Craniopharyngioma

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Craniopharyngioma is an epithelial tumor, classified as WHO grade 1. Although histologically benign, craniopharyngiomas are aggressive neoplasms, and recurrences are common because complete surgical resection is difficult. They represent approximately 5 % of all intracranial tumors. Craniopharyngiomas are derived from remnants of the Rathke pouch and can occur anywhere along the course of the craniopharyngeal duct, from the nasopharynx to the third ventricle. They are usually sporadic. Rare cases of adamantinous craniopharyngiomas have been associated with Gardner syndrome, a variant of familial adenomatous polyposis. Craniopharyngiomas are characterized by a bimodal age distribution, with a main peak in children (5–14 years) and a second peak in adults (fifth to seventh decade). They most commonly involve the suprasellar and sellar area (75 %), where they are either anterior or posterior to the optic chiasm. In fewer cases, their location is purely suprasellar (20 %) or entirely intrasellar (5 %). There are two histologic subtypes: the adamantinous type, most common form, typically seen in children and adolescents; and the papillary type, seen almost exclusively in adults. Adamantinous craniopharyngiomas are usually lobulated and cystic tumors, containing a dark greenish brown fluid. These cysts contain variable amounts of cholesterol, keratin, protein, methemoglobin, and necrotic debris, which account for their variable appearance on MRI. They are usually associated with calcification and often demonstrate local brain invasion, being adherent to adjacent vessels and nerves. Papillary craniopharyngiomas are generally well-circumscribed, predominantly solid tumors. The solid component may rarely contain small cystic areas or calcifications. The third ventricle is a common location of this subtype. Clinical symptoms are variable, on account of the variable location. Headache, visual field defects, decreased visual acuity, and hormone disturbance are the common clinical symptoms in the suprasellar location. The most common endocrine dysfunctions encountered in children are growth retardation and delayed or precocious puberty. Global hypopituitarism, hyperprolactinemia, and diabetes insipidus are the other endocrine disorders encountered. Less commonly, patients may present with cognitive impairment or personality change.

Adamantinous craniopharyngiomas classically occur in childhood or adolescence and typically present with three components: solid, cystic, and calcified portions, which occupy the suprasellar cistern (Figs. 22.1, 22.2, 22.3, and 22.4). The cystic component is single or multiple, and usually hyperintense on T1, T2, and FLAIR weighted images because of the presence of proteinaceous liquid. Less commonly, a CSF-like signal pattern can be seen in huge craniopharyngiomas with a thick wall enhancement. The solid component has variable signal intensities and shows contrast enhancement. Small necrotic areas account for its inhomogeneous signal before and after contrast administration. Calcifications

J.-F. Bonneville et al., *MRI of the Pituitary Gland*, DOI 10.1007/978-3-319-29043-0_22

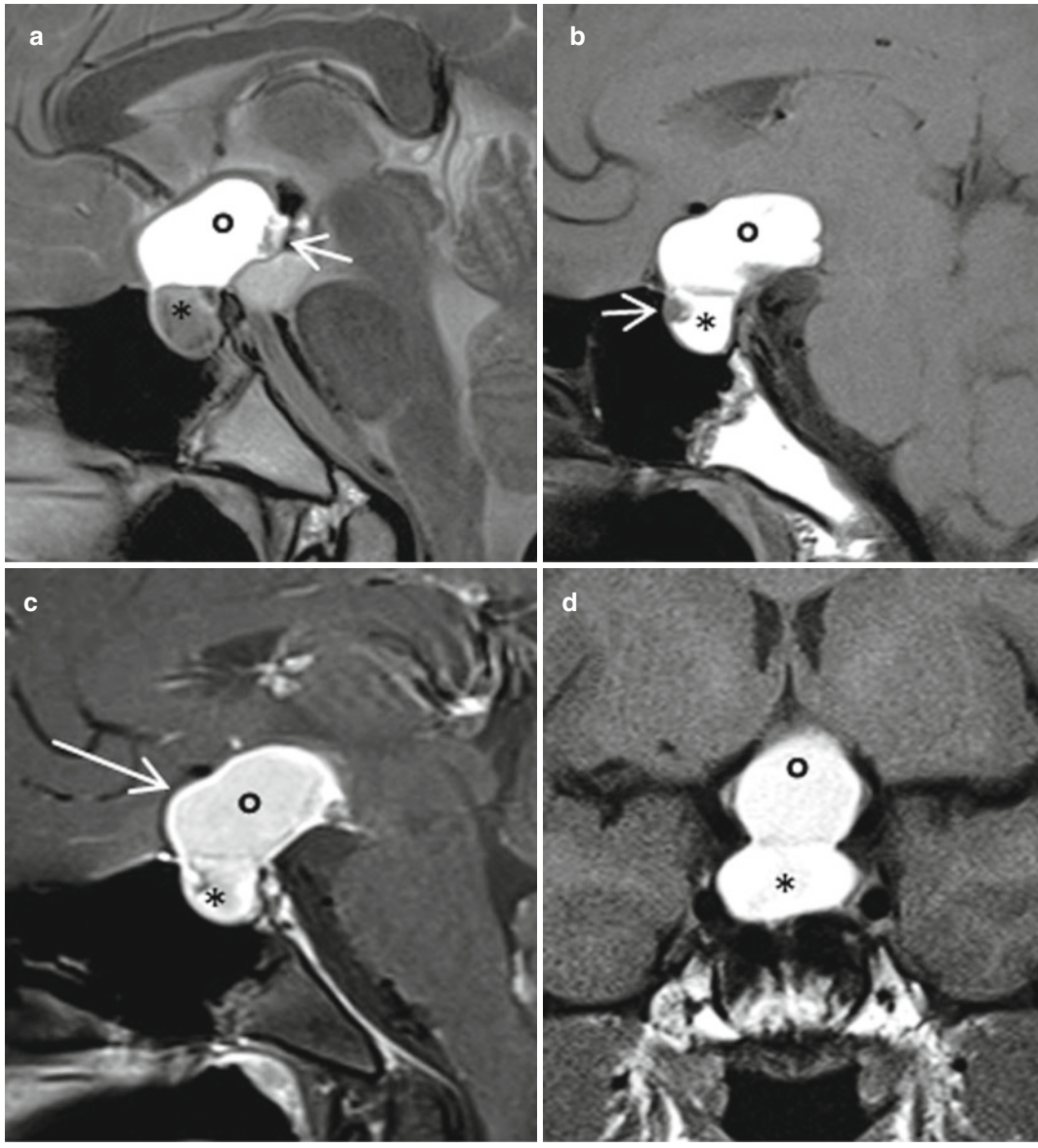
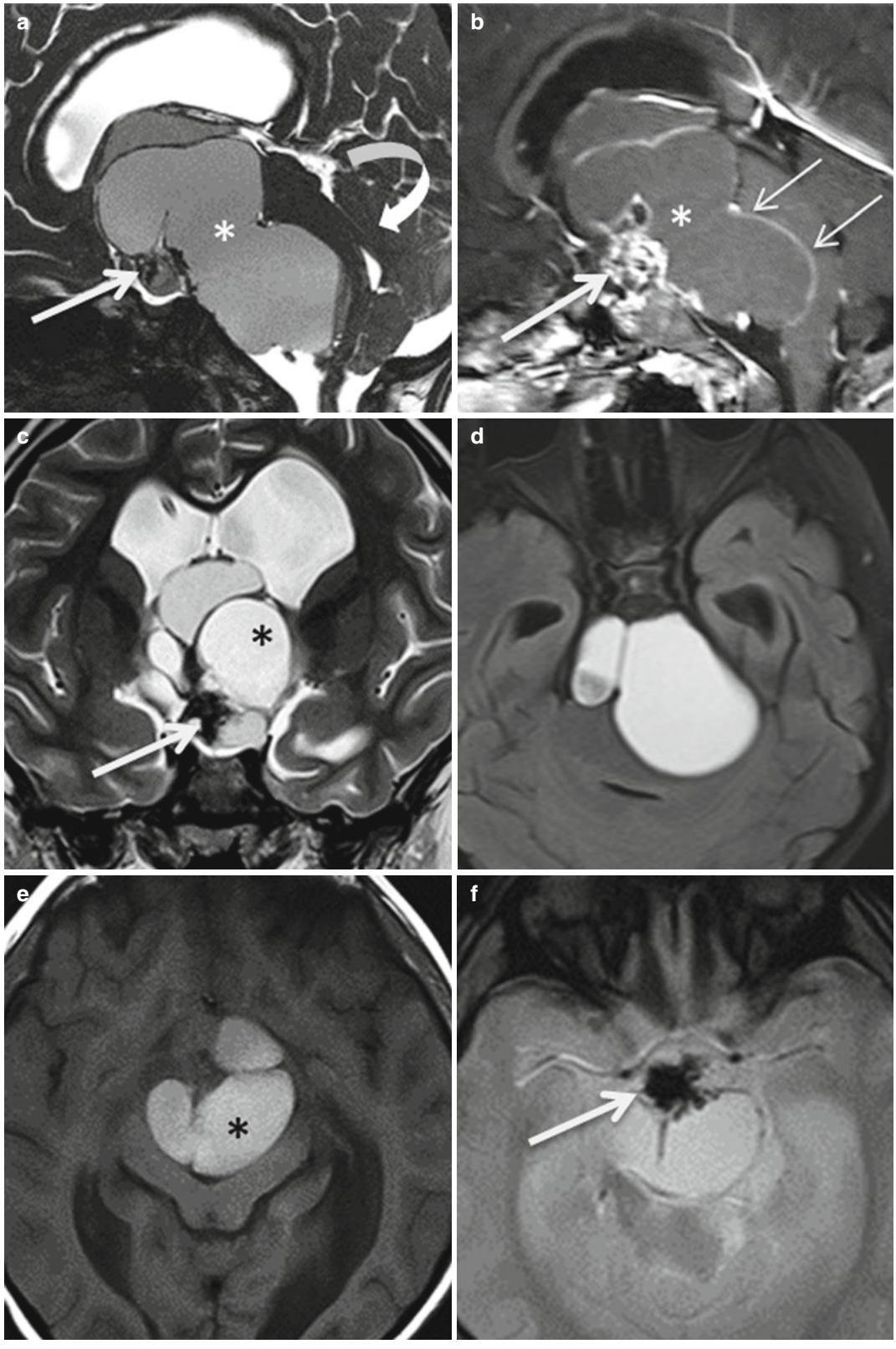


Fig. 22.1 A 16-year-old girl with headaches, secondary amenorrhea, and left hemianopsia. Intra- and suprasellar cystic adamantinous craniopharyngioma. (**a**–**c**) Sagittal T2, T1 and CE T1 fat-saturated WIs. Intrasellar heterogeneous cyst without sellar change (*asterisk*). Suprasellar hyperintense cyst (*circle*) with thin peripheral enhancement (*long arrow*). Nodular calcifications in both portions (*short arrows*). (**d**) Coronal T1WI. Superior extension with optic chiasm compression

Fig. 22.2 A 7-year-old boy with headaches and vomiting. Suprasellar adamantinomatous craniopharyngioma. (**a**, **b**) Sagittal heavily T2 and CE T1 WIs. (**c**, **d**) Coronal T2 and axial FLAIR WIs. Suprasellar multiloculated mixed mass with varying mainly bright signal intensities (*asterisk*) and solid calcified component heterogeneously enhanced (*thick arrow*). Cyst wall enhancement (*thin arrows*). Note posterior fossa extension with brainstem and cerebellar compression (*curved arrow*). Hydrocephalus by interventricular foramen obstruction. (**e**, **f**) Axial T1 and T2*WIs. Spontaneous hyperintense cyst signal due to proteinaceous content (*asterisk*). Markedly hypointense calcifications (*thick arrow*)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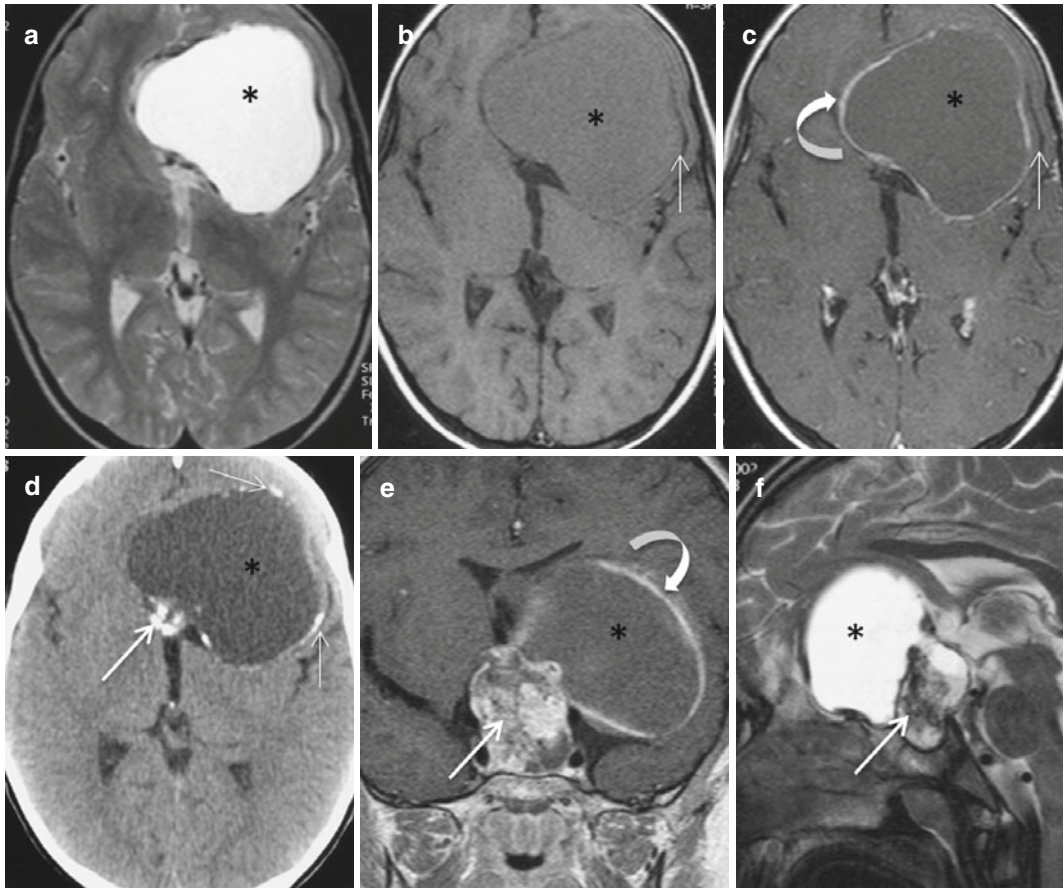

Fig. 22.3 An 8-year-old boy with afebrile seizures, bilateral optic atrophy, reduced visual acuity, and left temporal hemianopia. Intra- and suprasellar adamantinomatous craniopharyngioma. (**a**–**d**) Axial T2, T1, CE T1 WIs and CT scan. (**e**) Coronal CE T1WI. (**f**) Sagittal T2WI. Intra- and suprasellar mixed mass with cystic (*asterisk*) and calcified tissular component (*arrow*). The cyst wall is T1 hypointense and hyperdense with discontinuous calcifications (*thin arrows*). Heterogeneous enhancement of the solid component (*arrows*). Cyst wall enhancement (*curved arrows*)

are common (90 % of cases) and are better demonstrated by T2* sequence. However, their confirmation by a CT scan is sometimes necessary. Lipid resonances are usually detected in proton MR spectroscopy. Huge craniopharyngiomas can extend in all directions. The sella is often enlarged and may be eroded.

Papillary craniopharyngiomas usually appear as a solid or mixed, predominantly solid and cystic, spherical tumor in the suprasellar region (Fig. 22.4). They are characterized by a more upward growth toward the third ventricle. The solid parts classically show isointense signal intensity on T1 and T2 WI, with a hypointense signal on DWI with increased apparent diffusion coefficient reflecting the low-grade character. They present homogeneous or reticular enhancement because of their small necrotic areas. They less frequently contain calcifications. Nevertheless, their imaging features are nonspecific, and differential diagnosis is based on the age of the patient and the location of the tumor. Surgical removal is easier in this subtype than in the adamantinous type. Peritumoral edema spreading along the optic tracts was initially considered to be specific to craniopharyngiomas. Similar optic tract edema has been described since then in other parasellar tumors such as ade-

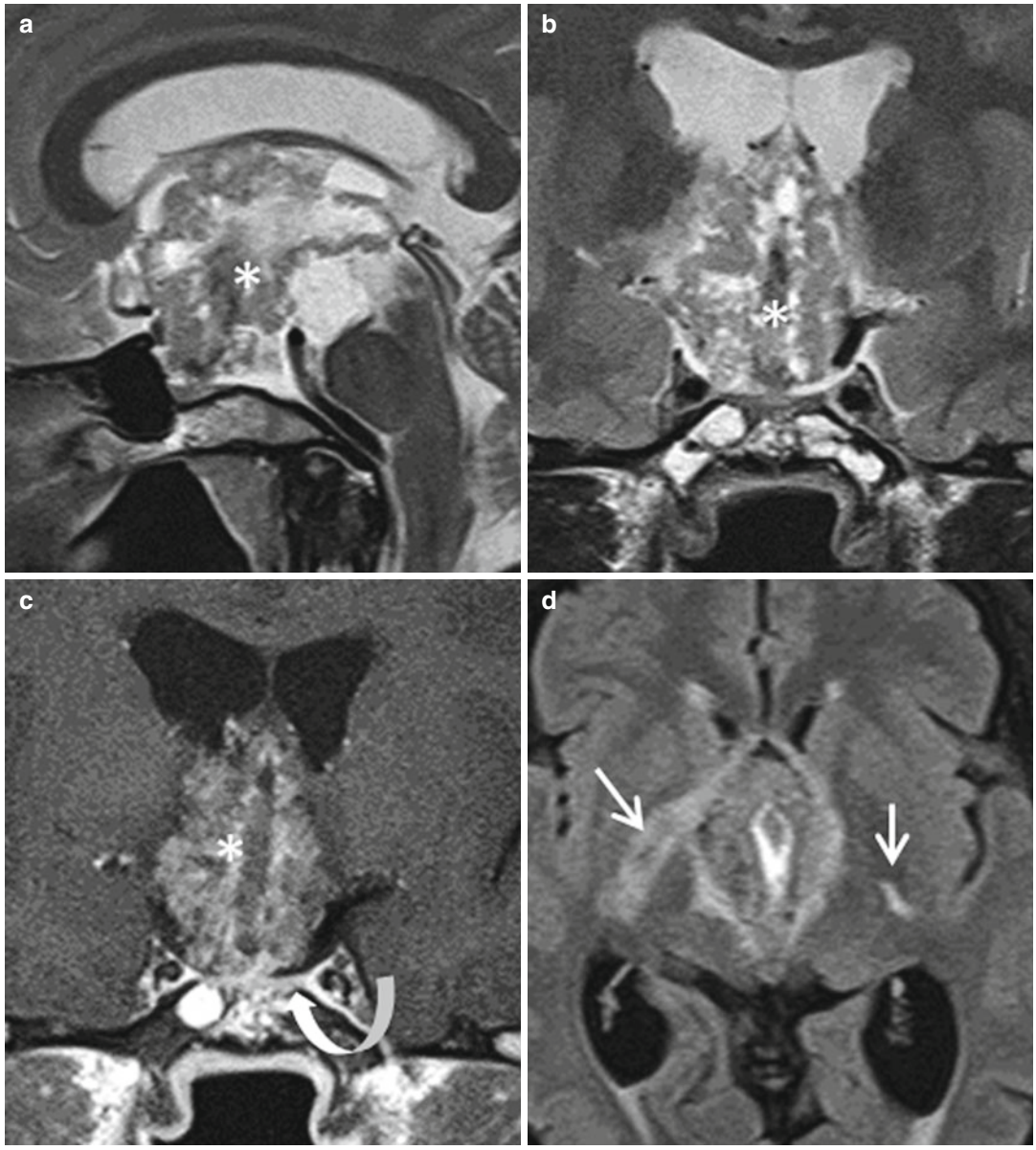

Fig. 22.4 A 28-year-old man with hypogonadotropic hypogonadism and diabetes insipidus. Suprasellar papillary craniopharyngioma. (**a**, **b**) Sagittal and coronal T2WIs. Mixed predominantly solid and cystic suprasellar mass with iso- to hyperintense signal (*asterisk*). (**c**) Coronal CE T1WI. Heterogeneous enhancement (*asterisk*), flattened pituitary gland (*curved arrow*). (**d**) Axial FLAIR WI. Bilateral edema along optic tracts (*arrows*)

nomas, germ cell tumors, gliomas, meningiomas, and lymphomas. This could be related to distension of perivascular spaces, which represent a drainage route of interstitial fluid into the subarachnoid space, their outflow pathway into the subarachnoid spaces being blocked by suprasellar tumor (Figs. 22.4 and 22.5). Occasionally, craniopharyngiomas are located in the third ventricle (Fig. 22.6). Rare locations include posterior fossa, sphenoid bone, and nasopharynx. Distant spread of craniopharyngioma is a rare complication that occurs independently of the subtype. Locations include cerebral hemispheres, posterior fossa, brainstem, basal ganglia, and lumbar

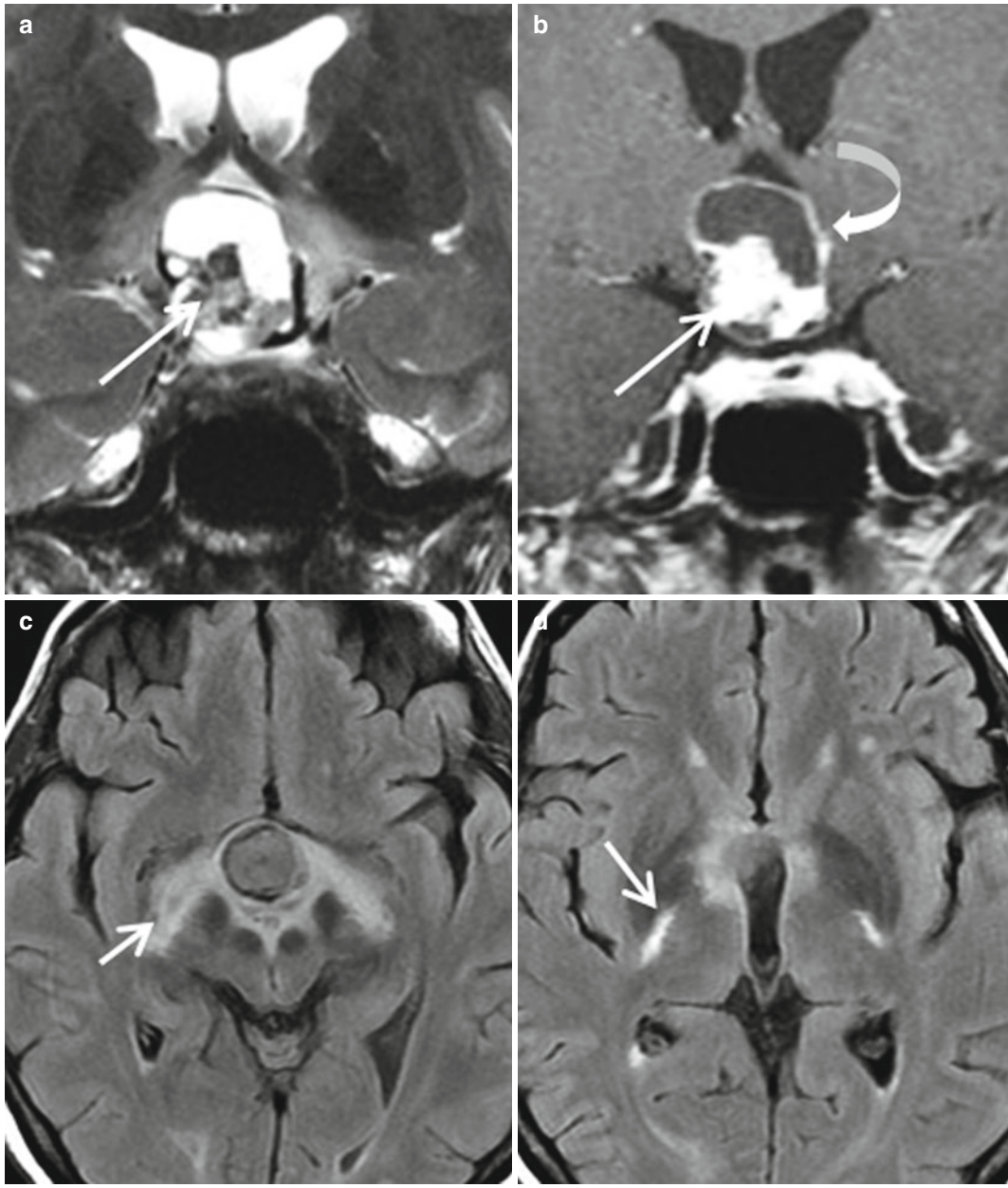

Fig. 22.5 A 22-year-old woman with reduced visual acuity. Suprasellar adamantinous craniopharyngioma. (**a**, **b**) Coronal T2 and CE T1WIs. Suprasellar mass with cystic component enhancing peripherally (*curved arrow*) and solid component enhancing heterogeneously (*long arrow*). (**c**, **d**) Axial FLAIR WI. Bilateral marked edema along the optic tracts (*short arrows*)

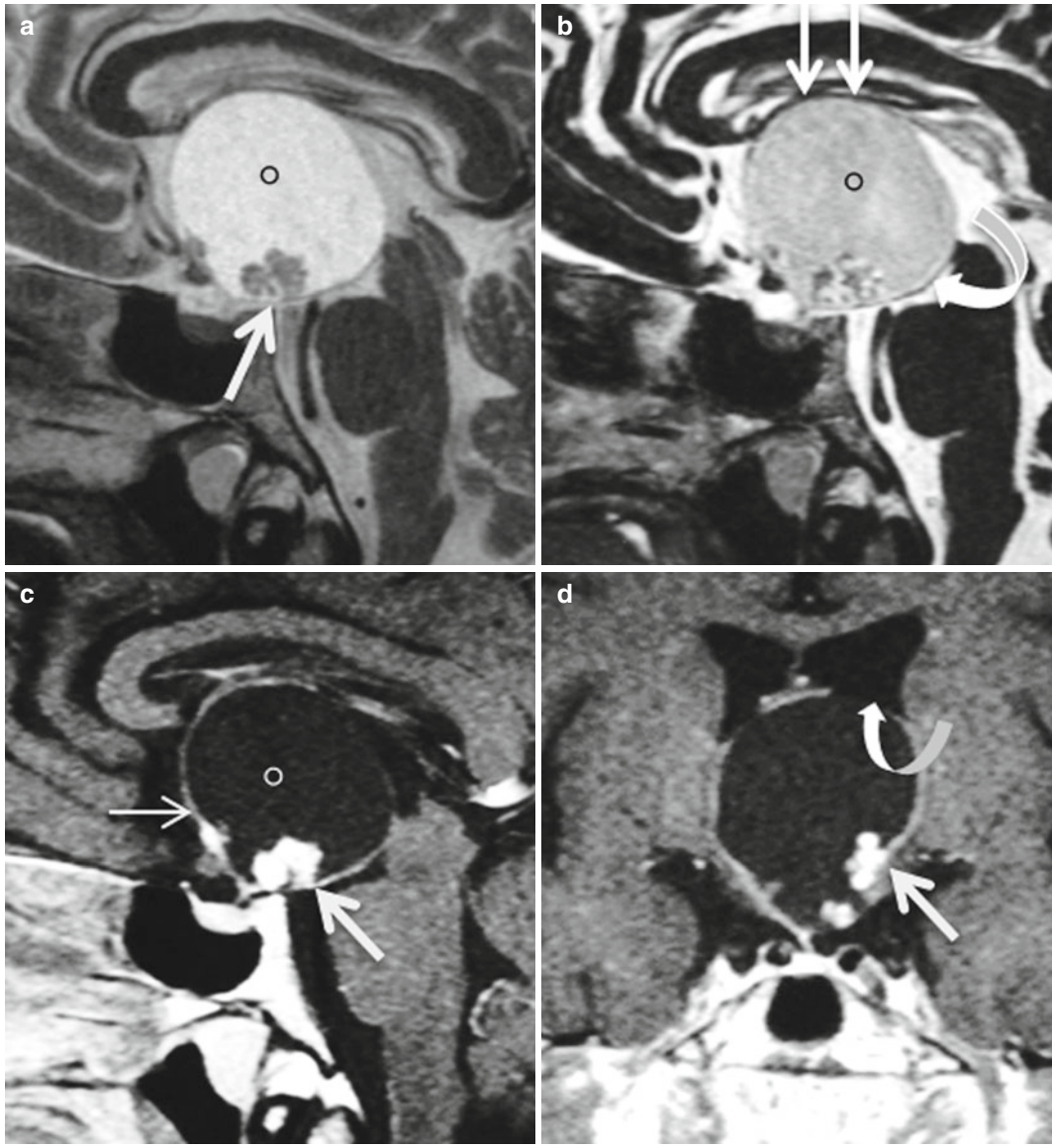

Fig. 22.6 A 54-year-old man with headaches, visual disturbance, difficulty in walking, and memory disorder. Third ventricular craniopharyngioma. (**a**–**c**) Sagittal T2, heavily T2, and CE T1 WIs. Mixed cystic (*circle*) and tissular (*thick arrow*) mass within the third ventricle. Tissular portion (*thick arrow*) and thin wall cyst (*thin arrow*) enhancement. Note the downward bowing of the floor of the third ventricle by the mass (*curved arrow*) and fornix displacement (*double thick arrow*). (**d**) Coronal CE T1WI. Interventricular foramen obstruction (*curved arrow*). Note normal pituitary gland

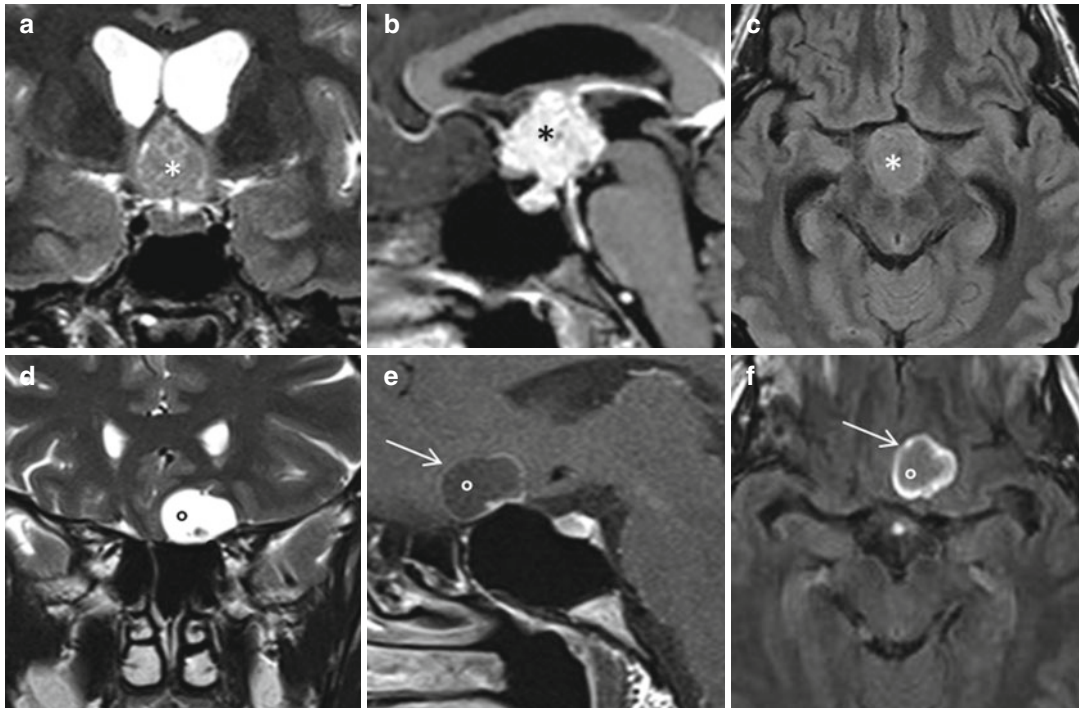

Fig. 22.7 A 43-year-old woman with headaches. Suprasellar papillary craniopharyngioma with recurrence 1 year after surgery away from suprasellar region, along the surgery path. (**a**) Coronal T2WI. Isointense suprasellar mass (*asterisk*). (**b**) Sagittal CE T1WI. Reticular diffuse enhancement (*asterisk*). (**c**) Axial FLAIR WI. Homogeneous isointense signal (*asterisk*). One year after complete surgical resection: (**d**) Coronal T2WI. Extra-axial left parasagittal and presellar hyperintense cystic mass (*circle*). (**e**) Sagittal CE T1WI. Peripheral enhancement (*thin arrow*) sparing the central area (*circle*). No suprasellar tumor relapse. (**f**) Axial FLAIR WI. Hyperintense cyst wall (*thin arrow*) and hypointense central area (*circle*)

spine. The mechanism is poorly understood, but most cases are thought to result from spread along the surgical path (Fig. 22.7). Metastatic leptomeningeal craniopharyngiomas have been reported, as a result of dissemination along CSF pathways (Fig. 22.8). Craniopharyngiomas with multiple recurrences and malignant transformations have been reported. The key factor associated with craniopharyngioma recurrence is the quality of surgical resection. Adhesion or encasement of adjacent cranial nerves or vessels (Fig. 22.7) significantly increases surgical morbidity. Differential diagnosis includes Rathke cleft cyst (Fig. 22.9), pituitary adenoma (Fig. 22.10), dermoid cyst, epidermoid cyst, teratoma, and pilomyxoid astrocytoma. Treatment usually consists of surgical resection with or without adjuvant radiation therapy.

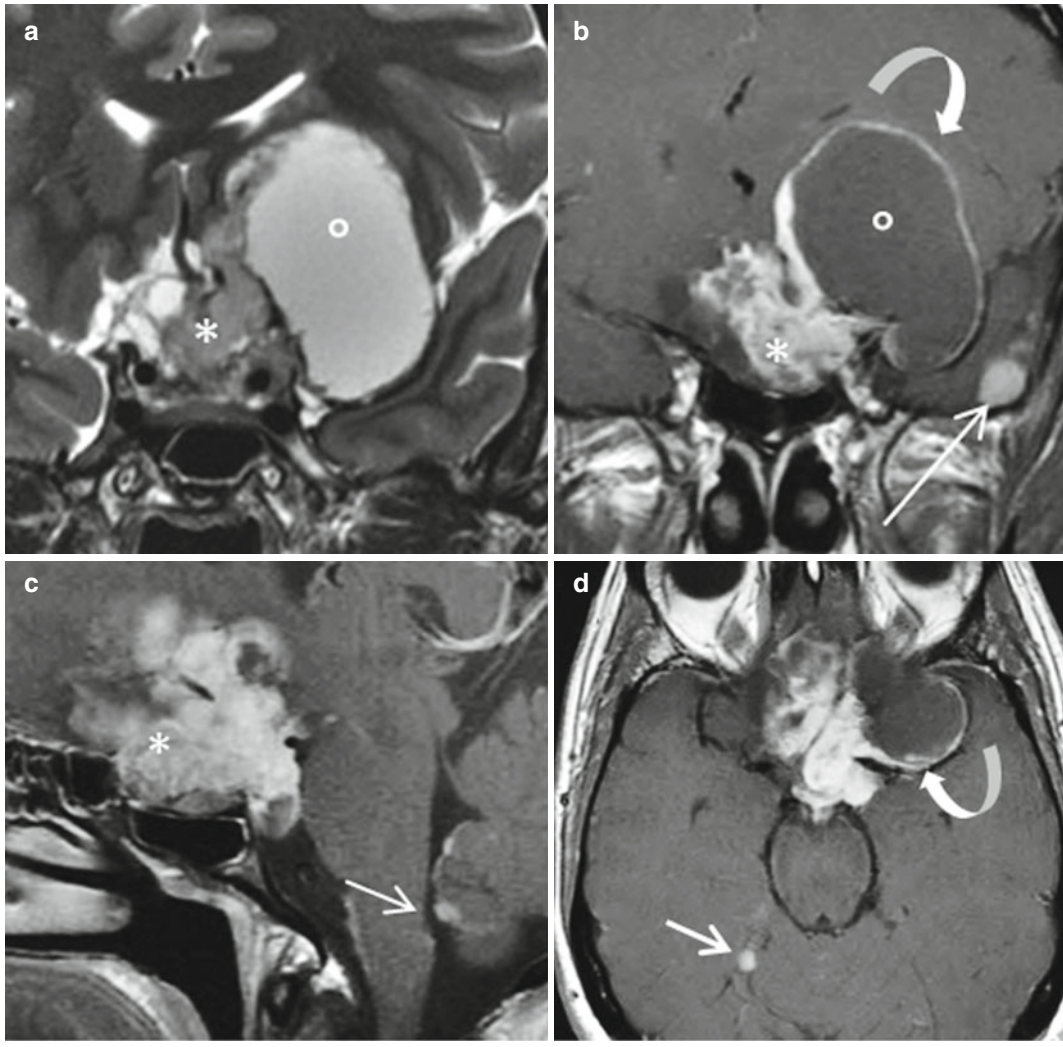

Fig. 22.8 A 32-year-old woman with headaches. Intra- and suprasellar adamantinous craniopharyngioma with concomitant leptomeningeal metastasis. (**a**, **b**) Coronal T2 and CE T1 WIs. Solid heterogeneous enhanced portion (*asterisk*), cystic portion (*circle*) with ring enhancement (*curved arrow*). Extra-axial left temporal nodular enhancement (*long arrow*). (**c**, **d**) Sagittal and axial CE T1WIs. Leptomeningeal enhancement in posterior fossa involving medulla and inferior roof of fourth ventricle (*short arrow*). Nodular enhancement in tentorium cerebelli (*thick arrow*). Note the encasement of the left middle cerebral artery

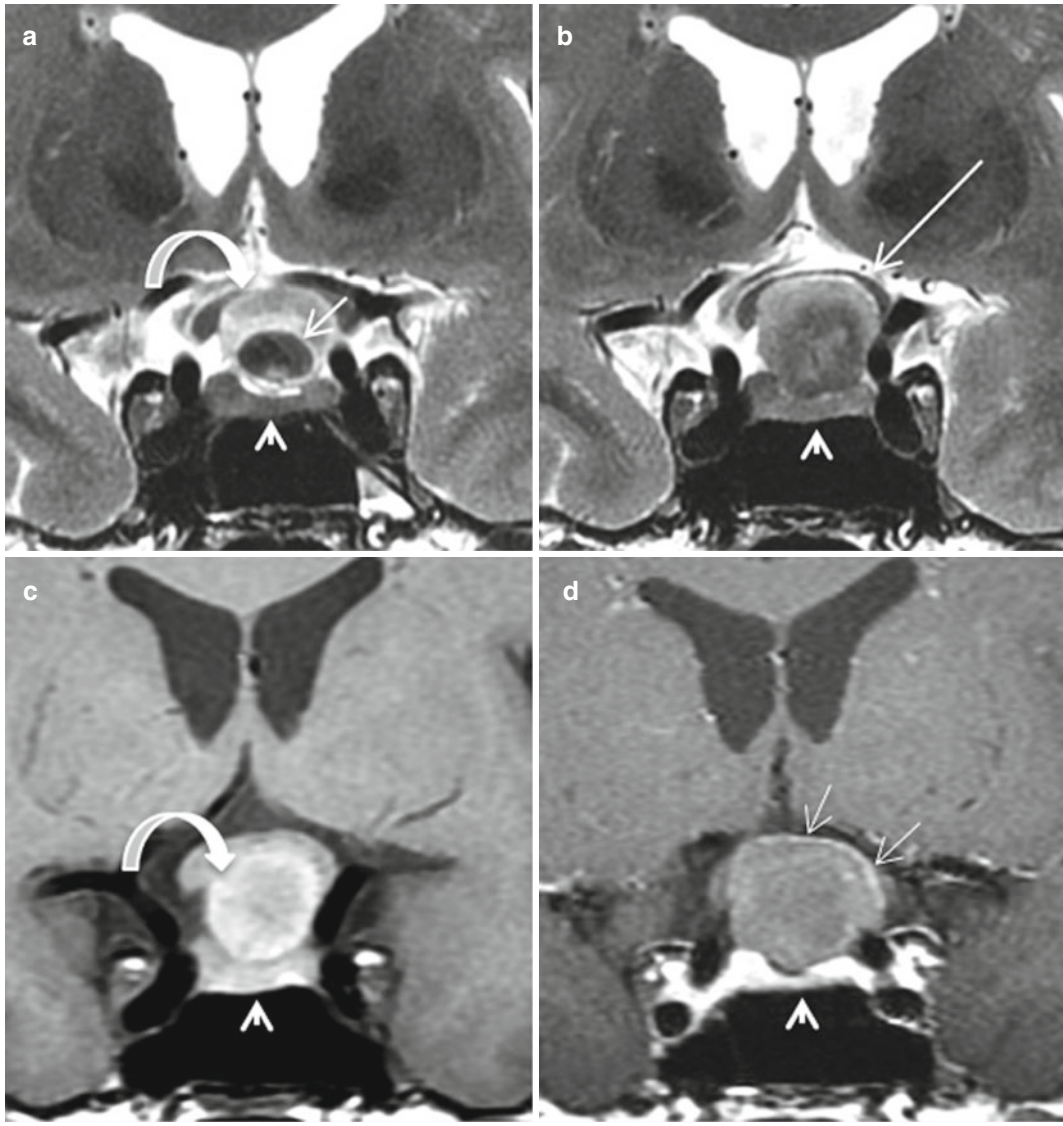

Fig. 22.9 A 57-year-old woman with decreased visual acuity. Suprasellar adamantinous craniopharyngioma mimicking a Rathke cleft cyst. (**a**, **b**) Coronal T2WIs. (**c**, **d**) Coronal T1 and CE T1 WIs. Round suprasellar mass with heterogeneous slightly T2 and T1 hyperintense content (*curved arrow*) and central T2 hypointense nodule (*thin arrow*). The mass is located above the pituitary gland (*short arrow*). Upward displacement and thinning of the optic chiasm (*long arrow*). Peripheral enhancement (*double arrow*) spearing the central area. Homogeneous enhancement of the pituitary gland (*short arrow*)

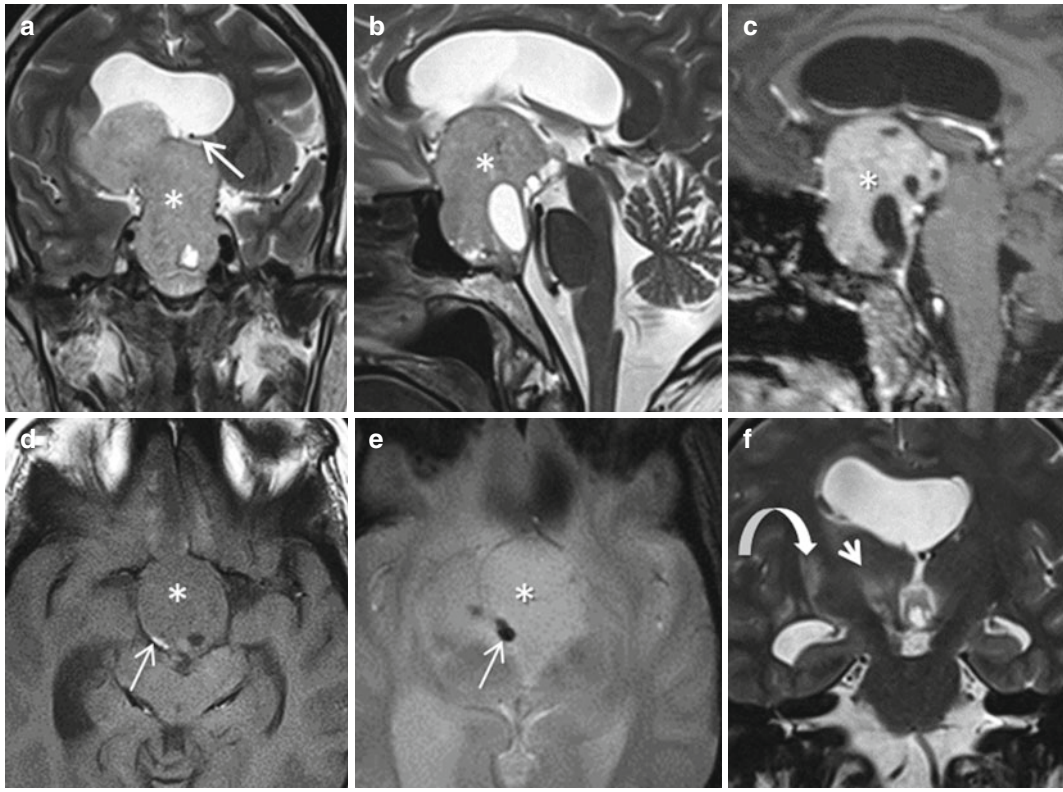

Fig. 22.10 A 54-year-old woman with headaches, visual disturbance, and left hemiparesis gradually increasing for 2 years. Intra- and suprasellar papillary craniopharyngioma mimicking a pituitary macroadenoma. (**a**, **b**) Coronal and sagittal T2WIs. Solid heterogeneous mass (*asterisk*) with hyperintense cystic areas and superior extension giving a figure-of-eight configuration. Hydrocephalus by obstruction of interventricular foramen (*thick arrow*). (**c**) Sagittal CE T1WI. Enhancement of the mass sparing intralesional cyst components (*asterisk*). (**d**, **e**) Axial T1 and T2* WIs. The mass is isointense on T1 with peripheral calcification (*thin arrow*). (**f**) Coronal T2WI. transependymal edema extending to right internal capsule (*curved arrow*). Edema along the optic tracts (*short arrow*)

Further Reading

Choi SH, Kwon BJ, Na DG et al (2007) Pituitary adenoma, craniopharyngioma, and Rathke cleft cyst involving both intrasellar and suprasellar regions: differentiation using MRI. Clin Radiol 62:453–462

Lee HJ, Wu CC, Wu HM et al (2015) Pretreatment diagnosis of suprasellar papillary craniopharyngioma and germ cell tumors of adult patients. Am J Neuroradiol 36:508–517

Saeki N, Nagai Y, Matsuura I et al (2004) Histologic characteristics of normal perivascular spaces along the optic tract: new pathogenetic mechanism for edema in tumors in the pituitary region. Am J Neuroradiol 25:1218–1222