

Interdisciplinary treatment for ectodermal dysplasia

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Ectodermal dysplasia is a hereditary disease characterized by a congenital dysplasia of one or more ectodermal structures. Characteristic manifestations include scanty hair and eyebrows, pigmented and hyperkeratinized skin around the eyes and mouth, frontal bossing with prominent supraorbital ridges, nasal bridge depression and dental anomalies. Hyperthermia or unexplained high fever as a result of the deficiency of sweat glands is common medical history. Findings of intraoral structures are anodontia or oligodontia with conical crowns. Consequently, generalized spacing and loss of vertical dimension of occlusion. Interdisciplinary approach has been performed to treat a 10-year old boy with ectodermal dysplasia. Orthodontists and a prosthodontist worked together on this case, and the result was satisfactory.

Key words : ectodermal dysplasia, oligodontia, interdisciplinary treatment, loss of vertical dimension

Ectodermal dysplasia is known as an inherited disease characterized by dysplasia of tissues of ectodermal origin(hair, nails, skin, and teeth). It is divided into two main clinical forms : (1) the X-linked hypohidrotic form characterized by the classical triad of hypohidrosis, hypotricosis, and hypodontia and by characteristic dysmorphic facial features(Christ-Siemens-Touraine syndrome) ; (2) the hidrotic form that usually spares the sweat glands but affects teeth, hair, and nails and is inherited as an autosomal dominant trait(Clouston's syndrome).¹⁾ The incidence is one to seven per 100,000 live births.²⁾ The diagnosis of ectodermal dysplasia is made on the basis of history and clinical examination. The affected child has an abnormal dentition, keratinized skin, sparse hair, heat intolerance, an inability to perspire and significant epis-

odes of hyperthermia in infancy or early childhood. The most common oral characteristics are hypodontia or anodontia and resultant loss of vertical dimension. Incisors, canines, and premolars, when present, often have conical crowns and the oral mucosa often appears dry. The pharyngeal and laryngeal mucosa may be atrophic, resulting in dysphonia. The most common complaint of childhood and adolescence is concern about dental abnormalities and facial appearance. Early extensive dental treatment is needed for not only improvement of masticatory and phonetic function but also positive psychological impact on the child. Removable partial dentures and/or complete dentures are the most frequent treatment choice for hypodontia or anodontia.³⁻⁵⁾ Recently, the use of endosseous implants has been reported for the treatment of children and adolescents with ectodermal dysplasia.⁶⁻⁸⁾ But in the hypodontic case, the existing teeth show abnormal shapes and aligned improperly. The prosthodontic treatment only cannot give a good result. The orthodontic realignment before prosthetic treatment can give better circumstances to make well functioning and esthetic prosthesis. A multidisciplinary team approach to treatment

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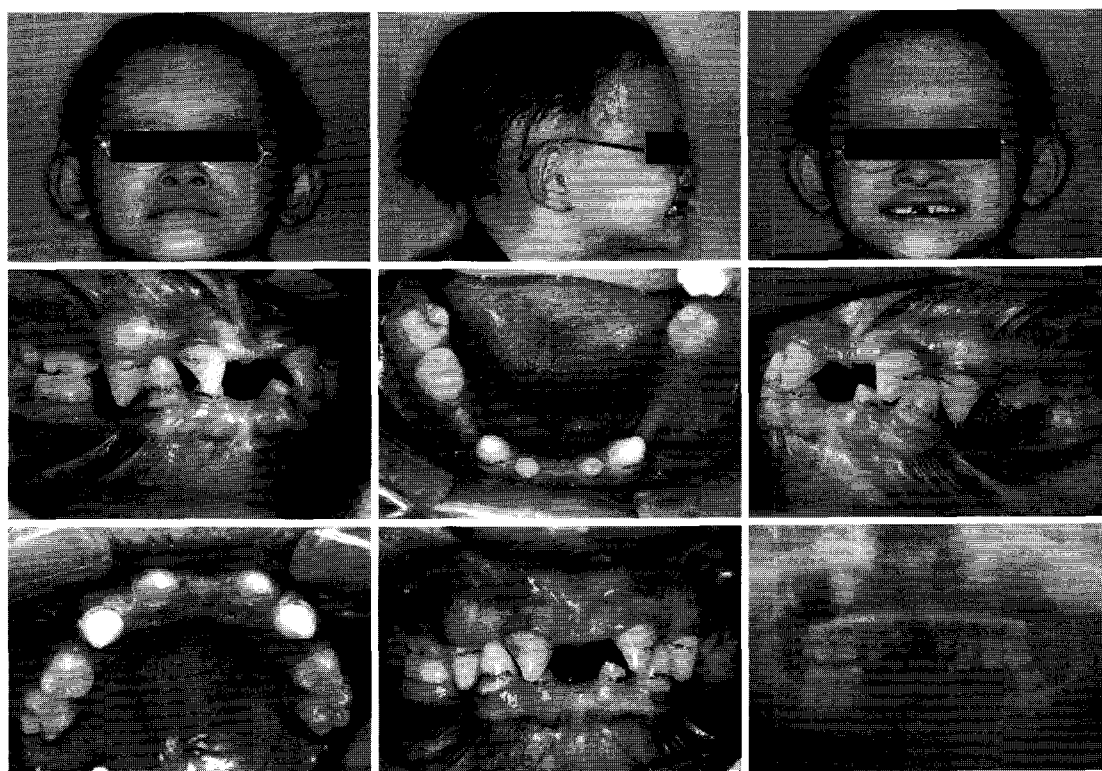


Fig 1. Facial and intraoral photographs before treatment showing typical stigmata of ectodermal dysplasia.

is recommended.

In this article, a case with satisfactory results from orthodontic treatment and following fixed and removable prosthetic treatments for an ectodermal dysplasia will be presented and considerations for orthodontic treatment will be discussed.

CASE

A 10-year-old boy visited our clinic, complaining of his ugly teeth and difficulties in chewing. He showed typical characteristics of ectodermal dysplasia including sparse hair, scant eyelash and eyebrow, pigmented and hyperkeratinized skin around the eyes and nose, depressed bridge of nose, protuberant lip, and dental anomalies. He had history of hyperthermia and was not able to bear warm temperature in summer. His parents did not exhibit signs of ectodermal dysplasia but his maternal cousin also had ectodermal dysplasia. We presumed that the ectodermal dysplasia in this child

had been originated from his mother. In the intraoral examination, he showed oligodontia. He had many congenital missing teeth (#12,14,15,17,22,24,25,27,31,32, 34,35,37,41,42,44,45,47) and retained primary teeth (#55, 65,72,75,82,85). All anterior teeth (#11,13,21,23) had conical crowns and molars also showed abnormal crown morphology. He did not have any prosthesis in his mouth. His lower facial height was short because the mandible was overclosed due to absence of anterior vertical stop. His face looked older than he was.(Fig 1)

Diagnosis

This patient case was diagnosed as X-linked hypohidrotic type ectodermal dysplasia with oligodontia. The cephalometric analysis of lateral head film showed Class III pattern with ANB of -2.6° and Wits Appraisal of 4.6mm.(Table 1) There was 100% overbite and mandibular plane angle was very low as 15.1°

Table 1. Cephalometric measurements of pretreatment and posttreatment

	<i>Norm</i>	<i>Initial</i>	<i>Final</i>
SNA (dg)	82.0	81.5	82.7
SNB (dg)	79.8	84.1	81.5
Facial Angle (dg)	89.0	93.2	90.8
ANB (dg)	2.3	-2.6	1.2
Wits Appraisal(mm)	-2.5	4.6	1.4
Palatal Plane Angl(dg)	1.1	-6.7	-6.2
FMA(dg)	23.5	15.1	20.5
Y - Axis (dg)	61.4	53.0	57.5
Gonion Angle(dg)	117.9	112.7	119.4
Lower Ant Fac Height(mm)	73.8	48.5	57.1
Post. FH / Ant. FH(%)	68.5	72.7	70.5
Mx 1 to FH (dg)	116.2	118.7	114.7

Treatment Objectives

Based on the clinical examination and consultation with the patient and his mother, the following treatment goals were established :

- to create space for a prosthesis
- to increase vertical dimension of occlusion for normal face profile
- to establish acceptable anterior esthetics
- to create a stable functional occlusion

The extensive orthodontic and prosthetic treatments were required to achieve these goals.

Treatment Plan

The orthodontic and prosthodontic team approach for this patient was recommended and treatment procedure was as follows :

1. Extraction of #72,#82
2. Fabrication of a 6-unit provisional restoration from #43-to-33
3. Fabrication of a posterior bite block
4. Distalization of maxillary central incisors for a restoration of 6-unit FPD(Fixed Partial Denture)(#13-11-x-x-21-23)
5. Fabrication of a 6-unit provisional restoration from #13 to #23 with anterior vertical stops to increase posterior vertical space for extrusion of molars.

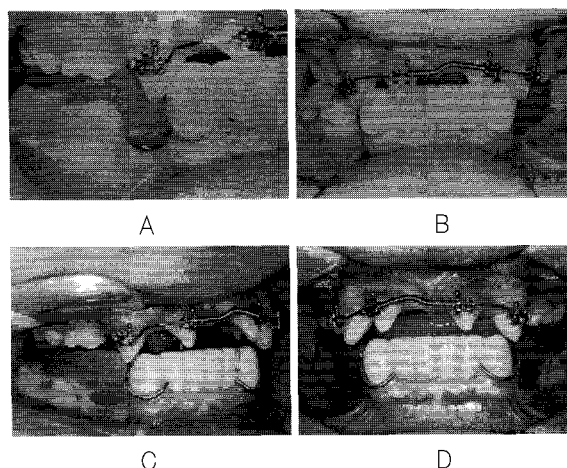


Fig 2. Intraoral photographs during treatment : mandibular anterior provisional restoration disturbed maxillary central incisors from moving into the position of lateral incisors with open coil spring(a,b). A bite block was delivered to allow that movement(c,d).

6. Forced eruption of posterior teeth
7. Delivery of transitional lower RPD(Removable Partial Denture)
8. Final prostheses
 - 1) FPDs: #13-11-x-x-21-23, #33-x-x-x-x-43
 - 2) Mandibular RPD

Treatment Progress

Primary mandibular lateral incisors were extracted for the convenience of provisional restoration. A 6-unit provisional restoration from #33 to #43 was cemented and a lower posterior bite block was delivered to allow the distal and lingual movements of maxillary central incisors by bite-opening.(Fig.2-c,d) The maxillary central incisors were distalized with open coil spring on the 019x025 SS wire for a restoration of 6-unit FPD on tooth #13, 11, 21 and 23.(Fig.2-a,b)

A 6-unit provisional restoration from #13-to-23 with anterior vertical stop was fabricated and cemented after finishing of distal movements of maxillary central incisors. These provisional upper and lower anterior prostheses increased the posterior vertical space for extrusion of the molars.(Fig.3-b) Direct bracket bondings on lower provisional restoration were done pas

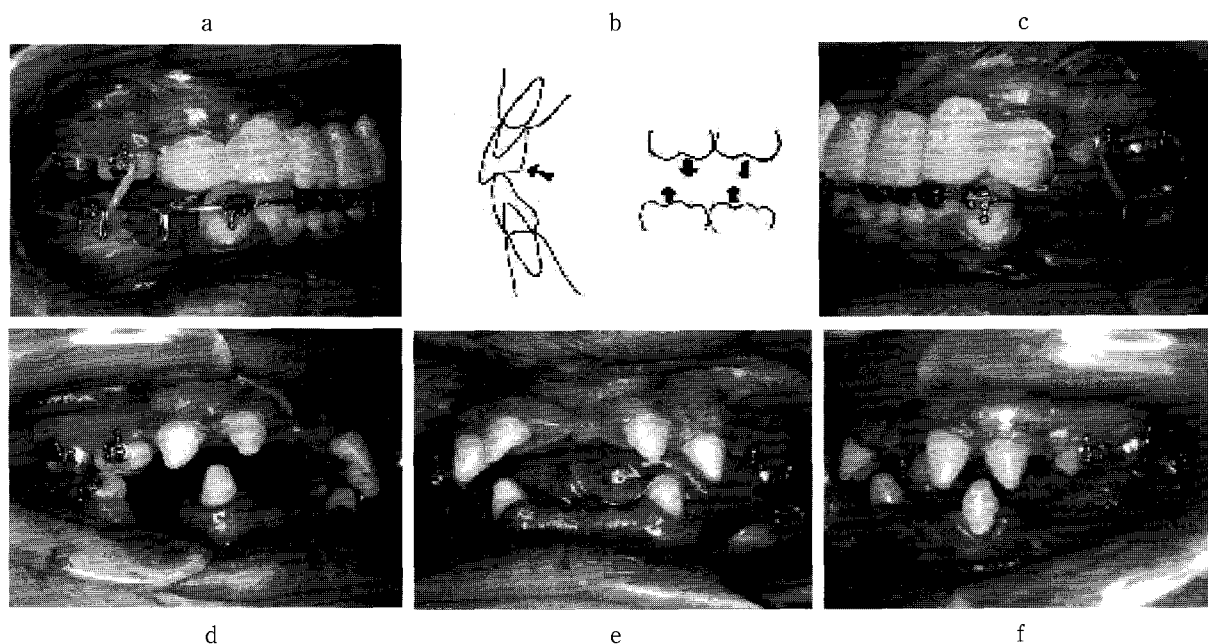


Fig 3. Intraoral photographs during treatment: Up and down elastics were used to extrude the posterior teeth(a,c), the lingual part of maxillary provisional restoration had anterior vertical stops(b), and the amount of increase of VDO(vertical dimension of occlusion) was confirmed after removal of provisional restorations(d,e,f).

sively and 018x022 SS wire with vertical loop was inserted on the lower arch. Up and down elastics to extrude upper and lower molars were delivered for 24 hours per day except during meals.(Fig.3-a,c) It took 4 months to extrude the molars.(Fig.3-d,e,f)

After extrusion of molars, anterior overjet was increased, and additional retraction of the maxillary anterior teeth was needed to reduce this overjet. Total time of orthodontic treatment was 14 months. After finishing of orthodontic treatment, new maxillary and mandibular anterior provisional restorations without vertical stops and a lower transitional Removable Partial Denture were delivered and maintained for 2 months to evaluate the adaptability of patient on the restorations. Final prostheses (FPDs:#13-11-x-x-21-23, #33-x-x-x-x-43 and a mandibular RPD) were delivered.

RESULTS

The masticatory and phonetic functions were restored with maxillary and mandibular FPDs and a mandibular RPD. And the patient was satisfied with his

improved esthetics due to anterior FPD and increased lower facial height.(Fig 4) ANB was improved to 1.2° and FMA was increased to 20.5° . Lower anterior facial height was increased to 57.1mm.(Table 1, Fig 5)

DISCUSSION

Ectodermal dysplasia syndrome is a hereditary disease but it may occur in a family without previous history of the disease because of gene mutation. Early prosthetic treatment is strongly recommended by some authors because the esthetic and psychologic problems as well as the functional problems make the affected children depressed.⁹⁾ Enhancement of facial appearance in children who have general normal intelligence helps to build self-confidence. Complete dentures, RPDs or implant-supported prostheses can be chosen for treating ectodermal dysplasia with anodontia or hypodontia. Major problems with early placement of prostheses are related to growth changes and to difficulties in achieving good retention and stability. Frequent adjustments of removable dentures are needed to allow

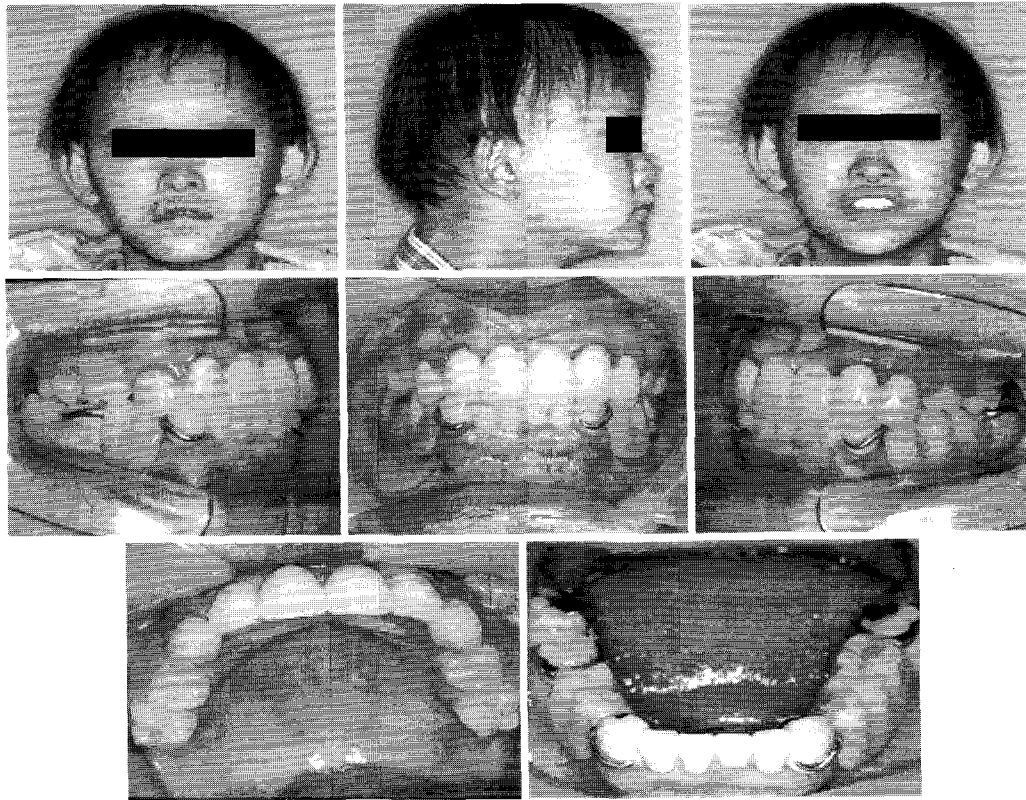


Fig 4. Facial and intraoral photographs after treatment showing increased lower facial height and improved facial esthetics.

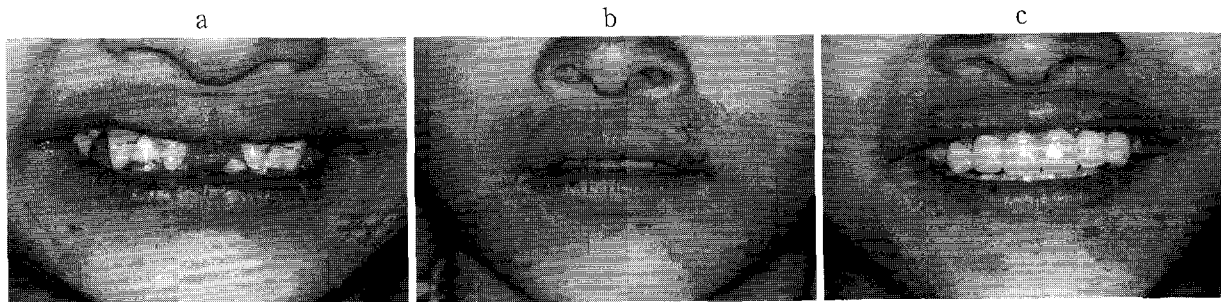


Fig 5. Lower anterior facial third photographs: deep overbite and diminished vertical dimension can be seen(a), vertical dimension was increased and esthetic was improved(b,c)

alveolar bone growth in growing children. Consequently, retention and stability of denture are diminished. To improve retention and esthetics, FPDs can be chosen in hypodontia. Recently, the placement of endosseous implants in growing children has been recommended not only improve retention and stability of prostheses

but also to stimulate alveolar bone growth by transmission of masticatory force. Smith et al¹⁰⁾ reported a success case related to placement of a single mandibular endosseous implant in a 5-year-old patient with hypohidrotic ectodermal dysplasia and oligodontia. There are several instructions to place endosseous



Fig 6. Superimposed cephalometric tracings of pretreatment and posttreatment.

implants in growing patients but those will not be mentioned in this article. In this case, FPDs and a lower RPD were delivered because of economic concern. Fixed prostheses, however, have been rarely used due to remaining growth.¹¹⁾ In the growing patient, the use of FPD across of the midpalatine suture is not recommended. Bishara et al¹²⁾ reported that intercanine and intermolar widths after the complete eruption of the permanent dentition, slightly decrease, more in the intercanine than in the intermolar widths and mandibular intercanine width, on the average, was established by 8 years of age, i.e., after the eruption of the four incisors. Ross-Powell and Harris¹³⁾ also reported that intercanine widths increase significantly during the deciduous phase but exhibit no systematic change after emergency of the permanent canines. After the eruption of the permanent dentition, the clinician should either expect no changes or a slight decrease in arch widths. In this patient, therefore, the change of the intercanine width may not be influenced by the FPD. The major problems treating this patients were abnormal functional tongue thrust and frequent detachment of brackets. The absence of 2 maxillary anterior teeth caused retained abnormal tongue thrust. Consequently, tongue thrust disturbed the retraction of maxillary central incisors and pushing out of the arch wire caused detachment of brackets. After myofunctional therapy, i.e., training of the way of positioning of tongue, this problems were solved. The extrusion of

posterior molars was easily achieved with up and down elastics and anterior vertical stops. This anterior vertical stop was constructed on the lingual surface of provisional restoration of maxillary anterior teeth and vertical dimension was determined by facial appearance and physiologic rest position. The position of the mandible at the beginning of swallowing act was used as a guide to the vertical dimension of occlusion and the extrusion of of molar needed in this patient to get a normal vertical dimension of occlusion and good facial appearance. Increased vertical dimension of occlusion with the provisional restoration was evaluated during the treatment and the patient was well adapted for increased vertical dimension of occlusion. Definitive occlusion was established by the final prostheses. Frequent detachment of brackets extended total period of treatment. In this case, complicated problems were solved one by one because there were not enough teeth to be anchorage. The mechanics used for this orthodontic treatment were not difficult, but other factors, like strong tongue thrusting or inadequate anchorage made the treatment difficult. During the treatment, the orthodontists and the prosthodontist had discussed about each procedures. Interdisciplinary approach on the treatment of ectodermal dysplasia can give better results and satisfy the patients.

REFERENCES

1. Lowry RB, Robinson GC, Miller JR. Hereditary ectodermal dysplasia.

- Symptoms, inheritance patterns, differential diagnosis, management. Clin Pediatr 1966 : 5 : 395-402.
2. Buyse M. Birth defects encyclopedia. St Louis : CV Mosby 1990 : 597-8.
 3. NaBadalung DP. Prosthodontic rehabilitation of an anhidrotic ectodermal dysplasia patient : a clinical report. J Prosthet Dent 1999 : 81 : 499-502.
 4. Ohno K, Ohmori I. Anodontia with hypohidrotic ectodermal dysplasia in a young female : a case report. Pediatr Dent 2000 : 22 : 49-52.
 5. Franchi L, Branchi R, Tollaro I. Craniofacial changes following early prosthetic treatment in a case of hypohidrotic ectodermal dysplasia with complete anodontia. ASDC J Dent Child 1998 : 65 : 116-21.
 6. Kearns G, Sharma A, Perrott D, et al. Placement of endosseous implants in children and adolescents with hereditary ectodermal dysplasia. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 1999 : 88 : 5-10.
 7. Escobar V, Epker BN. Alveolar bone growth in response to endosteal implants in two patients with ectodermal dysplasia. Int J Oral Maxillofac Surg 1998 : 27 : 445-7.
 8. Guckes AD, Roberts MW, McCarthy GR. Pattern of permanent teeth present in individuals with ectodermal dysplasia and severe hypodontia suggests treatment with dental implants. Pediatr Dent 1998 : 20 : 278-80.
 9. Boj JR, Duran J, Cortada M, Jimenez A, Golobart J. Cephalometric changes in a patient with ectodermal dysplasia after placement of dentures. J Clin Pediatr Dent 1993 : 17 : 217-20.
 10. Smith RA, Vargervik K, Kearns G, Bosch C, Koumjian J. Placement of an endosseous implant in a growing child with ectodermal dysplasia. Oral Surg Oral Med Oral Pathol 1993 : 75 : 669-73.
 11. Itthagarun A, King NM. Ectodermal dysplasia : a review and case report. Quintessence Int 1997 : 28 : 595-602.
 12. Bishara SE, Jakobsen JR, Treder J, Nowak A. Arch width changes from 6 weeks to 45 years of age. Am J Orthod Dentofac Orthop 1997 : 111 : 401-9.
 13. Ross-Powell RE, Harris EF. Growth of the anterior dental arch in black american children : A longitudinal study from 3 to 18 years of age. Am J Orthod Dentofac Orthop 2000 : 118 : 649-57.

국문초록

외배엽 이형성증환자의 협진치료

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외배엽 이형성증(ectodermal dysplasia)는 외배엽에서 분화되는 조직의 일부에 선천적인 이형성이 나타나는 유전적 질환으로 모낭 및 땀샘의 결여로 인한 성긴 모발과 눈썹, 눈과 입주위 피부의 착색 및 각화와 치아형성의 이상 등이 특징적으로 나타나는 질환이다. 구강내 소견으로 무치증 혹은 부분 무치증을 보이며 부분 무치증의 경우 치관이 원추형으로 나타나 결과적으로 전반적인 치아간극 및 수직고경의 감소로 저작 및 발음기능의 저하 뿐 아니라 심미적이지 못한 안모로 사회적 및 정서적 우울감에 빠지기 쉬우므로 조기치료가 요구된다. 바람직한 보철물 제작 및 안모의 개선을 위해서는 교정과 및 타 전문과의 협진이 요구되며, 본 증례에서는 ectopic dysplasia로 진단된 10세 남아를 교정의사와 보철의사의 긴밀한 협진으로 저작 및 발음기능의 회복과 안면고경의 증가로 심미성이 개선된 좋은 결과를 얻었기에 보고하고자 한다.

주요 단어 : 외배엽 이형성증, 부분 무치증, 협진, 수직고경