Hypodontia in a Patient With Actinic Prurigo: A Case Report

Caroline Margaret Campbell, MSc, BDS, MFDS, RCPS, M Paed Dent Jillian Margaret Phillips, BDS, MFDS, RCPS Marie Therese Hosey, DDS, MSc, BDS, FDS, RCPS

ABSTRACT

Actinic prurigo is a rare form of light sensitivity not previously described with hypodontia in the dental literature. The purpose of this report was to describe both the medical and dental management of a 12-year-old boy presenting with actinic prurigo, hypodontia, crowding, caries, and tooth tissue loss. The phases of dental treatment and the multidisciplinary treatment of hypodontia are discussed. (J Dent Child 2009;76:156-60)

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ctinic Prurigo (**AP**) is an idiopathic photodermatoses. It is one of a group of skin disorders primarily caused or exacerbated by exposure to UV or visible light.¹ It affects mainly sun-exposed areas of the skin and can present as erythematous papules and macules that may merge to form plaques, crusts, hyperpigmentation, and lichenification.² Lower lip involvement is common and often severe², and may be the condition's only manifestation.³ The vermillion may show swelling or edema, scaling, fissures, ulcers, and crusting.^{2,4} A lip biopsy can show distinctive histological features.^{2,3}

AP tends to run a chronic course and occurs most commonly in the Mestizo (mixed Indian and European ancestry) populations of Latin America.⁴ There are few cases seen in Britain, with a prevalence of 3.3 per 100,000 in Scotland.⁵ AP has an early onset and is more common in females, with a ratio suggested of 2:1.⁴ A strong association of human leukocyte antigen (**HLA**) and AP has been reported, suggesting an immunological element to the disease. A study by Menage et al 1996 showed a highly significant association of Class II antigen HLA-DR4 in Caucasian AP patients.⁶ Among British Caucasian AP patients, HLA-DR4 is present in 90% with the DRB1*0407 subtype present in 60%, compared to 30% and 4% to 6%, respectively of the general population.⁷

Treatment is difficult and several regimes have been suggested, such as avoidance of sun and use of sunscreens, antihistamines, Vitamin E, beta-carotenes, systemic corticosteroids, and antibiotics.²⁻⁴ Thalidomide has been shown to provide a dramatic improvement. In fact, this response to thalidomide may be an indicator in the diagnosis of AP.^{2,4}

Hypodontia is a dental anomaly, and is one of the most common anomalies found in the human dentition, with an incidence ranging from approximately 4% to 7%, excluding third molars which are missing in 9% to 30% of individuals.⁸ Commonly missing teeth are the mandibular second premolars, maxillary lateral incisors, and maxillary second premolars: This is known as incisor-premolar hypodontia.⁹ The permanent mandibular central incisors are less commonly missing, with a prevalence of less than 1%.¹⁰

Dr. Campbell is specialist registrar, Dr. Phillips is Senior House Officer, and Dr. Hosey is reader/honorary consultant, all in the Pediatric Dentistry Department, Glasgow Dental Hospital and School, University of Glasgow, Glasgow, Scotland, UK. Correspond with Dr. Campbell at <u>caroline.campbell@</u> northglasgow.scot.nhs.uk

CASE REPORT

In July 2006, a 12-year-old boy was referred to the Department of Child Dental Health at Glasgow Dental Hospital and School, Glasgow, Scotland, UK, for treatment regarding the congenital absence of his permanent mandibular central incisors. He was not especially concerned about his teeth's appearance. He did, however, prefer an improvement in the appearance of his front teeth in the long term. Medically, he has AP. He attended his local district hospital under the care of a consultant dermatologist. Additionally, on an annual basis he also attended a regional specialist photobiology unit under the care of a professor in dermatology. He is the second eldest of 4 siblings with no positive family history of either AP or hypodontia.

MEDICAL MANAGEMENT

AP was initially diagnosed at the age of 4 years, after a family holiday abroad. The patient has been mainly affected over the face and forearms, with some involvement of the upper lip. There has been no involvement of the nose. His treatment has been avoidance of sunlight, wearing SPF50 E45 sunscreen and long-sleeved clothing, with moderate use of topical steroids on his head and neck, and occasional use of strong topical steroids on his arms. In the past, a course of desensitization with artificial light has helped his condition considerably. He was not aware of exacerbating factors or of triggering through window glass or clothing. The patient has developed a rash a few hours after exposure to sunlight, which has lasted for days. The rash has consisted mainly of itchy papules with no evidence of blistering. The patient's AP is improving with time, although it is unclear whether this is due to desensitization light therapy or to spontaneous clinical improvement.

DENTAL MANAGEMENT

Extraoral examination demonstrated AP involvement of the face, with minimal lip involvement (Figure 1). Intraoral examination revealed that the patient had the following teeth missing: the permanent mandibular central incisors; permanent maxillary right second molar; and all permanent third molars. From an esthetic viewpoint, a midline diastema was noted in the maxillary arch. In the mandibular anterior region, the retained primary mandibular left central incisor was evident (Figures 2 and 3). Caries was evident clinically in the permanent maxillary right first molar, permanent maxillary left second molar and first premolar, permanent mandibular left second molar, and first premolar. Mild, noncarious tooth tissue loss was evident on the maxillary anterior teeth's palatal surfaces from the canine-to-canine region. Crowding was noted in the maxillary left and mandibular left quadrants, with the mandibular left second premolar lingually placed. A panoramic radiograph showed the maxillary



Figure 1. Actinic prurigo of the face.



Figure 2. Maxillary and mandibular arches prior to treatment.

left second premolar to be unerupted and confirmed caries and missing teeth as noted (Figure 4). A periapical radiograph showed that the retained primary mandibular left central incisor had a small root, which confirmed a poor long-term prognosis (Figure 5).



Figure 3. Missing permanent mandibular central incisors.



Figure 4. Panoramic radiograph.



Figure 5. Periapical radiograph of primary mandibular left central incisor.

In view of the clinical findings, the following treatment plan was formulated. Phase 1 involved prevention, including oral hygiene instruction, dietary counseling in the form of a 3-day dairy diet, and the daily use of a topical fluoride mouthwash (0.05%). Fissure sealant of

the maxillary and mandibular right first premolars and the mandibular right second premolar. Phase 2 involved placing preventative resin restorations in permanent maxillary and mandibular left first premolars and restoring the carious teeth as charted. Phase 3 consisted of referral to a multidisciplinary hypodontia clinic for assessment and long-term management of the dentition and the missing teeth in the mandibular anterior region.

Upon attendance at our clinic, the patient again stated that, although not unduly concerned, he desired an improvement in the appearance of his front teeth. In view of this and after explaining the options, a simple treatment plan was discussed and agreed. The final restorative phase of treatment included a Kesling set up, using a wax mock-up to demonstrate the esthetic possibilities prior to starting treatment. This showed the reduction of the midline diastema with restorative materials and extraction of the retained primary tooth and placement of a 1-unit bridge (Figure 6). A 1-unit bridge was possible, as mesial drift was anticipated upon extraction of the retained primary mandibular left central incisor. This mesial drift both alleviated the crowding in the mandibular left quadrant and reduced the space in the anterior region (Figures 7-9).

DISCUSSION

This patient's initiation of AP agrees with other reported cases in that it started in early childhood during a family holiday, when the patient was exposed to sunlight. Most AP patients are young and genetically susceptible, requiring short periods of sun exposure for development of intense pruritus and new lesions.²

AP's clinical expression depends on the interaction of the genetic susceptibility with definite environmental factors. In our case, there was no family history. This is not uncommon, with a family history only reported previously in 5% of published cases.⁵ The HLA Class II antigen DR4 appears to be strongly associated with AP in British Caucasian patients and may, therefore, have a causal role in the disease's pathogenesis by modifying the disease's response to a light-induced antigen.⁶

Most authors agree that AP's treatment is difficult, with several regimes such as antihistamines, antimalarials, tetracycline, vitamin E, B carotene, Psoralen ultraviolet therapy, systemic corticosteroids, and topical corticosteriods, antibiotics, and sunscreens producing only partial relief of symptoms. Our patient's experience would agree with these findings—he found some relief with desensitization with artificial light. From a dental viewpoint, lip involvement and anterior gingival overgrowth, due to mouth-breathing associated with intense pruritus, has been previously described as relating to AP.² These were not found to be associated



Figure 6. Kesling setup of anterior incisors.



Figure 7. Facial view following treatment.

with this patient's case. Of interest, however, was the missing permanent mandibular central incisors and permanent maxillary right second molar. Congenital absence of mandibular central incisors has previously been associated with a proportion of patients having allergies, atopic skin conditions, and asthma.¹¹

Although this patient did not have oligodontia, dry skin is also reportedly more prevalent in oligodontia patients (57%) than in healthy control persons.¹² There are also published reports of conditions associated with missing mandibular incisors (ie, Witkop syndrome). Additionally, more than 60 syndromes categorized in On-line Mendelian Inheritance in Man are associated with tooth anomalies, implying that common molecular mechanisms are responsible for tooth and other organ development.¹³

This patient demonstrated 2 dental anomalies associated with hypodontia: taurodontism of maxillary molars, and a permanent maxillary right rotated premolar. Taurodontism has been described previously in the literature relating to 35% of patients with hypodontia.¹⁴ Both hypodontia and taurodontism are defects of the ectoderm. Rotated premolars are seen with higher frequency (18%) in hypodontia patients.⁹ Opinions vary in terms of the degree of symmetry presented in the dentition with tooth



Figure 8. Maxillary and mandibular arches following treatment.



Figure 9. Frontal view of anterior teeth.

agenesis. Most patterns are bilaterally symmetric, with the exception of permanent maxillary lateral incisors, where the left is more frequently missing than the right one. The absence of the maxillary right second molar in an older patient might be due to a previous dental extraction. When charting missing teeth, it is always important to obtain an accurate past dental history to account for previous dental extractions. This was undertaken with this patient, who had previously undergone dental general anesthesia for the removal of primary teeth. The patient's notes verified this.

The multidisciplinary hypodontia clinic incorporates consultants in pediatric, restorative, orthodontic, and maxillofacial (surgical) dentistry. The importance of a team approach to the management of hypodontia cannot be overemphasized. It is essential to provide an overall treatment plan to ensure appropriate timing and management at each stage of treatment.¹⁵ The option of utilizing the crowded permanent maxillary left second premolar to facilitate an autotransplant to the mandibular anterior region was not feasible for 2 reasons. First, the premolar tooth was too large for the area intended. Secondly, the patient lacked the required commitment for this complex treatment option. The treatment plan discussed reflected the patient's attitude and lack of interest in more complex treatment. Should the patient change his mind at a later date, however, more complex treatment options, such as implants, will still be available.

It was noted that the same wavelength of light that initiates the AP reaction is used in the dental curing light (350-500 nm). In this patient case, no reaction was noted, but that the authors could, if presented with a similar patient, use a self-curing sealant in the future for preventative work.

This is the first known case of AP and hypodontia being discussed in the same patient. No genetic links have been reported between AP and hypodontia in the literature.

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