Pancreatoblastoma and Paediatric Dental Management - A Case Report

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Abstract: Pancreatoblastoma originates from the epithelial exocrine cells of the pancreas. An 8 year old boy attends AKUH dental clinic with a toothache, pancreatoblastoma with an immunocompromised health status, early mixed dentition showed maxillary, UR1, UR2, UL1, UL2, LL1, LL2, LR1, LR2, first permanent molars UR6, UL6, LL6, LR6 with taurodontism, compromised oral hygiene, chemotherapy induced gingivitis in the mandibular Central and lateral incisors, mucositis, grossly carious URE, ULE, LLE and LRE, retained root of LLD, mobile LRC and missing URC, ULC, URD.

The treatment plan included preventive advice, non-pharmacological behavior management, dental prophylaxis, fissure sealants for first permanent molars, GIC temporization of LLE, extractions of URE, ULE and LLD under local anesthesia and Duraphat fluoride varnish application. A follow up was advised after the second cycle of chemotherapy.

This case report focuses on the oral health effects of pancreatoblastoma and its dental management in a child patient

Keyword: Pancreatoblastoma, Dentition, Mixed chemotherapy, Adjuvant, Toothache, Immunocompromised host.

1. INTRODUCTION

Childhood cancers are the 4th most common cause of mortality in children younger than 15 years of age and account for nearly 1% of all population cases [1].

Pancreatoblastoma or infantile pancreatic carcinoma is a very rare, malignant, slow growing childhood tumor encompassing 0.5% of non-endocrine pancreatic tumors [2]. East Asian and Africans commonly present with mixed solid and cystic lesions [3]. Cancer therapy is a complete surgical resection with initial neoadjuvant chemotherapy. Follow up and monitoring with history, physical examination and imaging is important for treating recurrence [4].

Oral cavity in immunocompromised cancer patients is vastly prone to the effects of chemotherapy and radiation therapy [5]. Various acute oral sequelae with associated systemic complications include salivary gland dysfunction (xerostomia), taste dysfunction, oral ulcers, pain, mucositis, dental caries, secondary infections (candidiasis, herpes simplex virus), neurotoxicity, soft tissue necrosis, mucosal fibrosis, postradiation osteonecrosis, temporomandibular dysfunction (trismus), oral graft versus host disease (GVHD) and craniofacial and dental developmental anomalies [6,7].

The multidisciplinary team approach of childhood cancer patient management integrates the skills of the primary care physician, pediatric medical oncologists/hematologists pediatric surgeons, pediatric nurse specialists radiation oncologists, rehabilitation specialists, dentists, social workers, dieticians and other related health related professionals to warrant that children receive treatment, supportive care and rehabilitation for achieving an optimal survival and quality of life [8].

Dental evaluation is performed after the diagnosis of cancer and before, during and after the cancer therapy. This aids the diagnosis, prevention, stabilization and treatment of oral and dental manifestations secondary to the underlying systemic medical conditions and treatment of childhood cancers. It is important to educate patient and caregivers about the importance of good oral hygiene and to tailor a dental treatment plan in conjunction with the medical team [9].

2. CLINICAL CASE

An 8 year old boy with a medical history of pancreatoblastoma and an immunocompromised status was referred by the paediatric oncologist / hematologist for caries assessment. He was to undergo a second cycle of chemotherapy with cisplatin and doxorubicin followed by a surgery.

The first antineoplastic chemotherapy (Injection or infusion of cancer therapeutic substances) was elective. Prior investigations included a Biopsy, abdominal CT scan, lab investigations (CBC, ESR, LFT, calcium and Phosphate, Albumin, SGPT) and electrolyte imbalance. Pic line accessed, intravenous fluids, antiemetics and cisplatin with pre and post hydration were administered. Electrolyte imbalance was corrected and the patient was discharged in a stable condition.

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On the first dental visit an intraoral and radiographic assessment (OPG) indicated early mixed dentition, fair oral hygiene, completely erupted permanent central and lateral incisors- UR1, UR2, UL1, UL2, LL1, LL2, LR1, LR2 and all the first permanent molars UR6, UL6, LL6, LR6, chemotherapy induced gingivitis, mucositis, grossly carious URE, ULE, LLE and LRE, LLD roots, mobile LRC and missing URC, ULC and URD. Clinical pictures were taken following father’s written consent (Figures 1-4).

On the second visit a non-pharmacological behavior management approach was followed by prophylaxis, fissure sealing of the first permanent molars and a duraphat fluoride varnish (22,600 ppm) application.

On the third visit URE, ULE and LLD were extracted under Local anesthesia. The postoperative instructions provided included dietary advice (soft and cold for 24 hours) and analgesics. Preventive advice included dietary advice, brushing techniques with 1500 PPM fluoride toothpaste on a soft bristled toothbrush and saline rinses three times a day for one week.

The patient did not follow up after the second cycle of chemotherapy since he went to his village.

3. DISCUSSION

3.1. Cancers

Cancers are a multistage, multifactorial diseases arising secondary to environmental and genetic factors interplay [13]. Primary stage causes irreversible DNA damage due to chemical, infection or radiation. Secondary/promotion stage damages all the regulatory DNA processes. Third/progression phase is tumor metastases [10].

Detection, prognosis and monitoring rely on detection of molecular changes through DNA markers. Childhood cancers exhibit a variation in histological types

![Figure 1: An orthopentamogram showing the primary and permanent dentition.](image1)

![Figure 2: Intraoral clinical picture-facial view.](image2)
with few etiological connections whereas adult carcinomas have strong etiological associations [11].

3.2. Pancreatic Tumors

Pancreatic tumors are rare malignancies affecting 1-8 year olds with a mean age of 5 years and an incidence of 0.46 cases per 1 million under 30 years. Primary pancreatic tumors include endocrine and exocrine epithelial/non epithelial tumors. Relative survival rate for pancreatic cancer combined is 20% and five-year survival rate is 6% [12].

3.3. Pancreatoblastoma

Pancreatoblastoma is the most common childhood malignant exocrine pancreatic tumor arising from the pancreatic epithelium in upper abdomen with hemorrhage and fibrous peripheral capsule and diameter between 7 to 18cm. Lobules and nests of acinar and gland like cell formation are found along with squamous cell nests.

Ration for male to female is 1.14:1 and dominates in men and Asians. It approximates 0.5% of all exocrine pancreatic tumors, incidence less than 1% with 5 year median age; age ranges from 0-68 years with a 48 month median survival rate. Metastasis at initial presentation is seen in advanced stages [2,13].

Beckwith-Wiedemann syndrome and Familial Adenomatous Polyposis syndrome are screened with ultrasound and cross sectional imaging. Genetic alterations exhibit allele loss on chromosome 11p15 and mutations in the path of edematous polyposis coli/β-catenin. Alpha-fetoprotein elevates and symptoms include large abdominal mass, distention with upper abdominal pain, anorexia, vomiting, weight loss and failure to thrive [14].

3.4. Diagnosis

Diagnosis A definitive diagnosis is ascertained by biopsy and histology after distinguishing it from other differential causes of pediatric upper abdominal mass-
es like wilms tumor, neuroblastoma, hepatoblastoma or Non-Hodgkin lymphoma [15].

3.5. Treatment

Treatment of pancreatoblastoma includes surgical procedures for removal of pancreas and duodenum or part of the pancreas (head, body or tail). Complete surgical resection depends on the location, local extent and presence or absence of metastasis. Chemotherapy may be advantageous for managing localized unresectable tumor, metastatic or recurrent pancreatic carcinoma with a dual drug combination of cisplatin and doxorubicin. Radiation is indicated after incomplete resection, tumor spillage or positive surgical margins [16].

3.6. Childhood Cancer in Pakistan

Third world country prevalence ranges from 4.38% to 12.6%. 74% of the world’s total population lives in developing countries out of which 39% of the developing countries population whereas 23% of the total population in west constitutes Children under15 years of age. The total population of children less than fifteen years of age in Pakistan is almost 44% [17].

4. DENTAL AND ORAL CARE FOR CHILDREN RECEIVING CANCER THERAPY

The corner stone of effects of post-surgical, post chemotherapy post radiotherapy on the head and neck structures surrounding the tumor are an initial patient evaluation, hematological considerations, preventive advice, dental clinical procedures either before the cancer therapy initiates, during the therapy with immunosuppression or after completion of therapy.

A paediatric dentist liaises with an oncologist to provide dental care from the time of diagnosis, during the initial stages of chemotherapy till completion and remission phase for a careful consideration of the patient specific underlying systemic condition. The nature and degree of acute and chronic complications correlate with age, extent and location of malignancy, time and dose of chemotherapeutic agent and initial oral health status and level of care before, during and after the cancer therapy [18].

A medical history informs about the type of disease with the stage and prognosis, the type of treatment being instituted; medicines, allergy status, hematological and immunosuppression status and information about primary care physician/ oncology team.

A dental history gathers information on the oral hygiene, dietary habits, preventive practices, fluoride exposure, symptomatic teeth, previous care, and trauma. Clinical and radiological dental assessment of the head and neck with intraoral examinations aids assessment of oral hygiene status [19].

A dental assessment identifies and stabilizes or eradicates sources of infection which may either be predominant or probable, communicate the patient’s oral health status and treatment plan to the oncology team and to educate the patient and parents about the importance of optimum oral care in all the stages of cancer therapy [19].

4.1. Hematological Considerations

Assesses neutrophil and platelet count. In dental emergencies consult the physician supportive measures like platelet transfusions, bleeding control, hospital admission and care. Hemostasis in local procedures (microfibrillar collagen, topical thrombin) and medications recommended by the hematologist/oncologist (aminocaproic acid, tranexamic acid) may help control bleeding [19].

4.2. Preventive Strategies

Preventive strategies comprise of oral hygiene instructions, diet advice, teeth and tongue brushing with a soft nylon brush or electric toothbrush two/three times a day with a fluoridated toothpaste or gel, flossing, sucrose in pediatric medication, fluoride varnish for caries risk and xerostomia, [4] Lip care with Lanolin-based creams and ointments.

4.3. Dental Clinical Procedures

Treatment is completed before the cancer therapy initiates since blood counts start to fall five to seven days after each cycle begins and resumes normal levels few days after until the next cycle starts. If it cannot before cancer therapy; place temporary restorations with a delay in non-acute dental treatment until hematologically stable [1,19].

Prioritize infections, extractions, periodontal care, rectification of sources of tissue irritation before treating caries, root canal for permanent teeth and replacement of faulty fillings [3]. Use fluoride and/or sealants to treat incipient to small carious lesions until definitive care [19].
For pulpally affected teeth extraction is definitive treatment followed by an antibiotic therapy (penicillin or, for penicillin-allergic patients, clindamycin) for one week since pulpal infections threaten in immunosuppression. Two weeks prior to the cancer therapy extract root fragment, manage periodontal pockets more than 6 mm in size, treat symptomatic impaction, bone loss and mobility or furcal involvement. Delay endodontic treatment for asymptomatic non-vital permanent teeth until the patient is hematologically stable [19,20].

Manage mucositis with oral hygiene, analgesics, non-medicated oral rinses (eg, 0.9 percent saline or sodium bicarbonate mouth rinses four to six times/day), parenteral nutrition and mucosal coating agents like Amphogel®, film-forming agents like Zilactin® oral cryotherapy as prophylaxis, Palifermin [20].

Oral bleeding secondary to thrombocytopenia, disorder of coagulation factors and compromised vessel integrity is managed by pressure packs, antifibrinolytic rinses or topical agents, gelatin sponges locally and platelet transfusions and aminocaproic acid systemically. Dental sensitivity subsides after the chemotherapy concludes.

Xerostomia can be managed by sugar free chewing gums, sucking tablets, special dentifrices, saliva substitute for oral dryness, frequent sips of water, alcohol-free oral rinses, and/or oral moisturizer and bedside humidifier at night. Saliva stimulating drugs are contraindicated in children [20].

Orthodontic treatment may commence or resume after the therapy concludes and after at least two-year disease-free survival when the immunocompromised drugs stoop and the risk of relapse diminishes.

Oral surgery requires consultation with a periodontist, an oral surgeon, physician and an oncologist for non-elective oral surgical and invasive periodontal procedures but avoid elective invasive procedures.

Elective dental care should be deferred if a dental emergency arises. The treatment plan should be discussed by the dentist with the patient’s physician for supportive medical therapies like antibiotics, platelet transfusions and analgesia [20].

5. CONCLUSION

Hematological profile during the stages of cancer therapy dictates oral health management for the extent and severity of dental caries (number of affected teeth and those needing immediate treatment), endodontic procedure (pulpal versus periapical infection), periodontal status, extractions, soft tissue pathology and any other urgent needs.

Hence diagnosis and management of post-treatment oral sequelae of pancreatoblastoma; a rare childhood tumor; requires a multidisciplinary approach whereby the pediatric dentist liaises with the pediatric oncology/hematology team to tailor a comprehensive oral care plan and provide dental care from the time of diagnosis, during initial chemotherapy stages till the completion and remission phase specific to the needs of every child.

CONFLICT OF INTEREST

As the sole author I declare that I have read the authorship guidelines and have no financial or any other affiliation that may influence this case report.

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