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Clinical Record

Parapharyngeal abscess in a previously tonsillectomised child with periodic fever, aphthous stomatitis, pharyngitis and adenitis syndrome: first reported case

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Abstract

Objective: we present the first reported case of parapharyngeal abscess in a child with periodic fever, aphthous stomatitis, pharyngitis and adenitis syndrome, an uncommon syndrome of recurrent, self-limiting fever in children.

Method: Case report and review of the literature to date concerning periodic fever, aphthous stomatitis, pharyngitis and adenitis syndrome.

Results: Periodic fever, aphthous stomatitis, pharyngitis and adenitis syndrome is an incompletely understood syndrome which requires careful and thorough investigation in order to distinguish it from other causes of recurrent fever. There has been much recent debate in the literature over the merits of various treatment strategies, including tonsillectomy.

Conclusion: To the authors' knowledge, this is the first reported case of a parapharyngeal abscess in a child with periodic fever, aphthous stomatitis, pharyngitis and adenitis syndrome, made more significant by the fact that the child had undergone tonsillectomy one year prior. This case provides evidence that tonsillectomy does not protect against one of the more serious complications of oropharyngeal infection in children with this syndrome.

Key words: PFAPA Syndrome (Periodic Fever; Aphthous Stomatitis; Pharyngitis; Adenitis); Periodic Fever Syndromes; Paediatric Fever Syndromes; Parapharyngeal Abscess

Introduction

The PFAPA syndrome (Periodic Fever, Aphthous Stomatitis, Pharyngitis and Adenitis) is a recently described and incompletely understood disease. It is characterised by recurrent fevers which arise suddenly and last several days, before spontaneously remitting with equal suddenness. The above-mentioned symptoms are commonly seen, although not all are present in each episode, and others may also feature. Episodes characteristically begin in early childhood, and do not persist beyond the teenage years. The recent literature has contained much debate regarding the potential causes of this condition and the effectiveness of various treatments, particularly the role of tonsillectomy.

Case report

A five-year-old girl presented to her local general hospital with a one-week history of fever, malaise and right ear pain, unresolved with antibiotic treatment commenced by her primary care physician, together with a 24-hour history of painful, stiff neck held turned to the right.

The child had been extensively investigated one year previously at Great Ormond Street Hospital, London, where she had been given a diagnosis of PFAPA syndrome; she had also undergone adenotonsillectomy. Since that time, she has been free of PFAPA syndrome type episodes.

The patient was admitted under the care of the paediatricians for intravenous antibiotics and investigation.

Laboratory results were consistent with an infectious process. Ultrasound scanning of the neck was performed, showing a probable parapharyngeal abscess. A computed tomography scan of the neck was undertaken, for further anatomical evaluation, and showed an abscess lying mainly in the right parapharyngeal space and tracking into the retropharyngeal space (Figure 1).

The patient was transferred to the regional ENT service, where examination under anaesthesia showed a characteristic bulge in the oropharynx (Figure 2). Incision and drainage of the abscess was therefore undertaken via the trans-oral approach and approximately 10 ml of pus obtained. A nasogastric tube was placed in theatre, and the patient was nursed nil-by-mouth with intravenous hydration for 24 hours.

Recovery was unremarkable, and the patient was discharged home three days after the procedure.

Discussion

First described by Marshall *et al.* in 1989,[1] PFAPA syndrome is one of the less common paediatric recurrent fever syndromes.[2] The cardinal symptom is recurrent, abrupt episodes of fever lasting several days, followed by an entirely asymptomatic interval period.[1] Other

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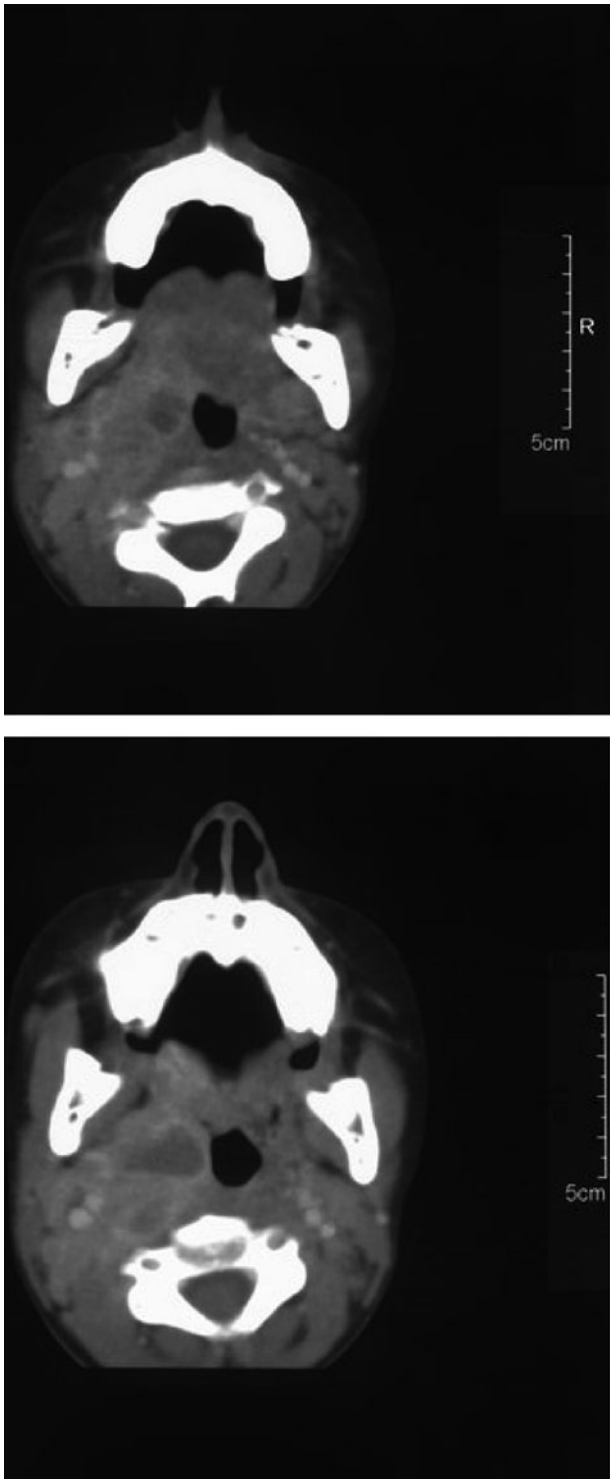

FIG. 1
Axial computed tomography images showing abscess formation in the right parapharyngeal space.

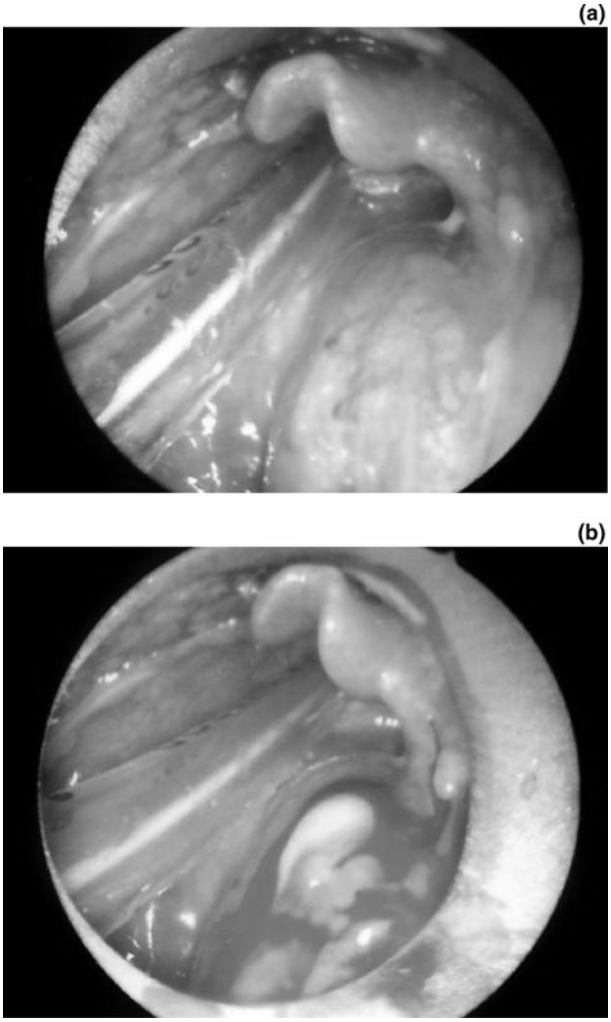

FIG. 2
Operative photographs showing oropharyngeal abscess (a) before and (b) after incision.

common symptoms include aphthous stomatitis, pharyngitis, cervical lymphadenopathy, abdominal pain, headache, nausea, myalgia and diarrhoea.[3] Classically, recurring episodes begin before the fifth birthday[1] and recurrences occur every three to eight weeks,[1,4,5] initially with clockwork regularity in early childhood but tending to become less frequent and less severe over time.[4] The syndrome rarely persists into adulthood.[1,6–8] The child's growth and development are normal, and there are no recognised long term sequelae.

The diagnosis is generally one of exclusion, requiring differentiation from other recurrent fever syndromes, including cyclical neutropenia, familial Mediterranean fever and hyper-immunoglobulin D (IgD) syndrome. Cyclical neutropenia is a disorder associated with fever, malaise, adenopathy and abdominal pain, and is characterised by a severe fall in peripheral neutrophil count every 21 days,[1,2,4,5] making it readily distinguishable from PFAPA syndrome. Familial Mediterranean fever is an autosomal recessive disease largely limited to four Mediterranean ethnic groups (Turks, Arabs, Armenians and non-Ashkenazi Jews) and characterised by recurring episodes of self-limiting fever.[2,4,5] A common gene, MEFV, has been identified.[5,9] Hyper-immunoglobulin D syndrome is a recessively inherited condition typified by recurrent fevers, arthritis, adenopathy and rash, in association with persistent, polyclonal elevation of serum IgD levels.[2,4,5,10] Other, less common periodic fever syndromes (familial Hibernian fever, familial periodic fever, etc) must also be excluded. Due to this diagnostic complexity, the diagnosis is generally made in the secondary or tertiary paediatric

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setting, often by those with an interest in infectious disease. Our patient had been extensively investigated to rule out the above conditions before the diagnosis of PFAPA syndrome had been made.

The cause of PFAPA syndrome remains undetermined. Initial theories based around genetic mutation and chronic infection have been largely unsupported by subsequent work.[4,9–12] More recent literature has raised the possibility of immune dysregulation as a significant factor.[13–16]

The PFAPA syndrome may come to the attention of the otolaryngologist through referral for recurrent stomatitis or for recurrent pharyngitis with a view to tonsillectomy,[4,17] with or without a definitive diagnosis.

There has been debate in the recent literature over the benefit of various treatment strategies; steroid treatment,[3,18] cimetidine[2,7,8] and thalidomide[15] all have their proponents and detractors. Most recently, however, particular attention has focussed on debating the role of tonsillectomy.[3,4,8,17] Various studies have quoted success rates from 0 to 100 per cent,[16] and a corresponding spectrum of opinion has been voiced within the otolaryngology community regarding the benefits of this approach.

Whilst several of the case series cited above have shown impressive results for one or other of these therapeutic strategies, their numbers have been small, their results not easily reproducible, and there has been a paucity of properly controlled studies. As a consequence, the majority of recommendations for various treatment modalities – tonsillectomy included – are based on low levels of evidence.

Deep neck space infection is well known as a complication of acute tonsillar and para-tonsillar infection, and it is generally accepted that removal of the tonsils substantially reduces the risk of this condition developing. Our patient had undergone adenotonsillectomy one year prior to the episode described above, and had not suffered a PFAPA syndrome type episode in the meantime. Why she developed a recurrence (and subsequently an abscess) after this time is not known.

Deep neck space infection has not previously been reported in connection with PFAPA syndrome. The current report widens the spectrum of potential complications of this syndrome, as well as adding to the ongoing debate over the merits of tonsillectomy in the treatment of this poorly understood condition.

- **The PFAPA syndrome is an incompletely understood syndrome of recurrent, self-limiting fever in children**
- **Current opinion is divided over the merits of tonsillectomy in the treatment of this condition**
- **Deep neck space infection is a recognised complication of tonsillar and parapharyngeal infection**
- **This report represents the first published case of deep neck space infection in a child with PFAPA syndrome**
- **By showing that tonsillectomy does not prevent one of the more serious complications of oropharyngeal infection in children with this syndrome, this report adds weight to the debate over the merits of tonsillectomy in this condition**

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