Oral Rehabilitation of a Patient with Diffuse Lymphangiomatosis Affecting the Maxilla: A Case Report

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A case is presented in which diffuse lymphangiomatosis resulted in the complete loss of the maxillary dentoalveolar complex and underlying basal bone. The complex investigation and treatment of this patient over a 10-year period is presented, and the importance of a multidisciplinary team approach in providing a functional and esthetic rehabilitation is highlighted. The use of a vascularized bone graft based on the deep circumflex iliac artery and subsequent restoration with an implant-supported prosthesis is described. (Case Report) INT J ORAL MAXILLOFAC IMPLANTS 2006;21:459–464

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Many pathologic processes can result in the loss of the dentoalveolar complex and underlying bone. The patient may subsequently suffer a gross functional and esthetic deficiency requiring complex reconstruction and oral rehabilitation. Treatment requires a multidisciplinary approach and the use of advanced surgical and restorative treatment modalities. The following case presentation illustrates an example in which the treatment of a rare hamartomatous condition resulted in a severely atrophic maxilla in an otherwise healthy teenage patient. *Lymphangiomatosis* is a term used for diffuse or multifocal involvement of bones, parenchymal organs, or soft tissues by lymphangiomas.[1] The treatment of this complex problem is also presented.

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CASE REPORT

A fit and well 15-year-old male adolescent was referred by his general dental practitioner to the Pediatric Department of the Liverpool University Dental Hospital with a 2-week history of mild discomfort and increasing mobility of the maxillary left premolars. The patient was seen for his first consultation in October 1994.

Clinical examination revealed a healthy-looking adolescent patient with no extraoral abnormalities. Intraoral examination revealed a well-maintained dentition with no obvious mucosal changes; however, complete segmental mobility affecting teeth 9(21), 10(22), 11(23), 12(24), 13(25), 14(26), and 15(27) and the associated bone was noted (Fig 1a). Radiographic examination revealed widening of the periodontal ligament spaces of all the teeth in the posterior segment of the maxilla with disruption of the normal architecture of the bone in this area (Fig 1b). The patient was subsequently referred urgently to the maxillofacial department of the local children's hospital.

The maxillary left canine and first premolar, which were extremely mobile, were extracted in an inpatient procedure under general anesthesia, and the associated soft tissue was sent for histopathologic investigation. Initial histologic examination revealed the presence of "intense chronic inflammatory tissue," but a definitive diagnosis was not made at this stage. During the following weeks, the remaining dentition in the left maxilla showed increasing mobility, leading to

Fig 1a Patient's oral situation at initial presentation.

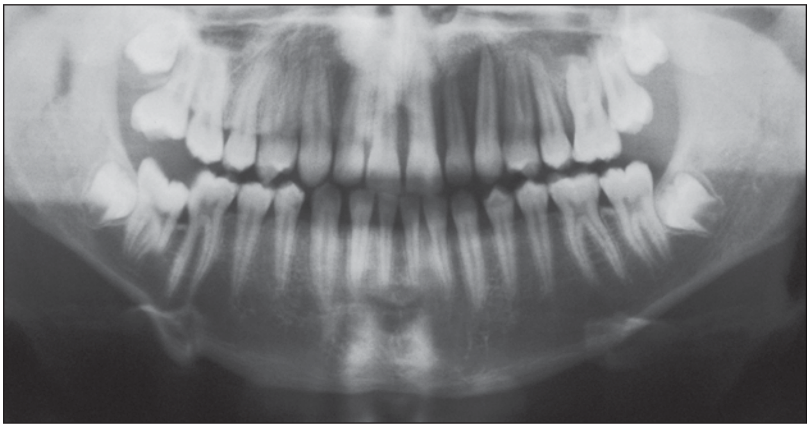

Fig 1b An orthopantomogram taken on initial presentation showing widening of the periodontal membrane in the maxillary left premolar region and disruption of the architecture of the maxillary bone in the maxillary left quadrant.

the removal of teeth 8(11), 9(21), 10(22), 13(25), 14(26), and 15(27). Removal of the associated soft tissue lesion resulted in an oroantral communication, which was closed immediately with a buccal advancement flap. Further histologic examination of the new pathologic specimen at the Liverpool University Dental Hospital and review by a second independent consultant histopathologist resulted in a definitive diagnosis of diffuse lymphangiomatosis.

Systemic examination, including the use of radiographs and computerized tomography (CT), showed no other foci of disease. Restoration of the missing units at this stage consisted of a removable mucosa-supported acrylic resin partial denture with Adams clasps around teeth 3(16) and 5(14) for retention. Over the next 3 years, the patient was examined regularly; his condition remained stable.

Subsequent to this period of stability, the patient developed rapid-onset mobility of the right maxillary premolar teeth and lost the remaining maxillary teeth and associated bone in a manner similar to that observed on the contralateral side. At this stage the patient was considered for implant-based restoration.

The patient underwent a bilateral sinus augmentation procedure and placement of a block corticocancellous onlay graft to the premaxilla using bone harvested from the iliac crest in preparation for implant placement. Healing following this procedure was uneventful. Six months later, as a secondary procedure, 6 Frialit-2 implants (Dentsply Friadent, Hanau, Germany) were placed in the 3(16), 4(15), 5(14), 12(24), 13(25), and 14(26) regions. Two implants failed to integrate satisfactorily and were removed prior to fabrication of the definitive prosthesis. A bar-retained maxillary overdenture was fabricated using a resilient lining material over milled bars for retention (Molloplast-B; Detax, Ettlingen, Germany) (Figs 2a and 2b). At this stage the patient was happy with both the appearance and function of the prosthesis (Fig 2c).

Despite the placement of a block corticocancellous onlay graft to the premaxilla, reducing the severity of the Class III malocclusion, this reconstruction was considered biomechanically unfavorable because of the posterior fulcrum of rotation about the maxillary implants and the difficulty in obtaining balanced articulation of a complete maxillary prosthesis opposed by intact mandibular dentition in a class III skeletal relationship (Fig 2d). Although there was concern about the long-term prognosis of the implant-supported restoration, the patient declined further treatment because of educational commitments.

The patient returned approximately 3 years later requesting further treatment to improve both his appearance and function. Further CT imaging was carried out, and following the fabrication of a stereolithographic model, a vascularized free flap of bone and muscle based on the deep circumflex iliac artery (DCIA) was harvested and transferred to the premaxillary region. This improved the skeletal relationship while also providing bone of satisfactory quantity and quality for implant placement (Fig 3a).

Refinement of the bone graft and sulcoplasty was carried out, followed by the placement of 6 Frialit-2 implants in the premaxillary region (Fig 3b). Following implant exposure and the placement of healing abutments 6 months later, a temporary overdenture was fabricated and relined in the premaxillary and milled bar region with a resilient lining material. At this stage the patient reconfirmed his satisfaction with esthetics, retention, and function.

The definitive removable prosthesis was then fabricated. Six separate anterior milled telescopic copings were cemented to screw-retained abutments for prosthesis retention. The posterior milled bars were


Figs 2a and 2b Initial rehabilitation showing the complete maxillary prosthesis with resilient lining and posterior implant-supported milled bars.


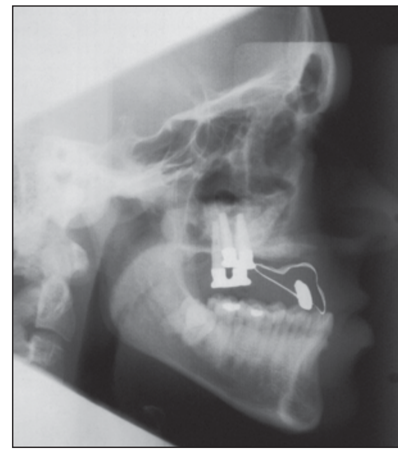

Fig 2c *(Left)* Anterior view of the prosthesis in intercuspal position.

Fig 2d *(Right)* Lateral cephalogram showing marked class III malocclusion and the resulting position of the prosthesis in an unfavorable position in relation to the mandibular arch.

replaced by 4 separate magnetic keepers (MAGFIT-IP; Davis, Scottlander & Davis, Herts, England) to allow better access for posterior cleaning (Figs 3c and 3d). The removable prosthesis housed 6 laboratory-milled gold alloy secondary telescopic copings and 4 magnets and was of a horseshoe design (Fig 3e).

After fabrication of the definitive prosthesis the patient had a Class I relationship between the maxilla and mandible, which resulted in a more stable and esthetic prosthesis (Figs 4a and 4b). The patient also showed improved soft tissue profile and improved upper lip support.

PATHOLOGY

A review of the literature shows no previously reported cases of diffuse lymphangiomatosis resulting in the complete loss of the maxillary dentition and associated bony structures. *Angiomatosis* is a term used to describe a spectrum of rare hamartomatous conditions characterized by fibrous tissue containing thin walled ectatic vascular channels, lined with endothelial cells, and containing lymph or blood.[1] By convention, the term *lymphangiomatosis* is reserved for lesions with predominantly, if not exclusively, lymphatic differentiation. Diffuse lymphangiomatosis is a rare condition that can affect soft tissue, viscera, or bone. This condition predominantly occurs in childhood and rarely manifests after the age of 20 years. It has no gender predilection. If bone infiltration occurs, it is most commonly seen in the pelvis, shoulder, spine, ribs, long bones, or occasionally, the skull. Eighty percent of lymphangiomatosis patients present with multifocal lesions.

Prognosis is dependent on the extent of disease and organ involvement. The success of surgical resection is impaired by an inability to separate lymph collections from tissue structures, leading to a high rate of recurrence. Patients with liver, spleen, and lung infiltration have a poorer prognosis. Patients with soft tissue involvement with or without bone invasion enjoy a much better prognosis.[2]

Radical surgery or radiotherapy has been used with varying degrees of success. Histologic differential diagnosis includes angiomatosis, acquired progressive lymphangioma, and angiosarcoma.

RECONSTRUCTION AND REHABILITATION

Reconstruction and oral rehabilitation of patients with gross tissue loss in the jaws and face requires a well-coordinated multidisciplinary team approach. Advanced surgical techniques together with the use of osseointegrated dental implants have allowed

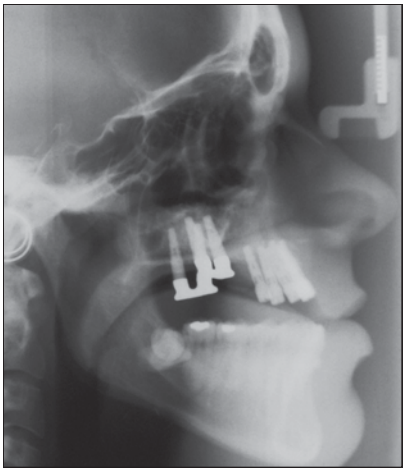

Fig 3a The restoration of a Class I skeletal relationship following the use of a DCIA vascularized graft.

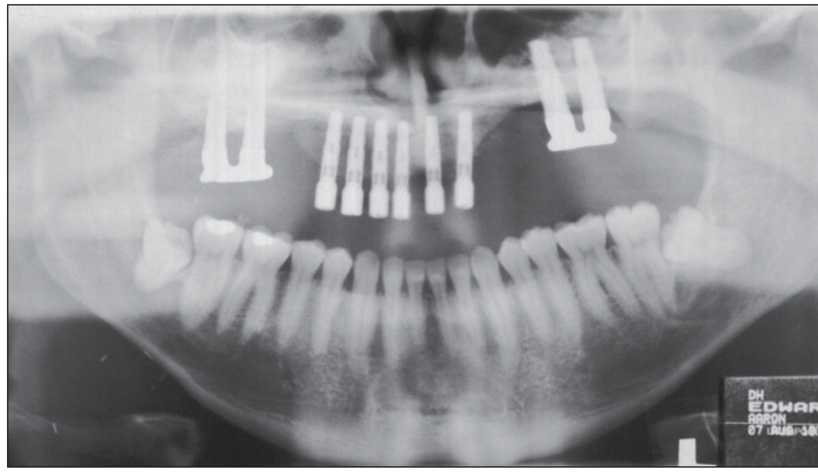

Fig 3b Orthopantomogram showing anterior 6 Frialit-2 implants in situ.

Fig 3c Anterior view of milled telescopic copings.

Fig 3d Intraoral view of milled telescopic copings and magnetic keepers.

Fig 3e Definitive prosthesis housing 6 anterior copings and 4 posterior magnets.

Figs 4a and 4b Anterior intraoral view of definitive prosthesis and prosthesis in situ.

great progress to be made in the full oral rehabilitation in this group of patients. The surgical treatment of this patient's pathology resulted in a defect similar to a severely atrophic maxilla (Cawood and Howell Class VI).[3] Severe Class III malocclusion existed, and any restoration was to be opposed by a dentate mandible. The use of bilateral antroplasty and anterior onlay bone grafting to allow the placement of an implant-supported prosthesis is a well-recognized treatment modality for such patients. Jaw reconstruction with a combined approach of bone grafts and implants is well accepted by patients and often provides a much-improved quality of life.[4,5] Osseointegration of dental implants in reconstructed jaws is now considered a reliable procedure, with reported long-term success rates between 81% and 98% in nonirradiated tissues.[6–8] The success rate of implant osseointegration in the present case was within these limits.

For an implant-supported prosthesis to be a viable option, bone must be present in both adequate quantity and quality. Many grafting techniques provide bone of sufficient quality for implant placement, but few provide sufficient quantity of bone. Microvascular composite free flaps from the ilium or fibula are routinely used for primary bone reconstruction following extensive maxillectomy. Urken and associates described the use of a free flap based on the DCIA for oromandibular reconstruction with minimal donor site morbidity after more than 1 year of follow-up.[9] The DCIA free flap is favored for reconstruction of the maxilla in the authors' regional unit.[10] This large bulk of tissue transfer allowed improvement in both the patients' extraoral profile and skeletal relationship while providing bone of satisfactory quantity and quality for implant-based rehabilitation.

Many factors should be considered when choosing between fixed or removable prostheses.[11,12] Firm criteria for treatment planning do not exist at present. Most factors precluding the use of a fixed prosthesis are related to deficiencies in soft tissue and bone quantity. In the present case, a removable prosthesis was considered preferable. Improved lip and facial tissue support can be better achieved by the use of a flange if a tissue discrepancy remains.[13]

For rehabilitation to be successful, it must restore function and esthetics while being comfortable. Zitzmann and Marinello found no significant differences between fixed and removable prostheses with respect to patient satisfaction with comfort, function, esthetics, speech, or self-esteem in their nonrandomized cohort study.[14] However Heydecke and associates showed in their clinical crossover trial that patient-based outcome measures supported the use of removable prostheses for improved general satisfaction, speech, and cleansibility.[15] The use of a removable prosthesis usually results in fewer speech errors compared to a fixed prosthesis; however, the presence or absence of palatal coverage appears to have no adverse effect.[16]

Various attachment types can be employed for use with an overdenture. These can be splinted (bar-and-clip design) or nonsplinted (ball/stud attachments, magnetic attachments, or telescopic copings). Consideration needs to be given to many factors when deciding which attachments to use. In this case vertical space, which can have implications for attachment choice, was not an issue. Nonsplinted attachments facilitate oral hygiene measures and may reduce moment loading on the implants. In vivo studies have shown ball attachments and bar-and-clip attachments to give similar retention.[17] Magnetic attachments have reduced retention but may aid reseating of a prosthesis and reduce horizontal load transmission to the implant.[18] Other factors to consider in attachment choice are initial time and cost, together with the long-term cost and maintenance implications for the restorative team and patient.[19,20]

The need for a multidisciplinary team approach in complicated rehabilitation cases such as the case presented is paramount if a satisfactory long-term outcome is to be achieved. This requires excellent communication between restorative clinicians, maxillofacial surgeons, and laboratory technicians to ensure a successful outcome. The use of a 3-dimensional stereolithographic model is helpful when planning this type of complex case.

SUMMARY

The reconstruction and rehabilitation of patients with gross destruction of maxillofacial tissues requires a well-coordinated multidisciplinary approach to optimize the chance of a successful outcome. Each member has an important role to play; the prosthodontist leads the team. The use of vascularized bone grafts and implant-retained prostheses greatly improved the quality of treatment and life in the individual under consideration.

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