# PROPTOSIS: MANAGEMENT OF 22 PATIENTS

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**ABSTRACT**

*The clinical features and management of 22 cases with proptosis is highlighted. Proptosis was unilateral in fifteen and bilateral in seven cases. The common etiologic factors were neoplasms, infections and bleeding diatheses. Of the ten cases with neoplasms as many as 7 expired, whereas of nine cases due to infections only one expired and one developed phthisis bulbi. All three patients with proptosis due to bleeds recovered completely without sequelae. Orbital CT scan was done in fourteen and ultrasound in eight cases, while in five cases both investigations were done.*

*As neoplasms are a frequent cause of proptosis in children and there is a high mortality in this group, a thorough systemic examination and hematological profile is essential to arrive at a speedy diagnosis. CT and USG are useful imaging modalities of the orbit, the latter being particularly useful for follow up of the lesion.*

**Key words:** *Proptosis, Etiology, Management, Orbital imaging.*

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Proptosis, which is defined as passive protrusion of the eyeball(1) though at times physiological is usually secondary to some underlying pathology and may lead to vascular congestion resulting in papilledema, optic atrophy and visual loss if not treated early(2). We have earlier reported cases of proptosis occurring in adults(3-5) and children(6,7). In this report we present our continued experience in the management of proptosis in 22 children, and stress the role of imaging of the pediatric orbit and the importance of multidisciplinary evaluation of such patients.

## Material and Methods

Twenty two patients aged 2 days to 12 years(14 males/8 females) who presented with proptosis were seen by us between 1984 and 1991. The cases were divided into 3 groups, according to the etiology of the proptosis. *Group A* consisted of 10 cases with neoplasms, *Group B* of 9 cases with infections and *Group C* of 3 cases due to bleeding diatheses. Diagnosis of proptosis was made by simple observation and confirmed by exophthalmometry. A difference of more than 2 mm in exophthalmometry reading between two eyes was considered as significant for unilateral proptosis(2). Visual acuity was tested in all patients (except the very young, or uncooperative/unconscious children) using Snellen's chart. All patients were subjected to an otorhinological, ophthalmological and neurological evaluation, and other relevant investigations carried out as required. Fourteen patients were subjected to CT scan of the orbit, 8 to orbital ultrasonography (USG), while in 5 both CT and USG were done.

## Results

Proptosis was bilateral in seven cases, of which six were due to neoplasms and

one as a result of an orbital bleed. In all, eight patients expired. In Group A, seven patients succumbed to their disease, of which only one was subjected to post-mortem (Case no. 9). In Group B four had facial or nasal furuncles and two had septicemia. One patient in this group expired of sepsis and meningitis, one was left with a residual hemiparesis, and one with a phthisis bulbi. All cases in Group C recovered without sequelae (*Tables I-III*).

**Discussion**

As the orbit forms the confluence between the eye, nose, paranasal sinuses and the contents of the anterior cranial fossa, varied etiologies can result in proptosis. This therefore necessitates not only local ocular but also thorough systemic examination and a multidisciplinary evaluation of the child. While we have previously reported(3-6) the surgical management of proptosis, most patients in the present series were treated medically.

Of the 22 cases, neoplasms were responsible for proptosis in 10. Of 10 cases in Group A, 8 developed proptosis due to metastatic infiltrates, while 2 were due to locally situated neoplasms, of which one was malignant(Case no. 10, *Fig. 1*).

The stages of orbital cellulitis have been well documented(8,9) and most cases resolve with suitable antibiotic and antiinflammatory therapy. In this group clinical examination, especially visual acuity and its hourly followup is critical in managing the patients and on which rests the decision of intervention although CT scan and USG aid in the diagnosis. A white blood cell count(WBC) clearly demonstrates the presence of an infection(10). All our patients with infections had WBC $>20 \times 10^9$/L. In Group B, only one case expired, while one case with cavernous sinus thrombosis (CST) was left with a residual hemiparesis, and one case developed phthisis bulbi. Of the three patients in Group C, one followed trauma to the orbit, one was due to scurvy, and one due to neonatal hepatitis. Orbital bleeds are rarely seen in scurvy or hepatitis. Whereas, in scurvy bleeding classically occurs in the gums, in hepatitis bleeds usually present as ecchymosis or hematemesis(11). Bleeding in scurvy is a result of increased capillary fragility while in hepatitis it is due to decrease in vitamin K dependent clotting factors.

Of a series of 21 cases of proptosis reported in the Indian literature(12), 12 were due to neoplasms and 6 due to infections, while orbital bleeds were not described. Retinoblastoma was the commonest cause of proptosis in that series, however, we did not see any such case.

Although CT scan is an established imaging method of the orbit(13), the use of orbital USG has also been described and recommended(14). We suggest that a USG of orbit be done first, followed by a CT scan, as the scan can then help confirm

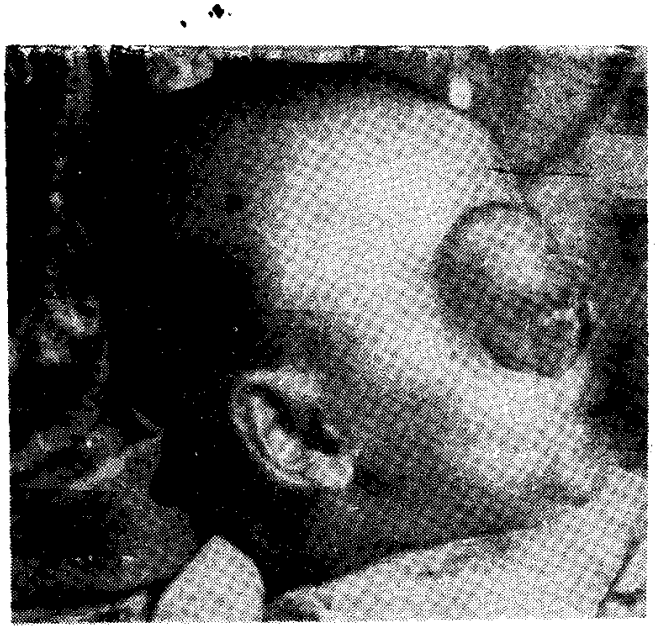

*Fig. 1. Large embryonal rhabdomyosarcoma causing gross proptosis.*

**TABLE I**—*Salient Features of Group A (n = 10)*

| Sr. No. | Sex/Age Diagnosis | Relevant investigations | Treatment and outcome |
|---|---|---|---|
| 1. | F/3 months Capillary hemangioma | USG – Solid homogenous intraorbital mass (L) eye CT– Contrast enhancing homogenous intraorbital mass Biopsy – Capillary hemangioma | Prednisolone *Lesion regressed* Vision –Normal Duration of follow-up – 1 year |
| 2. | M/5 years AML (Type M-4) | USG – Intraorbital deposits PS – Auer bodies present BM -- AML Type M-4 (FAB Classification) | IV -- Daunorubicin IV & IT Ara-C *Expired* |
| 3. | M/10 years AML | CT – Homogenous deposits (L) orbit and temporal bone PS – Auer bodies present | Treatment same as Case no.2 *Expired* |
| 4. | M/10 years AML | BM -- AML Type M-4 (FAB Classification) PS -- Auer bodies present | Treatment same as Case no. 2 *Expired* |
| 5. | M/8 years ALL | USG -- Dense homogenous orbital deposits BM – ALL (Type L 2) | Expired before therapy |
| 6. | M/5 years ALL | USG – Normal CT– Lacrimal gland and bulbar infiltrates BM – ALL | In Remission Duration of follow-up – 6 months |
| 7. | F/5 years Histiocytosis-X | *X*-ray Skull-multiple punched out areas Biopsy Scalp lesion – Histiocytosis-X | Steroids, Vincristine, Bleomycin Expired |
| 8. | M/10 years Neuroblastoma | *X*-ray Skull multiple punched out areas BM – Rosettes of Neuroblastoma | Refused therapy Expired |
| 9. | M/2 years Neuroblastoma | BM -- Rosettes of neuroblastoma PM – Histopathology confirmed diagnosis | No therapy offered *Expired* |
| 10. | F/6 months Embryonal rhabdomyosarcoma | CT – Homogenous mass (R) orbit with gross proptosis Biopsy – Diagnosis confirmed | Exentration (R) eye *Survived* Duration of follow up 1 year |

USG = Ultrasonography; AML = Acute myeloid leukemia; ALL = Acute lymphatic leukemia; IT = intrathecal; BM = Bone marrow; PS = Peripheral smear; ARA-C = Cytosine arabinoside; PM = Post-mortem.

**TABLE II**—*Salient Features of Group B (n = 9)*

| Sr. No. | Sex/Age/Diagnosis | Relevant Investigations | Outcome |
|---|---|---|---|
| 1. | M/7 years<br>Rt CST with meningitis | Fundus – Congestion<br>CT-CST with gross proptosis<br>CSF – Purulent meningitis<br>Blood Culture - No growth<br>WBC = $22 \times 10^9$/L | Vision –Normal<br>*Survived* |
| 2. | F/3 years<br>Rt CST with meningitis | Fundus – Normal<br>CT-CST, Retro-orbital edema infarct (R) cerebral hemisphere<br>USG - Sonolucent retrobulbar mass<br>Isotope brain scan - absent perfusion (R) MCA<br>Blood & CSF culture -- No growth<br>WBC = $25 \times 10^9$/L | Survived with residual hemiparesis<br>Vision – Normal |
| 3. | F/5 months<br>RT orbital cellulitis | Fundus – Hazy<br>*X*-ray Orbit-Haziness<br>Blood culture--Staph aureus<br>WBC = $23 \times 10^9$/L | *Survived* |
| 4. | F/12 years<br>Orbital cellulitis with maxillary sinusitis | CT – Orbital cellulitis with Maxillary sinusitis<br>Blood Culture - Staph aureus<br>WBC = $27 \times 10^9$/L | *Survived* |
| 5. | M/1 month<br>RT orbital cellulitis with intraconal abscess | Fundus -- Hazy<br>USG -- Intra bulbar orbital abscess<br>Blood Culture – Staph aureus<br>WBC = $25 \times 10^9$/L | Survived with phthisis bulbi |
| 6. | M/7 years<br>Orbital cellulitis with Intraconal abscess | CT– Orbital cellulitis with small intraconal abscess<br>Blood and CSF – Culture negative<br>WBC = $29 \times 10^9$/L | Survived |
| 7. | M/7 years<br>Lt orbital cellulitis | CT– Retrobulbar inflammatory mass<br>USG – Retrobulbar abscess with cystic area<br>WBC = $26 \times 10^9$/L | Survived |
| 8. | M/10 years<br>Orbital abscess with meningitis | CT – Orbital abscess, no brain abscess CSF -- Purulent meningitis<br>WBC = $25 \times 10^9$/L | Expired |
| 9. | M/9 years<br>Pseudotumor orbit | CT– Homogenous nonenhancing orbital mass<br>Normal hematological investigations | Lesion regressed<br>Vision – Normal |

CST = Caverous sinus thrombosis; CSF = Cerebrospinal fluid; MCA = Middle cerebral artery.

**TABLE III--** *Salient Features of Group C (n = 3)*

| Sr. No. | Sex/Age/ Diagnosis | Relevant investigations | Treatment and outcome |
|---|---|---|---|
| 1. | F/9 months<br>Traumatic orbital bleed | CT– Orbital hematoma<br>Normal hematological investigations, Fundus – normal | No treatment<br>Survived<br>Vision – Normal |
| 2. | M/3 months<br>Neonatal hepatitis | CT – Large retrobulbar clot<br>USG – Large retrobulbar clot<br>PT – 60 secs (control - 15 sec)<br>PTT – 80 secs (control - 45 sec)<br>SGPT – 400 IU<br>(Normal – 13 -35 IU)<br>SR bilirubin total = 204 umol/L<br>Fundus – Normal | IV Vitamin K 5 mg/day × 3 days<br>FFP = 10 ml/kg/day × 3 days<br>Survived<br>Vision – Normal<br>Duration of follow-up – 6 months |
| 3. | F/15 months<br>scurvy | CT – Bilateral retrobulbar clots<br>Normal hematological investigations<br>Fundus -Normal<br>*X*-ray – knees -- Scurvy | Oral Vitamin C 500 mg/day × 6 weeks<br>Survived<br>Vision – Normal |

FFP = Fresh frozen plasma; PT = Prothrombin time; PTT = Partial thromboplastin time.

the USG findings, demonstrate the vascularity of the lesion and determine the status of the intracranial cavity. Once the CT scan demonstrates the lesion to be limited to the orbit follow-up of lesion can be done by orbital USG. While there are few studies comparing the reliability of orbital USG with that of the CT scan of orbit, Levine(14) found orbital USG to be a reliable diagnostic tool, that confirms, and comple-

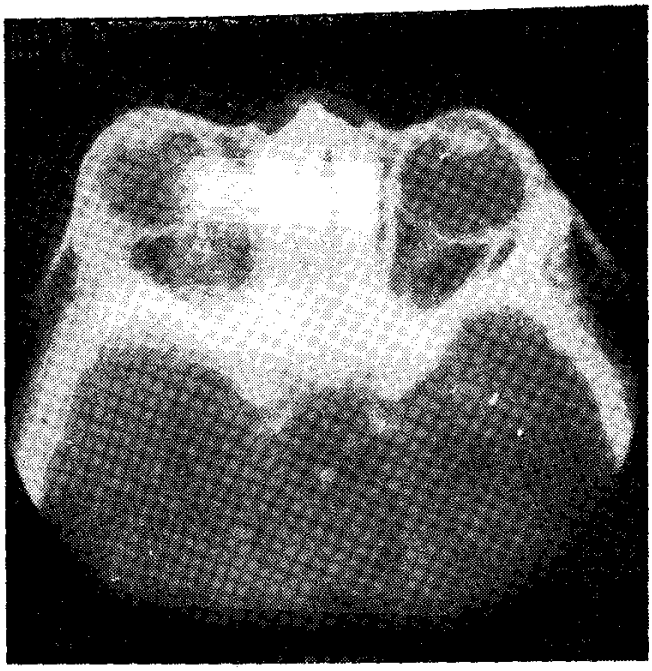

*Fig. 2a. CT scan showing retrobulbar mass.*

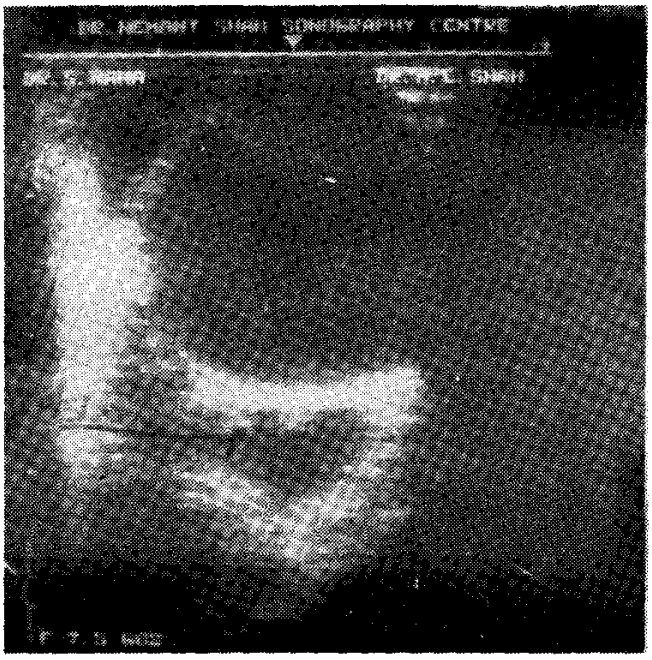

*Fig. 2b. USG orbit of same patient showing cystic retrobulbar mass.*

ments information derived from orbital CT scan. In addition, it is non-invasive and is cost effective. Of five patients in this series who had both USG and CT scans done, the USG findings correlated with those of the CT scan (*Fig. 2a, b*). All these cases were followed up with the aid of orbital USG for three to six months.

As neoplasms are a frequent cause of proptosis in children and there is a high mortality in this group, a thorough systemic examination and hematological profile including a peripheral smear, total white cell count, and, if required, a bone marrow examination may be essential. A multidisciplinary approach is necessary to arrive at a quick and correct diagnosis. CT and USG are useful imaging modalities of the orbit, the latter being particularly useful for follow up of the lesion.

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