SUNTEXT REVIEWS

SunText Review of Pediatric Care
ISSN: 2766-5216

Open Access
Review Article
Volume 1:2

Malignant Neoplasms of Head and Neck in Children

Shunmugavelu K* and Javid Shaikh SM

Consultant Dental Surgeon, Department of Pediatric Dentistry, Kanchi Kamakoti Childs Trust Hospital, Chennai, India

***Corresponding author:** Shunmugavelu K, Consultant Dental Surgeon, Department of Pediatric Dentistry, Kanchi Kamakoti Childs Trust Hospital, Chennai, India; Tel: 0091-9789885622; E-mail: drkarthiks1981@gmail.com

Received date: 25 November 2020; **Accepted date:** 10 December 2020; **Published date:** 17 December 2020

Citation: Shunmugavelu K, Javid Shaikh SM (2020). Malignant Neoplasms of Head and Neck in Children. SunText Rev Pediatr Care 1(2): 110.

DOI: https://doi.org/10.51737/2766-5216.2020.010

Abstract

Head and neck neoplasms are uncommon in pediatric patients and affect 12% of children. Head and neck masses may occur from a variety of causes, including infection such as enlarged lymph nodes, congenital cysts or masses that formed during development, or tumors.

Keywords: Lymphomas; Neoplasms; Tumors; Salivary gland

Introduction

Head and neck neoplasms are uncommon in pediatric patients and affect 12% of children. Head and neck masses may occur from a variety of causes, including infection such as enlarged lymph nodes, congenital cysts or masses that formed during development, or tumors. Approximately 1 in 10 childhood neoplasms involve the head and neck region. The children with diseases of the head and neck region are most frequently identified by pediatricians and otorhinolaryngologists. The presenting symptoms in the children with head and neck neoplasm are indistinct, which makes the diagnosis difficult. The tumor size, primary site of involvement, etiology, and presence of regional or distant metastases determines the prognosis of head and neck tumors. The early diagnosis of the tumors favors good prognosis. The most common pediatric head and neck malignancies are lymphomas (27%), neural tumors (23%), thyroid malignancies (21%), soft tissue sarcomas (12%), nasopharyngeal carcinoma, skeletal malignancies, and salivary gland malignancies.

Lymphomas

Lymphomas are characterized by the proliferation of mature lymphoid cells and are the heterogeneous group of malignant tumors of the hematopoietic system [1]. It is the second most common malignancy in the world [2]. The diagnosis of lymphoma is mainly based on the histhopathological examination of tissue from enlarged lymph nodes. They are broadly classified into Hodgkin's lymphoma and Non-Hodgkin's lymphoma.

Non-hodgkin's Lymphoma

Non-Hodgkin lymphoma is the fourth most common malignancy in children [2]. It most commonly occurs in children older than 10 years but can also occurs in children less than 5 years [2]. Sixty percent of lymphomas are Non-hodgkin type.

Typical symptoms can include painless swelling of lymph nodes in neck, chest, abdomen or groin, fatigue, occasionally such as fever $>38\,^{\circ}C$, night sweats and weight loss ($>10\%$ within 6 month), susceptibility to infections and changes in the hemogram. Other symptoms include abdominal pain, diarrhea, dyspnea, and dysphagia [1-4].

Treatment depends on type and staging. It includes chemotherapy, radiation therapy, surgery, use of monoclonal antibodies, high dose chemotherapy and supportive care [4].

Hodgkin's Lymphoma

Hodgkin lymphoma is characterized by the presence of typical multinucleated Reed-Sternberg cells, mononucleated cells known as Hodgkin Reed-Sternberg cells and they are derived from B

cells. It affects 6 % of the pediatric population and it occurs most commonly in male [5,6].

Most common symptoms include painless adenopathy in supraclavicular or cervical areas while some children show B symptoms – unexplained fever, drenching night sweats and weight loss [6,7].

Treatment includes chemotherapy, radiotherapy, and surgical excision of tumors. The risk of relapse is high, and the primary complication is development of secondary malignancy which remains a cause of concern [6].

Rhabdomyosarcoma

Rhabdomyosarcoma, the mesenchymal malignant neoplasm with skeletal muscle differentiation is the most common soft tissue sarcoma in the pediatric population [8]. It most commonly occurs in 1 to 4 years old and least common in children older than 10 years of age [9]. In head and neck region, orbit is the most site of involvement and in the oral cavity, the tongue followed by the soft palate, hard palate, and buccal mucosa are most are most involved. Etiology of Rhabdomyosarcoma is unknown but may be caused due to in utero radiation exposure, accelerated in utero growth, low socioeconomic status, and parents using recreational drugs during pregnancy. It presents as poorly circumscribed, white, soft, or firm, infiltrative masses. The tumor at the time of diagnosis is around 5cm in diameter [10].

Clinical manifestations of RMS include small cutaneous nodule on the face or an extensive fast-growing facial swelling, trismus, paresthesia, facial palsy, and nasal discharge. The pain, paresthesia, loss of teeth, and trismus are caused due to advanced tumor stage, infiltrative growth, and tumor location.

The treatment of RMS is a multidisciplinary approach, surgical removal of the tumor followed by chemotherapy with or without radiotherapy since RMS tends to metastasize to bone marrow. Prognosis of RMS is poor when compared to other oral soft tissue malignant lesions [9].

Nasopharyngeal Carcinoma

Nasopharyngeal carcinoma (NPC) is a malignant neoplasm arising from the epithelial cells lining the nasopharynx and rare malignancy in the pediatric age group [11-13]. Nasopharyngeal carcinoma affects around 5% of pediatric population [11]. NPC is a rare tumor in children younger than 10 years of age, and the incidence rises gradually with age of the patient. It is a common head-and-neck tumor seen in adults [13]. The median age of nasopharyngeal occurrence in children is 13 years with male predominance [11].

The genetic susceptibility, environmental factors such as exposure to chemical carcinogens, and infections with Epstein–Barr virus (EBV) are etiological factors related to NPC. The clinical symptoms of NPC include epistaxis, nasal obstruction, deafness, tinnitus, headache, and neck swelling. Bone erosion at the skull base region with or without affecting cranial nerves can also been seen [13].

The standard treatment for NPC is chemotherapy with surgery [13]. Treatment related toxicity includes mucositis, skin erythema, xerostomia, neck fibrosis, dental caries, trismus, hypopituitarism, stunted growth, and hypothyroidism [12].

Thyroid Carcinoma

Thyroid carcinoma is rare in the pediatric population, children more often present with aggressive and advanced stage of disease. It occurs in the second decade of life with female predominance (male female ratio is 1: 6) [14]. It is the 5th most common tumor and affects around 6 % of the pediatric population [14,15].

Signs and symptoms are thyroid nodule with asymptomatic neck mass, with or without cervical lymphadenopathy, trouble breathing and hyperthyroidism [14]. In pediatric thyroid cancer multiple nodules are present and they also have a higher risk of recurrence than adults [14].

The treatment of choice is total thyroidectomy along with radioactive iodine therapy and a near-total thyroidectomy is also performed in some cases, in which a small amount of thyroid tissue near the recurrent laryngeal nerve or superior parathyroid glands may be spared to decrease the possibility of damage to adjacent structures [14]. Pediatric patients have better prognosis and significantly lower mortality rates than adults [15].

Mucoepidermoid Carcinoma

Mucoepidermoid carcinoma (MEC) is the most common primary epithelial salivary gland malignancy [16]. Mucoepidermoid carcinomas account for 50% of malignant salivary gland tumors [18]. Most common in the parotid gland, only a few pediatric and adolescence cases have been reported in minor salivary glands [17]. The most common site for minor salivary gland tumors is the palatal region, followed by buccal mucosa. It occurs around 10-16 years of age with female predominance [16,17].

When the major salivary glands and tongue are involved, pain, paresthesia, and difficulty with swallowing are also present. Intraoral lesions appear as a localized fluctuant nodule with a bluish or reddish-purple, smooth, mucosal surface. In some case mucus may be discharged from the tumor through a small sinus tract. High grade lesions may be quite firm along with ulceration, resorption of bone and numbness of adjacent teeth [16].

Treatment for MEC is typically surgical resection. Complete excision (superficial or total parotidectomy) with preservation of facial nerve is the treatment of choice. Neck dissection should be considered when there is clinical evidence of regional metastasis, high TNM stage, high histologic grade, and involvement of regional nodes [18-20].

Neuroblastoma

Neuroblastoma is the most common extracranial soft tissue tumor affecting children less than 12 months of age and the second most common neoplasm after Rhabdomyosarcoma. The tumor arises from primitive neuroectodermal cells, which are derived from neural crest cells. Most cases are caused by sporadic mutation, in recent studies autosomal dominant are reported, with ALK and PHOX2B oncogenic mutations.

The palpable abdominal mass generalized bone pain, malaise, fever, and irritability are the most common clinical presentation. Primary cervical neuroblastoma should be ruled out in any child less than one year of age with a persistent neck mass. Horner's syndrome was the most common presentation in the head and neck neuroblastoma.

Diagnosis is usually made based on imaging and biopsy. Urine catecholamines plays an important role in diagnosis. Neuroblastomata are characterized by defective catecholamine synthesis. This leads to an accumulation and excretion of the catecholamine metabolites homovanillic acid (HVA) and vanillylmandelic acid (VMA). These levels are high in children with neuroblastoma.

Treatment options for neuroblastoma are determined by tumor stage, patient age, tumor location and resect ability, tumor histopathology, and MYCN gene amplification. Surgery with clear margins is the important prognostic factor. Multidrug chemotherapeutic regimens can be used in the preoperative or postoperative setting.

Conclusion

Incidence of head and neck neoplasms in the pediatric age group is rare but the incidence is increasing. Awareness about the malignancies and careful follow up with proper diagnosis in suspicious cases is mandatory so that proper treatment can be provided to such patients and the risk of metastasis is reduced which in turn helps to improve the prognosis. Therefore, it is important to have these masses evaluated if they persist.

References

1. Storck K, Brandstetter M, Keller U, Knopf A. Clinical presentation, and characteristics of lymphoma in the head and neck region. Head Face Med. 2019; 15: 1-8.
2. Minard-Colin V, Brugières L, Reiter A, Cairo MS, Gross TG, Woessmann W, et al. Non-Hodgkin lymphoma in children and adolescents: progress through effective collaboration, current knowledge, and challenges ahead. J Clin Oncol. 2015; 33: 2963.
3. Mărginean CO, Meliţ LE, Horvath E, Gozar H, Chinceşan MI. Non-Hodgkin lymphoma, diagnostic, and prognostic particularities in children–a series of case reports and a review of the literature (CARE compliant). Medicine. 2018; 97.
4. Song J, Li S. Hodgkin Lymphomas. In: Wang E., Lagoo A. (eds) Practical Lymph Node and Bone Marrow Pathology. Practical Anatomic Pathology. Springer, 2020; 189-208.
5. Sherief LM, Elsafy UR, Abdelkhalek ER, Kamal NM, Elbehedy R, Hassan TH, et al. Hodgkin lymphoma in childhood: clinicopathological features and therapy outcome at 2 centers from a developing country. Medicine. 2015; 94.
6. Nagpal P, Akl MR, Ayoub NM, Tomiyama T, Cousins T, Tai B, et al. Pediatric Hodgkin lymphoma–biomarkers, drugs, and clinical trials for translational science and medicine. Oncotarget. 2016; 7: 67551.
7. Abbas AA, Almaghraby HQ. Hodgkin lymphoma in children and adolescents: Advances in pathology, diagnosis, and treatment strategies. Indian J Med Paediatric Oncol. 2020; 41: 492.
8. Shern JF, Yohe ME, Khan J. Pediatric rhabdomyosarcoma. Critical Reviews™ in Oncogenesis. 2015; 20.
9. Shrutha SP, Vinit GB. Rhabdomyosarcoma in a pediatric patient: A rare case report. Contemporary Clin Dentistry. 2015; 6: 113.
10. Kaseb H, Babiker HM. Cancer, Rhabdomyosarcoma. StatPearls Publishing, 2019.
11. González-Motta A, González G, Bermudéz Y, Maldonado MC, Castañeda JM, Lopéz D, et al. Pediatric nasopharyngeal cancer: case report and review of the literature. Cureus. 2016; 8.
12. Khalil EM, Anwar MM. Treatment results of pediatric nasopharyngeal carcinoma, NCI, Cairo University experience. J Egyptian National Cancer Institute. 2015; 27: 119-128.
13. Swain SK, Samal S, Anand N, Mohanty JN. Pediatric nasopharyngeal carcinoma. Int J Health Allied Sci. 2020; 9: 1.
14. Paulson VA, Rudzinski ER, Hawkins DS. Thyroid cancer in the pediatric population. Genes. 2019; 10: 723.
15. Azhar Y, Achmad D, Lukman K, Hilmanto D. Pediatric differentiated thyroid carcinoma risk factor for analysis for disease free survival. Indian J Med Paediatric Oncol. 2018; 39: 153.
16. Flaitz CM. Mucoepidermoid carcinoma of the palate in a child. Pediatric Dentistry. 2000; 22: 292-293.
17. Ritwik P, Cordell KG, Brannon RB. Minor salivary gland mucoepidermoid carcinoma in children and adolescents: a case series and review of the literature. J Medical Case Rep. 2012; 6: 182.
18. Rahbar R, Grimmer JF, Vargas SO, Robson CD, Mack JW, et al. Mucoepidermoid carcinoma of the parotid gland in children: a 10-year experience. Arch Otolaryngol Head Neck Surgery. 2006; 132: 375-380.
19. Kaufmann MR, Camilon PR, Janz TA, Levi JR. Factors associated with the improved survival of head and neck neuroblastomas compared to other body sites. Annals Otol Rhinol Laryngol. 2019; 128: 241-248.
20. Su SY, Bell D, Ferrarotto R, Phan J, Roberts D, Kupferman ME, et al. Outcomes for olfactory neuroblastoma treated with induction chemotherapy. Head Neck. 2017; 39: 1671-1679.
21. .