

INTRODUCTION

Ewing Sarcoma is an aggressive and highly malignant undifferentiated round cell tumor. Ewing Sarcoma is commonly found in hips, chest wall, and legs of children between 10-20 years old coinciding with puberty. Cases of congenital Ewing sarcoma in the head and neck region are extremely rare. Side effects of chemotherapy and radiation treatments of pediatric cancer can have long-term side effects on the developing dentition.

Odontogenesis is a complex process and occurs over a long period of time. As each tooth goes through the developmental stages of initiation, proliferation, histodifferentiation, and morphodifferentiation it is susceptible to intrinsic and extrinsic factors. Chemotherapy and radiation has shown to cause dental developmental defect including agenesis, microdontia, blunted roots, and enamel hypoplasia. Craniofacial growth disturbances include reduced vertical growth, mandibular length, and height of alveolar process. The severity of the dental developmental anomaly will depend on the age and stage of development during exposure to cytotoxic agents or ionizing radiation. Craniofacial, skeletal, and dental developmental issues usually develop when children received cancer therapy under 6-years-old. This case report will describe dental manifestations of congenital Ewing sarcoma treatment, including immediate and long-term treatment.

CASE REPORT

A 5-year-old female presented to University Hospitals Rainbow Babies and Children for comprehensive dental care under general anesthesia in November 2024. Patients past medical history is significant for congenital Ewing Sarcoma of the right masseter region, 14 cycles of chemotherapy (8 days old), head radiation of 55.8 cGY in 31 fractions (7 months old), gastrostomy tube, and previous tracheostomy. Patient was last seen for dental examination in October 2022, and since developed sensitivity of the maxillary anterior.

Extra oral examination revealed brachycephalic facial profile with reduced lower anterior facial height and retrognathic mandible (*Figure 2A and 2B*). Clinical exam revealed class 2 malocclusion with moderate maxillary crowding and severe mandibular crowding(*Figure 1A and 1B*). Generalized gingivitis present secondary to plaque accumulation. Full primary dentition present with grade 1 mobility of molars, grade 2 mobility of anterior dentition, and non-restorable caries present on tooth D. Radiographic exam revealed normal bone levels, blunted roots and constricted cementum-enamel junction of all primary teeth, suspected agenesis of all permanent teeth with the exception of permanent maxillary central incisors, mandibular incisors, and first molars present at the calcification stage (*Figure 3*). Non-restorable caries was noted on tooth D with remaining dentition intact (*Figure3B*).

CLINICAL AND RADIOGRAPHIC PRESENTATION

Clinical Presentation



Figure 1: Intra Oral Photos



Figure 2: Extra Oral Photos

Radiographic Presentation

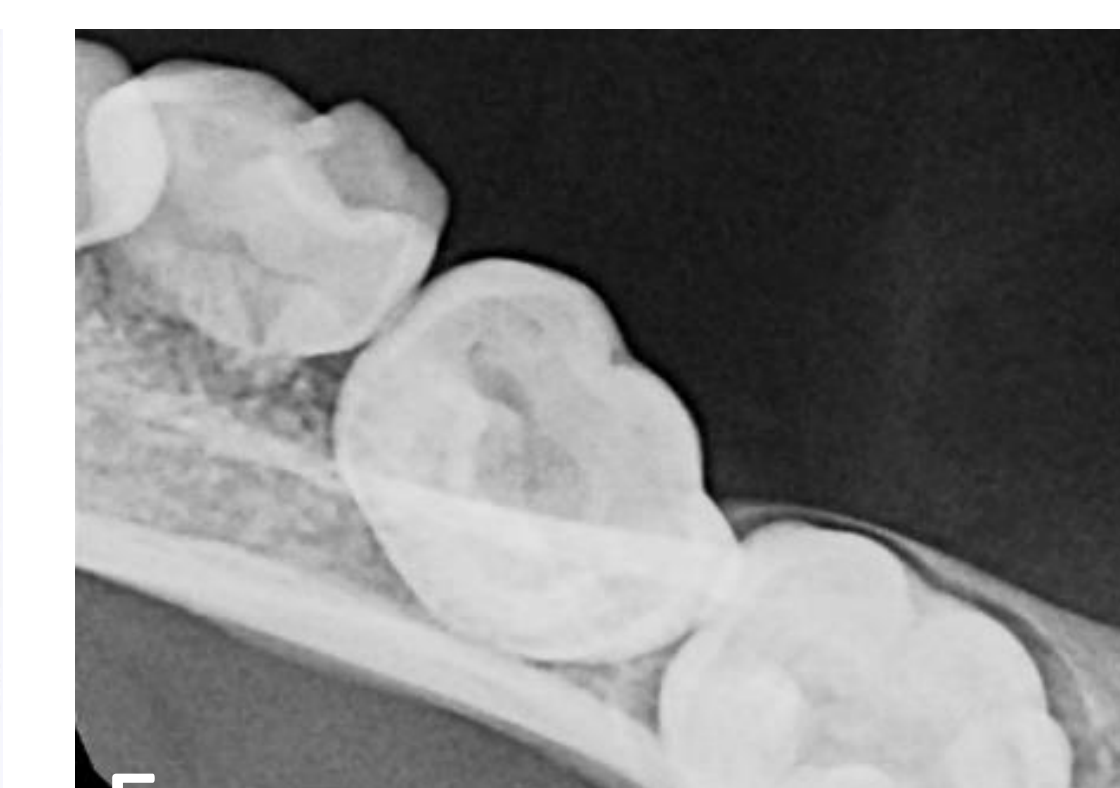
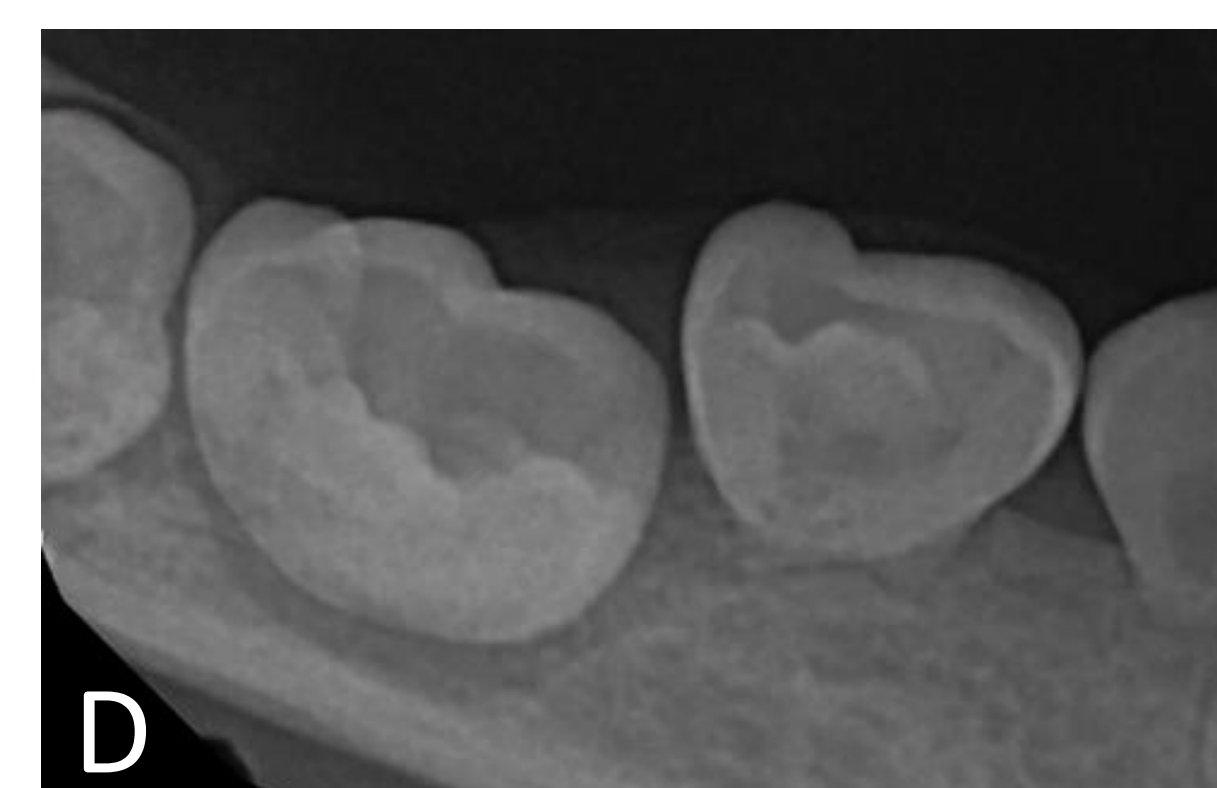
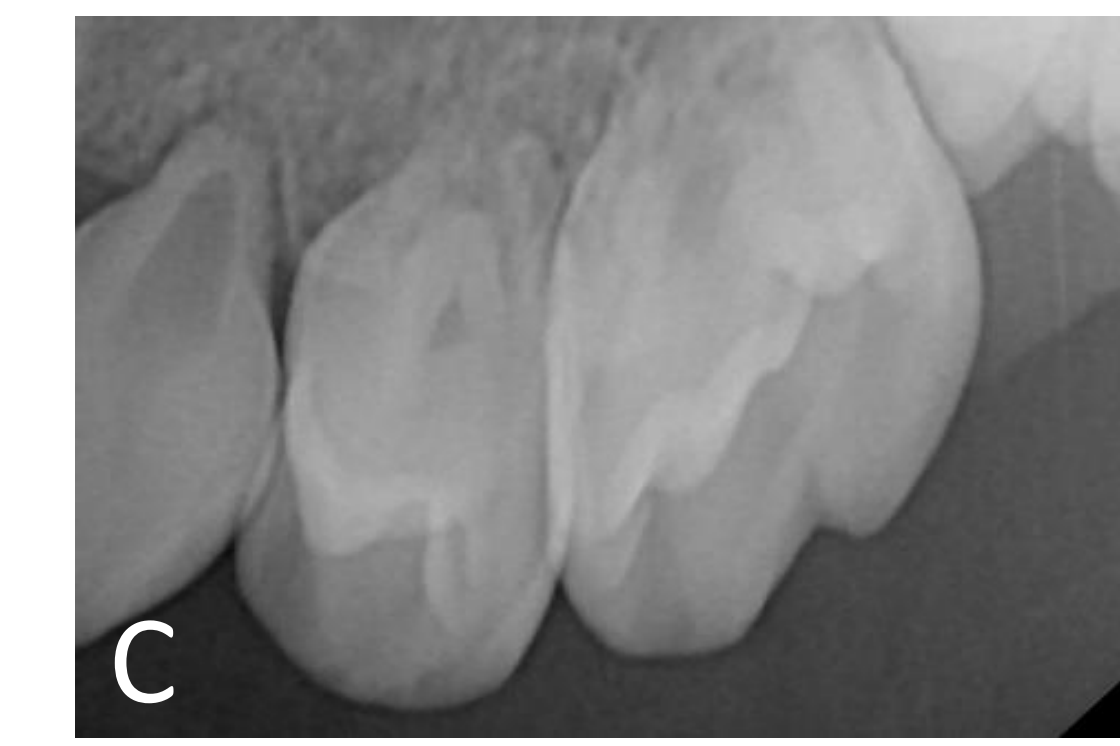
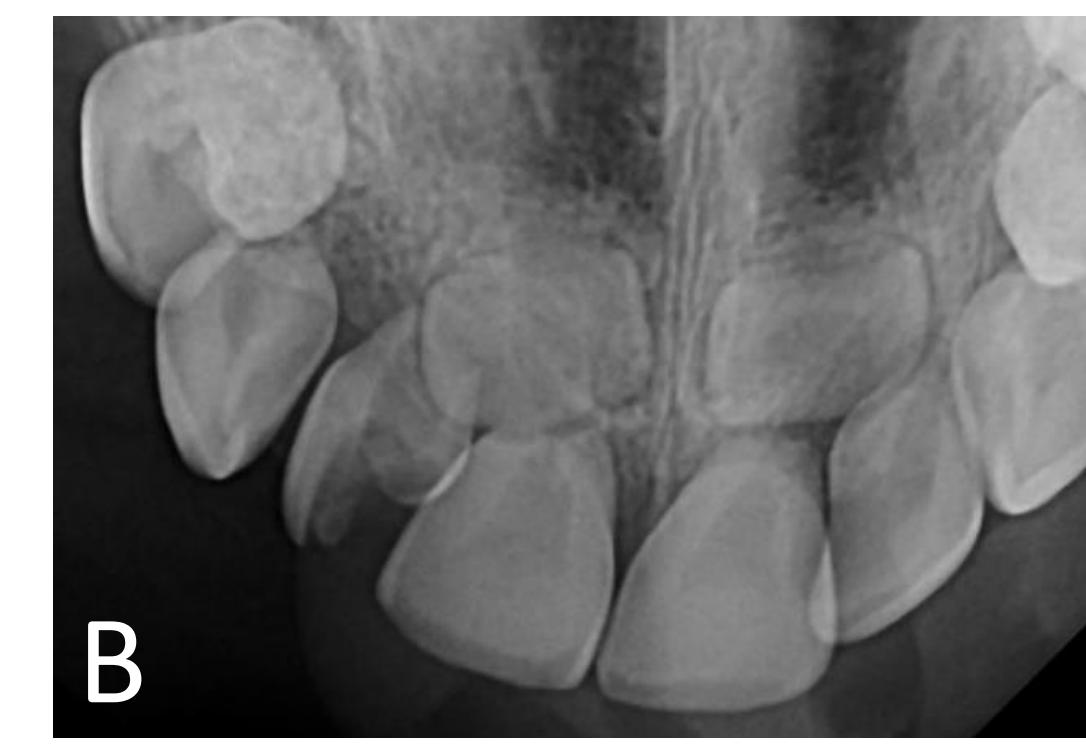
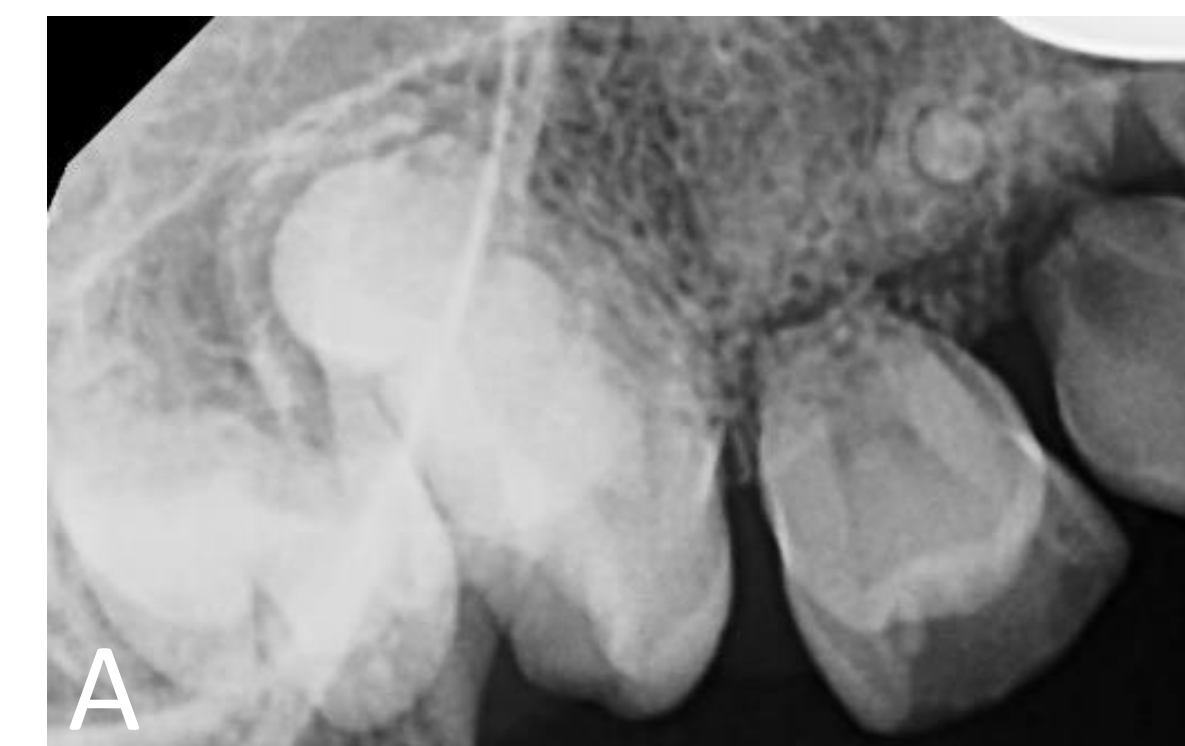


Figure 3: Radiographic examination

DISCUSSION

The dental management of children who have undergone chemotherapy and head/neck radiation early in life is extremely challenging. This case demonstrates the dental manifestations from treatment of a rare congenital Ewing sarcoma of the masseter region in which the patient began chemotherapy at 8 days old and radiation at 7 months old. Patient had 10 months of treatment ending in April of 2020(10 months old). At 5 years old, evidence of the beginning of calcification would be expected on all permanent teeth with the exception of third molars. Therefore, the patient presented almost certainly has agenesis of #s 4, 5, 6, 7, 10, 11, 12, 13, 20, 21, 22, 27, 28, and 29. Although unlikely, current radiographic examination is unable to determine crown formation of the second and third permanent molars.

The immediate treatment plan in the case presented included extraction of tooth D and a dental prophylaxis using toothbrush and prophy paste.

The future dental plan includes retaining existing primary teeth indefinitely to keep level of alveolar bone for future prosthesis/Implants and continued follow up with craniofacial team. Patient will have continued follow up at six-month intervals on an outpatient basis with pediatric dental team and scheduled for treatment under sedation as indicated.

Studies have shown retaining deciduous teeth as appropriate treatments in children and adolescents, and higher success for implant and implant supported prosthesis after 13 years old. The literature examining long term treatment modalities of children having been treated for congenital head and neck cancer is extremely limited and further research is needed as survival rates of pediatric cancer increases.

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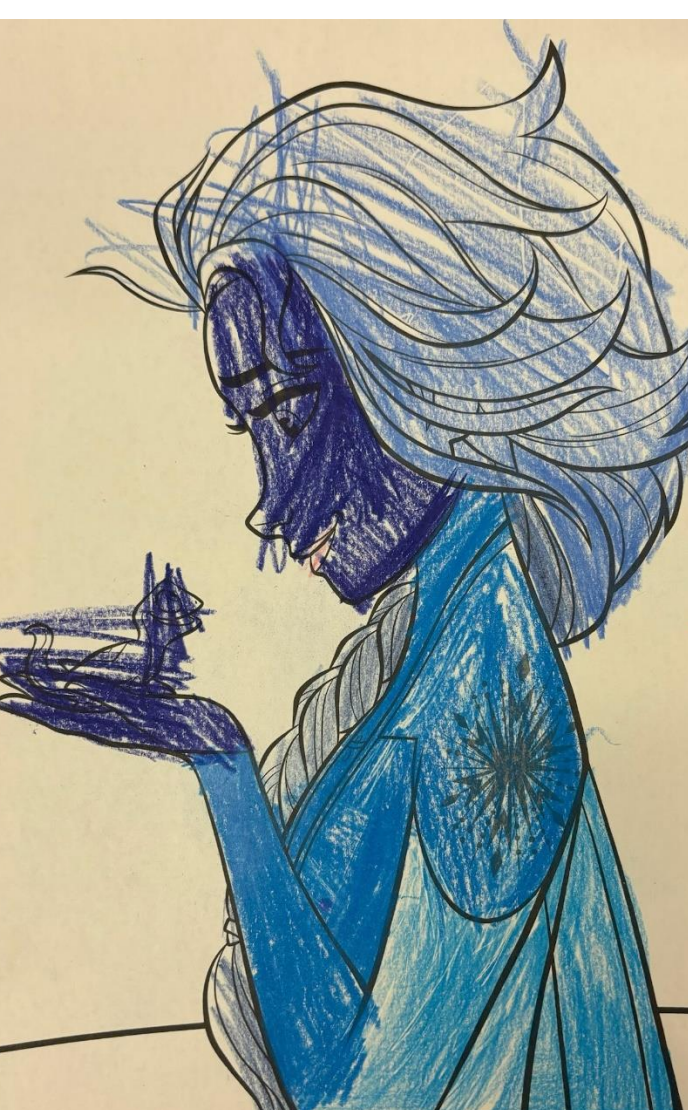


Figure 4: Patients coloring in pre-op prior to dental surgery