Anaesthesia and juvenile hyaline fibromatosis

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Summary

Juvenile hyaline fibromatosis is a rare autosomal recessive disease characterized by large cutaneous nodules, especially around the head and neck and often involving the lips. The effects become increasingly severe with age and also include joint contractures, gingival hypertrophy and osteolytic lesions. We describe the anaesthetic management of two sisters with this disease. Safe maintenance of a patent airway is the principal anaesthetic challenge. ([Br. J. Anaesth. 1996, 76: 163–166])

Key words

Anaesthesia, paediatric. Complications, juvenile hyaline fibromatosis.

Two sisters with juvenile hyaline fibromatosis (JHF) presented for surgery several times at our hospital. The older sister had received her first anaesthetic at 14 months of age and subsequently had three further anaesthetics. On each occasion management of the airway had become more difficult. The younger sister had received one anaesthetic at 2 yr of age when tracheal intubation was relatively easy.

For both sisters other aspects of anaesthesia and postoperative care were straightforward. For each anaesthetic they were premedicated with EMLA cream and atropine 20 µg kg⁻¹ i.m. Despite the presence of joint contractures, venous cannulation was performed successfully. When the airway was secured, the lungs were easy to ventilate with normal airway pressures and oxygenation. With the exception of the elder sister’s second anaesthetic, when surgery was of short duration and a laryngeal mask was used, the sisters were given atracurium 0.5 mg kg⁻¹ i.v. and the lungs ventilated. The patients remained in a stable cardiovascular state during anaesthesia. After anaesthesia the trachea was extubated with the patients awake without difficulty and they required only mild postoperative analgesia.

Case reports

PATIENT NO. 1

The older sister had her first anaesthetic at 14 months of age to enable tissue biopsies to be performed. At that age she already had very abnormal facies with polyplloid masses around the mouth and limited mouth opening. She also had large irregular growths on her head and back so that padding with soft packing material was required to position her for anaesthesia and surgery. Anaesthesia was induced with oxygen and halothane, and suxamethonium 1 mg kg⁻¹ was given i.v. and her trachea intubated without difficulty. Her subsequent anaesthetic was uneventful.

At 20 months of age she presented for further tissue biopsies. Anaesthesia was induced with oxygen and halothane and maintained with spontaneous breathing. Laryngoscopy was performed under deep halothane anaesthesia to assess the difficulty of intubation. It was impossible to view the epiglottis as the jaw, tongue and pharynx were infiltrated and rigid. A size 2 laryngeal mask was inserted with some difficulty; the mouth opened sufficiently to admit the mask but it took several attempts to pass it downwards behind the tongue. After insertion the laryngeal mask was used to maintain the airway with spontaneous ventilation for the remainder of the anaesthetic.

At 3 years 3 months she presented for removal of lesions around her mouth and trimming of deposits on the trunk which were beginning to ulcerate. On examination she had a grossly abnormal face with marked protuberance of her maxilla caused by infiltrations (fig. 1). Her mouth was open and would admit one finger but the tissues in and around the mouth were rigid. Her lower jaw was hard with infiltrations. She had a good air flow through her right nostril. Anaesthesia was induced with oxygen and halothane. A size 3.0-mm tracheal tube was introduced gently into the right nostril and used as a nasal airway to maintain inhalation anaesthesia while examination with a fiberoptic intubating bronchoscope was performed via the left nostril. The left nasal passage was grossly abnormal and very narrow, with hyaline deposits throughout. The epiglottis was infiltrated and looked abnormal, as did the larynx which was narrowed. It was impossible to identify the arytenoids. Intubation was performed successfully by passing a 4.0-mm tracheal tube over the fiberoptic bronchoscope.

Five months later she again presented for anaesthesia for surgical treatment of sacral ulceration and extensive debridement of facial and mouth lesions. A computed tomography scan of the airway was obtained before operation and showed significant narrowing of the pharynx and trachea (figs 2, 3). The same anaesthetic technique was used as in the previous anaesthetic. The airway viewed with the
fibreoptic bronchoscope was much narrower and more tortuous than on the previous occasion. However, it did lead directly to the larynx. This was identified as a small black orifice which appeared and disappeared with breathing. There were no identifiable anatomical landmarks as these were obscured by hyaline deposits. It was clear that there had been significant progression of the infiltration since the last anaesthetic. As previously, intubation was achieved by passing a 4.0-mm tracheal tube over the fibreoptic bronchoscope.

PATIENT NO. 2

The younger sister presented for anaesthesia at 2 yr of age for surgical removal of lesions around the mouth. Although she had obvious facial evidence of hyaline deposits she had reasonable mouth opening. A potentially difficult intubation was anticipated as there was no indication of whether or not there might be deposits in and around the pharynx and larynx. Anaesthesia was induced with oxygen and halothane, and laryngoscopy was performed under deep inhalation anaesthesia. The lower jaw was stiff and heavily infiltrated. A straight blade enabled a view of the arytenoids but not the cords. The pharynx appeared normal. A size 3.0-mm oral tracheal tube was passed easily. Fibreoptic review of her nasal passages revealed no abnormality and a 3.0-mm nasal tracheal tube was passed while the oral tracheal tube was removed, so facilitating surgical access.

Discussion

A variety of names have been given to JHF which was first described by Murray in 1873 as “peculiar cases of molluscum fibrosum” [1]. Other names include puretic syndrome, disseminated painful fibromatosis and fibromatosis hyalinica multiplex juvenilis [2]. Infantile systemic hyalinosis is a similar condition but has an earlier onset [3]. JHF is a rare autosomal recessive disorder with variable penetrance. One-third of affected children...
are siblings and are often born to consanguineous marriages [2]. Clinical features develop in the first few years of life and include: multiple large slow growing cutaneous nodules on the head and neck, which involve the lips and are also found elsewhere on the body, joint contractures, gingival hypertrophy and osteolytic lesions [4–6]. Intellect is often normal [2]. Infiltration of areas such as around the mandible have, as in the two children described here, led to hard unyielding tissues with limited mouth opening. Histologically the lesions consist of an eosinophilic hyaline matrix surrounding well differentiated fibroblasts [4, 5]. Previously described postmortem findings in the related disease infantile systemic hyalinosis include hyaline deposits in the skin, trachea, throughout the gastrointestinal tract, the heart, lungs, skeletal muscle, thyroid and adrenal glands [7]. Death, which occurs in the first few decades of life, may be caused by infection and there is predisposition to staphylococcal infection of skin lesions [2, 3].

There have been two previous reports of anaesthetic management of JHF. Sugahara and colleagues [8] reported a 6-yr-old with JHF whose trachea was intubated successfully. Vaughn and colleagues [9] reported a 13-month-old child with JHF whose trachea was intubated with the aid of a guide wire passed through the suction port of a fiberoptic bronchoscope. Subsequent laryngoscopy revealed the epiglottis and posterior arytenoids; however, difficulty was noted in seeing the vocal cords.

Our case reports illustrate not only the airway difficulties that may be encountered with this disease, but also that as the disease progresses these are likely to become more difficult in subsequent anaesthetics. This is also the case in other diseases which are characterized by accumulation of abnormal material in the tissues. These diseases include the mucopolysaccharidoses such as Hurler’s syndrome [10].

Preoperative assessment of our patients included a search for any co-existing disease but systemic manifestations resulting from hyaline deposits in organs such as the heart, lung and kidney were not present. A computed tomography scan of the pharynx was found to be useful in predicting a difficult airway.

A variety of techniques were used in the anaesthetics reported here, both as the elder sister’s disease developed and as experience was gained in anaesthetic management of JHF. In all instances gaseous induction was used as it was considered the safest method to achieve intubation while the airway was unsecured.

For the elder sister’s first anaesthetic, suxamethonium was used for intubation because its rapid onset allowed the airway to be secured quickly. Its rapid offset would have been useful if neither intubating nor ventilating the patient’s lungs with a face mask had proved possible during paralysis. However, it was not found to be necessary to use suxamethonium in any subsequent anaesthetic. Atracurium and positive pressure ventilation were used to avoid hypoventilation. Atracurium was used also in subsequent anaesthetics for the same reason. The one exception was the elder sister’s second anesthetic. In this anaesthetic laryngoscopy was attempted to assess the difficulty of intubation. When laryngoscopy proved impossible a laryngeal mask was used with spontaneous ventilation because the surgical procedure planned was short (approximately 20 min) and did not involve the mouth or face. Spontaneous ventilation was adequate for this period of time.

For the elder sister’s third and fourth anaesthetics it was planned to use an intubating fiberoptic bronchoscope from the outset; laryngoscopy in the previous anaesthetic had proved impossible and use of a laryngeal mask was precluded because surgery on the mouth was planned. Under these circumstances the fiberoptic bronchoscope, used while a nasal airway allowed spontaneous ventilation, was found to be invaluable for intubation. For the younger sister’s anaesthetic, intubation using laryngoscopy was achieved although the fiberoptic bronchoscope was still used to examine the airway.

Although the fiberoptic intubating bronchoscope was useful to view the airway and to secure it by facilitating intubation, there are limitations to its use. With young children, such as our patients, it is not practical to use it for an awake intubation because of the distress caused. It cannot be used for railroading a tracheal tube into the trachea if the internal diameter of the tracheal tube required is less than the diameter of the bronchoscope available, although using a guide wire passed down the suction port is an option [9]. In addition, the fiberoptic bronchoscope needs to be used by an anaesthetist experienced in its use.

Alternatives to using a fiberoptic bronchoscope would have included blind nasal intubation and a bougie inserted down a laryngeal mask. However, blind nasal intubation may cause bleeding, especially in an abnormal airway, while intubating using a fiberoptic bronchoscope caused no identifiable haemorrhage in the cases described here. Although no attempt was made to pass a laryngeal mask in the elder sister’s last two anaesthetics, it is likely, in view of the findings with the fiberoptic bronchoscope, that a laryngeal mask would have been impossible to insert because of infiltrations in the airway.

Postoperative care was uneventful in the cases described and pain was controlled with mild analgesics. However, there is probably an increased risk of postoperative airway obstruction in patients with JHF and this may be enhanced if postoperative analgesia is required.

In conclusion, JHF, in common with other diseases associated with accumulation of abnormal material in the soft tissues, may cause increasingly severe airway problems with age. Anticipating and preparing for a difficult airway is probably more important than using any particular anaesthetic technique. However, it is recommended that the facilities for fiberoptic intubation and an experienced paediatric anaesthetist are present for all such cases.

References

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