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Original Article

Assessment of Large Orbital Tumor with Intracranial Extension in Paediatric Patients: An Institutional Based Study

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***ABSTRACT*:**
Background: Occurrence of Nonosseous, extraocular orbital tumors is quite uncommon. Knowledge of the clinical presentation and patient's age helps to limit the differential diagnosis and to determine the appropriate imaging modality. Hence; we planned the present study to assess pain management of large orbital tumor with Intracranial Extension in Paediatric Patients. **Materials & methods:** The present study included evaluation of pain management of large orbital tumor with Intracranial Extension in Paediatric Patients. A total of 50 cases of large orbital tumors with Intracranial Extension were included in the present study. Complete demographic and clinical details of all the patients were obtained. We also obtained findings of radiographic investigations of all the patients. Final diagnosis was confirmed based on clinical, histopathological and radiologic findings. All the results were analyzed by SPSS software. **Results:** Among 50 paediatric patients included in the present study, 28 subjects were males while remaining were females. Retinoblastoma was the most common intra-orbital tumours with Extracranial extension in the present study, found to be present in 36 percent of the patients. This was followed by Dermoid tumour, inflammatory pseudo-tumour found to be present in 16 percent and 10 percent of the patients. **Conclusion:** Intra-orbital tumours in paediatric patients consist of a wide range of morphologic and clinical diversities. Therefore, a paediatrician must be aware of different tumours responsible for causing intra-orbital swellings.

Key words: Intracranial tumour, Orbital, Paediatric

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INTRODUCTION

Among paediatric subjects, occurrence of Nonosseous, extraocular orbital tumors is quite uncommon. However; they represent a wide range of histologic spectrum in comparison to those seen in adult subjects.[1] Mesenchymal lesions comprises of a majority of these pathologies. Rhabdomyosarcoma is the most common mesenchymal tumor of childhood. It might arise in or invade the orbit in young paediatric subjects. Other group lesions invading the orbit in paediatric subjects are the vasculogenic lesions. Hemangiomas are true neoplasms that are found in infants and are distinguished by their high-flow feeding vessels.[2- 3] Knowledge of the clinical presentation and patient's age helps to limit the differential diagnosis and to determine the appropriate imaging modality. Certain orbital pathologies such as retinoblastoma and rhabdomyosarcoma (RMS) are typically found in children, whereas malignant uveal melanoma, lymphoma, and inflammatory orbital pseudotumour (IOP) are seen in adults. Clinical symptoms such as extraocular muscle palsies, diplopia, visual impairment, exophthalmos, and eye pain can serve as useful pointers to the likely pathology. However, a biopsy may be needed to provide tissue diagnosis.[4- 6] Hence; we planned the present study to assess pain management of large orbital tumor with Intracranial Extension in Paediatric Patients.

MATERIALS & METHODS

The present study was commenced in the department of Paediatrics of the medical institute and it included evaluation of pain management of large orbital tumor with Intracranial Extension in Paediatric Patients. We obtained informed consent

from all the subjects/guardians after explaining in detail after explaining in detail the entire the research protocol. A total of 50 cases of large orbital tumors with Intracranial Extension were included in the present study. Inclusion criteria for the present study included: Patients with less than 15 years of age, Patients with negative history of any other systemic illness, Complete demographic and clinical details of all the patients were obtained. We also obtained findings of radiographic investigations of all the patients. We did the gross examination of each available specimen followed by staining with H and E stain. Special staining was done in cases where ever required. Final diagnosis was confirmed based on clinical, histopathological and radiologic findings. All the results were analyzed by SPSS software.

RESULTS

In the present study, a total of 50 paediatric patients with different intra-orbital tumours with extra-cranial extension were included in the present study. Among these patients, 28 subjects were males while remaining were females. Retinoblastoma was the most common intra-orbital tumours with Extracranial extension in the present study, found to be present in 36 percent of the patients. This was followed by Dermoid tumour, inflammatory pseudo-tumour found to be present in 16 percent and 10 percent of the patients. Osteoma and Neurofibroma were found to be present in 4 patients each. Meningioma was found to be present in 4 patients among which 2 were males and 2 were females.

Table 1: Different types of intra-orbital tumours with Extracranial extension

Individual type of tumour	Number of cases	Percentage
Dermoid	8	16
Retinoblastoma	18	36
Inflammatory pseudo-tumour	5	10
Osteoma	4	8
Neurofibroma	4	8
Meningioma	4	8
Schwannoma	4	8
Others	3	6
Total	50	100

Graph 1: Different types of intra-orbital tumours with Extracranial extension

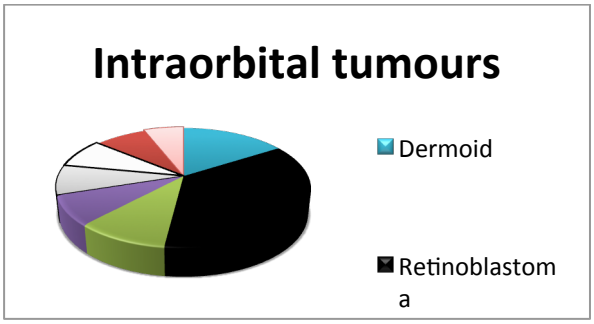
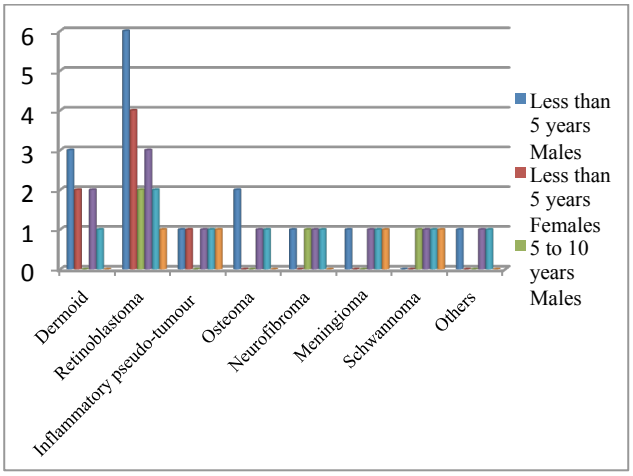


Table 2: Age wise and gender wise distribution of paediatric subjects with different types of intra-orbital tumours with Extracranial extension

Individual type of tumour	Less than 5 years		5 to 10 years		More than 10 years		Number of cases	
	Males	Females	Males	Females	Males	Females	Males	Females
Dermoid	3	2	0	2	1	0	4	4
Retinoblastoma	6	4	2	3	2	1	10	8
Inflammatory pseudo-tumour	1	1	0	1	1	1	2	3
Osteoma	2	0	0	1	1	0	3	1
Neurofibroma	1	0	1	1	1	0	3	1
Meningioma	1	0	0	1	1	1	2	2
Schwannoma	0	0	1	1	1	1	2	2
Others	1	0	0	1	1	0	2	1
Total	15	7	4	11	9	4	28	22

Graph 2: Age wise and gender wise distribution of paediatric subjects with different types of intra-orbital tumours with Extracranial extension



DISCUSSION

The wide varieties of rare intraocular and orbital neoplasms differ in presentation in the pediatric population when compared to these same lesions in adults. While most pediatric ophthalmic tumors are benign, they may have a significant impact on vision and may result in significant morbidity and mortality. Because of the superficial location of the globe and its cystic nature, ultrasound (US) with Doppler allows accurate depiction of a variety of pathologic conditions of the globe and orbit, and enables non-invasive and cost-effective follow-up. The technique is well

tolerated, easy to perform, and has a high accuracy for the characterization of vascular lesions in skilled hands.[7-9]

In the present study, a total of 50 paediatric patients with different intra-orbital tumours with extra-cranial extension were included in the present study. Among these patients, 28 subjects were males while remaining were females. Retinoblastoma was the most common intra-orbital tumours with Extracranial extension in the present study, found to be present in 36 percent of the patients. Viswanathan S et al analyzed 47 specimens comprising biopsies, excision specimens, and FNAC of extraconal pediatric orbital masses (excluding retinoblastoma) accessioned in the pathology department over 5 years in a tertiary referral cancer center. Immunohistochemistry (IHC-74%) and molecular methods (one case) were done where necessary. The chief presenting symptom was proptosis in 55.3% patients and radiologically 53.8% malignant tumors showed extraorbital extension. A diagnostic algorithm was formulated to assess which cases needed pathology evaluation. Malignant round cell tumors (76.6%), chiefly embryonal rhabdomyosarcoma (51%), benign spindle cell neoplasms, and infectious lesions (tuberculosis, fungal infections), were seen. Of the malignant tumors, those confined to the orbit achieved good treatment response and had an event-free follow-up while those with extraorbital spread had poor outcome. Pediatric orbital masses range from completely treatable infectious lesions, surgically resectable benign neoplasms to aggressive malignancies requiring chemotherapy and radiotherapy.[10] Markowski J et al, in another study, investigate the distribution of different types of primary orbital tumors, histopathological diagnosis, and postoperative complications. They analyzed 122 patients (68 women and 54 men) with orbital tumors, hospitalized in the ENT Department of the Medical University of Silesia in Katowice during 1990–2013. The patients were characterized in terms of anatomic, topographical, histopathological, and clinical parameters. The role of diagnostic imagining such as CT, NMR, and fine-needle aspiration (FNB) in preoperative diagnostics is discussed. Results of FNB, cytological, and histopathological examination of the postoperative specimens were compared. There were 56 (46%) patients with malignant tumors, 42 (34%) with benign tumors, 19 (16%) with inflammatory tumors, and 5 patients (4%) had other tumors. In cases of malignant tumors, local recurrence up to 5 years was found in 36 (64.3%) cases. In the other 20 (35.7%) cases of malignant tumors, the patients remained under close follow-up in the outpatient clinic, without signs of local recurrence (follow-up 1–17 years). According to histopathological examination, malignant tumors were detected in 45.9% of patients and non-malignant tumor in 34.4% of patients. In 19.7% of patients, inflammatory and other types of tumors were diagnosed. They characterized the occurrence and pathological profiles of orbital tumors.[11] Neudorfer M et al evaluated the role of ultrasonography (US) and color Doppler imaging (CDI) in the diagnosis of orbital tumors in children. This study included 42 children with intraorbital and periorbital tumors who were evaluated in our clinic. All children underwent a complete clinical evaluation as well as orbital US and CDI. The children then underwent operation or were followed up, based on the clinical diagnosis and the findings on the imaging modalities. The mean age of the patients at diagnosis was 22.5 months (range 2 weeks-14 years old). Eighteen patients (12 female and 6 male) were diagnosed with hemangioma based on the findings in US and CDI; 16 patients (9 female and 7 male) were diagnosed with dermoid, 5 patients (4 female and 1 male) with lymphangioma, 2 patients with rhabdomyosarcoma, and 1 patient with a subperiostal abscess. Twenty-two patients underwent operation, and the diagnosis was confirmed on pathology. Twenty patients did not undergo operation and continued to be followed up in the clinic (mean follow-up period 38.2 months). The disease course in all patients who did not undergo operation was consistent with the working diagnosis. Both US and CDI are useful modalities in the diagnosis of intraorbital and periorbital tumors in the pediatric age group.[12]

CONCLUSION

Under the light of above results, the authors concluded that intra-orbital tumours in paediatric patients consist of a wide range of morphologic and clinical diversities. Therefore, a paediatrician must be aware of different tumours responsible for causing intra-orbital swellings. However; future studies are recommended.

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