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RESEARCH ARTICLE

MALIGNANT EYELID AND CONJUNCTIVAL TUMORS IN CHILDREN: EPIDEMIOLOGY AND REVIEW OF LITERATURE

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ABSTRACT

Epidemiological study of the malignant eyelid and conjunctival tumors in children is very important in identifying the relationship between various external and internal factors and incidence in order to determine methods of prevention and effective treatment. Advanced research in epidemiological aspects of these tumors can establish a theoretical and practical knowledge base to ensure steady progress in the treatment of the tumors, which pose a serious health risk for the patients, especially children. The basis for each epidemiological study is research into the characteristics of incidence, mortality and survival rates

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INTRODUCTION

Approximately 5-10% of all skin cancers occur in the eyelids followed by in conjunctiva. Incidence studies indicate that basal cell carcinoma is the most frequent malignant eyelid tumor, followed by squamous cell carcinoma, sebaceous gland carcinoma and malignant melanoma. Malignant neoplasms represent the leading cause of plastic reconstruction in orbital region, followed by cicatricial retraction, post-traumatic loss of tissue, congenital colobomas. The American Cancer Society's estimates for eye cancer in the United States for 2015 are: (1)2,580 new cancers (mainly melanomas) of the eye and orbit: 1,360 in men and 1,220 in women (2)270 deaths from cancers of the eye and orbit: 140 in men and 130 in women

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Malignant Eyelid and Coniunctival Tumors

Malignant eyelid and conjunctival tumors are very rare in children. For example, of the 302 cases of epibulbar tumors in children only three cases (1%) were squamous cell carcinoma (Cunha *et al*., 1987) Basal cell carcinoma is extremely rare in children and usually associated with a genetic defect, such as basal cell nevus syndrome, xeroderma pigmentosum, or nevus sebaceous, or following radiotherapy treatment (Le Sueur *et al*., 2000; Al-Buloushi *et al*., 2005). It is probably the result of a combination of UV radiation exposure and genetic background. However, there is report about three children of 8, 11 and 16 years old, from which were 2 boys and 1 girl with primary isolated basal cell carcinoma unassociated with any other disease or syndrome. (Le Sueur et al., 2000) Other three cases, reported of de novo is basal cell carcinoma of the eyelid in children, had no known genetic syndromes and had not undergone radiotherapy (Al-Buloushi *et al*., 2005) Malignant

melanomas of the conjunctiva are extremely rare in children (Croxatto *et al*., 1987). They are highly malignant tumor that derives from melanocytes (Polat *et al*., 2006). There are reports about 6 and 11-year-old children with malignant conjunctival melanoma (Croxatto *et al*., 1987; Polat *et al*., 2006). Strempel and Krollé (1999) described three cases of 3, 4 and 14-year-old children, who suffered from this tumors. Of all 5 children, 2 were girls and 3 were boys. Another rarely reported malignant tumor of conjunctiva is rhabdomyosarcoma, which was described in a 10-year-old girl, who had no history of trauma to the eye or other ocular disease (Brichard *et al*., 2003). Malignant eyelid and malignant conjunctival tumors are such a rare malignancies that of 800 children treated in the Vladimir Filatov Institute of Eye Diseases (Odessa) only 2 children (0.3%) were with malignant eyelid and conjunctival tumors (Barchash *et al*., 1971) Ocular adnexal lymphoma is also rare in children. Only (1.4%) were under 21 years of age and a few cases of pediatric ocular adnexal lymphoma have been described in the literature. According to author's opinion, follicular lymphoma has not been described previously in the ocular adnexa in children (Perry *et al*., 2007).

Xeroderma pigmentosum is one of the other rare abnormalities, which is autosomal recessive genetic disorder starting in early childhood and characterized clinically by cutaneous photosensitivity, pigmentary changes, photophobia, and propensity for early development of malignancy in sun-exposed mucocutaneous and ocular structures (Kraemer *et al*., 1987; Robbins *et al*., 1974 and Paches *et al*., 1980). In 1968, Cleaver (Cleaver *et al*., 1968) was the first to report that skin cells from patients with xeroderma pigmentosum have an impaired ability to repair ultraviolet radiation-induced DNA damage. Xeroderma pigmentosum has a prevalence rate of 1:250000. (Kraemer et al., 1987; Robbins et al., 1974) 45.0 percent of the patients with xeroderma pigmentosum had basal cell carcinoma or squamous cell carcinoma of the skin, 5.0 percent of the patients had melanoma, which occurred in 79.0 percent and 65.0 percent respectively on the face, head or neck. Neurologic abnormalities were found in 18.0 percent of the case reported (Kraemer *et al*., 1987) Xeroderma pigmentosum occurred in 0.4 percent (3 of 758 patients with epithelial eyelid and conjunctival tumors) and 2 of 3 patients were sisters (Paches *et al*., 1980) Ocular abnormalities were reported in 40.0 percent of the patients and included ectropion, corneal opacity leading to blindness, and neoplasmsm (Kraemer *et al*., 1987) Blindness was noted in 26.0 percent of patients with malignant tumors (Touzri *et al*., 2008) Malignant tumors included squamous cell carcinoma, basal cell carcinoma, and malignant melanoma was seen in 30.0 percent (3 of 19 patients) (Goyal *et al*., 1994) or in 31.3 percent (10 of 32 patients). (Touzri *et al*., 2008) As a rule, the first symptoms of xeroderma pigmentosum noted by parents of children with a median age between 1 and 2 years (Kraemer *et al*., 1987; Robbins *et al*., 1974) Patients died at second or third decade of life from cachexia, haemorrhoidal meningitis or from dissemination of malignant tumors to the brain. 70.0 percent probability of survival was attained at age 40 years, a 28 years reduction in comparison with the US general population.

In Uzbekistan, malignant eyelid and conjunctival tumors in children are also rare malignancies. Information about patients with malignant ocular tumors in Uzbekistan has been collected only since 1978. Before 1978, there was no systematically collected information about the number of patients with malignant ocular tumors, therefore, epidemiologic aspects of these malignancies was investigated at first time in Republic. In joint collaboration with the IOR's Resource Department and Regional Oncology Dispensaries, we gathered malignant ocular tumors related information from 1978 to 1999. The information includes children's name, age, ethnic background, address. parents' occupation, diagnosis, method of treatment, results of treatment, date and cause of death. Only new cases with biopsy and histopathologically confirmation of the malignant tumors were registered. Histopathologically confirmation has been done in the Department of Histopathology of Institute of Oncology and Radiology (IOR) of Uzbekistan's Academy of Science.

For statistical purposes, the patients were divided into 0 to 4, 5 to 9 and 10 t0 14 age subgroups. They have calculated crude incidence, age adjusted incidence and standardized incidence per 100000 children male and female populations. For standardized incidence, Standard World Population was used. Population data were obtained from Uzbekistan's Statistic Committee. Incidence in different ethnic groups was compared to the number of the same ethnic groups in Uzbekistan. All rates were statistically tested and considered statistically significant if intersecting the 95 percent confidence interval (CI 95%, P< = 0.05). To analyze malignant ocular tumor incidence, the International Classification IX, 1984, was used: eyelid skin (172.1, 173.1), conjunctiva (190.3), retina (190.5), uveal tract (190.6), orbit except for bones (190.1), and orbital bones (170.0). For statistically purposes, all malignant ocular tumors were divided into four subgroups: eyelid, conjunctiva, intraocular and orbit. To calculate mortality rates, only those cases were considered whether death has been caused by a malignant ocular tumor, i.e. tumor-related death.

Table 1. Number and standardized incidence of children with malignant eyelid and malignant conjunctiva tumors per 100,000 male and female children population in urban and rural areas

Number of pa tents and standardized incidence	Eyelids		Conjunctiva	
Number of patients Standardized incidence	Urban	Rural	Urban	Rural

Malignant eyelid tumors occurred more frequently than conjunctiva tumors. So, there were 21 children with malignant eyelid tumors including 3 children with xeroderma pigment sum (14 males and 7 females) and only 11 patients with malignant conjunctival tumors including 2 patients with xeroderma pigment sum (7 males and 4 females). *Incidence*: In three age subgroups (0-4, 5-9 and 10-14) the highest age-adjusted incidence of malignant eyelid tumors was in males but only in age subgroup to 4 this incidence (0.5 per 100,000 children population) in males was statistically significant. In children with malignant conjunctiva tumors age-adjusted incidence was also highest in males but in age subgroup 10 to 14 years (0.4 per 100,000 children population). Comparing age-adjusted incidence of children with malignant eyelid tumors in urban and rural areas (Table 1) we found that the highest age-adjusted incidence was in urban children both

sexes (0.4 per 100,000 urban and rural children population). Because of small number of patients we calculated annual age-adjusted incidence tor both sexes of children with malignant eyelid tumors together with malignant conjunctival tumors, which ranged from 0.0 to 0.002 per million children population per year. *Mortality*: Overall mortality of children from eyelid tumors in urban areas for males and both sexes is 0.01 per 100,000 urban males and females children population and 0.0004 for rural males, females and both sexes per 100,000 rural males, females and both sexes children population, i.e. mortality of urban children from malignant eyelid tumors 2.5 fold higher than mortality from this tumors of rural children. Overall mortality of urban children from malignant conjunctival tumors for both sexes (0.004) is 2.0 fold higher than that for rural children (0.002) per 100,000 urban and rural both sexes children population.

Percentage of children died from malignant eyelid tumors less than 1 year, at 1 year, at 2 years, at 3 years and at 4 years was 0.0 percent, at 5 to 9 years was 9.5 percent (2 died patients) and at 10 years from initial diagnosis was also 9.5 percent (2 died patients) including 3 died patients with Xeroderma pigmentosum. Thus, during 10 years there were 19.1 percent died children (4 died of 21 patients with malignant eyelid tumors). Percentage of children died from malignant conjunctival tumors less than 1 year, at 1 year, at 2 years, at 3 years, and at 4 years was 0.0 percent at 5 to 9 years the rate was 9.1 percent (1 died patient) and at 10 years from initial diagnosis that was also 9.1 percent (1 died patient). Thus, during 10 years there were 18.2 percent died children (2 died from 11 children with malignant conjunctival tumors). Survival of patients with malignant eyelid tumors less than 1 year, at 1 year, at 2 years, at 3 years and at 4 years was 100.0 percent, at Sto 9 years the rate was 90.5 percent (19 survived patients) and at 10 years from initial diagnosis that was 81.0 percent (17 survived of 21 patients with malignant eyelid tumors). Survival of patients with malignant conjunctival tumors less than 1 year, at 1 year, at 2 years, at 3 years, and at 4 years was 100.0 percent, at 5 to 9 years the rate was 90.9 percent (10 survived patients) and at 10 years from initial diagnosed that was 81.8 percent (9 survived of 11 patients with malignant conjunctival tumors). All of our data concerning age-adjusted incidence, annual age-adjusted incidence, survival and mortality from malignant eyelid and conjunctival tumors in children require more epidemiologic investigations. Our work could be only a basis to compare of our and other future epidemiologic investigations in Uzbekistan.

Conclusion

Basal cell carcinomas are the most frequently encountered kind of malignant tumor affecting the eyelid and conjunctiva, making up about 85% to 95% of all malignant eyelid tumors. Squamous cell carcinoma is the second most common kind of malignant tumor of eyelid and conjunctiva (occurring in approximately 5% of malignant eyelid tumors). Sebaceous cell carcinoma originates in glands of the eyelid in elderly individuals. It is relatively rare but still accounts for 1% to 5% of malignant tumors of eyelids and conjunctiva. These are highly malignant tumors that may recur, invade the eye socket, or spread to lymph nodes. Malignant melanoma makes up almost 1% of all malignant eyelid and conjunctiva tumors but accounts for many of the deaths from malignant eyelid and conjunctival tumors.

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