in mid-western Ireland. Furthermore, the group identified is made up predominantly of young, female patients, despite distribution of gender being reported as equal for BD, with more aggressive disease progression is typically attrib-

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uted to the young, male, middle eastern BD patient cohort. Despite this, our patient group demonstrate noteworthy structural changes on examination with flexible laryngoscopy.

No previous description of assessment of this kind in a BD patient cohort has been reported in the literature, with all previous description of laryngeal disease appearing in symptomatic patients. Further investigation of the laryngeal manifestations of BD is required, therefore, to assess its prevalence in the asymptomatic BD patient population, as well as geographical variations in presentation and disease progression. Consideration should be given to the screening of all new-diagnosed BD patients using flexible laryngoscopy to assess for laryngeal manifestations of the disease given the potential for later airway compromise. Patients should be counselled regarding potential laryngeal manifestations of BD. Otolaryngologists should form part of the multi-disciplinary team involved in the investigation and management of BD in tandem with Rheumatologists.

Conclusion
Limited descriptions of the laryngeal manifestations of BD have, heretofore, been reported in the literature. Broader study of these disease manifestations should be undertaken by rheumatologists and otolaryngologists. Otolaryngologists should form part of the multi-disciplinary team in the management of BD, with flexible laryngoscopic screening forming part of initial investigations.

Key messages
• Laryngeal changes may be more prevalent in patients with Behçet’s disease than previously thought
• All newly diagnosed Behçet’s Disease patients should undergo assessment with otolaryngology including flexible laryngoscopy
• Otolaryngologists should form part of the multi-disciplinary team in the treatment of Behçet’s disease

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