Emergency dental management of a paediatric patient with Fanconi anaemia

Sanghvi, R. M., de Souza, M.
Paediatric Dental Department, Guy's and St Thomas' NHS Foundation Trust

Introduction

Fanconi anaemia is a rare genetic condition which is mostly inherited through an autosomal recessive pattern. The worldwide prevalence is 1:160,000.[1]

It is characterised by progressive bone marrow failure and is the most common inherited form of aplastic anaemia. Due to this, patients often present with anaemia, neutropenia and thromboyctopenia.[2]

Patients commonly display congenital abnormalities and may present with hypopigmentation, café-au-lait spots, cardiac anomalies, malformed thumbs, malformed ears and hearing loss.[2]

Patients with Fanconi anaemia are also predisposed to cancers, with a 10-30% risk of malignancy.[1] In particular, patients are at risk of developing acute myeloid lymphoma and head, neck or skin tumours amongst others.[2]

Presenting complaint

A 11-year-old boy presented to the paediatric department complaining of recurrent lip sores. Medically he was diagnosed with Fanconi anaemia.
He had no previous experience of dental treatment and was dentally anxious.

Extra-oral examination revealed generalised pallor (Figure 1). Angular chelitis was evident on the corners of the mouth. The lips appeared dry and cracked secondary to a lip licking habit.

Intra-oral examination revealed extensive caries affecting the LLE (Figure 2). Microdontia was also evident affecting both primary and permanent teeth.

Clinical Management*

Visit 1 10/03/20	• Saliva sampling • Lanolin cream recommended • Letter sent to haematology team for extraction protocol
Plan for next visit (cancelled due to Covid-19)	• Separator placement for proximal caries assessment between primary molars to avoid radiographs • Extraction LLE under LA (local anaesthetic)
Emergency A&E telephone triage 26/05/20	• Parent contacted department via telephone due to severe pain LLE • History taking conducted over telephone • Photographs sent via email to rule out any abscess or spreading infection which would require urgent attention in light of neutropenia • Covid-19 screen conducted • Discussion regarding social distancing protocols. Patient advised to travel to hospital via car and await call from department before entering hospital • Urgent contact made with haematology team to review up to date blood profile and provide extraction protocol • Contact with general medical practitioner (GMP) for prescription of pre and post-operative tranexamic acid
Visit 2 02/06/20 (during Covid-19 pandemic)	• Extraction LLE under LA using non-pharmacological behavioural management techniques • Local haemostatic measures implemented
Telephone follow up 09/06/20	• Telephone follow up to assess for any complications to avoid visit to hospital

Figure 1.** Extra-oral view of patient demonstrating pale skin and lip tone representing anaemia. Lip drying and cracking is also evident.

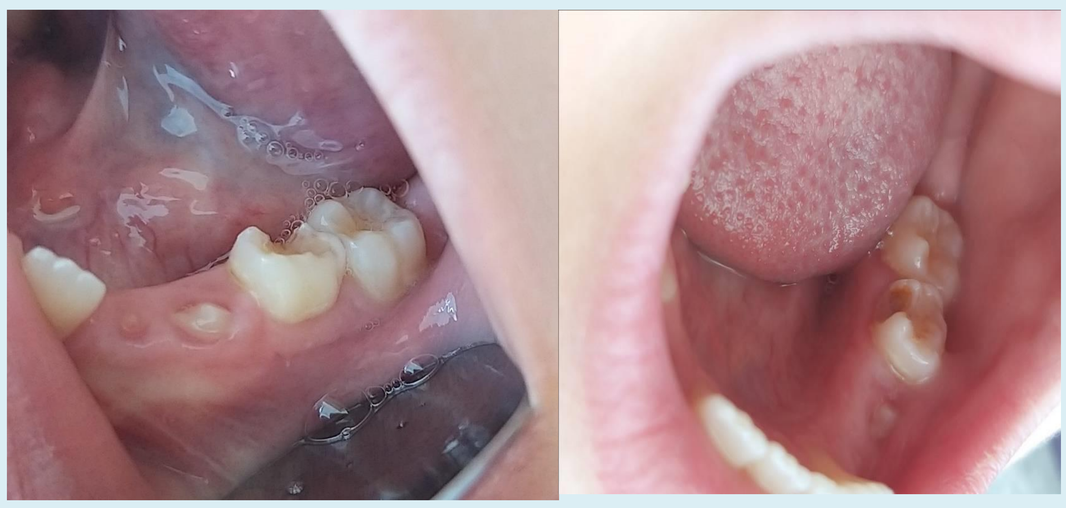

Figure 2.** Intra-oral images showing occlusal-lingual caries affecting the LLE with no evidence of gingival swelling or abscess.

Discussion

Patients diagnosed with Fanconi anaemia are at increased risk of infections and bleeding. This alongside their increased cancer risk means that precautions for dental treatment are required.

Alternative methods for diagnosis of proximal caries in the form of temporary tooth separation were implemented in this case to avoid radiographs. Furthermore, it was ensured that an extraction protocol was in place through teamwork with haematology and the GMP prior to carrying out any treatment

The Covid-19 pandemic has required cancellation of elective dental treatment. Due to the patient's symptoms and risk of infection, emergency treatment was required. Measures were taken to reduce the Covid-19 risk in a shielding patient including history taking over telephone, reducing time spent in the department, implementing social distancing and appropriate PPE use.

The patient was extremely dentally anxious and due to restrictions at the time on the use of inhalation sedation, non-pharmacological behavioural management techniques, particularly tell-show-do, positive reinforcement and enhancing control were used.

Conclusion

This case demonstrates safe and successful urgent management of a patient with Fanconi anaemia. It demonstrates a staged treatment planning approach using methods to ensure patient safety. Furthermore it highlights the importance of multidisciplinary teamwork and implementation of altered management strategies due to Covid-19.

References

1. Genetics Home Reference. Fanconi anemia. https://ghr.nlm.nih.gov/condition/fanconi-anemia#:~:text=Fanconi anemia occurs in Spain%2C and black South Africans. Published 2020. Accessed July 14, 2020.
2. Auerbach AD. Fanconi anemia and its diagnosis. *Mutat Res*. 2009;668(1-2):4-10. doi:10.1016/j.mrfmmm.2009.01.013

***** Management of this case was carried out by the lead author
** Parental consent for photographs gained