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The Investigation of Proptosis in Paediatric Practice.

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Case Series

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Abstract

A retrospective review of 65 cases of paediatric proptosis presenting to a rural teaching hospital in India, from February 2006 to June 2008. There were 17 cases with orbital cellulitis and 15 with retinoblastoma. We report the history, clinical examination, radiological findings and diagnosis. Most cases had a characteristic history and pathognomonic eye signs. Computer Tomography (CT) correlated with histopathology in most cases. CT is widely available, even within a remote setting, and is a convenient investigation. Thus, a detailed history, clinical examination and CT scan were the most fruitful approach to the diagnosis of childhood proptosis in this series.

Background

Proptosis (exophthalmos) is defined as an abnormal protrusion of the eyeball. The aetiology of proptosis can be inflammatory, vascular, or infectious. Protrusion >18 mm from the interzygomatic line has been adopted as the definition of proptosis in our study [1]. There is a substantial list of differential diagnosis for proptosis in children. The importance of early diagnosis leads to timely treatment and confers a better prognosis for malignancies [2]. While in the Developed World presentations are early, in a country like India, the realities of poverty means that many cases present late and require more complex management with a poorer prognosis. We seek to explore the diagnostic value of CT scan relative to other investigations histopathology or other investigations to become the sole diagnostic modality for paediatric proptosis.

Methods

A retrospective review of 65 case records of paediatric proptosis from Bankura Sammilani Medical College and Hospital, India from February 2006 to June 2008. There were 38 males and 27 females. The patients were aged between 27 Days and 17 Years, the mean age being 7.5 years. The review included demographic details, history, physical examination, fundoscopic examination, X-RAY findings, CT scanning technique and findings, histological examination and or any other investigations performed.

A high speed CT dual scanner from GE-Healthcare was used for scanning all the patients. Axial sections were taken with patient supine position with slice thickness of 1 mm and interslice gap of 1 mm. Coronal 1-3 mm sections were obtained only when required with the patient in prone position. Almost all scans were taken without contrast medium; contrast media were administered intravenously in a limited number of cases. Scans were done at 80 kVp and 100mA (80mA was used in case of infants less than a year old). Total duration of scan time for each patient was 45-60 seconds.

Results

55% of all cases were five year old or younger. Orbital cellulitis and Retinoblastoma represented the most common pathologies. The diagnoses of all of the cases are listed in Table 1. Orbital Cellulitis was documented as a complication of sinusitis in most cases. It was accompanied by fever, discharge and signs of local inflammation. Retinoblastoma was the second most common cause and was associated with leukocoria and strabismus as well as proptosis. Thirteen percent of cases had a positive family history and bilateral disease.

Most of the cases presented with recognised clinical features. Other than proptosis visual defects were present in 35 cases. Only rhabdomyosarcoma presented with rapidly progressive proptosis. Most cases were unilateral (41 cases- 63%). History of proptosis in 1st degree relatives was reported in one case of retinoblastoma and in all patients with neurofibromatosis. There was fever in orbital cellulitis, leukaemia and Ewing's sarcoma and pain in orbital cellulitis, neuroblastoma and pseudotumor. Echymoses and hemorrhagic spots were features of leukaemia, while those with neurofibromatosis had Café-au-lait Spots. Lymphoma, leukaemia and malignancies presented with cervical lymphadenopathy. Grave's disease featured palpitations and tremors. Various congenital anomalies were present in Sanderson Fraser Syndrome.

On systemic examination, neurofibromatosis patients were found to have a pulsatile eye mass. Lid oedema, erythema and discharge was found in orbital cellulitis, periorbital ecchymoses was found in neuroblastoma, lid swelling was present in rhabdomyosarcoma, Grave's disease had lid lag, while Sanderson Fraser featured an absent lid. Acute leukaemia was associated with a subconjuctival haemorrhage, while patients with retinoblastoma had a white pupil (leukocoria). On fundoscopy cases with retinoblastoma were found to have retinal detachment. Neurofibromatosis was

associated with Lisch nodules and disc atrophy, while the leukemias and Ewing's sarcoma presented with papilloedema.

CT Scan reports

In orbital cellulitis CT showed - diffuse subperiosteal involvement with pre-septal extensions and increased soft tissue density. There was abscess formation in 5 cases [Figure-1]. In retinoblastoma osseous involvement was present in a few cases [Figure-2].

In Optic Nerve Glioma CT suggested fusiform enlargement of optic nerve. Rhabdomyosarcoma was associated with a homogenous ring enhancing lesion. In 2 cases it was also associated with bony destruction and nerve invasion [Figure-3].Cases of neuroblastoma presented with underlying bony changes and intracranial extensions.

Scans of patients with Neurofibromatosis showed dysplastic wings of sphenoid (Bare Orbit Sign). Neurofibroma was demonstrated with a well circumscribed ovoid mass. Our series included a patient with an orbital hemangioma in which the CT showed a hyperdense mass. The patient with Ewing's sarcoma had a scan which revealed a high density area, generalized soft tissue thickening as well as extraocular muscle and optic nerve thickening.

The case of lymphoma presented with a scan in which a homogenous mass covered both the intraconal and the extraconal compartments and the extra ocular muscles, but excluded the optic nerve. The child with AML (acute myeloid leukaemia) had a homogenous enhancing mass which was associated with infiltration of optic nerve and extraocular muscles.

Pseudotumor cases presented with a heterogeneous mass. Dermoid Cysts featured a mass with underlying bony changes. Cases of child abuse were associated with a fracture of the orbital wall.

Fibrous Dysplasia of orbital bones was characterized by involvement of the lateral wall of orbit on both sides associated with bone thickness as well as destruction on axial CT [Figure-4]. Graves Disease was demonstrated with enlargement of the inferior rectus muscle.

CT diagnosis correlated well with the histopathology in 29 out of 32 cases of benign, malignant or metastatic neoplasm.

Discussion

Proptosis in children is not an uncommon finding. The differential diagnosis is quite different for the same feature in adults [3]. Many of the cases reported here had unusual and atypical presentations however visual deterioration was present in almost 50% of patients.

The most common causes of paediatric proptosis were orbital cellulitis or retinoblastoma. For the evaluation of visual loss or suspected cranial nerve dysfunction, MRI is the procedure of choice in adults. However, MRI is not widely available. Moreover, MRI is technically challenging in children.

On the other hand, the availability, low cost and relative faster speed of CT scanning makes the CT scan a valuable diagnostic tool in paediatric orbital diseases. Our study showed that CT provided accurate diagnosis in 91% of the cases and this was consistent with the proportion reported by Sabharwal et al [4]. CT guided FNAC (fine needle aspiration cytology) can also be performed in cases of residual uncertainty. Thus CT has been considered the best investigation for proptosis in a child. The CT findings for many conditions replicated those reported by other experts [4-11].

Therefore a detailed history, careful examination and a CT scan may be the best approach to paediatric proptosis. However the optimization of CT scanning protocols in paediatric imaging is of particular importance in order to minimize the radiation hazards. This needs to be addressed in future studies.

Abbreviations used:

CT-Computed Tomography
AML-Acute Myeloid Leukaemia
MRI-Magnetic Resonance Imaging
FNAC-Fine Needle Aspiration Cytology

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AUTHORS' CONTRIBUTIONS
Both authors contributed equally to all aspects of the study.

PEER REVIEW
Not commissioned; externally peer reviewed

CONFLICTS OF INTEREST
None

TABLE-1: Causes of proptosis in 65 patients- table and graph

CAUSES OF PROPTOSIS	NO. OF PATIENT	PERCENTAGE
1. Orbital Cellulitis	17	26.2
2. Retinoblastoma	15	23.1
3. Optic Nerve Glioma	7	10.7
4. Rhabdomyosarcoma	4	6
5. Hematoma	4	6
6. Neurofibroma and Neurofibromatosis	4	6
7. Pseudotumor	3	5
8. Dermoid Cyst	3	5
9. Lymphoma	1	1.5
10. AML	1	1.5
11. Hemangioma	1	1.5
12. Neuroblastoma	1	1.5
13. Sanderson-Fraser Syndrome	1	1.5
14. Ewing's Sarcoma	1	1.5
15. Grave's Disease	1	1.5
16. Fibrous dysplasia	1	1.5

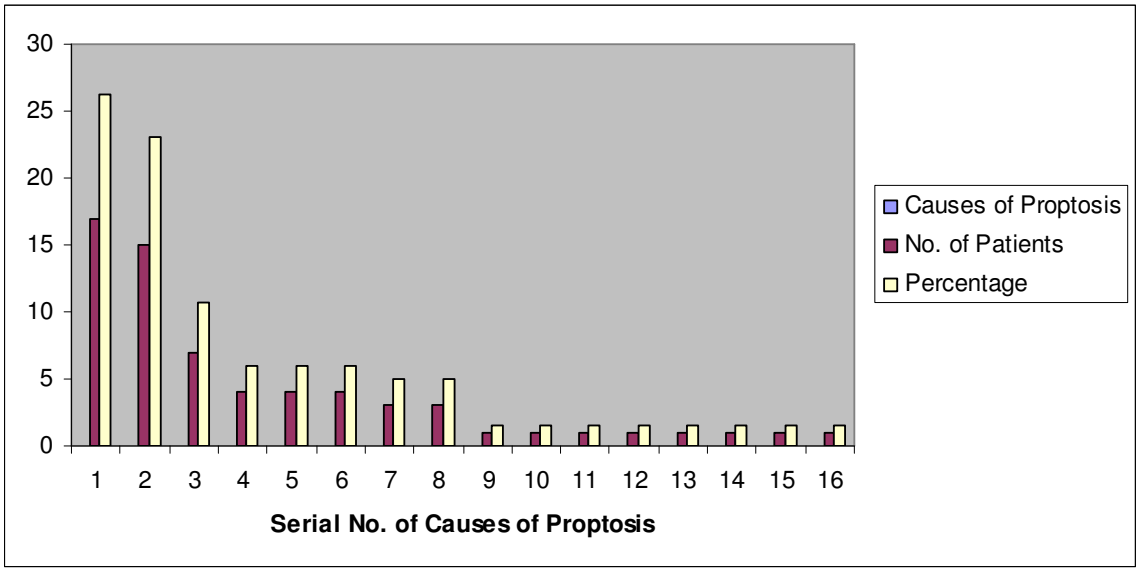

FIGURE-1: TITLE: CT Brain and Orbits (axial view) in a patient of orbital cellulites

CT shows diffused subperiosteal involvement with preseptal extensions and increased soft tissue density with probable abscess formation.

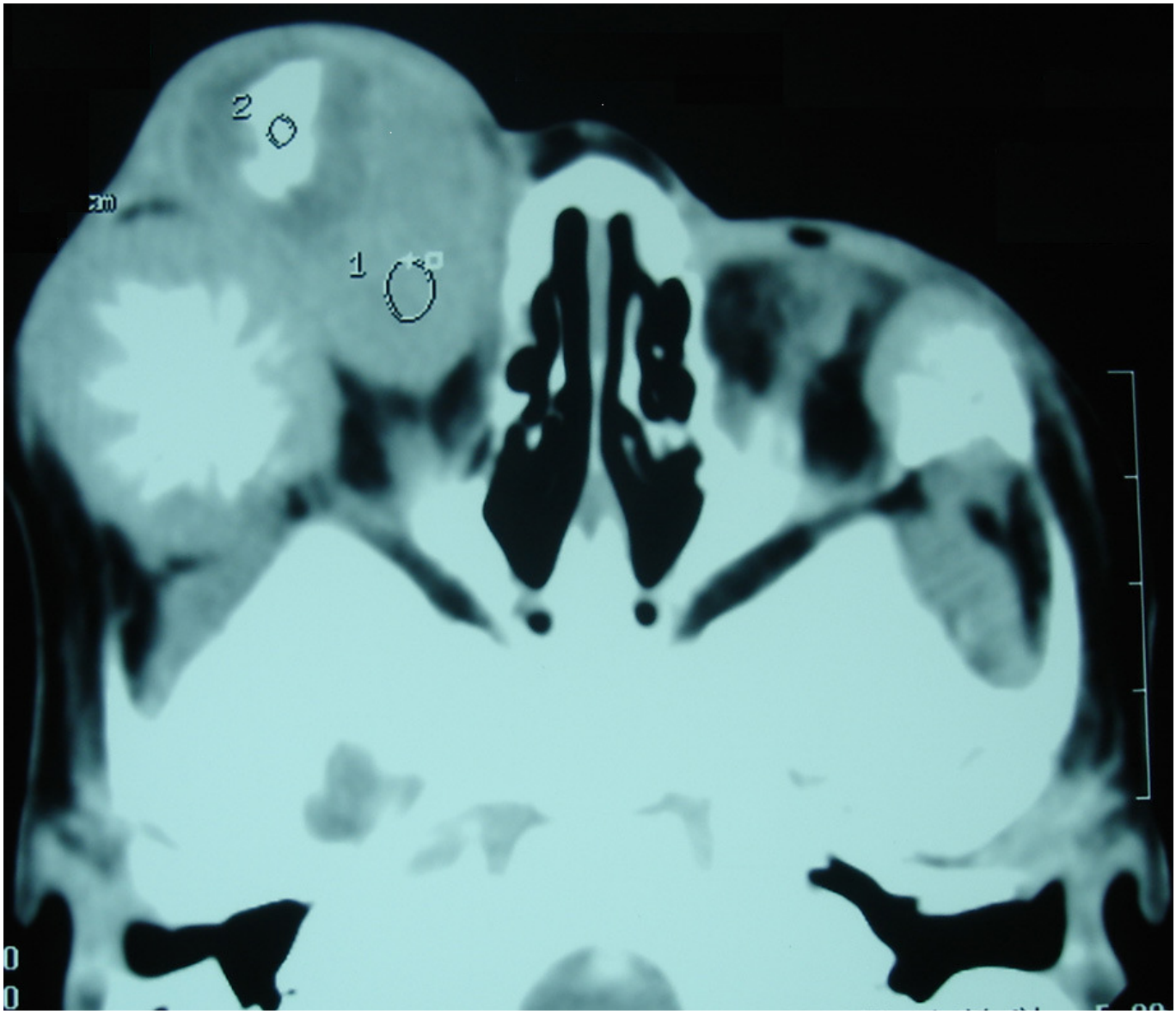

FIGURE-2: TITLE: CT Brain and Orbits (axial view) in a patient of retinoblastoma.
CT shows calcified infiltrating lesions in eyeball with osseous involvement.

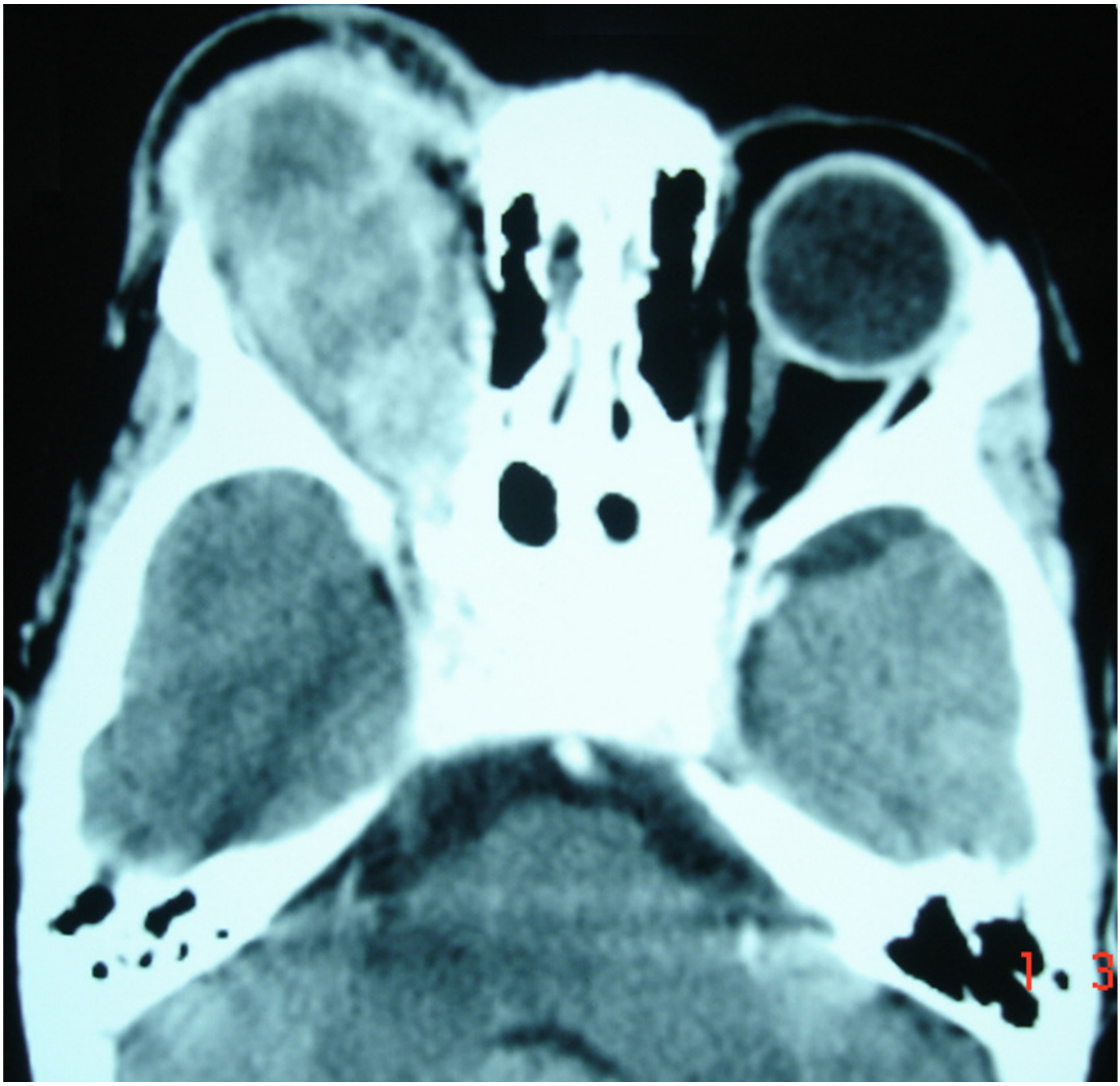

FIGURE-3: TITLE: CT Brain and Orbits (axial view) guided FNAC in a patient of rhabdmyosarcoma.
CT shows an FNAC needle inside a homogenous orbital mass. The mass has caused destruction to the bony orbit.

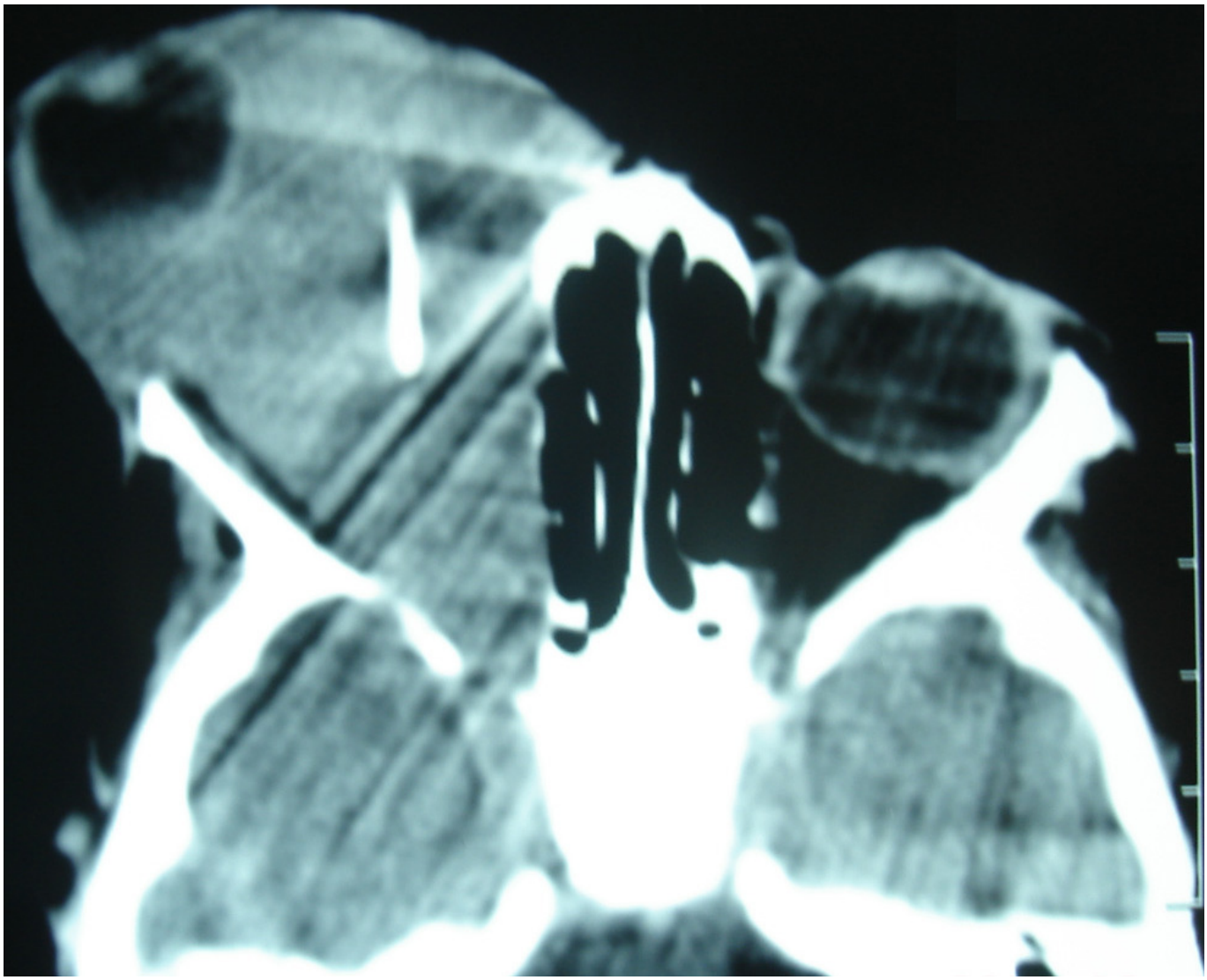

FIGURE-4: TITLE: CT (coronal view) in a patient of fibrous dysplasia.
CT shows bilateral mass arising out of lateral wall of orbit along with thickening and destruction of the underlying bone. The globes are displaced laterally.