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## Research Article

### PEDIATRIC SELLAR & PARASELLAR LESIONS: MRI CHARACTERISTICS

Nandha Kishore M Y\*<sup>1</sup>, Jeevika M U<sup>2</sup> and Siddesh M.B<sup>3</sup>

<sup>1,2,3</sup>Department of Radiodiagnosis, J.J.M Medical College, India

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#### ABSTRACT

Pediatric brain tumors have always been challenging as well as intriguing in their anatomical, surgical, and postsurgical management-related issues. They are a heterogeneous set of pathologies involving different age groups in childhood and also differ widely from their adult counterparts as far as adjuvant therapies are concerned. Our objective is to review the MR imaging characteristics and applications of sellar & parasellar lesions in the pediatric population. Specifically, we review 3 cases of craniopharyngiomas, 2 cases of hypothalamic hamartomas & 1 case of each pituitary colloid cyst, parasellar retinoblastoma, pns rhabdomyosarcoma with sellar extension.

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## INTRODUCTION

Central nervous system tumours are the most common childhood malignancy and leading cause of cancer related morbidity and mortality with incidence of 2.5–4 per 100,000 children[1]. Sellar and suprasellar tumors in pediatric form a separate entity as far as incidence, histology and responsiveness to therapy is concerned. They also differ in their management from the adults. While primary surgical management is the key in most of the adult tumors, chemo- and radiotherapy are the chief modality in some of these pediatric tumors like germinomas. We have reviewed 8 cases of sellar & parasellar lesions in pediatric age group and tried to focus on MR image characteristics.

### Objective

To know the characteristic MRI feature of sellar & parasellar lesions in pediatric population.

## MATERIALS AND METHODS

Prospective study of 8 pediatric patients referred for imaging having clinical features raising suspicion of sellar & parasellar lesions. They underwent magnetic resonance imaging of the brain-sella using 1.5 TESLA PHILIPS ACHIEVA MRI MACHINE, in the Department of Radiodiagnosis, Bapuji Hospital, Davangere.

## RESULTS

### Craniopharyngioma

In our study of 8 cases, there were 3 patients of craniopharyngiomas. Age range was from 3.5 to 9 year with 3 all cases seen below the age of 10 years, 2 cases were males and 1 case was female. The male to female ratio was found to be 2:1.

All 3 cases were presented with solid-cystic (mixed) morphology. In which solid component was iso-hypointense in 1 case, isointense in 1 case and iso-hyperintense in case on T1W images, isointense to slightly hyperintense on T2W images in all 3 cases. Cystic component were isointense on T1W & hyperintense on T2W images.

On post contrast study solid component showed heterogenous enhancement in all 3 cases. The postcontrast study was helpful in detecting cystic areas within lesions, depicting as nonenhancing or peripherally enhancing areas.

The presence of calcification is noted in all 3 cases.

Although all were primarily suprasellar in location, all cases of craniopharyngiomas in our study showed intrasellar extension. Bony sellar wall erosion was seen in 2 cases of craniopharyngioma. The sellar expansion was not as large as in pituitary adenomas. Only one case showed widening of sella on preliminary CT.

\*Corresponding author: *Nandha Kishore M Y*  
Department of Radiodiagnosis, J.J.M Medical College, India

Additionally hydrocephalus was found in 2 cases. This is related to the fact that the craniopharyngiomas tend to grow larger in size and cause more mass effect than any other lesion. Superior, lateral, posterior and inferior extensions of craniopharyngiomas are better demonstrated in contrast enhanced coronal sequences as in pituitary macroadenomas.

#### **Hypothalamic hamartoma**

In our study of 8 cases, there were 2 cases of hypothalamic hamartoma. 1 case was male aged 8yrs and 1 case was female aged 4yrs and both presented with features of seizures.

On MRI both were hypointense on T1W, 1 case was isointense and 1 case was slightly hyperintense on T2W and showed no enhancement on postcontrast study.

#### **Pituitary Colloid Cyst**

In our study of 8 cases, there was 1 case of pituitary colloid cysts was found in 17 years old female presented with headache.

MRI showed slight hyperintensity on T1W sequence, FLAIR sequence and isointensity on T2W sequence. Diffusion restriction and post contrast enhancement was absent.

#### **Retinoblastoma with parasellar extension**

In our study of 8 cases, there was 1 case of retinoblastoma with parasellar extension in 5 years old female child. Suprasellar / parasellar extension of retinoblastoma are very rare and sometimes are termed as quadrilateral retinoblastoma.

The patient was known operated case of left eye retinoblastoma with prosthetic non metallic left eye implant. Patient presented with headache, nausea and vomiting.

On MRI there was ill-defined intraconal lesion extending along the path of left optic nerve (left optic nerve was not visualized separately) with widening of optic canal and was invading optic chiasma, parasellar & suprasellar compartment. Prosthetic non metallic eye implant was noted. Lesion was slightly hyperintense on T1W images and hypointense on T2W images when compared with vitreous. Mass on post contrast T1W sequence showed diffuse intense enhancement and showed diffusion restriction (on high b value of 2500).

#### **PNS Rhabdomyosarcoma**

In our study of 8 cases, there was 1 case of paranasal sinus rhabdomyosarcoma in 4 years old male child with sellar extension. Rhabdomyosarcomas are the commonest paediatric soft-tissue sarcoma, constituting 3–5% of all malignancies in childhood.

Patient presented with headache, nausea, vomiting and nasal obstruction.

On MRI lesion was noted within the right maxillary sinus causing its expansion and was involving sphenoid sinus, infrasellar and sellar compartments. Superiorly lesion was eroding floor of orbit and invading into orbit. Lesion was appearing heterogeneous hypo-isointense on T1W images and iso-hyperintense on T2W images and was showing diffusion restriction. On post contrast T1W sequence lesion showed moderate heterogeneous enhancement.

Patient was operated and diagnosis was confirmed by histopathology.

## **DISCUSSION**

### **Craniopharyngioma**

Craniopharyngiomas arise from ectopic embryonic remnants of the craniopharyngeal duct, which connects the stomodeal ectoderm with the evaginated Rathke's pouch, which in turn forms the future adenohypophysis. This may explain why craniopharyngiomas arise not only along the migration route of the craniopharyngeal duct, through the nasopharynx and the sphenoid sinus in rare cases, but also, mainly, in the intrasellar and suprasellar regions. According to the second hypothesis craniopharyngiomas arise from squamous epithelial cells in the pars tuberalis of the adenohypophysis

Craniopharyngiomas account for 3 to 5% of all intracranial tumours. They are the most common nonglial brain tumors. They account for half of all suprasellar masses in this age group [2]. More than half of craniopharyngiomas occur in children and young adults.<sup>50</sup> Forty percent of craniopharyngiomas in children occur between age group 8 to 12 years. A second somewhat smaller peak occurs in middle aged adults. There is no gender predilection[2].

Location wise craniopharyngiomas are essentially confined to sellar region; only rarely do they occur in other locations. About 70% craniopharyngiomas are completely suprasellar with small intrasellar component, 20% are suprasellar only, and 10 % are intrasellar only [3].

**CT:** NECT scans typically show cystic appearing lobulated suprasellar mass with solid mural nodule. Nodular or rim calcification is present in nearly all pediatric craniopharyngiomas and is identified in 50% of adult population. Cyst content is typically slightly higher in attenuation than CSF. Postcontrast study shows rim or nodular enhancement in 90% cases.

**Magnetic resonance imaging:** Of all sellar region masses, the craniopharyngiomas have most heterogeneous MR imaging spectrum. Out of 10 cases evaluated by Johnsen *et al*[4], half of the craniopharyngiomas imaged showed roughly even amounts of high and low signal on T1-weighted images; of the remaining tumors, three were predominantly high in signal intensity and one was predominantly low. All lesions showed some degree of signal heterogeneity.

Optic tract edema seen on coronal T2 TSE images, is a useful MR finding for distinguishing craniopharyngiomas from other parasellar tumors with considerable sensitivity and high specificity [5].

Additionally ADC is a useful parameter for differentiating between craniopharyngiomas and pituitary adenomas and between craniopharyngioma and meningioma. The ADC of craniopharyngiomas was significantly higher than that of pituitary adenomas and meningiomas[6].

### **Hamartoma**

Hypothalamic hamartomas are developmental malformations consisting of tumorlike masses located in the tuber cinereum of the hypothalamus. Most patients present in the first or second decade of life, with boys being more commonly affected than

girls. These lesions have been divided into two main clinicoanatomic subsets: parahypothalamic hamartomas and intra-hypothalamic hamartomas. Parahypothalamic hamartomas are pedunculated masses that are attached to the floor of the hypothalamus by a narrow base.

These lesions seem more likely to be associated with precocious puberty than with gelastic seizures. Intrahypothalamic hamartomas are sessile masses with a broad attachment to the hypothalamus. They appear to lie within the substance of the hypothalamus itself and may distort the contour of the third ventricle. In addition, they seem to be associated more often with gelastic seizures than with precocious puberty [7].

At pathologic analysis, hypothalamic hamartomas contain nerve cells that resemble those of the normal hypothalamus, along with normal glial cells [8].

**MRI:** At MR imaging, they are seen as well-defined pedunculated or sessile lesions at the tuber cinereum and are isointense [10] or mildly hypointense [9] on T1-weighted images and iso- to hyperintense on T2-weighted images, with no contrast enhancement or calcification. The absence of any long-term change in the size, shape, or signal intensity of the lesion strongly supports the diagnosis of hypothalamic hamartoma [9,10].

#### **Pituitary Colloid Cyst**

Colloid cysts of the pituitary gland, contrary to Rathke's cleft cysts, are filled with a colourless or whitish, relatively thick colloid (mucopolysaccharide) substance. Their walls are built of connective tissue devoid of epithelial elements, which indicates to the fact that colloid cysts of the pituitary gland are not of epithelial origin [11]

Colloid cyst of the pituitary gland is a rare phenomenon [11,12]. The available medical literature does not include any numerical data on the incidence of this pathology. They are mostly asymptomatic, due to their small size, ranging from 6 to 21 mm [11], and from 5 to 15 mm, according to some other authors [12].

However, growing cysts lead to abnormalities of the endosecretory function, suggestive of sellar pathology. Hypopituitarism affects the gonadotropic cells mostly, leading to menstrual disorder (oligomenorrhea and galactorrhea) and hypogonadotropic hypogonadism in men [11,12]. Moreover, headaches are quite frequent as well – according to Nomikos *et al.*, they are experienced by 27% of patients [11]. There were also reported cases of diabetes insipidus or the pituitary stroke<sup>2</sup>. Apart from the disturbed pituitary gland function, the colloid cysts growing suprasellarly may compress the optic chiasm and cause visual disturbances [11].

**CT:** As far as the CT is concerned, the cysts are mostly (in two thirds of all examined cases) hyperdense in comparison to the grey matter, while one third of them are hypo- or isodense. These are mainly oval or round structures. After contrast administration, CT reveals a thin layer of enhancement, which may correspond to the capsule of the cyst [13]

**MRI:** On MRI, colloid cysts may produce diverse signal intensity. Some lesions are inhomogenous. Sometimes, it is possible to visualise the fluid level or the presence of an

additional structure, located centrally or peripherally within the cyst. Approximately 50% of these pathological lesions are hyperintense in T1-weighted images. The rest may be iso- or hypointense as compared to the gray matter. In T2-weighted images, most of the colloid cysts show decreased signal intensity, while FLAIR sequences reveal increased signal intensity. In DWI sequences, those cysts are shown to form areas of decreased signal intensity [13]. In T1-weighted images, the colloid cyst is hypointense and remains unenhanced after contrast administration [11,12].

#### **Retinoblastoma**

Retinoblastoma is the most common intraocular tumor in children. The incidence is one in 17,000 births. Mean age at clinical presentation is 2 years in unilateral forms (60% of cases) and 1 year in bilateral forms [14,15]. All bilateral forms, as well as 15% of unilateral forms are related to a constitutional (hereditary or de novo) mutation of the RB-1 gene, localized on chromosome 13q14 [15]. Usually the patients present with leukocoria (white pupil reflection) or a squint.

The growth pattern of retinoblastoma can be subdivided into three types: endophytic, exophytic, and combined endophytic and exophytic [16]. Extraocular extension of retinoblastoma develops in less than 10% of patients [17] and is associated with a considerably higher mortality rate. Retinoblastoma spreads by direct extension into the orbit along scleral emissary vessels through hematogenous dissemination, invasion of the optic nerve, and dispersion through the cerebrospinal fluid after tumor cells in the optic nerve invade the leptomeninges [18]

**MRI:** Retinoblastomas are intermediate to hyperintense intermediate signal intensity, hyperintense c.f. vitreous compared to vitreous on T1 [19,20] and hypointense on T2. On post contrast T1 small lesions enhances homogeneously and large tumours may show few non enhancing necrotic areas. The tumour shows restricted diffusion on DWI at high b values. It exhibits low ADC values in contrast to the high intensity of the vitreous in the ADC maps. ADC map can be used to differentiate viable and necrotic tumour [21]. DWI is valuable in evaluating the response to eye-preservation treatment [21].

#### **Rhabdomyosarcoma (RMS)**

RMS is a rare malignant tumor, originating in the conjunctive and derivative tissue. It is especially rare in adults, whereas it is the most frequent form of soft-tissue tumor in children [22]. RMS mainly develops in the thorax or extremities in adults, and in the ENT region or uro-genital tract in children [23]. Only 10–15% of adult ENT locations are in the paranasal sinuses [24,25].

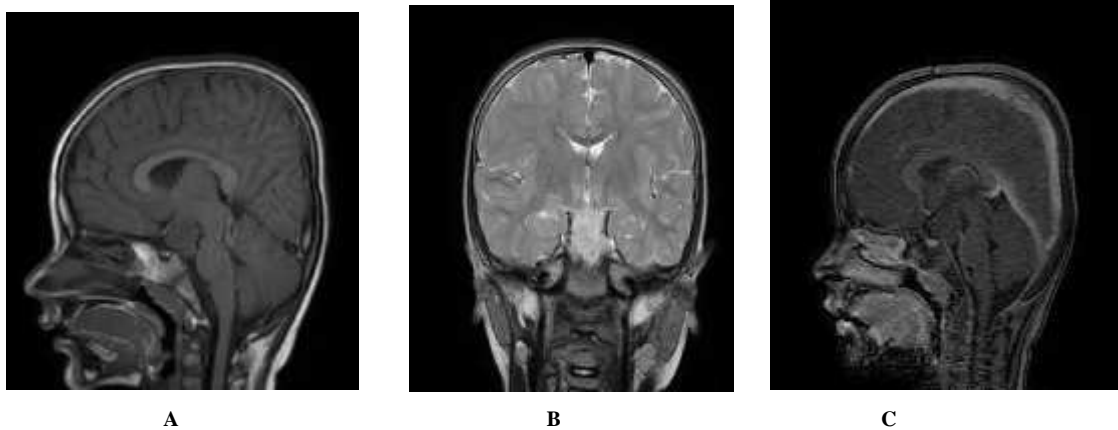
Histologically, three types are distinguished: embryonic, alveolar and pleomorphic. The embryonic type is the one most frequently encountered in ENT, mainly affecting children under 5 years of age; it is found in 25% of nasopharynx and sinonasal cavity RMS locations, and is the form associated with the best prognosis. The alveolar type mainly affects children over 5 years of age, adolescents and young adults; it may be located in the sinonasal cavities, and is of poorer prognosis. The pleomorphic form is rarer, mainly affects adults and is of poor prognosis [22].

**CT:** Lee *et al* [27] reported that 10 HNRMSs appeared as isodense (100%; 10/10) on pre-contrast CT and homogeneously enhanced (60%; 6/10) on post-contrast CT. Tumour may show adjacent bone destruction in 20% cases [28]

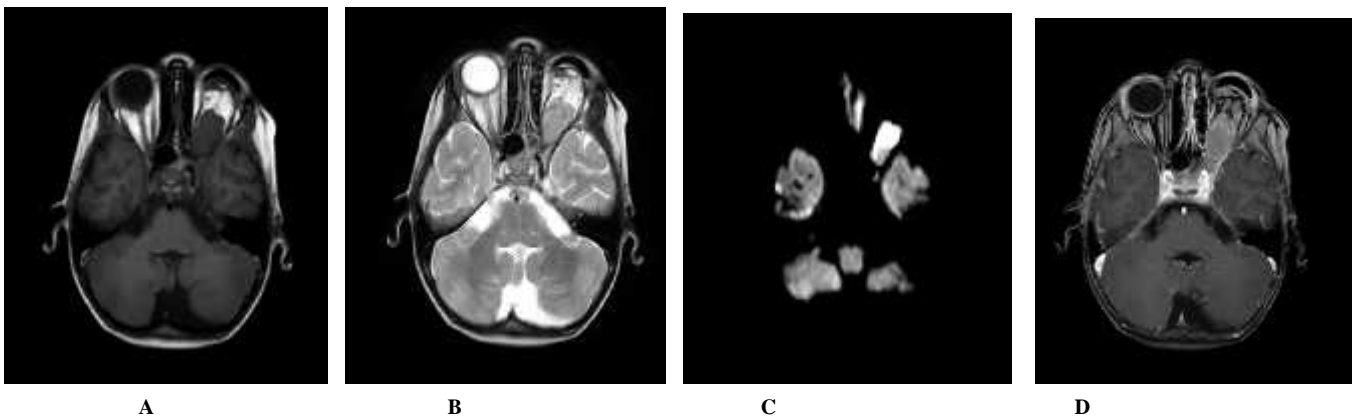
**MRI:** Hagiwara *et al* [26] presented eight HNRMSs with isointensity (37.5%) and slight hyperintensity (62.5%) on T1WI, and homogeneous (12.5%) and heterogeneous hyperintensity (87.5%) on T2WI, and heterogeneous enhancement (100%) on CE-T1WI.



**Case No.1: CRANIOPHARYNGIOMA:** A)T1W sagittal image, B)T2W coronal image and C)T1W post contrast sagittal images showing a large solid cystic lesion in suprasellar region with cystic component iso-hypointense on T1W, hyperintense on T2W and show peripheral enhancement on post contrast images. Solid component show mixed signal on T1W and T2W images and heterogenous enhancement on contrast study. D) Axila Gradient image (T2FFE) shows areas of blooming suggestive of calcification.



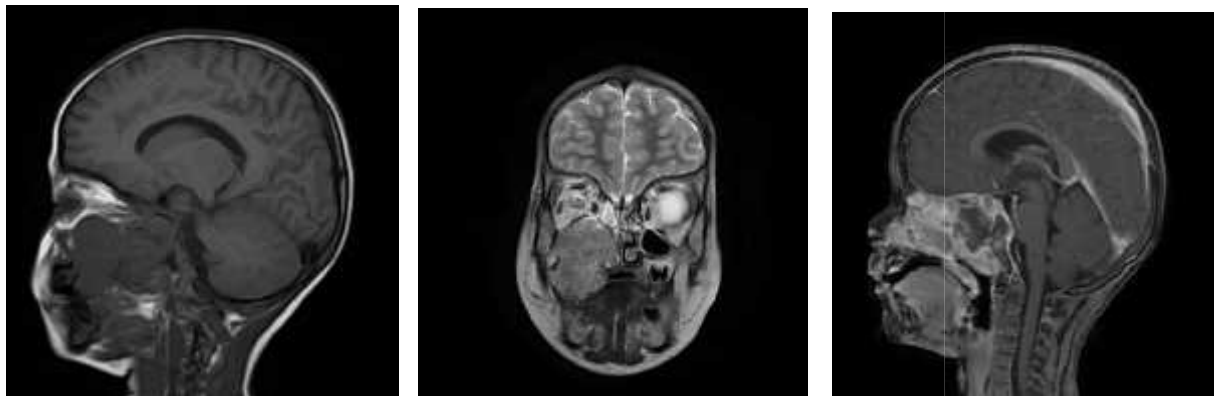
**Hypothalamic hamartoma:** A) T1W sagittal and B) T2W coronal images show a well defined mass in relation to tuber cinereum which is hypointense on T1W and hyperintense on T2W images. Note that pituitary is seen separately. C) Post contrast sagittal image showing no enhancement.



**Retinoblastoma With parasellar Extension**

A) precotrast T1W and B) T2W axial images shows a intraconal solid lesion in left orbit involving optic nerve extending through optic canal and involving optic chaisma, lesion is slightly hyperintense on T1W and hypointense on T2W images when compared to vitreous. C) Lesion is showing diffusion restriction. D) Post contrast T1W axial image show homogenous enhancement of the lesion. Left side non metallic prosthetic eye implant is seen.

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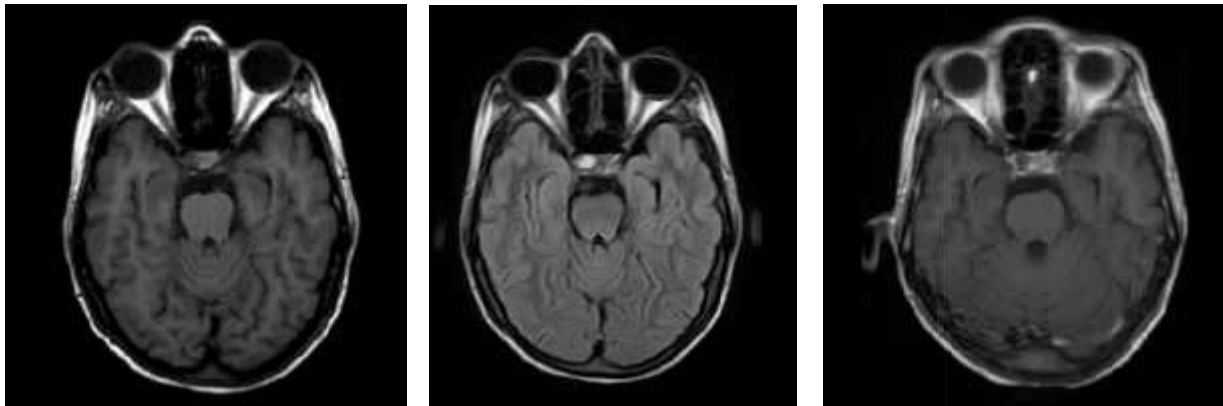


A

B

C

**PNS Rhabdomyosarcoma:** A) precontrast T1W sagittal and B) T2W coronal images shows a solid lesion in right maxillary sinus causing its expansion posterosuperiorly lesion is invading infrasellar and sella and superiorly eroding floor of orbit and invading into orbit, lesion is iso-hypointense on T1W and iso-hyperintense on T2W images. C) Post contrast T1W sagittal image show heterogenous enhancement of the lesion.



**Pituitary Colloid Cyst:** A) precontrast T1W and B) FLAIR axial images shows a well defined lesion in the pituitary gland on the right side, lesion is slightly hyperintense on T1W and hyperintense on FLAIR image. C) Post contrast T1W axial image, lesion is not showing any enhancement.

## CONCLUSION

MR imaging characteristics of the most common pediatric sellar & parasellar lesions like craniopharyngiomas and hypothalamic hamartoma were sufficiently distinct to allow them to be differentiated from each other and from most other entities. Other characteristics such as extrasellar versus intrasellar location, nature of contrast material enhancement, the presence of cystic components, and clinical findings permit further differentiation among the various other abnormalities. The superior resolution and multiplanar capacity of MR imaging best depicts the extent of sellar, parasellar and suprasellar lesions.

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