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Prosthodontic Treatment and Medical Considerations for a Patient with Turner Syndrome: A Clinical Report

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Abstract

This clinical report describes a multidisciplinary approach in the rehabilitation of a 23-year-old Caucasian woman affected with Turner's syndrome and subsequently diagnosed with T4 Giant cell reparative granuloma of the right maxillary sinus. The surgical treatment included a maxillectomy and infratemporal fossa dissection followed by a free fibula palatal reconstruction, fibula bone graft of the orbital floor, dental implant placement, and prosthodontic rehabilitation. Prosthodontic planning and treatment considerations in an adult patient with Turner Syndrome are discussed.

Turner's syndrome (TS) is the most common chromosomal abnormality in females, affecting 1 in 2500 live births.[1] It is characterized by anomalies of the X chromosome. The etiology still remains unknown with no specific risk factor.[1-5]

TS encompasses several conditions with no two patients presenting the same symptoms.[2-4] The characteristic physical abnormalities include Turner's triad, described as sexual infantilism, webbing of the skin of the neck, and deformity of the elbow.[5-7] Other common features include lymphedema, broad chest, low hairline, rotated and low-set ears, soft upturned nails, palmar creases, drooping of the eyelids and dry eyes, multiple melanocytic naevi, increased keloid scar formation, and short stature.[1,2,4,7-9]

TS is responsible for significant medical problems with mortality three times higher than in the average population, primarily due to cardiovascular disease.[2-4] Ninety-eight percent of patients are infertile, and 85% will have no pubertal development. Patients are at increased risk of developing osteoporosis, hypothyroidism, diabetes, obesity, and/or gastrointestinal disorders. Other possible symptoms include kidney abnormalities, vision problems, hearing loss, autoimmune diseases, and cognitive deficits.[2-4,6,10] Patients with TS are also at increased risk of developing malignancies.[2-4]

Patients can exhibit a wide variety of dental features mostly described in the pediatric and orthodontic literature. The most common include micrognatia, high-arch palate, narrow maxilla, reduced crown width, short roots, and increased root resorption.[5,9,11-14] Very few articles describe the impact of TS on dental care in adulthood. This clinical report describes the treatment of a 23-year-old woman with TS who was also diagnosed with a T4 Giant cell reparative granuloma of the right maxillary sinus.

Clinical report

History

The patient, a 23-year-old female Caucasian, was referred to the University of Texas M.D. Anderson Cancer Center (Houston, TX) in June 2008 complaining of congestion and maxillary swelling unsuccessfully treated with antibiotics. A multidisciplinary comprehensive evaluation was performed. The patient denied tobacco, alcohol, or recreational drug use. She was 5'1" tall, weighed 220 lbs, and morbidly obese (Fig 1). TS was diagnosed at age 16 during a workup for amenorrhea but was not further monitored. She reported a history of

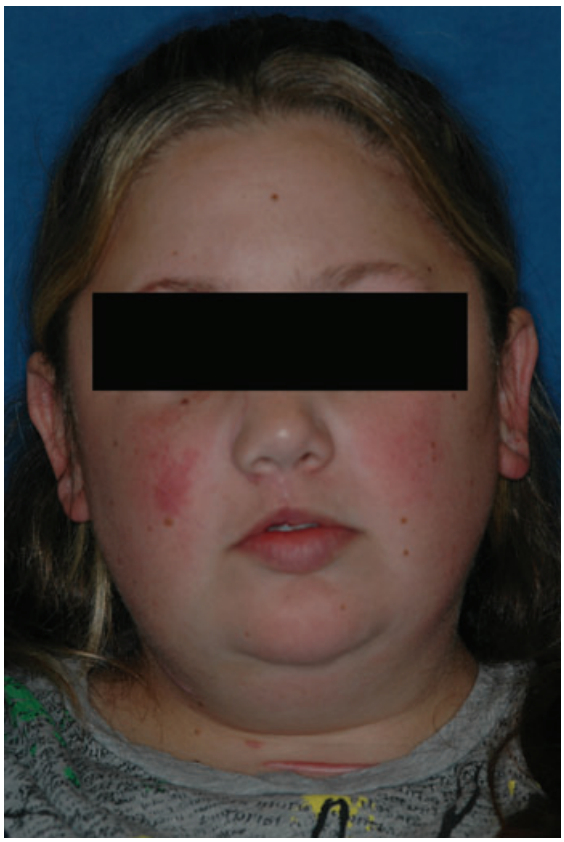

Figure 1 Initial presentation.

chronic sinusitis, myringotomy tube surgeries, and excision of cholesteatoma of the left ear. She was taking Seasonique (Teva Women's Health, Woodcliff Lake, NJ) as estrogen therapy and Percocet (Endo Pharmaceuticals, Chadds Ford, PA) as needed for pain.

Examination

The patient presented with facial asymmetry and a mass defect on the right hard palate and in the right buccal vestibule. She reported discomfort and pain in the malar, zygoma, and periorbital regions with associated trismus and paresthesia. She had increased epiphora and distorted vision from the right eye because of added pressure from the mass pushing against the eye. Intraorally, she had swelling of the gingiva and pain in the buccal vestibule and palatal regions. She had small areas of numbness and loss of motor innervations of the right V2 region. The alveolar bone around the molars and premolars was destroyed by the tumor, which caused displacement and loosening of teeth, leading to difficulty eating. A biopsy was performed in July 2008, and a diagnosis of T4 giant cell reparative granuloma of the right maxillary sinus was confirmed. Radiographs revealed a large radiodense area apical to the right maxillary teeth with erosion of the root tips of teeth #2 to 5 (Figs 2 and 3).

A multidisciplinary treatment plan was drawn. Surgery was performed in August 2008 involving a total right maxillectomy and infratemporal fossa dissection. The patient also had a tracheotomy to create a postoperative protected airway due to intraoral swelling, an anterior and posterior ethmoidectomy and sphenoidectomy to remove the tumor, and a dacryocystorhinostomy to provide a duct for the tears to drain. A microvascular free fibula myoosseous flap from the leg containing fibula bone, muscles, and blood vessels connected to the vasculature of her neck was used to serve as palatal reconstruction and to close the maxilla. Bone grafting was also performed to reconstruct the floor of her orbit and infraorbital rim to provide support for the eye, in conjunction with an open reduction and internal fixation of the orbital floor and a medial canthoplasty.

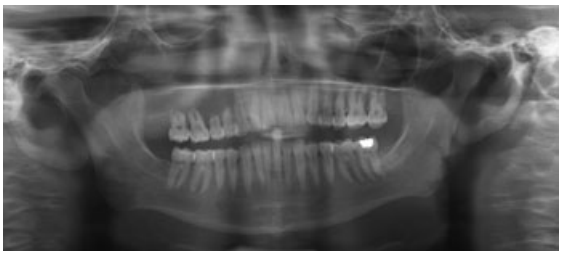

Figure 2 Initial Panorex.

Six months following these surgeries, the patient presented with hypertrophic scarring of incisions, slight bilateral tinnitus, and continuous tearing of the eye. She had a slightly enlarged right cheek, had lost teeth #2 to 8 to the surgery, and showed good healing of the flap. She had no buccal vestibule for the right side of her maxilla and showed very thick maxillary labial frenum. The prosthodontic treatment plan included an endosteal implant placement in the debulked fibula graft followed by an implant-supported maxillary prosthesis. Her medical treatment plan included revision of surgical scars and a dacryocystorhinostomy for her nasolacrymal duct obstruction concurrent with endosteal implant placement.

Three endosteal Astra Osseospeed implants were visually lined up and placed into the fibula at site #3 (3.5 × 11 mm), 5 (3.5 × 11 mm), and 6 (4.0 × 11 mm) (Astra OsseoSpeed TX, Astra Tech Inc., Waltham, MA). The fibula exhibited good bleeding, but was soft with poor cortical bone and offered weak primary stability on all implants. The implants were uncovered after 6 months of healing. Two of the three implants failed to integrate. A Locator abutment (4 mm height) was placed on implant #3 (Locator Abutment 3.5/4.0, Astra Tech Inc.). The failed implants at sites #5 and 6 were removed and immediately replaced with shorter but wider diameter implants to achieve primary stability (4.0 × 9 mm and 5.0 × 9 mm, respectively). A heat-cured acrylic resin oral surgical stent (Lucitone 199, Dentsply Trubyte, York, PA) was relined (Trusoft, H.J. Bosworth Company, Skokie, IL) and fixed to the dentition with ligature wires. Two weeks following the surgery, the oral surgical stent was removed. The patient was then referred to an endocrinologist for management of her TS. Bone density scans revealed osteopenia and osteoporosis, and recommendations were made to optimize calcium and vitamin D intake and exercise regularly. The patient was also referred to a cardiologist. She required an annual follow-up for metabolic syndrome and assessment of hypertension, diabetes, and hyperlipidemia. Her

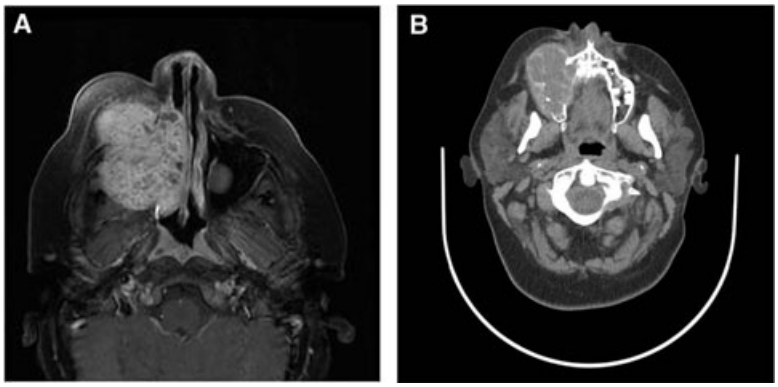

Figure 3 Diagnostic imaging: (A) Preoperative MRI; (B) Preoperative CT.

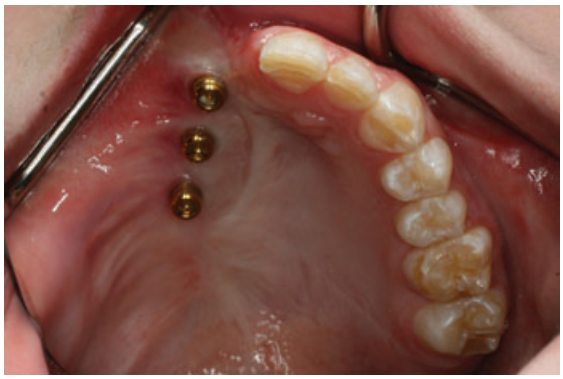

Figure 4 Maxilla without prosthesis: showing endosteal Astra Osseospeed implant placements at site #3 (3.5 × 11 mm), #5 (4.0 × 9 mm), and #6 (5.0 × 9 mm).

concern over the possibility of getting pregnant was addressed by referring her to a fertility clinic where it was assessed she would need an oocyte donor.

Following an additional 6-month healing period, the remaining implants were uncovered, and Locator abutments were selected to go through the skin paddle of the fibula flap and placed intraorally (3 mm height for #5; 4 mm height for #6, Locator Abutment 3.5/4.0) (Fig 4). Prosthetic rehabilitation was initiated with an interim heat-cured acrylic removable partial denture (RPD) (Lucitone 199) with clasp on tooth #15 and pick-up impression copings on implants #3, 5, and 6 (Locator Abutment Pick-up, Astra Tech Inc.) retained with light-cured material (Triad, Dentsply Trubyte). Preliminary impressions of the maxilla and the mandible were made using irreversible hydrocolloid (Jeltrate alginate, Dentsply Caulk, York, PA) and poured up with type III dental stone (Microstone, Whip Mix Corporation, Louisville, KY).

The maxillary cast was surveyed for appropriate RPD design. Rest seats were prepared on teeth #9, 11, and 15. A final impression in light vinyl(poly siloxane) (VPS) (Aquasil Ultra, Dentsply Caulk) was made for the maxilla. A maxillary framework was cast in cobalt-chrome alloy (Wironium, Bego USA, Lincoln, RI), tried in, and adjusted intraorally. Border molding was performed for the maxillary right distal extension, and a VPS-altered cast impression was performed. Jaw relation (centric and protrusive relations) and facebow were recorded using aluwax, and casts were mounted on an articulator set using the protrusive records (Hanau H2 XO, Whip Mix Corporation). High-angle cusp prosthetic denture teeth (Vivodent Blue line Shade B1 mold A12 and PU2, Ivoclar Vivadent Inc., Amherst, NY) were set up in cross-bite with group function. The wax setup was assessed intraorally. Esthetics, phonetics, and function were evaluated. The RPD was processed in original shade

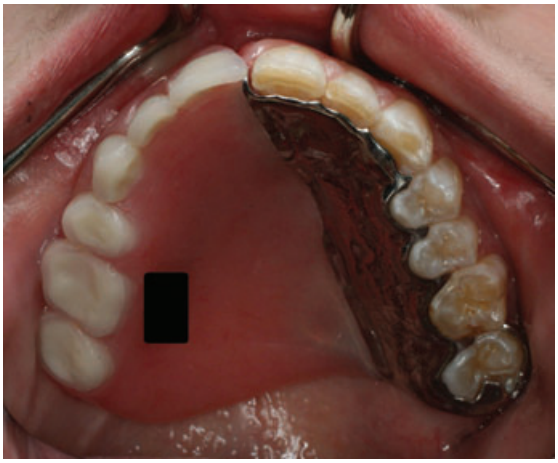

Figure 5 Maxilla with prosthesis.

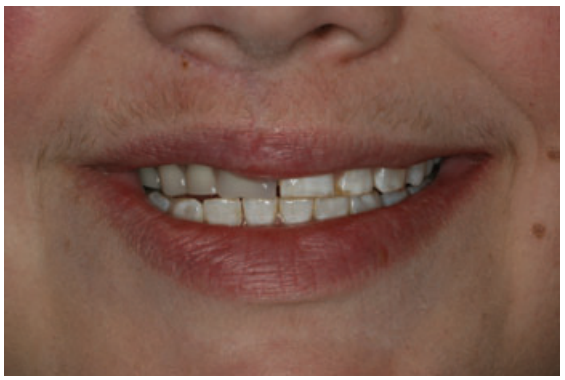

Figure 6 Close-up smile in "Sunday bite" with prosthesis.

heat-cured acrylic resin. The prosthesis was then tried in the patient's mouth using pressure-indicating paste. Adjustments were made on the intaglio surface, and the occlusion was refined intraorally. The prosthesis was then polished and the RPD was delivered to the patient (Fig 5).

The patient was seen at a 24-hour follow-up visit at which time she was asymptomatic. When smiling, the patient had a habit of protruding her mandibular jaw, bringing her teeth to an edge-to-edge position, a phenomenon common with class II patients and described as a "Sunday bite" in the orthodontic literature (Fig 6). The patient was still extremely pleased with the esthetics, function, and comfort of the prosthesis at the 3-month follow-up visit. Further follow-ups in prosthodontics will be scheduled simultaneously with her cancer follow-up appointments.

Discussion

Of the many symptoms of TS, the patient presented with short stature, obesity, ear infections, primary amenorrhea, and multiple melanocytic naevi. She was unaware of the significant medical complications associated with TS. Her cancer workup and treatments revealed further symptoms of poor cortical bone, considerable keloid scar formation, osteopenia, osteoporosis, and infertility. Osteoporosis is a common problem in adult TS patients with reports of increased fracture occurrence, especially in patients where puberty had to be induced.[10,15] Lower osseointegration rates of implants have been reported for osteoporotic and osteopenic bone in comparison to normal bone, and adjustment of surgical technique with a longer healing period is recommended.[16] This may be the cause of the two failed implants in this patient.

The medical and dental literature show a lack of follow-up of TS patients when they transition from childhood to adulthood.[2] Their management should include elaborate workups at the diagnosis of the syndrome with continuous follow-up. Conventional treatments include exercising, calcium and vitamin D supplements, growth hormone, and estrogen replacement therapy.[1,2,6-8] Our patient brought up her concern relating to pregnancy. Only 2% to 4% of patients will be able to have unassisted pregnancy, the vast majority being infertile. Fertility options include achieving in vitro fertilization with an oocyte donor, and current research aims toward the possibility of cryopreserving the follicles in ovaries of adolescent patients for future pregnancies.[2,3,17] Orthodontic evaluation and treatment might be necessary if malocclusion is present.[1] These patients should be closely followed up by a cardiologist and an otorhinolaryngologist throughout their life, as the higher risk of cardiovascular diseases is the main cause of reduction of life expectancy in TS. Antibioprophylaxis may be necessary prior to dental treatments if cardiac anomalies are present.

Conclusion

This clinical report shows the importance of elaborate workup and planning when treatment planning patients with TS as well as other congenital syndromes. In this report, the associated osteoporosis/osteopenia and poor cortical bone quality may have contributed to the early failure of endosteal implants. Extended healing times may be required when placing implants in this patient population. The management of patients with TS is a team effort requiring close interactions between geneticist, cardiologist, endocrinologist, otorhinolaryngologist, audiologist, fertility specialist, psychologist, pedodontist, orthodontist, and prosthodontist.

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