Salivary Gland Malignancies in Children

An Essay

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in General Surgery

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Abstract

Introduction: Salivary gland disease is managed by a number of specialties. There are three pairs of major salivary glands and several hundred minor salivary glands within the upper aerodigestive tract. Pathology is diverse, including infective, inflammatory and neoplastic diseases. Clinical presentation is usually with a lump within or swelling of the gland. Investigations combine clinical assessment, fine needle aspiration cytology and radiology.

Aim of the Work: To highlight the incidence, pathology, clinical presentation, investigations, and treatment of the malignant tumors of salivary glands in children.

Methodology: Epithelial salivary gland neoplasms are infrequent in both adult and children. The incidence of these tumors increase with age; most pediatric cases are more than 10 years of age and the presence of these tumors in newborn is exceedingly rare. Published data suggest a male predominance. The major salivary glands, the parotid and the submandibular, are the main site of occurrence. Malignant cases account for 49% of the published pediatric cases as compared to 37% reported in adult literature. In children it was found that malignant tumors affected the parotid gland in about 50% of published pediatric cases, while in the submandibular gland benign neoplasms were twice as frequent as malignant tumors. In adults, malignant tumors compromised 25-32% of tumors in the parotid gland, and 40-50% of the submandibular gland neoplasms.

Conclusion: The role of radiotherapy in salivary gland tumors remains controversial. Usually the presence of high-grade malignancies, a large and aggressive tumor, multiple level involvements of cervical lymph nodes and incomplete surgery are considered the main indication of irradiation. Overall, salivary gland malignant epithelial tumors in children usually have a good prognosis. Five-years overall survival ranges from 81% to 90%.

Keywords: Salivary Gland Malignancies in Children

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Aliaa Elhusseiny Naeim

Contents

List of Abbreviations

ACC : Acinic cell carcinoma

ADC : Apparent diffusion coefficient

ADCC : Adenoid cystic carcinoma

CCL : Chronic Lymphocytic Leukemia

CT : Computed Tomography

DCE-MRI : Dynamic Contrast-Enhanced Magnetic Resonance Imaging

DW-MRI : Diffusion-Weighted Magnetic Resonance Imaging

EPV : Epstein - Barr virus

FNAC : Fine –needle Aspiration Cytology

FNM : Facial nerve monitoring

HIV : Human Immunodeficiency Virus

HPV : Human Papilloma Virus

IgA : Immunoglobulin A

MALT : Mucosa-associated Lymphoid Tissue

MEC : Mucoepidermoid Carcinoma

MRI : Magnetic Resonance Imaging

PA : Pleomorphic Adenoma

PAS : Periodic Acid-Schiff reaction

PET : Positron Emission Tomography

PPS : Prestyloid Parapharyngeal space

SCC : Squamous Cell Carcinoma

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Introduction

Salivary gland disease is managed by a number of specialties. There are three pairs of major salivary glands and several hundred minor salivary glands within the upper aerodigestive tract **(Bradley& O'Hara, 2012)**.

Pathology is diverse, including infective, inflammatory and neoplastic diseases. Clinical presentation is usually with a lump within or swelling of the gland. Investigations combine clinical assessment, fine needle aspiration cytology and radiology **(Bradley& O'Hara, 2012)**.

With an annual incidence of less than 1 per million, salivary gland malignancies in children are rare, constituting less than 10% of pediatrichead and neck cancer **(Yoshida et al., 2013)**.

According to the surveillance, epidemiology, and end results public-access database, the annual incidence of salivary gland tumor between 1973 and 2006 was 0.8 and 5 per million in children/adolescents younger than 20 years old and adults, corresponding to 0.5% of all pediatric tumors and 0.3in adults, respectively. The clinical characteristics in children differ somewhat from those seen in their adult counterparts. In particular, a firm mass in the territory of a salivary gland is 2.5-fold more likely to be malignant that in adults (i.e. corresponds to malignancy in 50% of SGT in children VS 20% in adults) **(Thariat et al., 2013)**.

Although over 20 histological types of salivary gland cancer have been reported in adults, a smaller number have been observed in the pediatric population. Mucoepidermoid carcinoma is the most common histological type followed by acinic cell carcinoma. Since the majority of salivary gland carcinomas are diagnosed at an early stage, the overall prognosis is often favorable with complete surgical resection **(Yoshida et al., 2013)**.

Pleomorphic adenomas are the most common benign neoplasms in salivary glands. Salivary gland Pleomorphic adenomas in children and adolescents have different characteristics compared with their adult counterparts in regard to histologic subtype and location. Surgical removal is the best treatment option for Pleomorphic adenomas in children and adolescents **(Fu et al., 2011)**.

Most children initially present between the ages of 10 and 16 years with palpable swelling in the salivary gland region. A slow growing, asymptomatic mass is often the only presenting sign with an average time to presentation of about 12–24 months. Approximately half of patients will complain of recent onset of pain in the absence of infectious or inflammatory symptoms. Rarely, patients may present with cranial nerve palsy or tethering of the skin **(Yoshida et al., 2013)**.

Ultrasound examination is the imaging procedure with the best predictive diagnostic capability for the salivary

glands. Due to the salivary glands, relatively superficial anatomical location, clear boundary from surrounding tissue and comparatively typical echogenicity, therefore sonography is ideal for diagnosis. Sonography allows detection of obstructive salivary gland diseases such as stenosis or sialolithiasis, as well as sialadenosis such as Sjogren's syndrome. Ultrasound examination alone is sufficient to diagnose benign tumors. However, in the case of malignant tumors, computer tomography or MRI may be also required, especially to determine the question of infiltration of the skull base **(Zengel et al., 2013)**.

Preoperative FNAC is a useful and accurate adjunct for preoperative evaluation of pediatric parotid tumors. We recommend that preoperative FNAC should be part of the initial evaluation of pediatric patients with parotid masses **(Lee et al., 2013)**.

Management is medical or surgical dependent on pathology. Surgical intervention is commonly performed for chronic inflammatory disorders and neoplasms. Management requires a sound knowledge of anatomy and oncologic principles **(Bradley& O'Hara, 2012)**.

Although pediatric parotid masses are unusual, they can represent a variety of pathological diagnoses, including malignancy. The intralesional injection can treat parotid hemangiomas in pediatric population effectively. Parotidectomy remains the mainstay treatment for both pediatric parotid gland benign and malignancies of epithelial cell origin. Adjuvant radiotherapy should be used judiciously in pediatric patients due to the higher risk of post-irradiation complications **(Liu et al., 2012)**.

To date, no prospective or retrospective data comparing outcomes of surgery alone versus multimodality therapy in the management of salivary gland malignancies in the pediatric population exists. Consequently, management decisions are made on a case-by-case basis, taking prognosis, treatment-related morbidity, and long-term sequelae into account **(Yoshida et al., 2013)**.

Aim of the Work

To highlight the incidence, pathology, clinical presentation, investigations, and treatment of the malignant tumors of salivary glands in children.