Journal of Multidisciplinary Dental Research

Review Article

Pediatric Head and Neck Malignancies

R Sowmya[1], Sowndarya Gunasekaran[2,*], D Vinola[3], J Baby John[4], Basil M Mathew[2], Pradeep Daniel Gainneos[2]

[1]*CRI, Vinayaka Mission's Sankarachariyar Dental College, Vinayaka Mission's Research Foundation (Deemed to be University), NH 47 Sankari Main Road, Ariyanoor, 636308, Salem, India*
[2]*Assistant Professor, Department of Pediatric and Preventive Dentistry, Vinayaka Mission's Sankarachariyar Dental College, Vinayaka Mission's Research Foundation (Deemed to be University), NH 47 Sankari Main Road, Ariyanoor, Salem, 636308, India*
[3]*Professor and HOD , Department of Pediatric and Preventive Dentistry, Vinayaka Mission's Sankarachariyar Dental College, Vinayaka Mission's Research Foundation (Deemed to be University), NH 47 Sankari Main Road, Ariyanoor, 636308, Salem, India*
[4]*Professor and Dean, Department of Pediatric and Preventive Dentistry, Vinayaka Mission's Sankarachariyar Dental College, Vinayaka Mission's Research Foundation (Deemed to be University), NH 47 Sankari Main Road, Ariyanoor, Salem, 636308, India*

ARTICLE INFO

Article history:
Received 19.09.2022
Accepted 25.11.2022
Published 28.12.2022

* *Corresponding author.*
Sowndarya Gunasekaran
drsowndaryag@vmsdc.edu.in

https://doi.org/10.38138/JMDR/v8i2.22.42

ABSTRACT

Head and neck masses represents a common clinical entity in pediatric age group. In general, these masses are classified in to developmental, inflammatory, or neoplastic. Having a working knowledge of these lesions in head and neck region & conducting a thorough history & physical examinations generally enable the clinician to simplify an appropriate workup and establish a diagnosis. The differential diagnosis is wide-ranging, and expeditiously distinguishing the benign from malignant masses is difficult for instituting a timely multidisciplinary approach to the management of malignancy. Malignancy of the head and neck account for approximately 5% of all childhood carcinomas. This review aims to discuss the most common head and neck malignancies in childhood. The evidence for the diagnosis and management of childhood, head and neck malignancy is summarized. The rarity of these tumors are inevitably results in a paucity of high-level evidence to guide the treatment.

Keywords: Children; Head and neck malignancy; Lymphoma; Malignancy; Rhabdomyosarcoma

1 INTRODUCTION

Tumors of the head and neck in children comprise of heterogenous group of conditions. Some of which are true neoplasms (both malignant and benign) and others are hamartomas or congenital malformations.[(1)] It is often surprising to realize that many tumors which is arising in the head and neck region in childhood are malignant. Because of the complex anatomy and development of the head and neck, neoplasms during infancy and childhood arising at this site represent the most difficult challenges in clinical practice.[(2)] In majority of cases, tumors of head and neck in children are first seen by general practitioners or pediatricians with subsequent delays in investigation and diagnosis.

Studies of the clinical data on 778 children with malignant neoplastic disease showed that in 210 children the cancer involved primarily the anatomic structures of the head and neck.[(3)] The most frequent diagnoses are rhabdomyosarcoma, Hodgkin lymphoma, non-Hodgkin lymphoma, retinoblastoma, carcinoma of the thyroid, squamous cell carcinoma, nasopharyngeal carcinoma, neuroblastoma, salivary gland malignancy. Special age predilections were demonstrated by retinoblastoma for the infancy group & by lymphomas & thyroid cancer for the older children. With the notable exceptions of carcinoma of thyroid, children with head and neck cancer were more commonly affected are boys.

Cancer represents a significant cause of mortality among the pediatric patients. The frequency with which children between the ages of 5and 14 years die from cancer is second only to death from accidental trauma.[(4)] Despite improvements in managing many pediatric malignancies, the incidence of childhood cancer continues to rise.[(5)] Over

the past two decades, annual increases of 1% to 2% in the rate of pediatric malignancies have been reported in the United States. Similar increases have been reported in the Australia, Spain and Britain

2 MALIGNANCIES OF HEAD AND NECK

2.1 Rhabdomyosarcoma

It is the most common soft tissue sarcoma found in children.[2] it is a tumor on which malignant cells look like young, immature muscle cells. Almost three third of pediatric rhabdomyosarcoma cases are diagnosed in children under 6. It is a malignant tumor composed of neoplastic mesenchymal cells with varying degrees of striated muscle cells differentiation.[1] Common site is head & neck including the orbit, Para meningeal areas & non parameningeal areas [paramengieal areas-nasopharynx, nasal cavity, PNS, middle ear, pterygopalatine fossa; non Parameningeal –parotid, oropharynx, larynx]. Incidences decline with age the alveolar form peaks in childhood & adolescence. Usually treatment is chemotherapy & radiotherapy.[6] Clinically malignant condition followed by progressive course in about one third of the children and lead to death within three months.in another third the disease was slower in it but death occur in 12 -18 months.

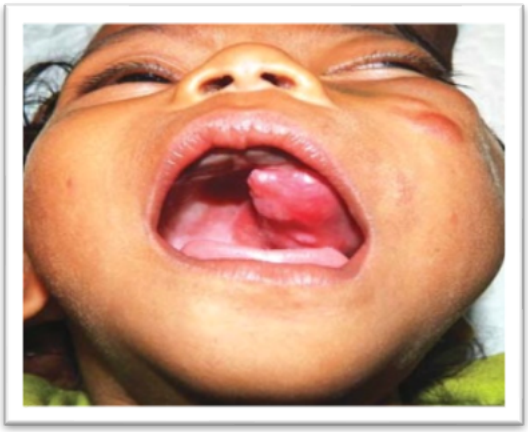

Fig. 1: Rhabdomyosarcoma

2.2 Thyroid cancer

It is the most common endocrine malignancy in children.[7] it occurs in patients under 20 years of age. Thyroid cancer arising in childhood rarely shows an undifferentiated histological pattern and, despite clinical features typical of a more aggressive behavior and tendency to metastasize.[8] Single nodule more likely to be malignant than multiple nodules. Ionizing radiations appear to be an important causal factor.[9] There was a steep rise in the incidence of papillary thyroid carcinomas in the young population following the 1986 Chernobyl nuclear incident.it is treated with radio iodine therapy, total thyroidectomy surgeries.

2.3 Retinoblastoma

It is the most common intraocular malignancy in children. Primary malignant neoplasm of retina arises from immature retina cells. Retinoblastoma affects both boys' and girls' equal frequency & has no known racial predilection.[10] Highest in first few months of life yearly incidence decrease steadily. It typically presents with leukocoria or strabismus. In later stages of retinoblastoma, the child may have proptosis, buphthalmos or hypopyon.[11] Children with intraocular retinoblastoma have an excellent overall & ocular survival rate. Retinoblastoma is treated by enucleation, radiation therapy, cryotherapy, laser therapy & chemotherapy.

Fig. 2: Retinoblastoma

2.4 Hodgkin Lymphoma (HL)

[12] It is one of the most curable forms of childhood cancer, with estimated 5-year survival rate.HL is inflammatory milieu with rare multinucleated giant cells (Reed-Sternberg cells) or large mononuclear cell variants (Hodgkin's or lacunar cells) R-S cell appears to arise from pre apoptotic germinal center B cells (no Ig production), although rarely may arise from T cells.[13] Signs and symptoms of Hodgkin's lymphoma may include Painless swelling of lymph nodes in, neck, armpits or groin, Persistent fatigue, Fever, Night sweats, Unexplained weight loss, Severe itching. Chemotherapy and especially radiotherapy will help to improve the therapeutic index.

2.5 Non-Hodgkin Lymphoma

[14] Non-Hodgkin lymphomas (NHLs) of childhood include high-grade mature B cell lymphoma [Burkitt lymphoma (BL), diffuse large B cell lymphoma (DLBCL), and primary mediastinal large B cell lymphoma (PMLBCL)], anaplastic

large cell lymphoma (ALCL), and lymphoblastic lymphoma (LL).symptoms of NHL are Enlarged lymph nodes (seen or felt as lumps under the skin). Abdominal (belly) swelling or pain, Shortness of breath, wheezing, or cough. Fever, Weight loss, Night sweats, Fatigue. Treatment will depend on the type and stage.[15] NHL can be treated with Chemotherapy, Radiation therapy, Surgery, High-dose chemotherapy with a stem cell transplant

2.6 *Squamous Cell Carcinoma*

Squamous cell carcinoma of the oral cavity occurs very rarely in children and is particularly rare during the first decade of life.[16] Most reported cases of these lesions arising in children occur in the respiratory tract, including the larynx and trachea; as well as the tongue, lips, and skin and present with variable symptoms based on tumor site. Most frequently, it is associated with previous malignancies, immunosuppressant therapy, and/or genetic conditions. Chemotherapy, radiation, surgery are the treatments.

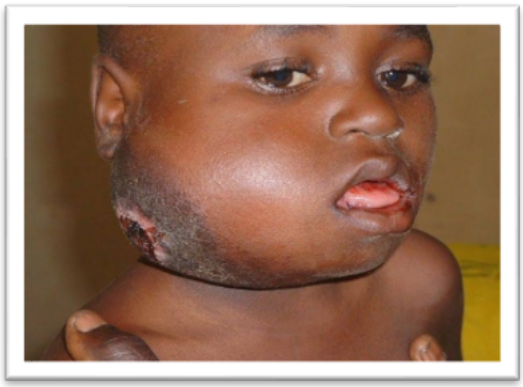

Fig. 3: Squamous cell carcinoma

2.7 *Neuroblastoma*

Neuroblastoma is the most commonly occurring extracranial solid tumor in childhood.[17] Neuroblastoma is a rare cancer which develops in nerve tissue. The tumor usually begins in the adrenal gland tissues found in the abdomen but may also begin in nerve tissue in the chest, neck, or spinal cord.[18] It usually affects kids under age 5. Symptoms include fatigue, decreased appetite and a lump in the chest, neck or belly Children with a family history of neuroblastoma may be more likely to develop the disease.[19] Yet, familial neuroblastoma is thought to comprise a very a smaller number of neuroblastoma cases. In most of the cases of neuroblastoma, cause is never identified. It is treated by Surgery (for tumor and/or metastatic resection, and removal of lymph nodes involved), Chemotherapy, Radiation, Bone

2.8 *Nasopharyngeal Carcinoma*

Nasopharyngeal carcinoma is a rare type of cancer that occurs in the nasal cavity the pharynx, and the upper part of the throat behind the nose. This cancer is called a carcinoma because it begins in the cells that line the nose and throat.[20] Childhood nasopharyngeal carcinoma is more common in adolescents than in children. Because this cancer is rare and has no distinguishing symptoms some symptoms include Nose bleeds, Nasal congestion, discharge and blockage, Frequent ear infections, enlarged or swollen lymph nodes in the neck and throat, Sore throat.[21] it may take time to detect and diagnose a tumor. The most effective treatment is a combination of chemotherapy and radiation therapy.[21] Depending on the stage of cancer. If left untreated, nasopharyngeal cancer can spread (metastasize) to other parts of the body and which lead to death.

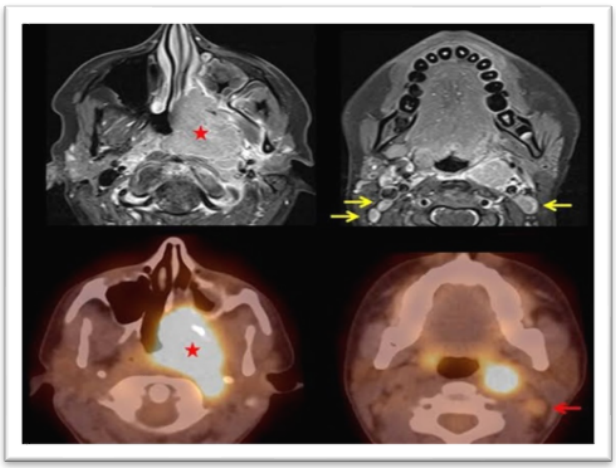

Fig. 4: Nasopharyngeal carcinomas

2.9 *Salivary gland Malignancy*

[22] Not more than 5%of all salivary gland neoplasm occur in children. lesions of the major salivary gland are uncommon in infancy and childhood.[23] In children, tumors of salivary gland origin are relatively more common after the tenth year of life.[24] Certainly below 7 yr. of age they are rare.[25] In children hemangioma is most common salivary gland tumor. The commonest lesion was the pleomorphic adenoma involving the parotid gland. Most congenital salivary gland tumors are carcinomatous in nature. The majority are highly malignant invasive tumors with anaplastic histologic features,[26] that a mass in the salivary gland of a child is malignant.[27] Certain features may increase the clinician's suspicion that a mass in the salivary gland of a child is malignant Features such as pain, rapid growth, facial nerve paresis, or lymphadenopathy.[2] The mainstay of treatment for salivary carcinomas is surgical resection with radiotherapy The extent of surgery is

dependent on the site and size of the tumor.

3 CONCLUSION

The incidence of head and neck malignancies among children younger than 15 years in the United States from 1973 through 1996 increased at a greater rate than childhood cancer in general. Incidence of head and neck tumors in pediatric patient is very rare. It is only 0.25% of pediatric patients attending the ENT department in 3 calendar years. Among the malignant lesions, lymphomas are most frequent carcinoma. Incidence of non-Hodgkin's lymphomas is more than Hodgkin's lymphoma. The Commonest age of the malignant diseases is above 10 years. There is overall male child predominance with a male: female ratio of 1.78:1. Awareness of a potential malignancy & careful follow-up of children with suspicious head and neck cancers is important so that more and more head and neck cancers in children are brought to treatment before it is too late.

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