Case Report

Anaesthetic management of a child with stone man syndrome: Look before you leap!

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ABSTRACT

Stone Man syndrome or fibrodysplasia ossificans progressiva (FOP) is an extremely rare (1 in 2 million) genetic disorder characterised by ectopic ossification of the skeletal and connective tissues leading to progressive fusion of axial and appendicular skeleton. Surgery and anaesthesia-induced trauma can lead to disease flare-up if due precautions are not taken and disable the patient further. However, rarity of the disease may lead to its common misdiagnosis and anaesthesiologist may be caught unaware. There is relative paucity of literature regarding anaesthetic management of children with FOP. Videolaryngoscopes (VLs) provide a non-line-of-sight view and require less anterior force to visualise the glottis, may provide an alternative to fibreoptic intubation for airway management in such cases. Use of VL has only been reported once in an adult with FOP for nasotracheal intubation. We describe the successful anaesthetic management of an 11-year-old child with FOP and anticipated difficult airway.

Key words: Fibreoptic intubation, fibrodysplasia ossificans progressiva, heterotopic ossification, videolaryngoscope

INTRODUCTION

Fibrodysplasia ossificans progressiva (FOP), also known as stone man syndrome, is a severely disabling and catastrophic-inherited disorder of connective tissue characterised by congenital malformation of the great toes, thumbs and vertebrae associated with progressive ossification of striated muscles.[1,2] In such patients, progressive fusion of axial and appendicular skeleton, temporomandibular joint (TMJ) ankylosis, associated restrictive lung disease and sensitivity to even trivial oral trauma make airway management challenging.[1-4]

CASE REPORT

An 11-year-old, 30 kg male child, suspected to be a case of cysticercosis, presented with multiple hard swellings over the nape of neck, paraspinal region, arms, thighs and legs. He was posted for excision biopsy of the back swellings. He had received antitubercular treatment and albendazole with no remission. Pre-operative airway evaluation revealed a Mallampati class III, restricted neck movement (45°) and mouth opening (MO) of 2.5 cm. Cardio-respiratory examination and all routine investigations including chest X-ray and electrocardiogram (ECG) were found to be normal. In the operation room, ECG, non-invasive blood pressure, end tidal carbon dioxide and pulse oximeter were attached. Intravenous (IV) access was secured and injection fentanyl 60 μg was given. The only fibreoptic bronchoscope (FOB) in the department was not working. Hence, Truview PCD™ video laryngoscope (Truview video laryngoscope [TVL], Netanya, Israel)-guided intubation was planned. Difficult airway cart was kept ready. Anaesthesia was induced with sevoflurane 5%–8% (1.5 minimum alveolar concentration) in oxygen and supplemented with propofol 30 mg. During laryngoscopy with TVL, a Cormack and Lehane (CL) Grade 3 of glottis was

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obtained which improved to CL 2b with optimum external laryngeal manipulation. A tube introducer could be inserted into the glottis, and a size 6.0 flexo metallic endotracheal tube was threaded over it. Thereafter, muscle relaxation was achieved with injection atracurium 10 mg, and the patient was positioned prone for surgery. Adequate padding was applied to eyes, bony prominences, deformities of upper limbs (shoulder and elbow) and swellings of lower limbs. During excision biopsy, the surgeon reported bony tissue in the nodules and got suspicious of FOP. The surgery was abandoned, the patient was made supine and his trachea was extubated after reversal of neuromuscular blockade. The patient was shifted to post-operative room for close observation. Injection diclofenac sodium 50 mg was given as slow IV infusion for post-operative pain. Subsequent histopathology of the biopsy tissue revealed mature osseous tissue, audiometry showed conductive hearing loss and skeletal survey revealed ectopic bone formation over the back, neck, upper limbs and lower limbs, thus confirming the diagnosis of FOP [Figures 1 and 2]. The patient was discharged home 2 days later.

DISCUSSION

FOP (myositis ossificans progressiva) is autosomal dominant connective tissue disorder characterised by extra skeletal endochondral ossification. It may be associated with mutation in bone morphogenic protein4 (BMP4) antagonist gene which leads to increased production of BMP.[2] It is characterised by progressive heterotopic bone formation in the connective tissue and skeletal muscle. The ossification usually starts in the neck, spine and shoulder girdle and progressively immobilises all the joints of the axial and appendicular skeleton during the first decade of life.[1,2] Conductive hearing loss due to ossification of bones of internal ear is also common.[2]

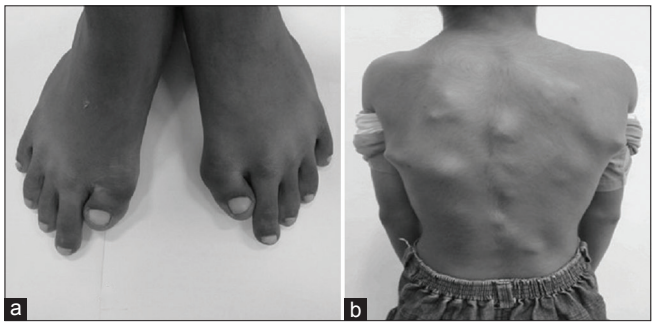

Figure 1: Classical signs of fibrodysplasia ossificans progressiva in our patient: Great toe malformation (a) and extensive heterotopic ossification on the back (b)

Minor trauma, biopsy, surgical intervention, etc., may flare up the disease.[3] Hence, one should avoid deep intramuscular (IM) injections, injections into jaws for dental procedures, invasive biopsy, excision procedures for heterotopic masses and manipulations of stiff joints to prevent disease progression.[2-4] These patients have a median life span of 40 years and usually die due to complications of thoracic insufficiency syndrome or pneumonia.[5]

Anaesthetic management of these patients is complicated due to anticipated difficulty in airway management (cervical spine fusion and TMJ ankylosis), restrictive lung disease and their extreme sensitivity to trauma.[6,7,8] The common interventions during anaesthesia such as traumatic IV and arterial cannulation, IM injections, overstretching of joints during positioning and regional blocks can precipitate iatrogenic heterotopic ossification and any such intervention should be avoided.[3,4,6]

In early stages, patients may not have any symptom, and since it is a rare entity, the diagnosis may be missed.[3] This patient had bony hard swellings with restricted movements and was misdiagnosed as calcified muscular cysticercosis initially. During surgery when hard bone-like tissue was found in the nodule during dissection, FOP was suspected. Subsequently, a diagnosis of FOP was confirmed on the basis of two classical features, namely congenital great toe malformations and heterotopic ossification in specific anatomic patterns [Figures 1 and 2] and mature bone tissue on histopathology of the biopsy tissue.

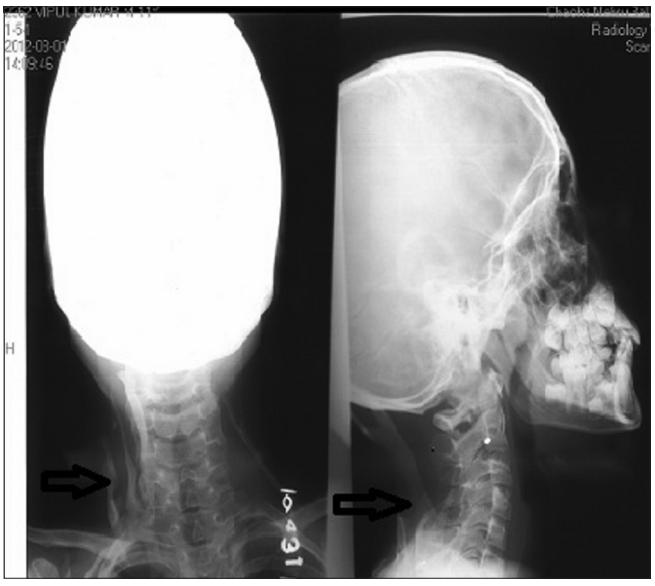

Figure 2: Head and neck X-ray with linear ossification of the paraspinal muscles

Difficult airway and cannot ventilate cannot intubate situation needing emergency tracheostomy have been reported in FOP.[9-12] Direct laryngoscopy and manoeuvres-like jaw thrust may cause ossification and ankylosis due to overstretching of TMJ.[1,2] Hence, overstretching of the oropharyngeal structures should be prevented by avoiding jaw thrust and ensuring that the MO during laryngoscopy and intubation remains well below the base line values obtained prior to the anaesthetic.[9,12] Awake fibreoptic intubation is considered the gold standard even in patients with adequate MO and jaw movement to avoid any stimulus to TMJ.[7-10] To prepare the airway for awake intubation, nebulisation, spray as you go or direct instillation of local anaesthetic is preferred over airway blocks. Elective tracheostomy should also be avoided as ossification of the incision site may result in airway obstruction on decannulation.Our patient had a potential difficult airway (restricted MO and neck movements), was uncooperative and the only FOB in our setup was not working. Anterior larynx with reduced MO invariably requires excessive force during conventional laryngoscopy, which may be disastrous in a patient with FOP.[13-15]

Successful use of Glidescope™ has been mentioned in one case for nasotracheal intubation of an adult FOP patient (having a normal MO) in a case series of 30 patients, but use of VL has never been described for a child.[9]

CONCLUSION

Anaesthetic management for FOP is extremely challenging, and clinical interventions that may put patient at a risk of new localisation of heterotopic ossification leading to substantial impact on their quality of life need to be diligently avoided. VL assisted with bougie may be considered as an alternative to FOB for airway management of these patients.

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Conflicts of interest

There are no conflicts of interest.

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