GLOBAL JOURNAL OF BIOLOGY, AGRICULTURE & HEALTH SCIENCES
(Published By: Global Institute for Research & Education)
www.gifre.org

RARE PEDIATRIC TUMORS IN WESTERN UTTAR PRADESH-A PROFILE WITH REVIEW OF LITERATURE

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Abstract

Objective: To find out the profile of rare childhood tumors in Western districts of Uttar Pradesh in India**.**

Material and Methods: The study included children upto 12 years of age with confirmed diagnosis of cancer by means of histological or cytological examinations, who presented in the out patients and in patients departments of pediatrics, surgery and orthopedic surgery with tumor or tumor associated sign and symptoms.

Result: Out of 252 confirmed cases of pediatric tumors,132 (52.4%) were benign and 120 cases (47.6%) malignant. Maximum numbers of cases were seen in 7-12 years of age,175 cases(69%) while 77 (31%) cases occurred in 0-6 years of age. 15 cases of very uncommon/rare tumors were found in the study, which included 3 cases each of adenocarcinoma rectum (mucin containing) and squamous cell carcinoma of the tongue,1 case each of dermato-fibrosarcoma protuberans, chondrosarcoma, nasopharyngeal carcinoma, basal cell carcinoma, pleomorphic adenoma and 4 cases of giant cell tumor of bone.

Conclusion: All the reported rare cases contribute to the medical literature with a need to remain vigilant about their diagnosis in pediatric patients. In children, these cases can be difficult to diagnose, as they may present differently from adults.

Key Words: *Pediatric age, Rare tumors, Histopathology.*

Introduction

Although child health continues to be the priority health issue, childhood cancer is not yet a major area of focus. Worldwide, the annual number of new cases of childhood cancer exceeds 200,000 and more than 80% of these are from the developing world (Arora et al, 2009). The incidence of childhood cancer and type vary greatly throughout the world. Few pediatric tumors are relatively uncommon in India as compared to the western world.

Primary gastrointestinal malignancies constitute only 1% of pediatric neoplasms and therefore, remain unsuspected in children, often presenting late with symptoms of intestinal obstruction. Less than 200 cases of carcinoma of large bowel have been reported in children (Usenius T et al,1965). Unlike adult colonic malignancies, the overall prognosis in pediatric age group is poor because of inadvertent delay in diagnosis, advanced stage of the disease at presentation and lack of histological differentiation.

Dermato-fibrosarcoma protuberans (DFSP) is a low to intermediate grade soft tissue sarcoma originating from the dermal layer of the skin. DFSP comprises roughly 0.01% of all malignant tumors and approximately 2 to 6 percent of all soft tissue sarcomas (C. Garcia et al, 1996). The incidence in children is even less frequent, although a proportion of those identified in adulthood may reflect a delay in diagnosis of childhood DFSP (J.M. Weinstein, 2003).Chondrosarcomas are uncommon malignant neoplasms of cartilage that can occur anywhere in the body but are most commonly found in the long bones and pelvis. Pediatric patients with chondrosarcoma are rare, with only a handful of case reports and small series (ranging from two to seven patients) in the literature (Medline 1966–1999).

Pleomorphic adenoma is a comparatively rare disorder, accounting for only 1% of all neoplasms of the head and neck region. Nasopharyngeal carcinoma constitutes only 1-5% of all cancers and its incidence in children is even rarer.

The peak incidence of basal cell carcinoma occurs in the seventh decade of life and is rare in children. Basal cell carcinoma in pediatric age group, is usually associated with a genetic defect, such as basal cell nevus syndrome, xeroderma pigmentosum, or nevus sebaceous (Leibowitz E et al, 1997).World literature includes only 14 case reports or remarks on squamous cell carcinoma of the tongue in children aged less than 15 years (Lindqvist C et al, 1982). Giant cell tumors of the bone are rare, locally aggressive lesions that primarily affect the epiphyses of long bones, typically present in the third to fourth decades of life and rarely occur under 20 years of age (James B et al, 2007).

Material and Methods

The study included children with cancer, aged 0 to 12 years attending the out patients and in patients departments of pediatrics, general surgery and orthopedic surgery at Jawaharlal Nehru Medical College Hospital, Aligarh, who presented with the tumor or with tumor associated sign and symptoms, diagnosed by means of cyto-histological examination.

The profile of childhood cancer was studied focusing on the prevalence of tumors according to nature of lesions (benign vs malignant), most common site of involvement of particular tumor, age, sex, and common or uncommon for that age. 3-5µm histopathological sections from paraffin embedded tissues were stained with Hematoxylin and Eosin stain. Special staining were done as per the requirement.

Results

Our study included 252 confirmed cases of pediatric tumors aged 0 to 12 years. Out of these cases, we found 15 very uncommon tumors which comprised of 3caseseach of adenocarcinoma rectum and squamous cell carcinoma of the tongue, 1 case each of dermato-fibrosarcoma protuberans, chondrosarcoma, nasopharyngeal carcinoma, basal cell carcinoma, pleomorphic adenoma and 4 cases of giant cell tumor of the bone.

Mucin secreting adenocarcinoma rectum was seen in 9,10 and 12 years old male patients who presented with bleeding per rectum and abdominal pain.Computed tomography revealed mass in lower third of rectum in the first and second case and stricture in the first part of rectum in the third case. Histopathology showed stratification of glandular lining epithelium with enlarged tumor cells having hyperchromatic nuclei, seen infiltrating into the stroma. Numerous signet ring cells and lakes of mucin were also seen in the background.(Figure 1)

Squamous cell carcinoma of the tongue was seen in three male patients, aged6, 8 and 11 years and histopathology revealed well, moderately differentiated and poorly differentiated squamous cell carcinoma respectively.

Dermato-fibrosarcoma protuberans was seen in a 10 years old female who presented with swelling in the right shoulder. She was clinically diagnosed as a case of vascular malformation but histopathologic examination revealed the diagnosis of dermato-fibrosarcoma protuberanswith monomorphic spindle cells arranged in a storiform pattern, and entrapped isolated subcutaneous fat.(Figure 2)

Chondrosarcoma(clear cell variant) was diagnosed in a female patient aged 12 years, who presented with swelling in lower end of femur. Histopathologic examination showed tumor cells with abundant clear or ground glass cytoplasm interspersed with small trabeculae of woven bones.

Nasopharyngeal carcinoma was seen in an 11 year male patient, who presented with complaints of nasal mass and cervical lymphadenopathy. Histopathology of the resected mass revealed nasopharyngeal carcinoma with undifferentiated tumor cells in nesting pattern, round to oval cells, vesicular nuclei and prominent nucleoli. (Figure 3)

Pleomorphic adenoma was seen in a 12 year female patient who presented with swelling in the pre-auricular region. Histopathology revealed biphasic appearance of tumor with an admixture of sheets and ducts like structures of epithelial cells in a myxomatous background and helped to confirm the diagnosis.

Basal cell carcinoma was seen in a 5 year male, who presented with a nodule under the left eye, which on histopathology showed nests of malignant tumor cells with peripheral palisading.

Giant Cell Tumor of the bone (Osteoclastoma) was reported in 4 cases with mean age of 9.7 years. 2 cases involved upper end of femur, 1 case each involved upper end of fibula and the talus that showed a lytic lesion on X-ray. On histopathology osteoclast like giant cells dispersed uniformly in a stroma of mononuclear oval to spindle cells and fibrous tissue were seen.

Discussion

A galaxy of benign and malignant tumors occur in infants and children and in a few cases may present diagnostic challenges. Pediatric tumors differ markedly from adult tumors in their nature, distribution and prognosis. Incidence of pediatric tumors is on the rise during the last few decades. The pattern of childhood tumors also shows wide variation amongst different sexes and age groups**.**

Colorectal cancer is rare in children with an incidence of 1.3 to 2 cases per million children as per National Cancer Institute Monograph Bethesda, USA. 1981. In India, colorectal carcinoma does not figure amongst the 10 most common malignancies (Rao DN, 1998).Most of these cases in children occur in the second decade of life, although we reported it in males aged 10 and 12 years. Rao et al,1998 noted a mucinous histology in most of the cases, which correlated with our study. The symptomatology of vague abdominal pain, constipation, nausea, vomiting and rectal bleeding etc., are usually not thought of as features of malignancy and patients are often treated for causes like amoebiasis and worm infestations, which are more prevalent in our country.(Dinesh K Sarda et al, 2004) Dinesh K Sarda et al, 2004 have asserted that patients with blood or mucoid discharge per rectum and chronic abdominal pain should undergo a careful abdominal and rectal examination, who reported a case of mucinous adenocarcinoma rectum in an 11 year old male, presenting with bleeding per rectum and pain abdomen.

Squamous cell carcinoma (SCC) although a rare neoplasm of pediatric age group, was seen in 6, 8 and 11 year old male patients. Histopathology revealed well, moderately differentiated and poorly differentiated squamous cell carcinoma respectively, similar to as reported by Usenius T et al, 1965 in patients aged 0 to 13 years. Although rare in pediatric population, squamous cell carcinomas do occur in this age group, so one should be very careful during reporting of ulcerative lesions in the pediatric patients.

Dermato-fibrosarcoma protuberans (DFSP) was reported in a 10 year old female with swelling in the right shoulder. A clinical diagnosis of vascular malformation was made but histopathological examination revealed DFSP, a finding very similar to the experience of Reddy C et al,2009 in 3 cases, aged 5, 10 and 11 years male of DFSP seen at The Children's Hospital at Westmead, Sydney.On clinical grounds, one of their cases was diagnosed as angiolipoma. Although relatively rare in the pediatric population, there is a need to remain vigilant about the diagnosis of DFSP in this age group, given its aggressive nature and high recurrence rates.

A 12 year female who presented with swelling in the lower end of femur was diagnosed as chondrosarcoma, which on histopathologic examination showed clear cell variant of the tumor. Nick Vertzyas et al, 2000 reported a case of chondrosarcoma, involving the head of the left 8th rib in an eight year female.

Nasopharyngeal carcinoma (NPC) has a bimodal age distribution. A small peak is observed in late childhood, and a second peak occurs in people aged 55-65 years (Ayan I et al,2003).We reported a case of nasopharyngeal carcinoma in an 11 years male, who presented with complaints of nasal mass and cervical lymphadenopathy. Our observation was similar to that of Y Noorizan et al 2008, who reported two cases of NPC in 11 year old males with cervical lymphadenopathy.

Salivary gland tumors are extremely rare in the pediatric population and pleomorphic adenoma accounts for fewer than 5% of all salivary gland tumors, occurring in patients below 16 years(Greer RO et al, 1983).We observed a case of pleomorphic adenoma in a female patient while Pratt, 1996 found equal incidence in males and females. Histopathology revealed a biphasic pattern with epithelial component of glandular nature and fibromyxoid tissue in our case, a finding consistent with those of David et al, 1980.There is a wide variation in the claimed incidence of malignancy in pediatric salivary gland tumors, which has prompted physicians who encounter these rare neoplasms to continue to critically review their true nature.

Basal cell carcinoma (BCC) was seen in a 5 year old male, who presented with a nodule under left eye, which on histopathology showed nests of malignant looking basal cells. Benjamin W et al, 2000 have reported a case of BCC in an 8 years old male child. As incidence rates of basal cell carcinomas continue to rise especially in areas of high-level UV radiation exposure, childhood cases may become more common.

Four cases of giant cell tumors (GCT) were seen by us with a mean age of 9.7 years. 2 cases involved the upper end of femur while 1 case each involved the upper end of fibula and talus that showed a lytic lesion on X-ray. Campanacci et al,1975 reported giant cell tumor as a rare tumor of childhood, commonly involving the lower end of radius, while James B. Elder et al,2007 reported two cases of GCT of the skull, in two pediatric patients aged 2 years male and 7 weeks female.

Conclusion

It is important to remember that although rare and apparently presenting in an inocuous manner in the pediatric population, there is a need to remain vigilant about the diagnosis in this age group, given its aggressive nature and high recurrence rates. Therefore high index of suspicion is required to detect early disease in children and prompt physicians who encounter these rare neoplasms to continue to critically review their true nature.

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Annexure

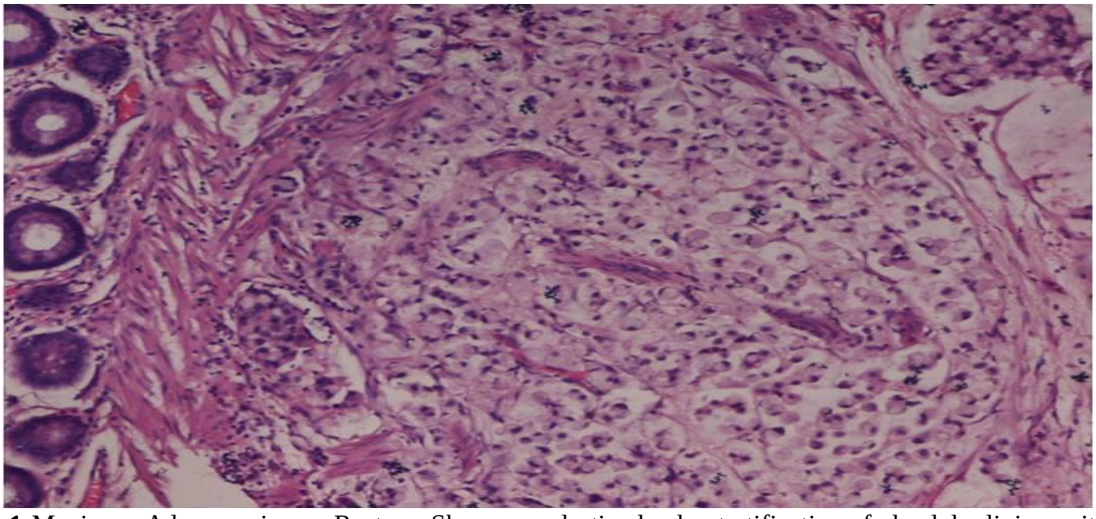

Figure 1: Mucinous Adenocarcinoma Rectum: Shows neoplastic glands, stratification of glandular lining epithelium with enlarged tumor cells having hyperchromatic nuclei, seen infiltrating into the stroma and signet ring cells (H& E, 10X).

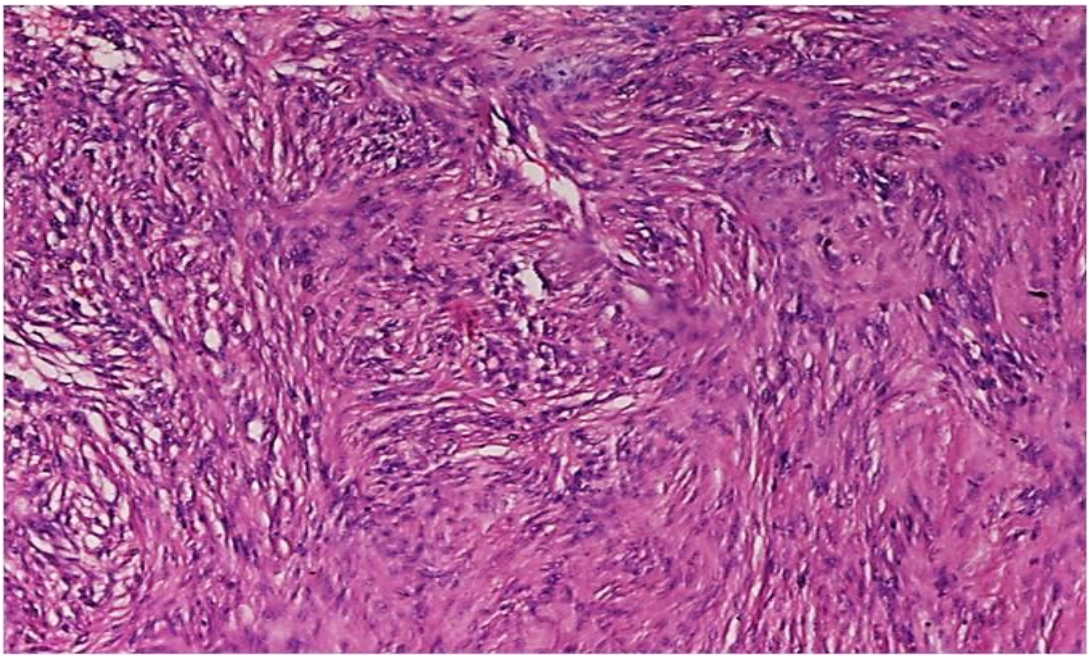

Figure 2: Dermatofibrosarcoma Protuberans: Shows monomorphic spindle cells with storiform pattern of growth (H& E, 10X).

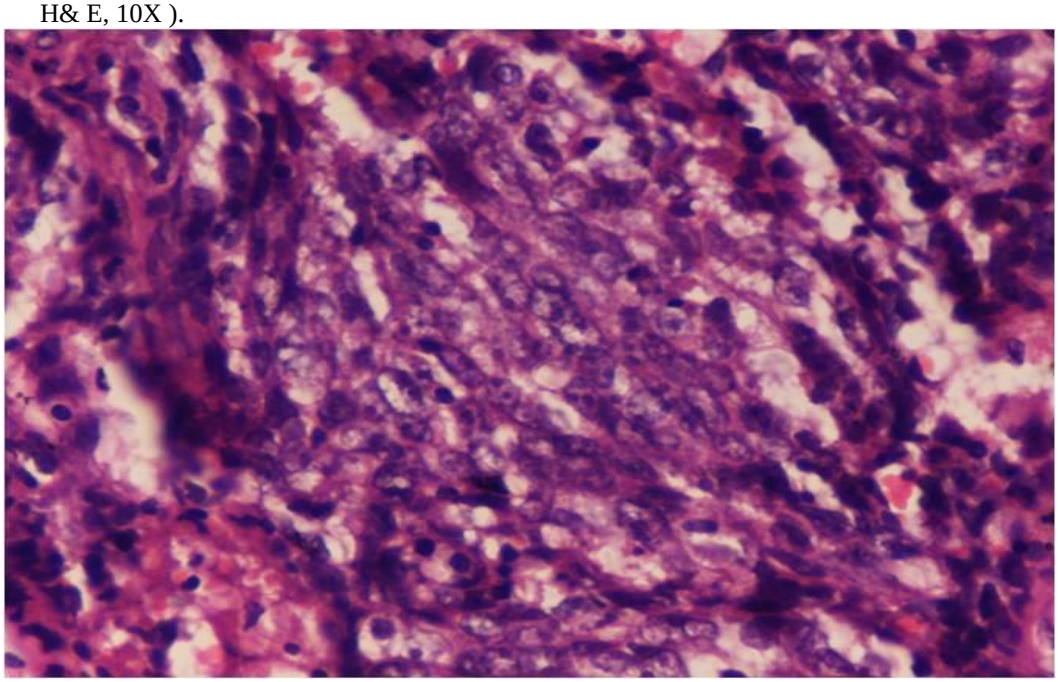

Figure 3: Nasopharyngeal Carcinoma: Undifferentiated tumor cells in nesting pattern, round to oval cells, vesicular nuclei and prominent nucleoli. (H& E, 40X).