CASE REPORT

Large laryngeal vascular malformation in a 5-year-old child

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SUMMARY

A 5-year-old boy was admitted with stridor, which was initially interpreted as subglottic laryngitis. He had a history of prolonged hoarseness and his voice was deep for his age. The stridor persisted despite treatment with epinephrine inhalations and intravenous glucocorticoids. A direct laryngoscopy and blood work up did not support the suspected diagnosis. A MRI was then carried out, followed by angiography. The latter revealed an arteriovenous malformation (AVM) involving the lingual and superior thyroid arteries bilaterally. Owing to the rareness and extent of such an AVM, the patient was referred to Paris for further treatment. This case presents the difficulties in diagnostics and emphasises the importance of diagnostics and multidisciplinary approaches with regard to treatment of AVMs.

BACKGROUND

Arteriovenous malformations (AVMs) are rare in infancy and adolescence. Some remain undetected until late adulthood. Lesions are not restricted to any particular location, however, the head and neck are the more common sites of predilection. According to a study performed by Kobayashi *et al*,¹ the oral cavity (59%) and nasal cavity (35%) were the most common locations for vascular malformations.

Lesions can be subdivided by their cellular and de novo behaviour. Regardless of classification, they remain a diagnostic challenge.

The presented case is an example of a very large AVM with a protracted diagnostic course due to misinterpretation of the symptoms. Furthermore, it emphasises the importance of a multidisciplinary approach to ensure adequate treatment.

We found no cases presenting children with similar malformations in the current literature.

CASE PRESENTATION

A 5-year-old boy was rushed to the local paediatric department due to respiratory problems. For a week he had suffered with a sore throat and a heavy cough. He was seen by the general practitioner, where a rapid antigen test against haemolytic group A *Streptococcus* was negative. Asthma medication was prescribed.

On the night of admission, the patient's respiratory problems had caused syncope and resulted in the parents giving him mouth-to-mouth resuscitation. At admission, he presented with inspiratory and expiratory stridor and the symptoms were initially interpreted as subglottic laryngitis (pseudo-croup). He was treated with epinephrine-inhalation and

glucocorticoids. A chest X-ray and blood tests showed no signs of infection. An echocardiography ruled out a vascular ring anomaly. Initially, the condition improved, but despite treatment the stridor recurred and the hoarseness persisted.

At the local ear, nose and throat (ENT) department, a flexible laryngoscopy was attempted, but was not possible. As general anaesthesia was needed, the boy was transferred to the regional university hospital where an examination under anaesthesia was required for further diagnostics. The initial direct laryngoscopy showed prominent lingual tonsils as well as oedema around the right vocal cord and the subglottic space. No biopsies were performed. Medical treatment continued with epinephrine inhalations, and intravenous hydrocortisone and cefuroxime. The boy was kept intubated and sedated.

A vascular malformation was suspected due to the absence of signs of infection. An MRI confirmed the diagnosis, with lesions involving the floor of the mouth and the larynx. An angiography revealed that the lesions were connected to the lingual and superior thyroid arteries bilaterally and an AVM confirmed as the correct diagnosis. No other malformations were found.

An MRI was performed 4 days after admission. T2-weighted fat-saturated images showed multiple flow-voids with diffuse oedema in the sublingual space continuing down towards the larynx. There was no obvious soft tissue mass. T1-weighted fat-saturated postgadolinium images showed diffuse enhancement in the sublingual space and some of the vascular structures at the same level. Digital subtraction angiogram verified the diagnosis of a high-flow arteriovenous malformation showing large dilated feeding arteries from external carotid branches, mainly the lingual and superior thyroid arteries bilaterally. The nidus was situated in the sublingual space with shunting into multiple dilated abnormal veins in the neck. Some of these veins communicated from the left side to the right, passing through the larynx. [Figures 1 and 2](#) illustrate the malformation.

Attempts to extubate the patient failed and a tracheostomy was necessary.

According to the family, the boy had recurrent hoarseness and a deep voice. Furthermore, his parents had noticed intermittent stridor.

He had allegedly suffered from recurrent tonsillitis and symptoms of mechanical obstruction leading to sleep apnoea. Initially, an adenoidectomy was performed, followed by a tonsillectomy a few months later and approximately 8 months prior to

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Figure 1 (A) Coronal MR T2-weighted fat-saturated image shows multiple flow voids in dilated vessels in the sublingual space (arrow). (B) Axial T2-weighted image shows diffuse high signal changes in soft tissue surrounding the flow voids in the sublingual space (arrow). (C and D) Coronal +axial MR postgadolinium T1-weighted images at the same level as (A and B) show diffuse enhancement in the soft tissue in the sublingual space and enhancement of some of the dilated vessels (arrows).

his admission. He had gone through all normal developmental milestones and had followed the Danish national vaccination programme.

TREATMENT

Owing to the rarity and extent of the malformation, the child was transferred to a neuroradiology department in Paris. The AVM was embolised over several occasions. The child recovered completely, but will require further embolisation, lifelong follow-up and, possibly, surgery.

DISCUSSION

Several types of vascular lesions exist, and treatment still depends on the classification as proposed by Mulliken and Glowacki.² The vascular malformations can be divided into different subgroups according to their origin, that is, capillary, venous and arterial. The malformations can also be divided by their vascular dynamics as 'high-flow' or 'slow-flow'.³

The disorder is congenital and will evolve as the patient ages; starting quiescently (level I), after which it expands (level II) and, finally, causes ischaemia (level III).³ Most AVMs will eventually require intervention.

There have been no previous published cases describing children presenting stridor caused by an AVM in the larynx. The incidence of children presenting stridor due to vascular malformations is not known.

This case clearly demonstrates the difficulties in the management of children with abnormal respiratory sounds, emphasising the risk of misinterpretation and prolonged diagnostics. When a child presents with noisy breathing in primary and secondary care, several considerations need to be made to ensure a correct diagnosis and avoid mistreatment. Boudewyns *et al*⁴ suggest a clinical guide for management of infants and children with noisy breathing. The article divides the different types of respiratory sounds by their origin into 'stridor', 'wheezing' and 'snoring'.

Stridor can represent anything from a benign self-limited disease to an acute life-threatening situation. A rapid evaluation

Figure 2 (A) Lateral right common carotid artery angiogram in the early arterial phase shows a dilated lingual artery (small arrow) and superior thyroid artery (large arrow) supplying a large nidus. (B) Lateral right common carotid artery angiogram in the late arterial phase shows fast shunting into dilated abnormal veins (arrows). (C) Anterioposterior left common carotid artery angiogram in early arterial phase shows dilated lingual artery (arrow) and a large surrounding nidus. (D) Anterioposterior left common carotid artery angiogram in venous phase shows shunting into dilated abnormal veins (long arrow) draining into the right jugular vein and the left jugular vein (short arrow).

of severity is crucial to determine the need for immediate intervention.

Stridor can be divided into acute and chronic. Underlying causes can be:

- ▶ Acquired: foreign bodies and infections (croup and epiglottitis)
- ▶ Congenital: laryngomalacia and vascular malformations such as AVM

Stridor in children is a symptom that should be thoroughly investigated and not a diagnosis.⁴

Inspiratory stridor suggests a laryngeal obstruction, while expiratory stridor implies an obstruction in the lower trachea. Biphase stridor suggests a subglottic or glottis lesion.

According to Boudewyns *et al*, an otolaryngologist should evaluate every child with acute and chronic stridor. This includes a clinical examination and a flexible fiberoptic endoscopy. If the endoscopy is normal or inconclusive, a diagnostic endoscopic procedure in general anaesthesia should be performed.

In this case, the patient's breathing sounds were worsened every time he had an upper respiratory tract infection. With hypertrophic tonsils and snoring, this may have led to misinterpretations and, perhaps, unnecessary procedures.

Among the vascular lesions, the AVMs as presented represent the most challenging cases regarding treatment. The preferred treatment today is embolisation, surgery or a combination of both.

According to Mulliken and Glowacki,² up to 90% of vascular malformations are recognised at birth, but as described by Visser *et al*,⁵ patients tend to be in their 20s when referred for active treatment due to growth of the AVM and progressing symptoms (mean age: 29 years).

Lesions are often external and visible, thus making the diagnosis easier, but internal lesions covered by mucosa can be more difficult to diagnose. In the presented case, the boy's deep voice, tendency to hoarseness and sleep apnoea were interpreted as chronic tonsillitis, and hypertrophy of the tonsils and adenoids. Fortunately, the adenotonsillectomy was performed

Learning points

- ▶ Arteriovenous malformations (AVMs) are rare in children, but should come to mind when dealing with recurrent symptoms that cannot be explained.
- ▶ AVMs are a rare cause of stridor in children, but should, as in other congenital abnormalities, be considered in the differential diagnostic process.
- ▶ Children with stridor may have an unstable airway and stabilising this should be the primary concern. Careful management of the airway requires a multidisciplinary team including anaesthetists, paediatricians and otolaryngologists. Diagnosis is secondary; flexible and direct laryngoscopy may be required.
- ▶ If an internal vascular malformation is suspected, MRI is useful for visualisation of potential lesions, and for further investigation an angiography should be performed to highlight involved blood vessels before the treatment modality is chosen.
- ▶ Treatment of head and neck AVMs should be discussed in a multidisciplinary setting including ear, nose and throat-surgeons and interventional radiologists.

without any complications. Accidental damage of the lesions could have led to severe, possibly fatal bleeding.

The prevalence of dysphonia in children is 6–9%.⁶ When treating children presenting with dysphonia, several aspects should be considered. Usually, the cause is benign and self-

limiting, such as an infection or inflammation, but in prolonged cases, a full ENT examination should be carried out. The chronology of symptoms is essential for the underlying diagnosis. If symptoms have persisted since birth, a congenital cause such as laryngeal cyst, cleft or web, is more likely.⁷

The fact that head and neck lesions remain undiagnosed for years may be due to the uncharacteristic symptoms, and is emphasised by the case presented by Chimona *et al*,⁸ where a 52-year-old woman was diagnosed with an AVM of the floor of the mouth after she presented with dysphagia and dysarthria of 12 months' duration.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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