Rathke Cleft Cyst: Asymptomatic

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Rathke cleft cysts (RCC) are the most frequent pituitary lesions revealed on MRI. They are intra-/suprasellar lesions believed to derivate from remnants of the Rathke pouch; therefore, one major characteristic is their strict and quite constant location on the midline. Nevertheless, RCC located off-midline may be encountered. They can originate either intra- or suprasellarly and their precise origin is best recognized if small, i.e., less than 10 mm in size. The displacement of the normal pituitary gland depends on the precise origin of the cyst. If initially intrasellar, RCCs are located on the midline, just in front and in close contact with the posterior lobe. Axial T1WI is in this case the most informative (Figs. 19.1 and 19.2), while a small hyperproteinic RCC may be mistaken for a large posterior lobe on sagittal view (Fig. 19.3). If initially suprasellar, RCCs originate from the pars tuberalis of the Rathke pouch. They are depicted on the upper surface of the pituitary gland or embedded in the pituitary with the appearance of an egg in an egg-cup, on the midline (Figs. 19.4 and 19.5). Asymptomatic RCCs involving the pituitary stalk (Fig. 19.6a–f) probably share the same origin; they must be differentiated from a deep infundibular recess of the third ventricle or from a pathological enlargement of the infundibulum. Rare RCCs can be encountered off-midline in the suprasellar cistern (Fig. 19.6g–i). Double RCCs may coexist. Concomitant pituitary adenomas are not exceptional (see Chap. 21). RCCs are classically described as pituitary cystic lesions with smooth contours, without calcification and no rim enhancement. "MicroRCCs," one to a few millimeters in diameter, are seen with increasing frequency with high-field MR scanners, either intrasellarly or at the distal end of the pituitary stalk; they have to be considered as anatomic variants (Fig. 19.5), since such microRCCs are present in routine autopsies in up to 33 % of normal pituitary glands. Larger RCCs, around 5–10 mm in size, are also frequent and nearly always asymptomatic (Fig. 19.7). In contrast to what is observed with pituitary adenomas, the sella turcica is unchanged. This is particularly remarkable on axial sequences where it can be observed that the anterior wall of the sella is not or scarcely distorted in front of RCCs even as large as 10 mm in diameter. RCCs larger than 12 mm may be symptomatic (for a fuller description see Chap. 20). RCCs can be stable, enlarge slowly or more rapidly, or shrink or even disappear, and can recur. Some changes have been depicted during pregnancy (see Chap. 8). From a general point of view, evolution of RCC is unpredictable. For this reason, serial imaging and visual field examination seem reasonable for those RCCs around 1 cm in diameter or less if the RCC is suprasellar in location. The cyst content is composed either of a thick mucus rich in protein and mucopolysaccharide or, much less frequently, of a CSF-like transparent fluid with low viscosity. The former is present in the vast majority of intrasellar cysts and is responsible for a more or less T1-hyperintense signal, depending

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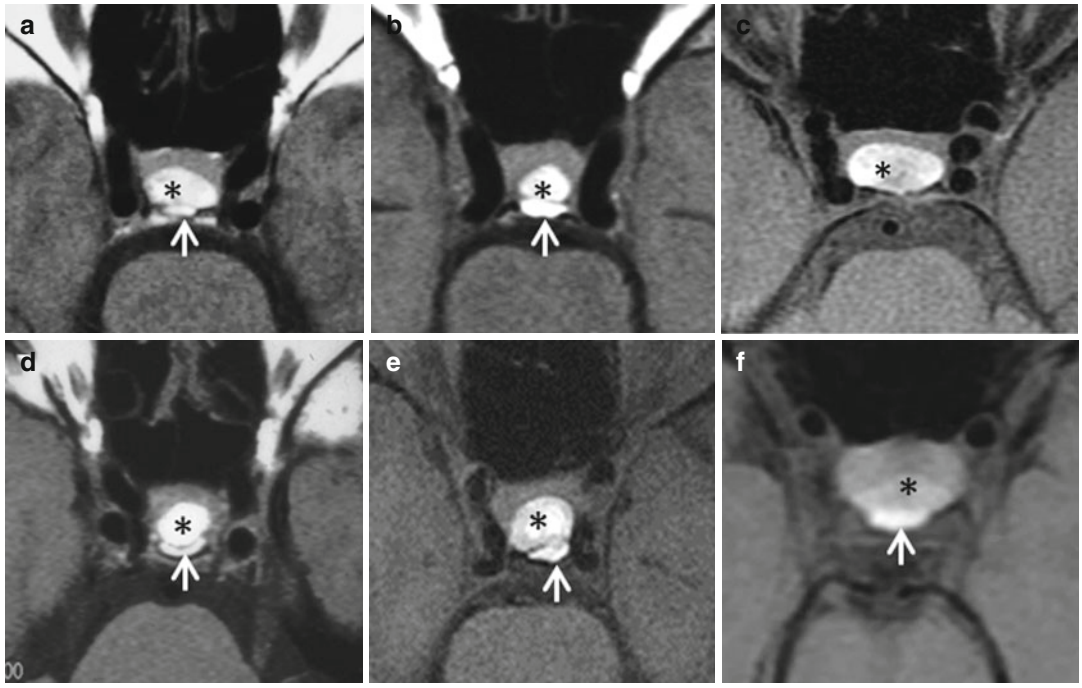

Fig. 19.1 RCCs on axial T1 (**a**, **b**, **d**) and axial T1 fat-saturated WI (**c**, **e**, **f**). RCCs (*asterisks*) are located between anterior and posterior lobes and are in close contact with the posterior lobe. The posterior lobe itself (*white arrows*) is in close contact with the dorsum sellae excepted in **c**

on the protein concentration and degree of dehydration of the cyst content. The usual absence of mass effect observed with RCC, together with their midline location, easily permits their differentiation from hemorrhagic pituitary adenomas. The less frequent CSF-like RCCs are T1 hypointense (Fig. 19.8) and are seen more frequently in suprasellar than in intrasellar locations. Spontaneous changes in signal can occur with time. Intracystic waxy nodules are observed in 75 % of the cases in T1-hyperintense RCC. The main components of these nodules are protein and cholesterol. Most are floating freely within the cyst and their position can change with time; a few are adherent to the cyst wall. The signal of these hyperproteinic nodules is isointense or slightly hyperintense on T1, and frankly hypointense on T2WI. They do not enhance or faintly enhance after contrast administration and are unique or, more rarely, multiple. They can be large, occupying almost the whole cyst, or extremely tiny (Fig. 19.9). In all cases, their recognition warrants the diagnosis of RCC. The activity of mucus-producing cells is variable, and responsible for variations of the cyst size together with absorption of cystic fluid. Factors influencing mucus secretion and, therefore, RCC size increase, are unknown. The cyst wall is very thin, lined usually by a single-cell respiratory-type epithelium often with goblet cells. Thus, the wall of asymptomatic RCC is not visible after gadolinium injection. A potential cause of errors is the enhancement of the normal pituitary gland surrounding the RCC, thus mimicking wall enhancement (Fig. 19.10). Rapid enhancement of the normal pituitary tissue early after contrast injection may help to avoid confusion with cyst wall enhancement that may be seen in symptomatic RCC. Nonenhancement of the cyst wall is essential to differentiate RCC from craniopharyngioma, in which the wall is thicker and stratified, and from complicated RCC.

MRI characteristics of most asymptomatic RCC, namely, no or faint sellar change, unique intrasellar midline location, close contact with the posterior lobe for intrasellar RCC, T1 hyper-

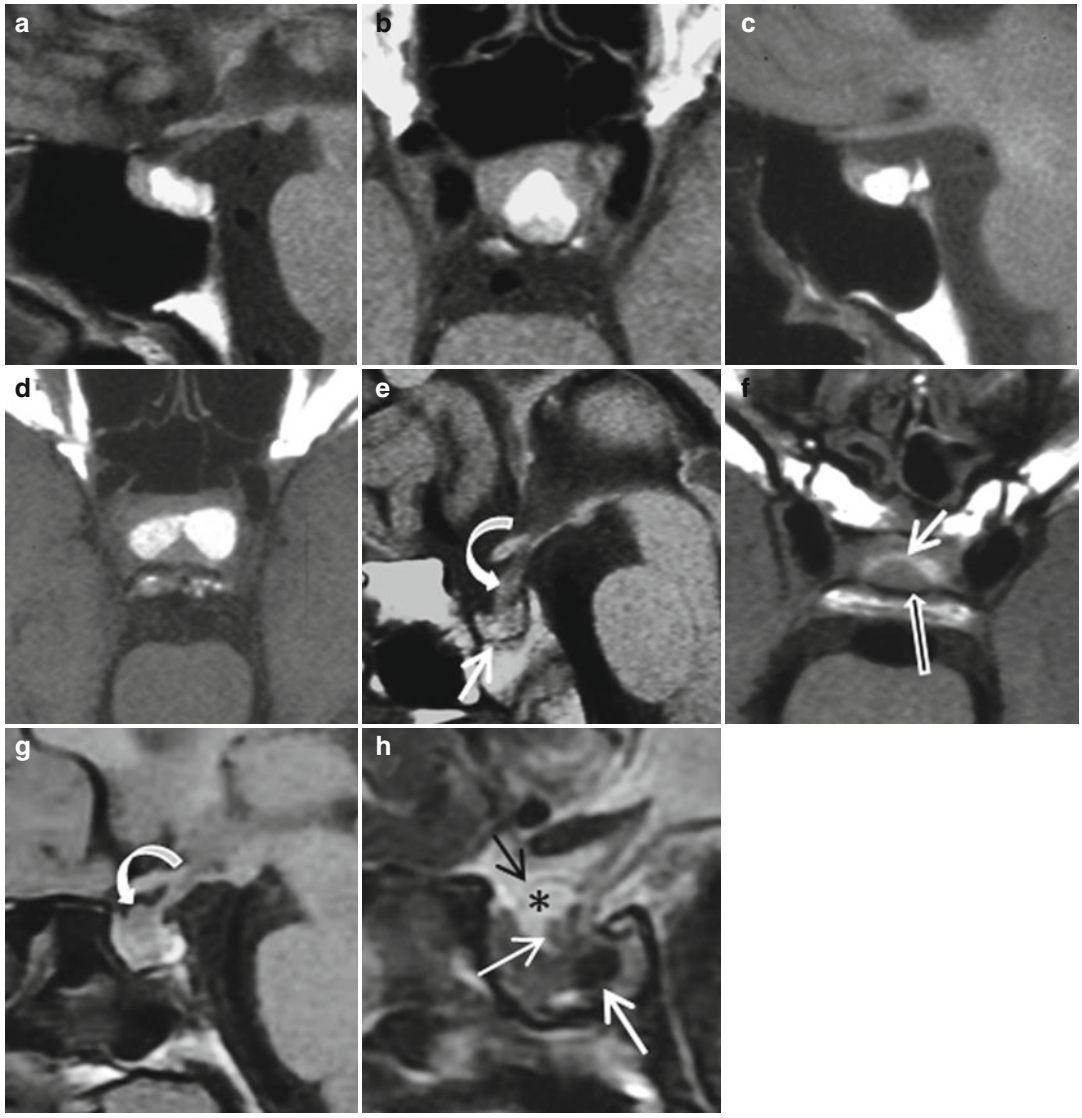

Fig. 19.2 RCCs on sagittal (**a**, **c**, **e**, **g**) and axial (**b**, **d**, **f**) T1WIs. (**h**) Sagittal T2WI. Particular features: (**a**, **b**) Elongated RCC in an elongated sella. (**c**, **d**) Bilobulated RCC. (**e**, **f**) Asymptomatic RCC and infundibuloneurohypophysitis in a 5-year-old boy; thickened pituitary stalk (*curved arrow*); enlarged T1 isointense posterior lobe (*black* and *white arrow*) outlined by an arch-like RCC (*white arrow*) compressed by the abnormal posterior lobe. (**g**, **h**) Double RCC on sagittal T1 and T2 WIs. An anterior T2-hyperintense RCC (*asterisk*) presents a hyperproteinic nodule (*thin white arrow*); the posterior RCC is T2 hypointense (*thick white arrow*). Both are almost invisible on T1WI

intensity without fluid level, T2-hypointense hyperproteinic nodules, and no cyst wall enhancement, usually make the diagnosis of most RCCs easy. These characteristic features should permit one to dramatically restrict the use of the vague term "incidentaloma" too frequently used in radiological reports. Although the vast majority of RCCs remain asymptomatic, some give rise to clinical manifestations. Rapid increase in cyst volume, change of signal, and post-gadolinium rim enhancement can be predictive markers of complications (see Chap. 20).

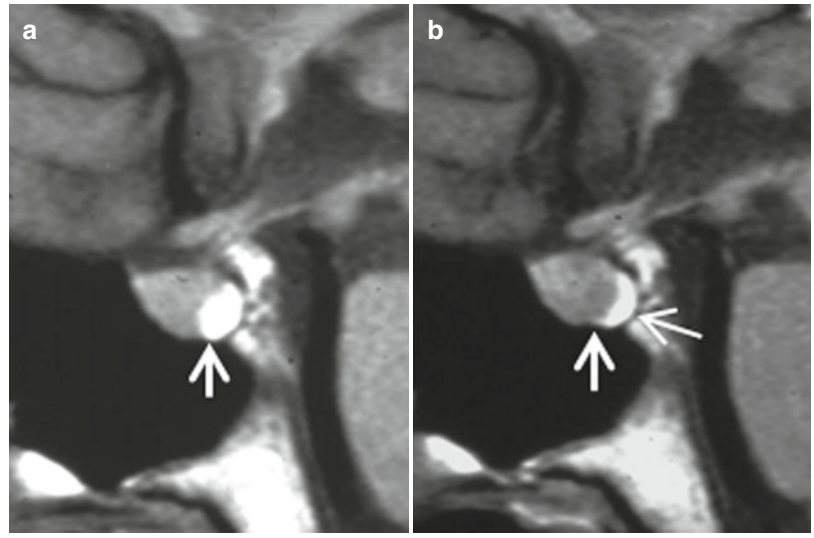

Fig. 19.3 (**a**, **b**) Sagittal T1WIs. RCC (*thick arrow*): spontaneous T1 signal change in a few months (from T1 hyperintense to T1 hypointense). The posterior lobe (*thin arrow*) cannot be distinguished from RCC in (**a**)

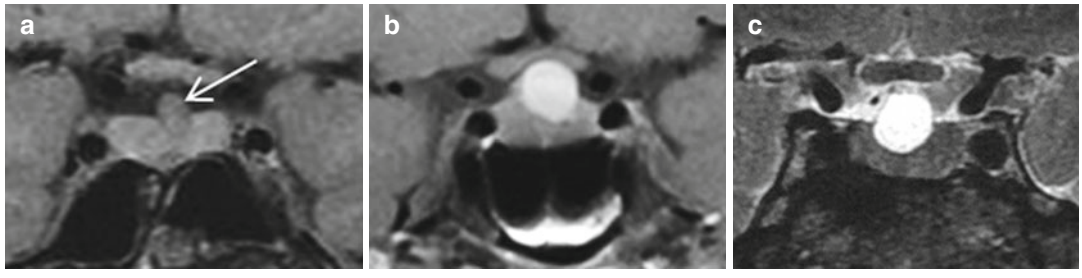

Fig. 19.4 Coronal T1 (**a**, **b**) and T2 WIs (**c**) in three patients with RCC. Egg in an egg-cup pattern. RCC is slightly T1 hypointense in (**a**) if compared with the normal anterior pituitary, markedly hyperintense in (**b**), and T2 hyperintense in (**c**)

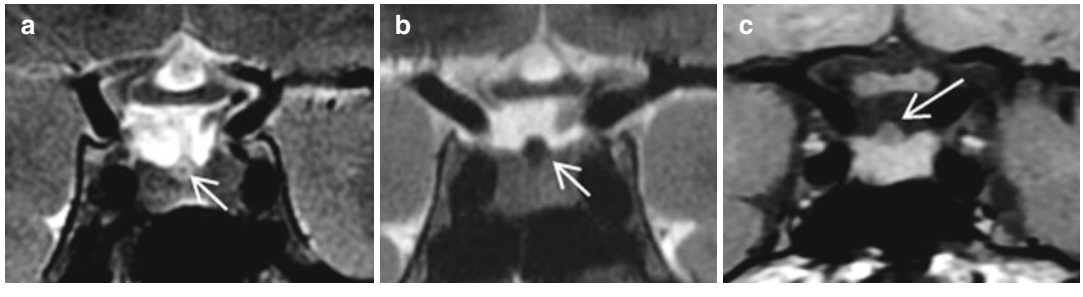

Fig. 19.5 MicroRCCs at the sellar diaphragm level. (**a**, **b**) Coronal T2WI and (**c**) T1WI. RCC is T2 hyperintense with a central hypointense hyperproteinic micronodule (*arrow*) in (**a**), T2 hypointense in (**b**), T1 hypointense in (**c**)

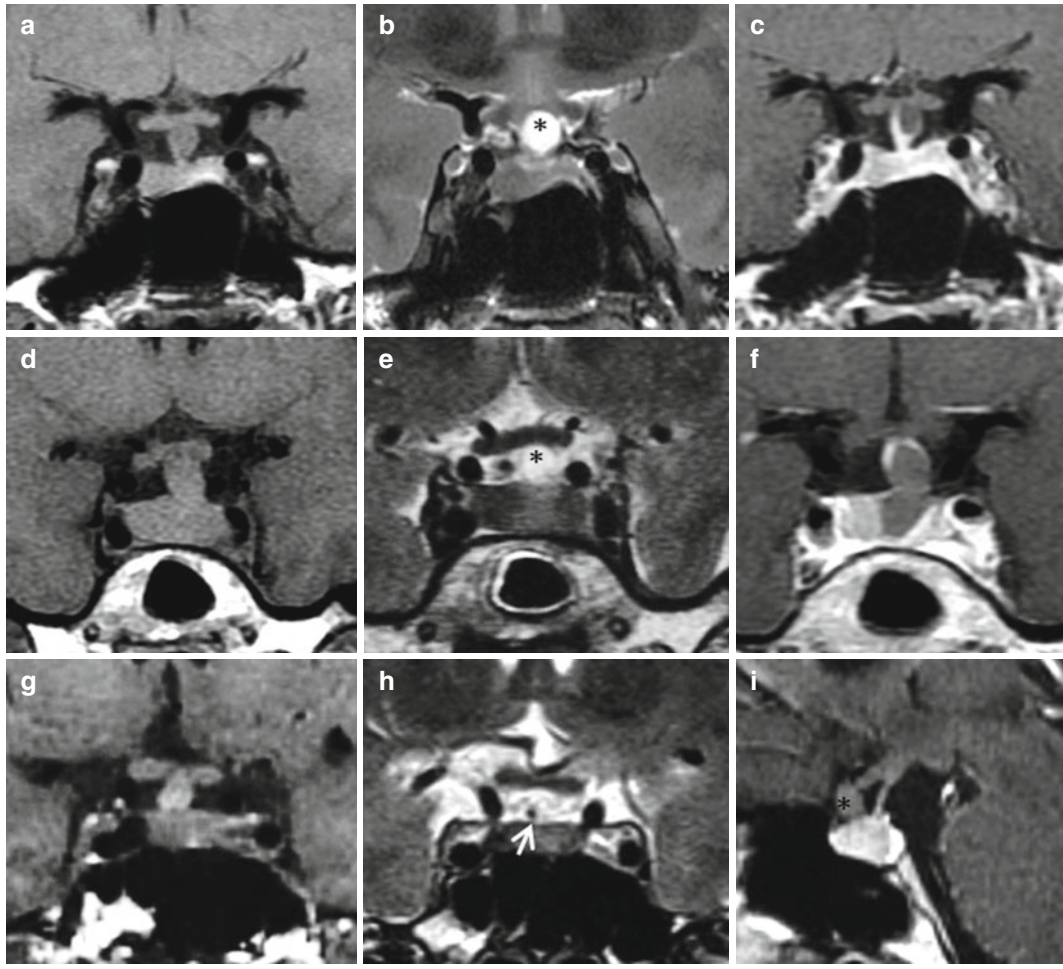

Fig. 19.6 (**a**–**c**) Coronal T1 (**a**, **d**, **g**), coronal T2 (**b**, **e**, **h**) and sagittal CE TI WIs (**c**, **f**, **i**). RCC of the pituitary stalk (**a**–**c**), intrasellar RCC extended to the pituitary stalk (**d**–**f**), and off-midline RCC of the suprasellar cistern (**g**–**i**). All RCCs are T2 hyperintense (*asterisk*) with a tiny T2-hypointense nodule in (**h**) (*arrow*). Postcontrast enhancement of a distorted pituitary stalk in (**c**) and (**f**) with incomplete and thin enhancement of RCC wall. In (**i**), the T1 nonenhanced RCC (*asterisk*) is located in front of the pituitary stalk

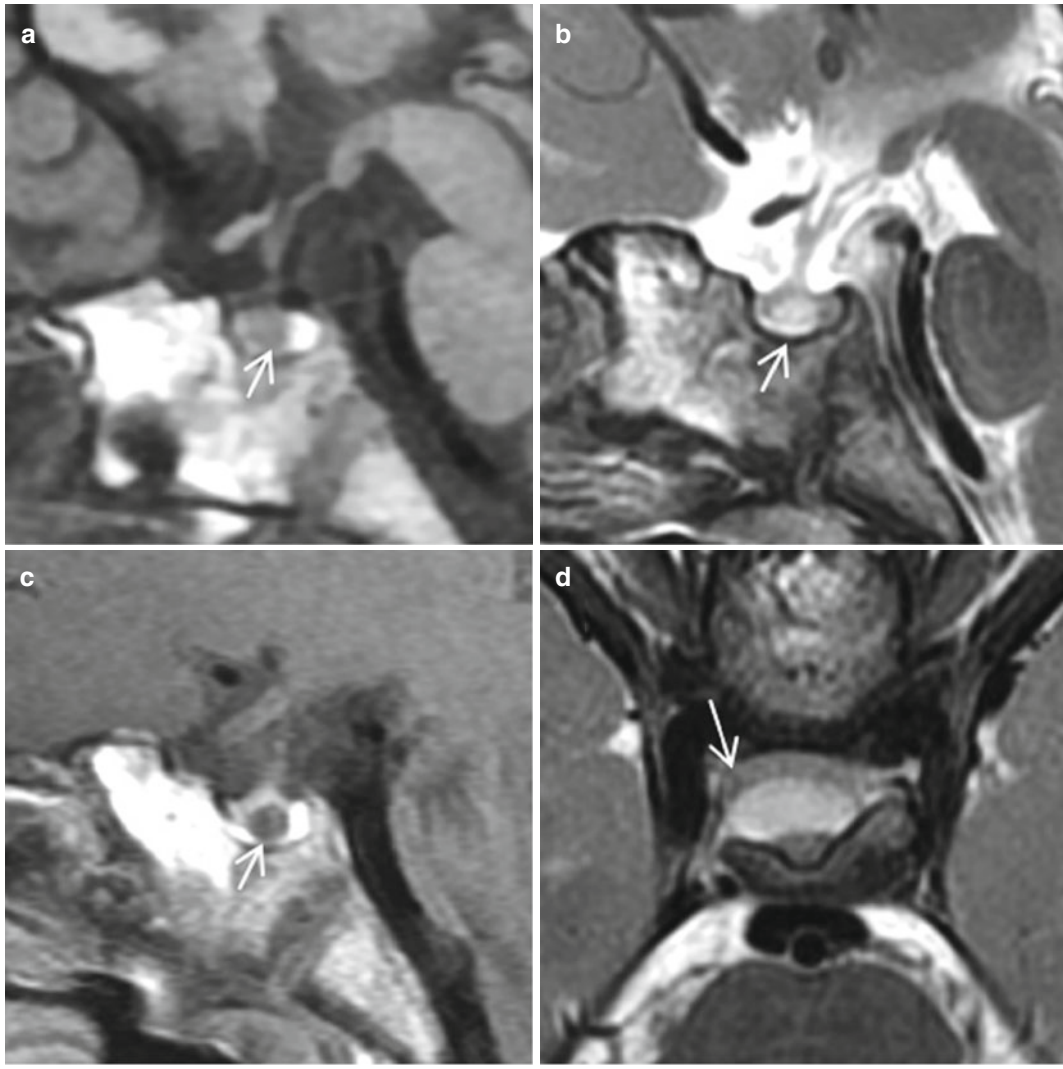

Fig. 19.7 Asymptomatic RCC in a 4-year-old boy. (**a**–**c**) Sagittal T1, T2, and CE T1 WIs. (**d**) Axial T2WI. Characteristic shape and location of an RCC with a CSF-like signal on sagittal and axial images (*arrow*)

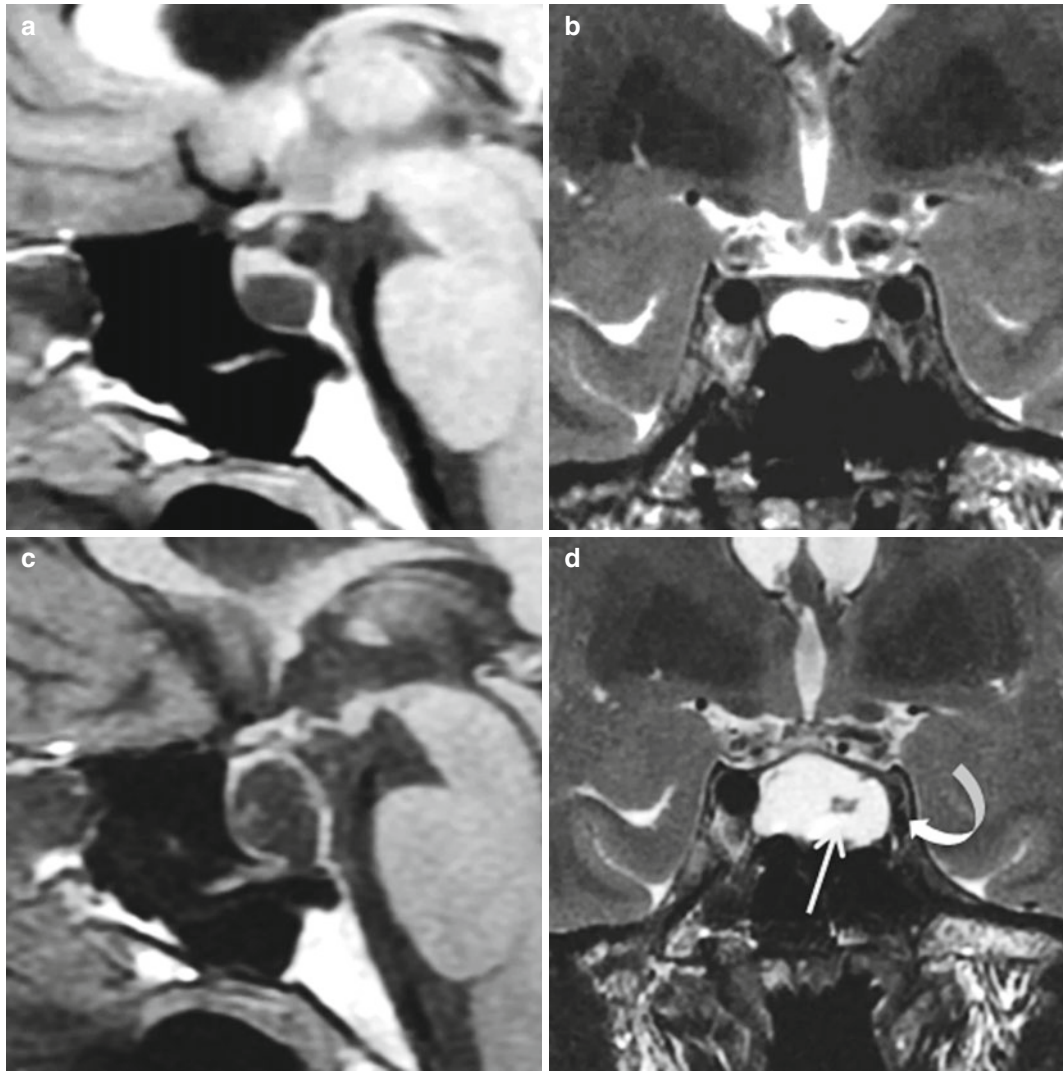

Fig. 19.8 Asymptomatic RCC in a 32-year-old woman followed yearly for 8 years. (**a**, **c**) Sagittal T1WI and (**b**, **d**) Coronal T2WI. RCC is markedly T1 hypointense and T2 hyperintense. Hyperproteinic nodule (*arrow*). Low-speed cyst volume increase in 8 years. The patient remains asymptomatic but a follow-up is necessary. Note the incidental finding of an asymptomatic thrombosis of left internal carotid artery in (**d**) (*curved arrow*)

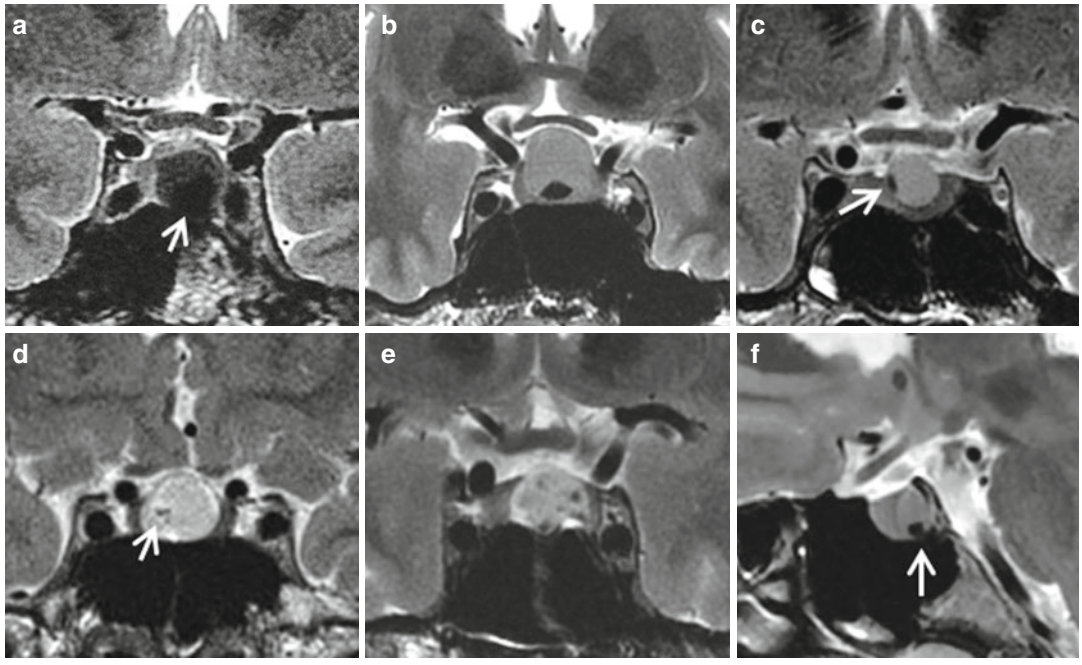

Fig. 19.9 RCCs with T2-hypointense hyperproteinic nodules. (**a**–**e**) Coronal T2WIs. (**f**) Sagittal T2WI. The nodules can be massive (**a**), small (**b**, **f**), tiny (**c**, **d**), or, more rarely, multiple (**e**)

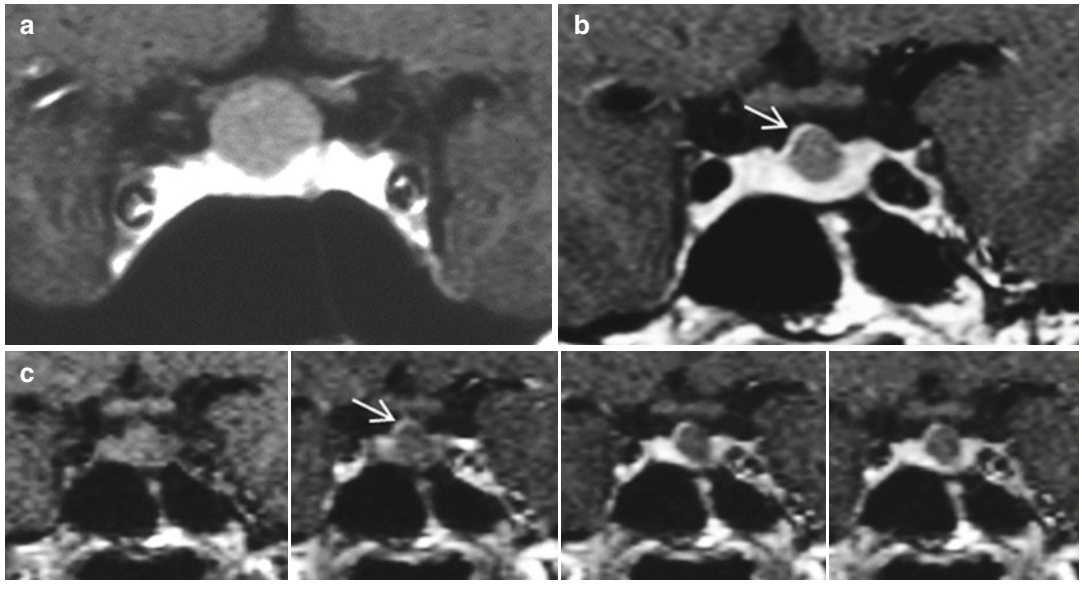

Fig. 19.10 RCCs after gadolinium injection on coronal CE T1WIs (**a**, **b**). Characteristic pattern with no rim enhancement in (**a**). Thin enhancement (*arrow*) of the displaced pituitary gland simulating RCC partial wall enhancement in (**b**). In (**c**), dynamic sequence confirms that the enhancement occurs early and involves the displaced anterior pituitary (*arrow*) and not the RCC wall which, if at all, should occur later

Further Reading

Bonneville F, Cattin F, Marsot-Dupuch K et al (2006) T1 signal hyperintensity in the sellar region: spectrum of findings. Radiographics 26(1):93–113

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