DIAGNOSTIC ROLE OF CT SCAN IN PROPTOSIS IN PAEDIATRIC AGE GROUP

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ABSTRACT

Objective: To analyse the diagnostic role of Ct scan in proptosis in paediatric age group.

Material and Methods: All Children with proptosis who were sent from eye unit for CT scan were included in the study. A proforma was where all the necessary information was entered. At the end of study these were analyzed.

Results: A total of 50 patients were evaluated. There were 34 males and 16 females. The mean age was 6.3 years. Besides proptosis (100%), visual deterioration (50%) was the most common symptom. Tumors were the most common (56%) cause of proptosis. Out of these retinoblastoma, optic nerve glioma were on the top of the list. This was followed by the inflammatory disease process (24%) in frequency.

Conclusion: CT was carried out in all the case and it gave excellent results in the evaluation of the disease process and further management of the patient.

Key words: CT Scan, Children, Proptosis,

INTRODUCTION

Proptosis is defined as an abnormal protrusion of the ocular globe. Owing to the rigid bony structure of the orbit, with only anterior opening for expansion, any increase in the orbital contents taking place from the side or from behind will displace the eyeball forward.[1] The management of orbital disease in pediatric patients differs greatly from management in adults, because many malignant lesions in this age group are treatable and prompt evaluation can lead to a tumor being treated at a smaller size than if delay occurs.[2] In the Western world help is sought very early in the course of the disease and the radiological findings in proper setting

can improve the outcome of these patients while in our set up misconception and poor socio-economic status leads to late diagnosis, difficulty in management and disastrous consequences like permanent proptosis.[3]

Hence an accurate clinical evaluation, carefully selected diagnostic imaging studies, laboratory evaluation and early initiation of treatment will lead to better prognosis for both vision and life.

As far as the radiological investigations are concerned, findings on plain x-ray and ultrasonography are not pathognomonic of most of the orbital disease process. Though some help can be obtained in characterization of the lesion in certain cases. For the evaluation of visual loss or suspected cranial nerve dystfunction, MRI is the procedure of choice due to lack of bony artifact and improved conspicuity of subtle lesions[4]. However, MRI is not cost effective and not widely available. Moreover, MRI would not be technically feasible in children due to longer duration of examination and motion artifacts. On the other hand, the easy availability and operability, good maintenance and speed makes CT scan as an affordable diagnostic tool in orbital diseases under existing circumstances and present setup. Moreover, the additional advantages of spiral CT have further cemented CT role as the screening examination of choice for the orbit[5].

MATERIAL AND METHODS

This study was conducted in the Radiology Department, Postgraduate Medical Institute, Hayatabad Medical Complex, in collaboration with the Department of Ophthalmology HMC from January 2000 – June 2002. A total of 130 patients with proptosis were analyzed out of which 50 cases were in pediatric group. Lesions were grossly classified as inflammatory, neoplastic, congenital and traumatic. CT scan was carried out in all the patients and help of other radiological investigations such as x-ray and ultrasound was taken when required. Clinical records were reviewed to help in determining the reliability of CT scan findings in patients management. Later CT scan findings were verified against operative findings, excisional/ incisional biopsy or clinical follow up. A special proforma was designed for data collection, history and record of subject patients.

Inclusion Criteria

Patients of both sexes with equal or less than 13 yrs of age presenting with proptosis and swelling in and around orbit.

Exclusion Criteria

Patients previously diagnosed / operated and now referred for re-assessment and follow up.

RESULTS

Age and Sex incidence

The age in our study ranged from 4 months old baby to 13 years of age with mean age of 6.3 years. 48% of patients were under 5 years of age. Out of 50 patients, 34 were males and 16 were females with a male to female ratio of 2.1:1.

CLINICAL FEATURES

	Clinical Feature	No. of cases	Percentage
1.	Proptosis	50	100%
	Unilateral	38	76%
	Bilateral	12	24%
	Painless	34	68%
	Painful	16	32%
2.	Visual Deterioration	25	50%
3.	Fever	9	18%
4.	Diplopia	12	24%

TABLE-1

Besides proptosis (100%) visual deterioration was present in 50%, fever in 18% and diplopia in 24% of cases.

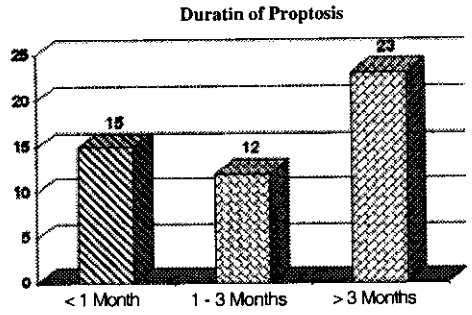

Fig. 1

15 cases had an acute onset i.e. duration of proptosis was < 1 month. 12 cases within 1 to 3 months and 23 cases presented with more than 3 months duration.

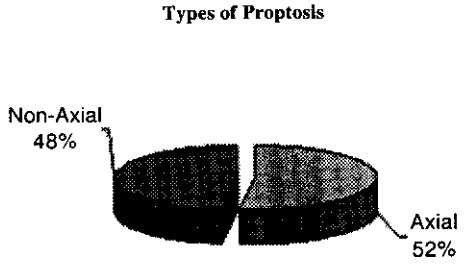

Fig. 2

CLASSIFICATION AND HISTOPATHOLOGY

(NO. OF CASES = 50)

S. No.	Orbital Lesion	No. of cases	%age of Total Orbital Lesions
1.	Neoplastic	28	56.0%
2.	Inflammatory	12	24.0%
3.	Congenital	7	14.0%
4.	Traumatic	3	6.0%
	Total	50	100%

TABLE–2

48% had non-axial proptosis and 52% had axial proptosis.

Among the 50 cases neoplastic lesions were 56%, inflammatory 24%, congenital 14% and traumatic 6%.

Retinoblastoma was the most frequent orbital tumor among the total orbital lesions i.e. 20% followed by optic nerve tumor 18%. Vascular tumors were 2% while muscular tumor and nervous tumors were 4% and 2% respectively. Hemopoitic reticuloendothelial system tumors formed 4% of total orbital lesions. Metastatic deposits were 6%.

NEOPLASIA

(NO. OF CASES = 28)

S. No.	Description	No. of Cases	%age of Total Orbital Lesions	%age Orbital Tumours
1.	Retinoblastoma	10	20.0%	35.7%
2.	Optic Nerve Tumor	09	18.0%	32.2%
3.	Vascular Tumor	01	2.0%	3.6%
4.	Muscular Tissue Tumor	02	4.0%	7.1%
5.	Nervous Tissue Tumor	01	2.0%	3.6%
6.	Hematopoietic RES Tumors	02	4.0%	7.1%
7.	Metastasis	03	6.0%	10.7%
	Total	**28**	**56%**	**100%**

TABLE–3

CONGENITAL LESIONS

(NO OF CASES = 7)

S. No.	Description	No. of Cases	%age of Total Orbital Lesions	%age of Congenital Lesions
1.	Encephalocele	3	6.0%	42.8%
2.	Crouzon's Syndrome	1	2.0%	14.3%
3.	Dermoid Cyst	2	4.0%	28.6%
4.	Neurofibromatosis	1	2.0%	14.3%
	Total	**7**	**14%**	**100%**

TABLE-4

INFLAMMATORY CAUSES

(NO. OF CASES = 12)

S. No.	Description	No. of Cases	%age of Total Orbital Lesions	%age of Inflammatory cases
1.	Infectious:			
	a. Sinus Related	7	14.0%	58.3%
	b. Local Infection	3	6.0%	25.0%
2.	Non-Infectious (pseudo tumor)	2	4.0%	16.7%
	Total	**12**	**24.0%**	**100%**

TABLE-5

POST-TRAUMATIC

(NO. OF CASES = 3)

S. No.	Description	No. of Cases	%age of Total Orbital Lesions	%age of Traumatic Causes
1.	Organized Hematoma	2	4.0%	66.7%
2.	Orbital Cellulitis	1	2.0%	33.3%
	Total	**3**	**6.0%**	**100%**

TABLE-6

Encephaloceles were 6% of total orbital lesions. Crouzon's 2%, dermoid cyst 4% and neurofibromatosis 2% of total lesions.

Inflammatory causes were divided into infectious and non-infectious groups. Among the infectious group 14% were secondary to sinusitis and 6% due to spread from local infection. Non-infectious group formed 4% of total cases.

Organized hematoma formed 4% of total lesion while orbital cellulites secondary to trauma formed 2% of the list.

DISCUSSION

50 patients up to the age of 13 years were investigated and they comprised 38% of total patients with proptosis. A study in Mayo Hospital showed this figure to be upto 40%[6].

Age at the onset of a condition is important in pediatric diagnosis because of the narrow age spectrum of some conditions, as well as the more limited number of lesions that may appear[2]. The age in our study ranged from 4 months to 13 years with mean age of 6.3 years. 48% of patients were under 5 years of age and male to female ratio was 2.1:1. A Moroccan study showed their mean age of involvement to be 4.2 years and male to female ration of 2:1[7]. As it was a hospital based study, there was a high chance that males get to reach for treatment more often in our setup while females are not that privileged.

In our study, majority of patients (70%) developed proptosis over a period of months. This is in agreement with Rootman[22] who reported chronic onset of disease in 60% of cases. Acute onset of proptosis was largely due to trauma and acute inflammatory lesions.

Besides proptosis, the 2nd most common presenting complaint was visual deterioration i.e. 50%. These figures are comparable to the study carried out by Cristante which showed proptosis and visual deterioration as primary symptoms[8]. Similar figures are also given by Asif et al[3].

The orbital tumors were the top most causes of proptosis in our study i.e. 56%. Khan AA et al showed this figure to be 54.5% in their study[6] which is quite comparable to our study.

Among the orbital tumors the retinoblastomas formed 20% of total orbital lesions. On comparison, this figure was 30% in a study by Khan AA et al while a Moroccan study on the epidemiological aspects of orbital diseases in childhood also showed retinoblastomas to be on the top of the list[7]. According to another study it is the most common ocular and orbital tumor in Pakistan[9]. Bilateral involvement of retinoblastoma was seen in 20% cases. Khan et al also showed bilateral involvement in 20% of cases[6]. Intracranial extension was seen in 60% of cases. Average age at presentation mentioned in literature is under 2 years but can occur in older children[10]. In our study, the age ranged from 2 to 6 years with mean age of 3.2 years. Retinoblastoma is the most common intraocular malignancy of childhood. It occurs in heredity and non-heredity form. The heredity form is usually bilateral[10].

Second in frequency in our study among orbital tumors were optic nerve tumors i.e. 18% of total orbital lesions 88.9% were gliomas and 11.1% Meningioma. Comparison with the study by Khan et al showed only gliomas[6] while other worker on orbital tumors sowed that in their series gliomas were 80% and Meningioma 20%[11]. The age at presentation mentioned in the literature is during first decade and loss of vision is the first symptom[10]. In our study, the mean age of the patients was 7 years.

In vascular tumor only one case of capillary hemangioma was detected making it 2.0% of total orbital lesion. In contrast to that Khan et al found it to be 5.4% of total orbital lesions[6]. Capillary hemangioma occurs primarily in infants during the 1st years of life[12]. In our study, the age of the patient was 10 months. These tumors are more compressible than those in adults, as connective tissue capsule does not develop till later in life[13].

Rhabdomyosarcoma is the most common primary orbital malignant in the pediatric age group with most presenting below 6 years of age. In our study, they formed 4.0% of total orbital lesions with mean age of 8 years at presentation. Intracranial spread was seen in both the cases at the time of presentation. Khan et al showed rhabdomyosarcoma to be 5.4% of all orbital lesions[6]. In contrast, this figure is slightly higher in 2 similar studies in Sydney and Morocco giving a value of 12.2 and 16% respectively[17,7].

There was a single case of isolated Schwanoma in 11 years old child. It was well-defined and encapsulated when seen pre-operatively. They are slow growing tumors and isolated variety is usually seen in adults[4]. According to a Chinese study isolated orbital Schwannoma most commonly occurs at the age of 20-40 years[14].

There were 2 cases of bilateral orbital leukemic infiltration making it 4% of total orbital lesion. Khan et al showed leukemic infiltrates to be 5.8% of all orbital lesions[6].

3 cases of retrobulbar metastatic deposits were identified i.e. 6% of total orbital lesions. All of them metastized from neuroblastomas and were bilateral in nature. There were also underlying bony changes and intracranial extension as evident on CT scan. Sindhu et al showed this figure to be 7.0% in their study[5].

After orbital tumors the 2nd most common orbital pathology in children was inflammatory disease process in our study i.e. 24% which was further divided into infectious and non-infectious groups. The infectious conditions usually occur secondary to direct injury or spread from an adjacent focus particularly the paranasal sinuses or face[4]. Reider et al demonstrated in their study the concomitant orbital and para-nasal sinuses involvement[15]. CT findings were of help in evaluating the extent of the disease processes. A study on unilateral proptosis due to sino-nasal pathology by Mumtaz et al also shows CT scan in axial and coronal plan to be the single most reliable investigation in evaluation of disease process[16]. In our study, 14% were secondary to paranasal infection. 4% were a complication of dacryo cystitis and 1 case was secondary to a large infected boil on the cheek which later on involved the orbit leading to proptosis.

In non-infections category, the 2 cases were due to idiopathic orbital inflammatory disease (pseudotumor) with bilateral involvement. Both of them showed improvement with steroids.

Khan et al showed inflammatory causes to be 19% of total orbital lesion[6] while Sindhu et al showed that the most common cause of proptosis is children presenting to their institute was infective orbital cellulites and the most useful initial investigation was an orbital computed tomography[17].

In congenital lesions, encephaloceles formed 6% of total cases. Swelling over the nose, hypertelorism and proptosis was seen in all of them. X-ray skull was carried out but the bony defects with contents of the herniated brain substance were best assessed on CT scan.

There was 1 case of crozoun syndrome. CT scan with reformations was done before maxillofacial surgery in this case. 2 cases of dermoid cysts were included forming 4.0% of total orbital lesions. Both of them presented with painless proptosis. Ultrasound was also carried out in these patients however CT scan showed the full extent of the disease process. Bony changes and density of the different tissues in the lesion were also checked. Dermoids are commoner between 3-10 years of age. Upper temporal site is the commonest and usually they are anteriorly located[19]. Dermoid cyst though congenital in origin becomes apparent in late childhood[20]. Orbital sonography plays an important role in their pre-operative diagnosis[21]. There was only one case of neurofibromatousis. X-rays showed enlargement of the affected orbit with absent sphenoid wing giving 'bare orbit' sign. CT scan was also carried out in this case.

Traumatic lesions also behave like tumors. In our study, 2 patients present with proptosis secondary to trauma displacement of eyeball was due to organized heamatoma in the orbit. Associated fractures of the walls and entraped soft tissues were well picked up on both x-rays and CT scan. 1 post-

traumatic patient developed proptosis secondary to orbital cellulites. Overall traumatic lesion formed 6% of the total orbital lesions. Khan et al showed this figure to be 5.5% in his study[6].

Finally, CT scan findings were verified against clinical features, operative findings and / or biopsy. Out of 50 patients in 40 patients the pre-operative diagnosis on the basis of clinical history and CT findings were found correct. Hence the diagnostic accuracy of CT scan in our study was 80% which is quite a remarkable percentage.

CONCLUSION

The list of orbital lesions causing proptosis in children is quite different from that in the adult. Most of the patients are under 5 years of age. Besides proptosis visual deterioration is present in almost half of the patients. The most common underlying cause is a neoplasm followed by inflammatory disease process. However orbital involvement from pathologies of adjacent structure such as paranasal sinuses is not uncommon. Many pathology had unusual and atypical presentation. Timely referral, early diagnosis and appropriate management cannot only be vision but also life saving.

Moreover, CT scan with contrast, axial and coronal views may be considered as a single, non-invasive diagnostic tool which will not only localize and characterize the lesion but will also show calcification, cystic changes and extent of disease process.

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