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Imaging and Histopathological Features of Pilocytic Astrocytoma Involving Various Locations of Central Nervous System– Series of Multiple Cases

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Abstract

Background: Pilocytic astrocytoma is low grade glial tumor which commonly occurs in pediatric population. It is rare in adult population. It is Grade 1 tumor according to 2016 world health organization classification and they generally have good prognosis.

Methodology: The cases of five patient were reviewed which were given the histopathological diagnosis of pilocytic astrocytoma, one patient was also diagnosed as the variant of pilocytic astrocytoma which is pilomyxoid astrocytoma. All of these tumors are involving different locations of central nervous system including cerebellum, fourth ventricle, suprasellar region and optic chiasm, cerebral hemisphere and brain stem in different patients.

Conclusion: Pilocytic astrocytoma has a wide spectrum of neuroradiological presentations. Besides its classical appearance as low-grade glioma, a more atypical presentation makes the diagnosis challenging. The best method to achieve the pre-operative diagnosis is the combination of morphological and non-morphological MR features as well as site base approach of this tumor.

Keywords: Pilocytic Astrocytoma; Magnetic Resonance Imaging; Pilomyxoid Astrocytoma; Intracranial Tumors; Low grade Glioma

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Introduction

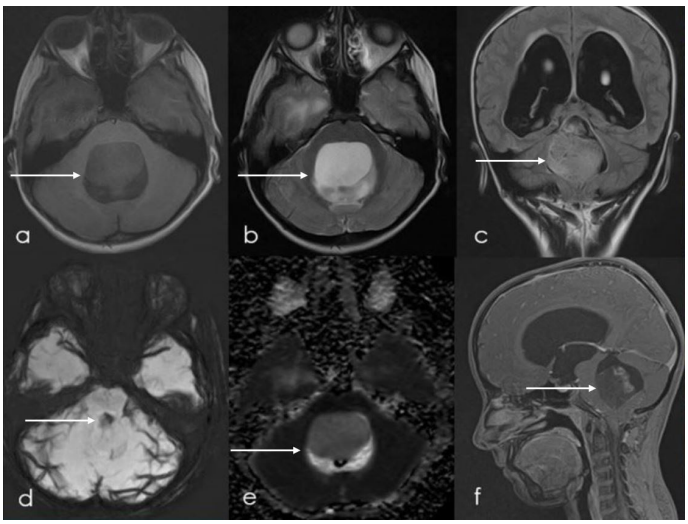
Pilocytic astrocytoma is the most common central nervous system (CNS) tumor in children and adolescence. It is the most common primary brain tumor in children between 5 and 14 years [1,2]. According to WHO classification, it is considered as low grade, WHO grade I CNS tumor [3]. We are going to discuss their clinical presentations, morphological features, histological features as well as their differential diagnosis with main focus on imaging feature.

Case 1

Pilomyxoid Astrocytoma of Fourth Ventricle

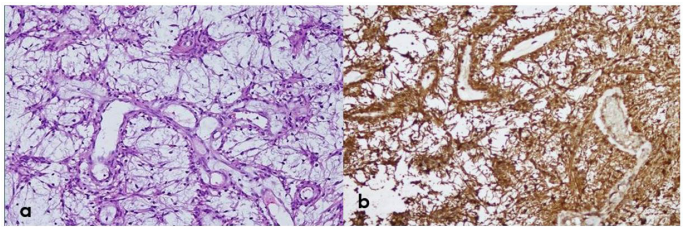
9-year-old male patient presented with complaint of disbalance, headache and vomiting for 3 months.

Magnetic Resonance Imaging (MRI) scan (Figure 1) was done which showed abnormal signal intensity space occupying lesion within floor of 4th ventricle with mass effect over brain stem and significant obstructive hydrocephalus. This lesion predominantly returns T2 hyperintense signals (Figure 1b) and hypointense signal on T1-weighted images (Figure 1a), without diffusion restriction (Figure 1e) or susceptibility focus (Figure 1d) and shows internal patchy post contrast enhancement (Figure 1f).



9-year-old male with pilomyxoid astrocytoma of fourth ventricle.
Findings: Well-defined solid space occupying lesion within fourth ventricle inseparable from its floor. It is hypointense on T1 (a), hyperintense on T2 (b) and FLAIR (c), without diffusion restriction (e), and shows internal patchy post contrast enhancement (f). It is not showing any focus of susceptibility on SWI sequence to suggest internal Hemorrhage (d). This lesion is causing significant proximal obstructive hydrocephalus and periventricular CSF seepage.
Figure 1: MRI Brain with contrast

The patient underwent posterior fossa craniotomy and debulking of space occupying lesion. Histopathology of this lesion was performed (Figure 2) which showed monomorphic population of bipolar cells with predominant angiocentric arrangement, and prominent myxoid background (Figure 2a). It was also showing diffuse, strong immunoreactivity for Glial fibrillary acidic protein (GFAP) immunostain (Figure 2b). These features are consistent with pilomyxoid astrocytoma.

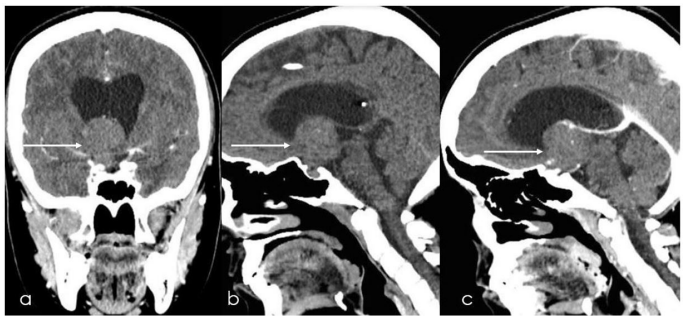


9-year-old male with pilomyxoid astrocytoma of fourth ventricle.
Findings: (a) Monochromatic population of bipolar cells with predominant angiocentric arrangement and prominent myxoid background (hematoxylin and eosin X 20). (b) Diffuse, strong immunoreactivity for GFAP immunostain. These features are consistent with pilomyxoid astrocytoma.
Figure 2: Microscopic examination

Case 2

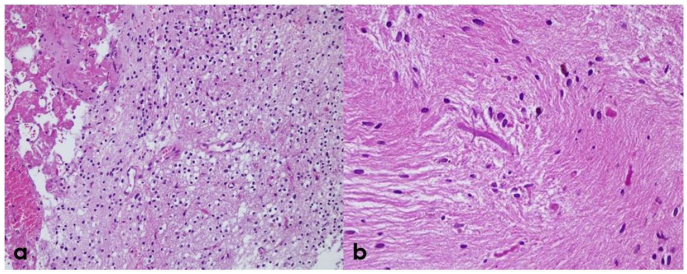
Pilocytic Astrocytoma of Suprasellar Region and Optic Chiasm

22-year-old female patient presented with complaints of drowsiness, vertigo and impaired vision for 4 months. Initially Computed tomography (CT) scan was performed (Figure 3) which showed isodense, non-enhancing lesion anterior to the third ventricle, bulging downwards towards the infundibular recess in the suprasellar area on the right side and resulting in obstructive hydrocephalus.



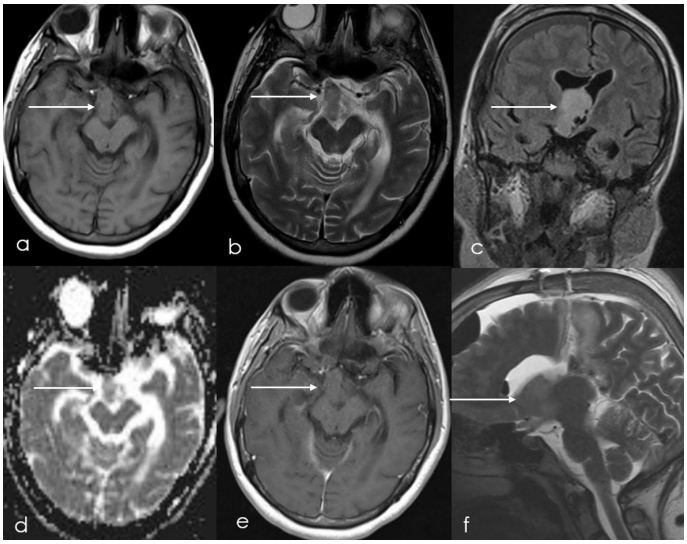
22-year-old female with suprasellar pilocytic astrocytoma.
Findings: Preoperative CT scan showing isodense non-enhancing lesion in suprasellar region resulting in obstructive hydrocephalus.
Figure 3: CT scan with contrast

After that patient underwent neuronavigation guided craniotomy, excision of the lesion. Histopathology was performed (Figure 4) which showed pseudo-oligodendroglial pattern with regular round cells, rounded nuclei and thin chicken-wire vasculature (Figure 4a). Bipolar cells were showing fibrillary processes with scattered Rosenthal fibers and eosinophilic granular bodies (Figure 4b). These features are consistent with pilocytic astrocytoma.



22-year-old female with suprasellar pilocytic astrocytoma.
Findings: Pseudo-oligodendroglial pattern with regular round cells, rounded nuclei, and thin chicken-wire vasculature (a). Bipolar cells showing fibrillary processes with scattered Rosenthal fibers and eosinophilic granular bodies (b). These features are consistent with pilocytic astrocytoma.
Figure 4: Microscopic examination

Approximately one week after the resection, the MRI brain was performed (Figure 5) which showed residual T1 isointense (Figure 5a) and slightly T2 hyperintense (Figure 5b) non-enhancing (Figure 5e) lesion in suprasellar cistern extending into third ventricle up to the Foramen of Monroe, this residual lesion was inseparable from optic chiasm. Unfortunately, patient was lost to follow up and further could not be assessed.

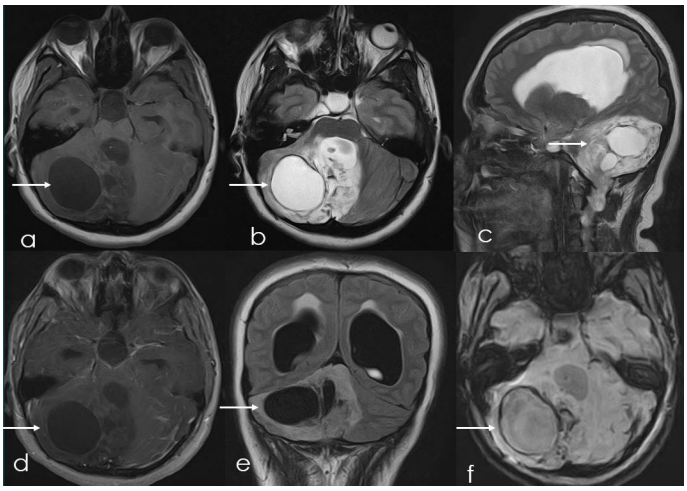


22-year-old female with suprasellar pilocytic astrocytoma.
Findings: Postsurgical resection MRI with residual T1 isointense (a) and slightly T2 hyperintense (b,f) lesion is identified in suprasellar cistern extending into third ventricle (f). This lesion is non-enhancing (e). This lesion is inseparable from the optic chiasm on right side (c).
Figure 5: MRI Brain with contrast

Case 3

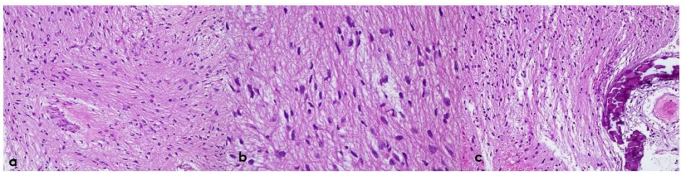
Pilocytic Astrocytoma of Cerebellum

23-year-old female patient presented with complaint of headache, vomiting and difficulty in walking. The Magnetic Resonance Imaging (MRI) scan was done (Figure 6) which showed large, heterogenous solid and cystic infiltrating mass lesion in posterior intracranial fossa, involving bilateral cerebellar hemispheres, predominantly on the right side as well as involving fourth ventricle, growing along the ventricular lining inferiorly. It showed heterogenous dropout signals on susceptibility weighted images (SWI) (Figure 6f), minimal patchy post contrast enhancement (Figure 6d) and no diffusion restriction. It was resulting in gross obstructive hydrocephalus with periventricular seepage (Figure 6c).



23-year-old female with pilocytic Astrocytoma of cerebellum.
Findings: Large, heterogenous infiltrating mass lesion in posterior intracranial fossa involving cerebellar hemispheres predominantly on the right side with involvement of fourth ventricle. It has solid (c) as well as cystic component (a). It shows minimal surrounding patchy post contrast enhancement (d). This lesion is resulting in gross obstructive hydrocephalus with periventricular seepage. It is showing heterogeneous signal dropout on SWI representing some hemorrhagic component.
Figure 6: MRI brain with contrast

Patient then underwent posterior fossa craniotomy and maximum safe resection of lesion. Histopathology results (Figure 7) showed bipolar cells arranged loosely in fibrillary background (Figure 7a) with long thin hair like processes (Figure 7b) as well as with focal calcification (Figure 7c). These features are consistent with pilocytic astrocytoma.



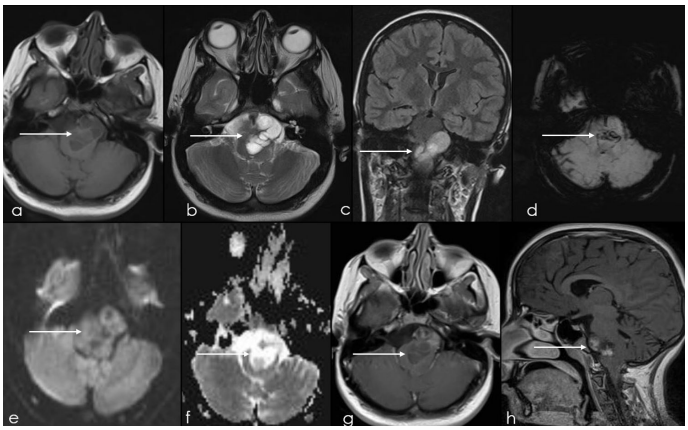
Findings: Bipolar cells arranged loosely in fibrillary background (a). Densely fibrillary areas composed of bipolar cells with long thin hair like processes (b). Bipolar cells arranged with fibrillary background with focal calcification (c). These features are consistent with pilocytic astrocytoma.
Figure 7: Microscopic examination

Case 4

Pilocytic Astrocytoma of Brain Stem

13-year-old female presented with dysarthria, dysphagia and ataxic gait. MRI brain was performed (Figure 8) which showed an irregular slightly lobulated lesion with few cystic ar-

eas noted at the pontomedullary junction. It was slightly effacing fourth ventricle without significant obstructive hydrocephalus. It was extending into the interpeduncular fossa slightly towards left of the midline. This lesion showed areas of inhomogeneous post contrast enhancement (Figure 8h), however no diffusion restriction was seen (Figure 8e). Few peripheral areas of signal dropout noted on SWI which could represent siderosis or calcification (Figure 8d). On histopathology it was proven to pilocytic astrocytoma.

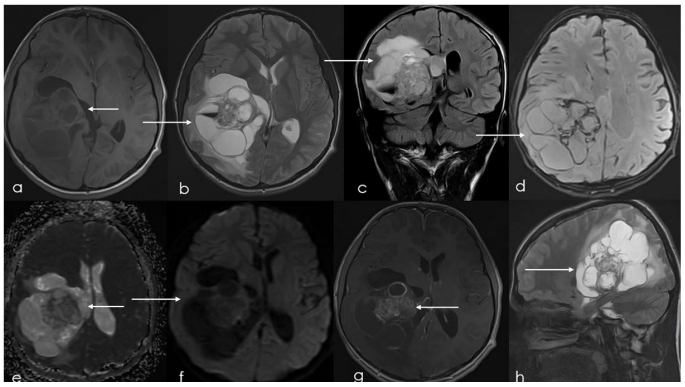


13-year-old female with pilocytic astrocytoma of brain stem.
Findings: Irregular slightly lobulated lesion with few cystic areas at pontomedullary junction. It is slightly effacing fourth ventricle without significant obstructive hydrocephalus. It is extending into the interpeduncular fossa slightly towards left of the midline. This lesion is showing areas of inhomogeneous post contrast enhancement (h), however no diffusion restriction is noted (e,f). Few peripheral areas of signal dropout noted on SWI (d), this may represent siderosis or calcification. On histopathology it was proven to pilocytic astrocytoma.
Figure 8: MRI brain with contrast

Case 5

Pilocytic Astrocytoma Of Cerebral Hemisphere

16 years male presented with headache, orbital pain and vertigo for 4 days. MRI brain was performed (Figure 9) which showed a large multicystic lesion in right cerebral hemisphere involving the parietal lobe and peritrigonal region. It was extending up to the superior cerebral peduncle. It was causing compression effect on third ventricle and occipital horn of right lateral ventricle. This lesion appeared hyperintense on T2 (Figure 9b), hypointense on T1 (Figure 9a) showing peripheral enhancement (Figure 9g). There was significant perilesional edema and focal midline shift. There were foci of signal dropout within this mass with some element of layering in the dependent portion suggesting hemorrhage (Figure 9d). Overall findings suggested cystic neoplastic lesion. Patient then underwent right temporoparietal Craniotomy with excision of this space occupying lesion. On histopathology it was proven to pilocytic astrocytoma



16 years male with pilocytic astrocytoma of cerebral hemisphere
Findings: Large multicystic lesion in right cerebral hemisphere involving the parietal lobe and peritrigonal region. It is causing compression effect on third ventricle and occipital horn of right lateral ventricle. This lesion appears hyperintense on T2 (b), hypointense on T1 (a) showing patchy enhancement of solid component (g). There is significant perilesional edema and focal midline shift. Foci of signal dropout noted within this mass on SWI (d) with some element of layering in the dependent portion suggesting hemorrhage within this mass. On histopathology it was proven to pilocytic astrocytoma.
Figure 9: MRI brain with contrast

Discussion

Demographics

Pilocytic astrocytoma is the most common central nervous system (CNS) tumor in children and adolescence and accounts for 15.5% of all cases. It is the most common primary brain tumor in children between 5 and 14 years [1,2]. Very rarely it occurs above the age of 30 and is the second most common in children between 0–4 and 15–19 years of age. According to WHO classification, it is considered as low grade, WHO grade I CNS tumor [3].

Location

Pilocytic astrocytoma can arise from anywhere though out central nervous cistern, however it most commonly arises from cerebellum, with second most common location of optic chiasm. Other less common location includes brain stem, hypothalamus, cerebral hemispheres and spinal cord [4]. In pediatric patients it is commonly seen in infratentorial region while in adults it is usually supratentorial. Pilocytic astrocytoma involving optic nerve and optic chiasm are also called optic pathway glioma (OPG), these are relatively rare and accounts for 2 to 5% of cases. 30% of OPG are associated with neurofibromatosis 1 (NF1) [5].

Clinical Presentation

Clinical presentation is variable and depends upon the site of involvement of tumor. When the tumor arises from cerebellum, it usually causes obstruction of fourth ventricle, resulting in intracranial hypertension. Symptoms includes headache, nausea/vomiting and cerebellar symptoms such as gait disturbance, slurring of speech and symptoms due to involvement of optic pathway such as blurred vision, diplopia, etc.

Table 1: Summary table for pilocytic Astrocytoma

CLINICAL PRESENTATION	**variable and depends upon the site of involvement of tumor**
Gender ratio	No recognized gender predisposition
Age predilection	children and adolescence
Location	Most commonly arises from cerebellum, second most common location is optic chiasm. Other less common location includes brain stem, hypothalamus, cerebral hemispheres and spinal cord
Known association	Strong association with neurofibromatosis type 1 (NF1), especially pilocytic astrocytoma involving optic nerves and chiasm
Imaging features	• Well-defined cystic lesion with solid some component. • Smooth margins with enhancing cystic wall. • Enhancing solid component are isointense to hypointense on T1 and hyperintense on T2/FLAIR images. • Very little or no perilesional edema. • Solid component may also show some calcification.
Histopathological Features	Bipolar cells with fibrillary processes, scattered Rosenthal fibers and eosinophilic granular bodies.
Prognosis	Good overall survival in pediatric patients, 10-year overall survival of more than 90% Survival rate decreases with increasing patient age. At the age of 40, the 10-year survival rate is closer to 70%.

Table 2: Differential diagnosis table for pilocytic astrocytoma

	Age predilection	**Typical Location**	**Clinical presentation**	**Differentiating Imaging Feature**
HEMANGIOBLASTOMA	adults	cerebellar hemisphere	Headaches Symptoms of raised intracranial pressure Cerebellar dysfunction:	Wall of cysts usually does not enhance. Usually shows smaller mural nodule with angiographic contrast blush.
MEDULLOBLASTOMA	younger age group (2-6 years of age).	vermis and roof of the fourth ventricle	Rapid clinical onset. Signs and symptoms of raised intracranial pressure	More solid in appearance with larger heterogeneously enhancing mass.
EPENDYMOMA	No recognized age predilection	the floor of the 4^{th} ventricle	Signs and symptoms of raised intracranial pressure	Usually solid in nature. Tends to fill the fourth ventricle and protrude through foramen of Luschka and foramina of Magendie
CEREBELLAR ABSCESS	Can occur in any age group	Eccentric cerebellar hemisphere	Symptoms of sepsis as well as symptoms of raised intracranial pressure, seizures and focal neurological deficits	, Well defined peripheral enhancing lesion with central restricted diffusion, associated daughter lesion and disproportionate edema is the usual presentation
CRANIOPHARYNGIOMA	Bimodal distribution (5-15) and above 40 years	Suprachiasmatic region	Headaches Visual symptoms Hormonal imbalances	Usually, mixed solid and cystic in nature with enhancing solid component, (suprachiasmatic pilocytic astrocytoma are usually solid).

Imaging Features

Typical imaging features of pilocytic astrocytoma consist of well-defined cystic lesion with solid some component. The cystic component shows smooth margins with enhancing cystic wall in majority of cases. The solid portion is usually isointense to hypointense on T1 and hyperintense on T2 FLAIR images and shows post contrast enhancement [6]. Due to slow growing nature of these lesions, usually there is very little or no perilesional edema. Solid component may also show some calcification which are easily appreciated on CT and T2*/SWI images.

Although in most cases the pilocytic astrocytoma usually has cystic component but it can give atypical presentation in which pilocytic astrocytoma is completely solid without any cystic component, these are usually supratentorial in location and are usually seen in adults [7] as seen in the second case.

Histopathological Features

The typical histopathological features of pilocytic astrocytoma includes bipolar cells with fibrillary processes, scattered Rosenthal fibers and eosinophilic granular bodies. Hyalinization of blood vessels is another feature of Pilocytic Astrocytoma.

Pilomyxoid astrocytoma is a variant of pilocytic astrocytoma which is characterized by monochromatic population of bipolar cells with predominant angiocentric arrangement and prominent myxoid background, they show diffuse and strong immunoreactivity for GFAP immunostain. These tumors are more aggressive than pilocytic astrocytoma showing local recurrence and cerebrospinal fluid (CSF) seepage [8].

Treatment and Prognosis

The overall survival of patients with pilocytic astrocytoma is good in pediatric patients, with 10-year overall survival of more than 90% [2]. The survival rate decreases as the age of patient increases at the time of diagnosis. In adults, at the age of 40 years, the 10-year survival rate is closer to 70% [2]. The Overall survival rate Also depends upon the site of tumor. Tumors that can be completely resected are usually curative have better overall survival rate, for example tumors involving cerebellum and cortex than those in the brainstem and optic pathway.

Differential Diagnosis

Some of the differentials that should be considered while giving the diagnosis of pilocytic astrocytoma includes:

Hemangioblastoma: It is usually seen in adults. If it occurs in children, then it is usually associated with von Hippel-Lindau disease. Compare to pilocytic astrocytoma, the wall of cysts usually does not enhance. It usually shows smaller mural nodule with angiographic contrast blush [9].

Medulloblastoma: Most commonly occurs in younger age group (2-6 years of age). Typically arise from midline (especially vermis and roof of the fourth ventricle) while pilocytic astrocytoma usually arises from cerebellar hemisphere. They are more solid in appearance with larger heterogeneously enhancing mass [10].

Ependymoma: No recognized gender and age predilection. Usually arises from the floor of the 4th ventricle. Usually solid in nature. Tends to fill the fourth ventricle and protrude through foramen of Luschka and foramina of Magendie [11].

Cerebellar Abscess: Usually presents with symptoms of sepsis as well as symptoms of raised intracranial pressure, seizures and focal neurological deficits, well defined peripheral enhancing lesion with central restricted diffusion, associated daughter lesion and disproportionate edema is the usual presentation.

Craniopharyngioma: If the location of pilocytic astrocytoma is suprachiasmatic region then possibility of craniopharyngioma should be considered. It has bimodial distribution, first peak occurs from age of 5 to 15 and second peak occurs in adults above 40 years [12]. They are usually of mixed solid and cystic in nature with enhancing solid component. While suprachiasmatic pilocytic astrocytoma are usually solid.

Teaching Point: Pilocytic astrocytoma has a wide spectrum of neuroradiological presentations. Besides its classical appearance as low-grade glioma, a more atypical presentation makes the diagnosis challenging. The best method to achieve the pre-operative diagnosis is the combination of morphological and non-morphological MR features as well as site base approach of this tumor.

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