Summary

Fourth branchial arch defect is a term used to describe a syndrome of congenital defects affecting the extrinsic structures of the larynx, due to a failure of development of structures that derive from the fourth branchial arch. This Case Report describes a 6-year-old Welsh section A mare presented for investigation of a chronic cough that was exacerbated on exercise. A fourth branchial arch defect was identified on resting upper airway endoscopy, with dorsal pharyngeal collapse evident on dynamic respiratory endoscopy, although full abduction of both the left and right arytenoid could be achieved and maintained at exercise.

Introduction

The extrinsic structures of the larynx have been shown to develop from the fourth branchial arch (Hast 1972). Fourth branchial arch defect (4-BAD) is a term that has been applied to a syndrome of congenital defects affecting the extrinsic structures of the larynx, resulting from either a complete or partial failure of development of structures that derive from the fourth branchial arch (Lane 1993). The syndrome has been reported as individual case reports or small case series, although sometimes under alternative descriptions such as cricopharyngeal laryngeal dysplasia (Dixon et al. 1993) and rostral displacement of the palato-pharyngeal arch (Cook 1974; Goulden et al. 1976; Wilson et al. 1986; Klein et al. 1989; Blikslager et al. 1999). The condition has been recorded in Thoroughbreds and larger breeds of horses, but does not appear to have been reported in small ponies. This Case Report describes a Welsh section A pony mare diagnosed with 4-BAD.

Case history

A 6-year-old Welsh section A pony mare, used as a child’s riding pony and for showing, that had been in the owner’s possession for 3 years, was presented for investigation of a chronic cough. The cough was exacerbated on exercise, in particular when the pony was asked to collect itself, and work on the bit, and had failed to respond to dust-free management and administration of oral clenbuterol. The owner also reported that the pony exhibited tachypnoea, dyspnoea and an adventitious respiratory noise when exercised. The pony had previously been diagnosed with a grade 4 pansystolic cardiac murmur, louder on the right; however, echocardiography had failed to identify any abnormalities.

Clinical findings and diagnosis

On initial examination the pony was bright and alert, had no nasal discharge and had a resting respiratory rate of 12 breaths/min. Thoracic auscultation did not reveal any pulmonary abnormalities, and confirmed the presence of a grade 4 pansystolic cardiac murmur, which was louder on the right hand side. When exercised at walk and trot there was no coughing or tachypnoea initially; however, when the pony was asked to work in an outline, a harsh inspiratory and expiratory noise was audible, along with a slight increase in respiratory rate (24 breaths/min) and sporadic cough.

Palpation of the larynx was grossly abnormal, with an enlargement of the cricothyroid notch on the right hand side. Upper airway endoscopy (Fig 1) revealed rostral displacement of the palato-pharyngeal arch (Cook 1974; Goulden et al. 1976; Wilson et al. 1986; Klein et al. 1989; Blikslager et al. 1999). The condition has been recorded in Thoroughbreds and larger breeds of horses, but does not appear to have been reported in small ponies. This Case Report describes a Welsh section A pony mare diagnosed with 4-BAD.
revealed mucus, plant material and bacteria, consistent with aspiration of feed material. A diagnosis of 4-BAD was made based on these findings.

Exercising endoscopy was performed using an Optomed dynamic respiratory endoscope. Overall, the pony was worked at walk, trot and canter under saddle for 45 min. This initially showed rostral displacement of the palatopharyngeal arch (Fig 3), although dynamic endoscopy revealed that the pony could achieve and maintain full abduction of both the left and right arytenoid cartilages almost immediately upon commencement of exercise (Fig 4). However, when asked to work in an outline dorsal pharyngeal collapse, obscuring approximately half of the cross-sectional area of the rima glottis, was evident.

Discussion

Fourth branchial arch defects consist of aplasia or hypoplasia of one or more of: the wings of the thyroid cartilage, the cricothyroid articulation, the cricothyroid muscles and the cricopharyngeal muscle (Lane 2003). The most common presenting clinical sign is the presence of abnormal noise on exercise (Lane 2003) although some horses may have recurrent aerophagia, sometimes confused with windsucking, resulting from dysfunction of the proximal oesophageal sphincter (Lane 2007).

Diagnosis of 4-BAD has most frequently been made in Thoroughbred horses, although it has been postulated that this may be due to the athletic nature of the breed (Lane 2007) as investigation of an abnormal respiratory noise may not be sought in horses used for less strenuous work unless it is perceived to adversely affect performance. Although a diagnosis of 4-BAD has been reported in other...
larger breeds such as Warmbloods and a Haflinger (Wilson et al. 1986; Deegen and Klein 1987; Klein et al. 1989), there appears to be no published report of small ponies being affected. However, this may be related to the fact that many small ponies are not used for strenuous activities, primarily being used as children’s riding ponies, as in the case reported above.

Diagnosis of a 4-BAD can be made on palpation of the larynx, where it is usually possible to identify a gross abnormality of the cartilaginous structures (Lane 2003). The most frequent finding, as was noted in the current case, is an enlargement of the cricothyroid notch, formed by the overlapping of the wing of the thyroid cartilage and the cricoid ring. In normal horses this space, filled by the cricothyroid ligament, is small; however, shortness of the wing of the thyroid cartilage on the affected side in horses with 4-BAD will result in a marked enlargement of this space.

Upper airway endoscopy performed at rest can be diagnostic in the majority of 4-BAD cases (Lane 2003). Endoscopy will visualise any reduction in arytenoid motility, along with the possible presence of rostral displacement of the palatopharyngeal arch. Although horses with reduced motility of the right arytenoid cartilage may be readily recognised and are likely to be affected with 4-BAD, in those horses affected on the left side, a reduction in left arytenoid motility may be confused with left recurrent laryngeal neuropathy. In addition, rostral displacement of the palatopharyngeal arch may not always be present in such cases, or only a small rim of the displaced palatopharyngeal arch may be visible above the apices of the corniculate processes (Lane 2007). In the current case a complete lack of motility of the right arytenoid cartilage was seen at rest, along with normal function of the left arytenoid. These laryngeal findings are most commonly caused by 4-BAD (Tulleners et al. 1996), and when accompanied by obvious rostral displacement of the palatopharyngeal arch where an open oesophageal aditus is visible, a diagnosis of a 4-BAD can confidently be made (Lane 2003).

If resting endoscopic findings are equivocal, diagnosis of a 4-BAD can also be made during high-speed treadmill endoscopy (Lane 2007). In the current case endoscopy at exercise was performed in order to establish what structures were collapsing into the airway at exercise, as the pony was reported to have exercise related dyspnoea and abnormal respiratory noises. The dynamic respiratory endoscope was used as this allowed the pony to be exercised under saddle at the clinic, in particular working the pony in an outline, as this action appeared to exacerbate the abnormal respiratory noise and tachypnoea. This endoscopic system allowed good visualisation of the pharynx and larynx at exercise, without the vibration commonly seen when performing high-speed treadmill endoscopy, although the current case was not exercised at the high intensity normally associated with this vibration. In addition it was also possible to visualise the upper respiratory tract whilst the pony worked at different paces, both when collected into an outline and when worked with the head and neck outstretched, demonstrating which structures were intruding into the airway at different phases of exercise. The use of dynamic endoscopy was able to demonstrate that despite having a grade 4 right-sided laryngeal dysfunction at rest, the pony was able to achieve and maintain full right arytenoid abduction at exercise. The most likely cause of the cough and increased respiratory rate at exercise in this case was therefore thought to be related to the aspiration of feed material at rest, due to the dysfunction of the proximal oesophageal sphincter, and the dorsal pharyngeal collapse.

**Manufacturer’s address**

1Optomed Endoscopie Veterinaire, Les Ulis, France.

**References**


