Suprasellar Germinoma

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The most common germ cell tumor of the suprasellar region is the germinoma. A suprasellar germinoma can be a primitive tumoral lesion or a metastatic lesion from a pineal germinoma. Germinomas are most commonly seen in adolescents and young adults: 90 % of patients are younger than 20 years. There is no gender preference for the suprasellar location, unlike the pineal location with male predominance. Clinical symptoms include diabetes insipidus, precocious puberty, or growth failure related to hypothalamic involvement and visual loss provoked by chiasmatic compression. Large lesions can cause hydrocephalus by obstruction of the interventricular foramens. Tumoral markers such as α-fetoprotein and human chorionic gonadotropin (HCG) can be found in the serum and/or CSF. In the case of suggestive clinical and radiological patterns with presence of tumoral markers, the diagnostic biopsy can be avoided. Infundibular thickening and absence of the posterior lobe bright spot on T1WI are the more precocious radiological signs, and at this stage can simulate an inflammatory process, Langerhans cell histiocytosis, or lymphocytic infiltration (Fig. 31.1). As it grows, the lesion appears as a well-delineated round or lobulated lesion, isointense or hyperintense to gray matter on T1WI and isointense to hyperintense on T2WI (Fig. 31.2), with hyperintense necrotic foci in some cases. Calcifications, better seen on CT than on MRI, are not unusual but less frequent than in the pineal location. After gadolinium injection there is a marked and mostly speckled enhancement. The origin within the optic nerve or optic chiasm is very rare. Intraventricular extension in lateral ventricle and infiltration of the basal ganglia and the corpus callosum can occur (Fig. 31.3). Multifocal germinomas usually involve the pineal gland and the suprasellar cistern, simultaneously or not (Fig. 31.4). Leptomeningeal spread with drop metastases along the spinal cord are reported. ADC is usually decreased. MR spectroscopy shows markedly elevated choline with diminished N-acetyl aspartate. Despite the aggressive MRS pattern, a good response is observed in most cases after radiotherapy and chemotherapy. Before treatment, a complete survey of the brain and spinal canal is required because germinomas can metastasize by subarachnoid seeding. Negative MRI in children with diabetes insipidus does not exclude germinoma (Fig. 31.5). A repeat MRI examination is required in 3–6 months. Early diagnosis may reduce the risk of dissemination and the morbidity of treatment. The limited differential diagnosis includes craniopharyngioma with a more heterogeneous appearance associated with cysts, solid components and calcifications, and hypothalamic-chiasmatic glioma rarely associated with diabetes insipidus (see Chap. 30).

J.-F. Bonneville et al., *MRI of the Pituitary Gland*, DOI 10.1007/978-3-319-29043-0_31

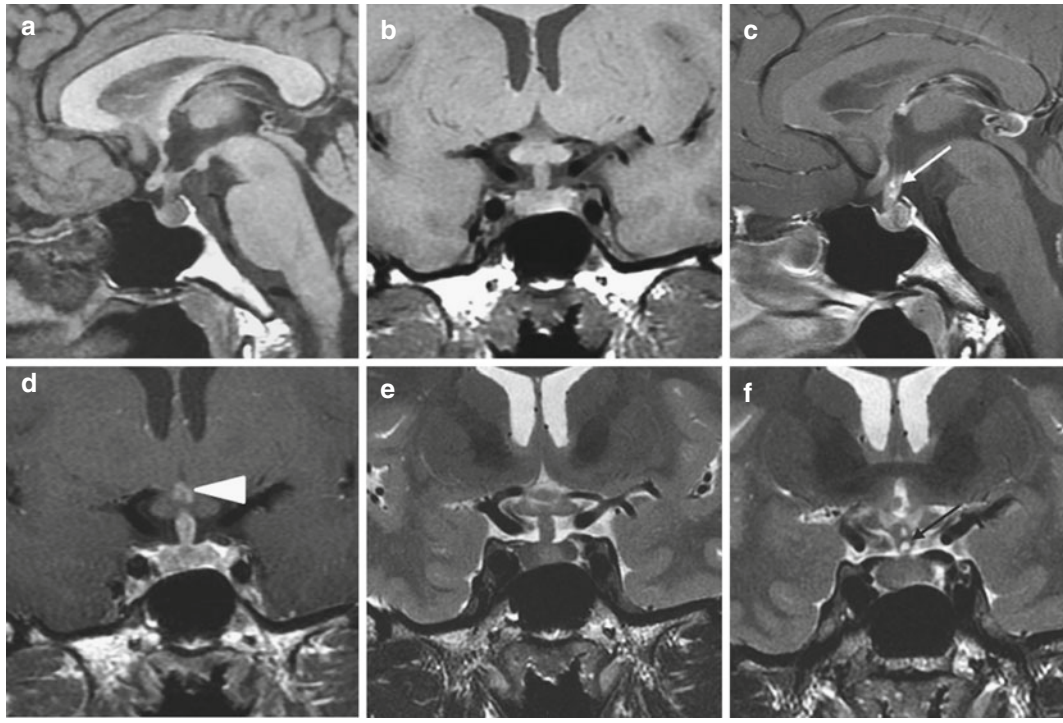

Fig. 31.1 Twenty-three-year-old man presenting with diabetes insipidus for 1 year. (**a**, **b**) Sagittal and coronal T1WIs. The hypersignal of the posterior lobe is absent and the pituitary stalk is enlarged. The anterior pituitary gland appears normal. (**c**, **d**) Sagittal and coronal CE T1WIs. Speckled enhancement of the pituitary stalk (*arrow*) and tuber cinereum (*arrowhead*). (**e**, **f**) Coronal T2WIs. Small hyperintense spots in the pituitary stalk (*black arrow*). At 3 months (not shown), there were no changes. (**g**, **h**) Sagittal and coronal CE T1WIs at 6 months. Presence of a hypointense intrasellar posterior lesion associated with significant increase in diameter of the pituitary stalk and large contrast-enhancing suprasellar sellar mass involving the optic chiasm and tuber cinereum. (**i**) Coronal T2WI. The lesion is quite homogeneous and isointense to gray matter. The diagnosis of germinoma was confirmed by the evidence of a high level of β-HCG in the CSF. (**j**) Coronal CE T1WI after corticotherapy and radiation therapy. Decrease in size of the pituitary gland. Complete shrinkage of the suprasellar tumor with marked atrophy of the pituitary stalk

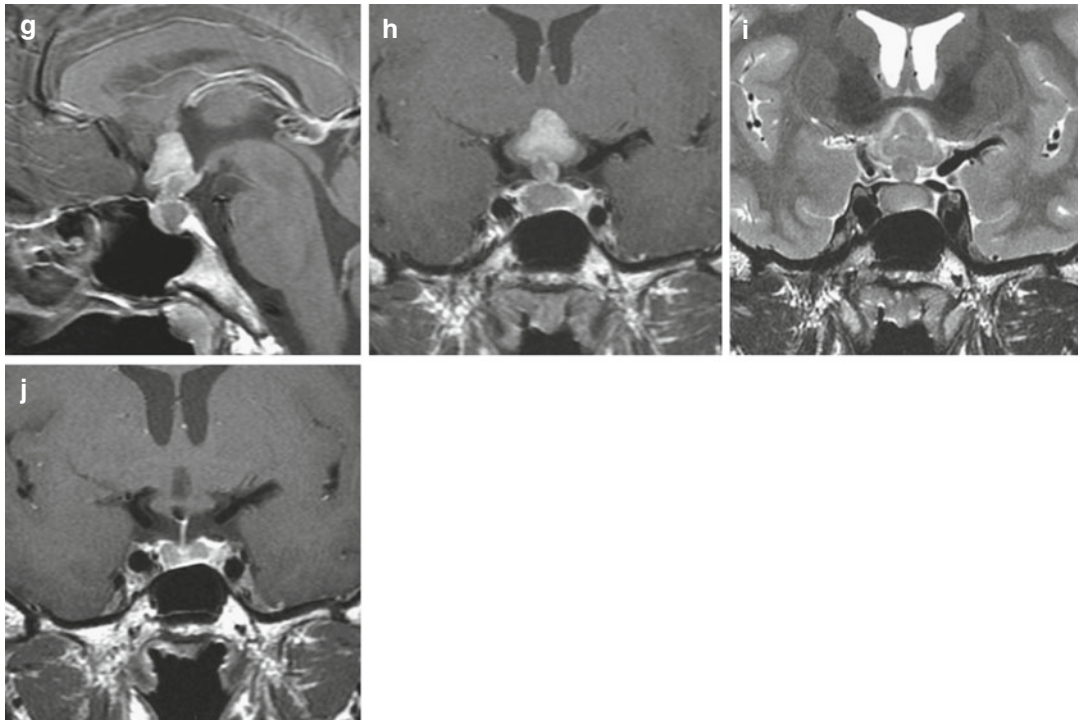

Fig. 31.1 (continued)

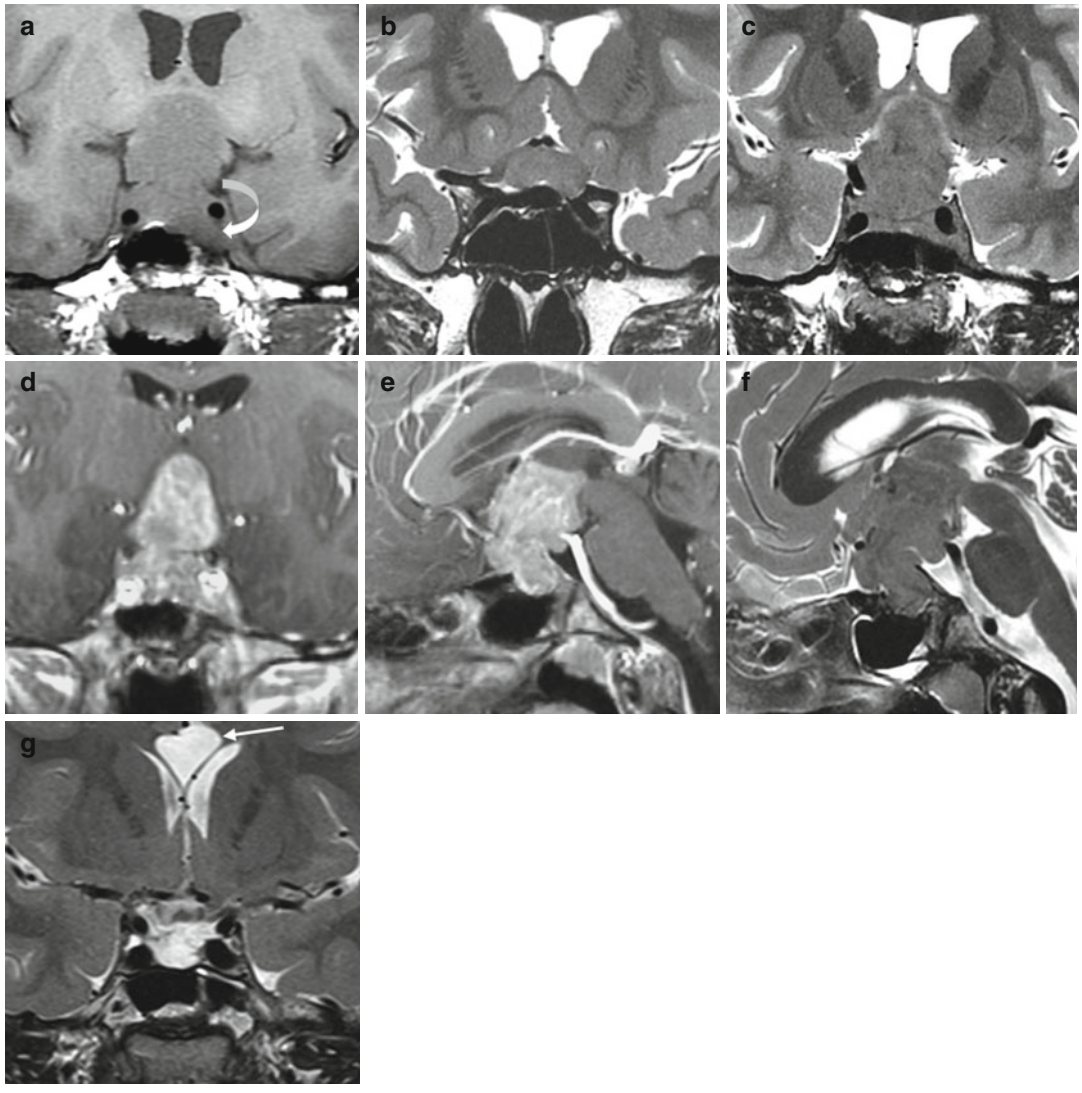

Fig. 31.2 Headaches, bitemporal hemianopia, visual loss, and diabetes insipidus in a 13-year-old girl. (**a**) Coronal T1WI. Large intra- and suprasellar tumor with invasion of the left cavernous sinus (*curved arrow*). (**b**, **c**) Coronal T2WIs. Involvement of the optic chiasm and complete obstruction of the lumen of the third ventricle. (**d**, **e**) Coronal and sagittal CE T1WIs. The enhancement is heterogeneous with multiple specks. (**f**) Sagittal T2WI. The signal is homogeneous. Note the polycyclic contours of the tumor. (**g**) Coronal T2WI. Shrinkage of the tumor after medical treatment and radiation therapy. Empty sella turcica, signal abnormalities of the optic chiasm and porencephalic cavity after transcallosal biopsy (*arrow*)

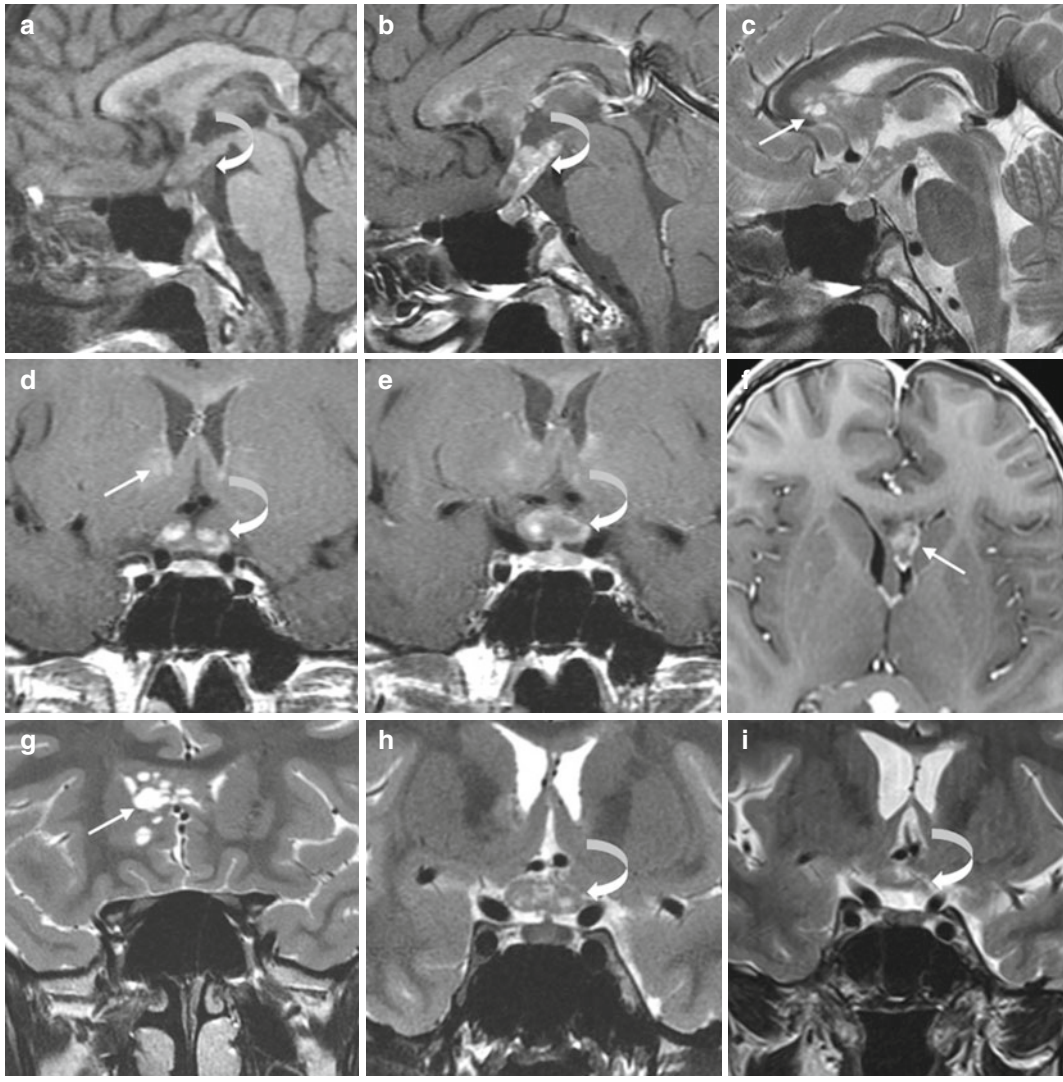

Fig. 31.3 Rapidly progressive visual loss and diabetes insipidus for 4 months in a 15-year-old boy. (**a**–**c**) Sagittal T1, and CE T1 and T2 WIs. The T1 hypersignal of the posterior lobe is missing. Suprasellar mass (*curved arrow*) with patchy enhancement and multiple hyperintense foci on T2WI associated with poorly delimited frontal periventricular lesions (*arrow*) presenting the same MR characteristics. (**d**, **e**) Coronal and (**f**) axial CE T1WIs. Demonstration of the contrast-enhanced intrachiasmatic lesions (*curved arrow*). The pituitary stalk is not thickened. Bifrontal periventricular tumoral infiltration (*arrow*). (**g**, **h**) Coronal T2WIs. The multicystic appearance of the optic chiasm (*curved arrow*) and periventricular lesions (*arrow*) is suggestive of germinoma. (**i**) Coronal T2WI after treatment: normalization of the optic chiasm (*curved arrow*)

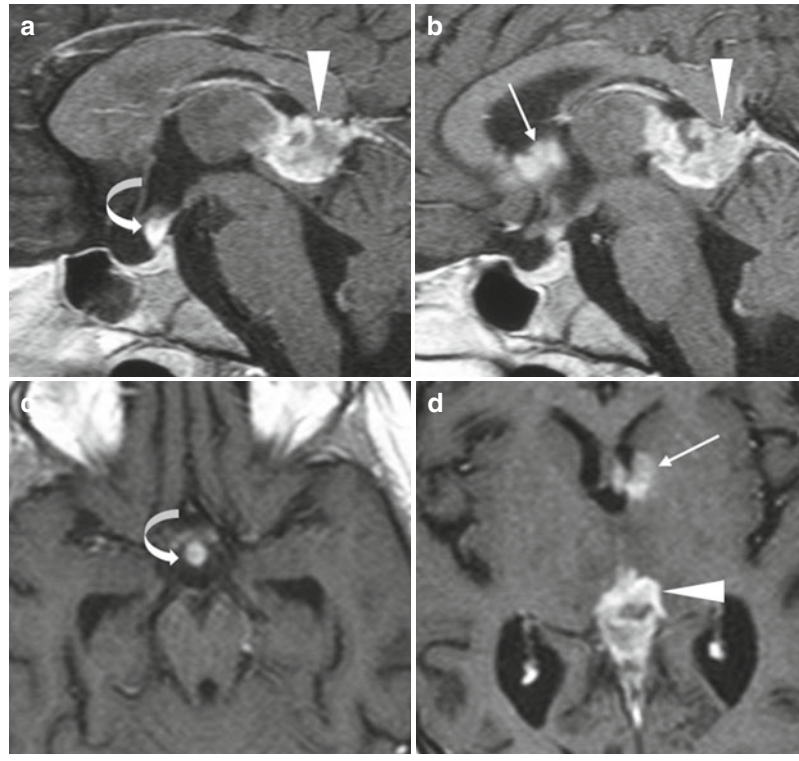

Fig. 31.4 Multifocal germinoma. (**a**, **b**) Sagittal and (**c**, **d**) axial CE T1WIs. Enlargement and marked enhancement of the superior part of the pituitary stalk (*curved arrow*), contrast-enhanced pineal mass (*arrowhead*), and involvement of the wall of the left frontal horn and surrounding parenchyma (*arrow*)

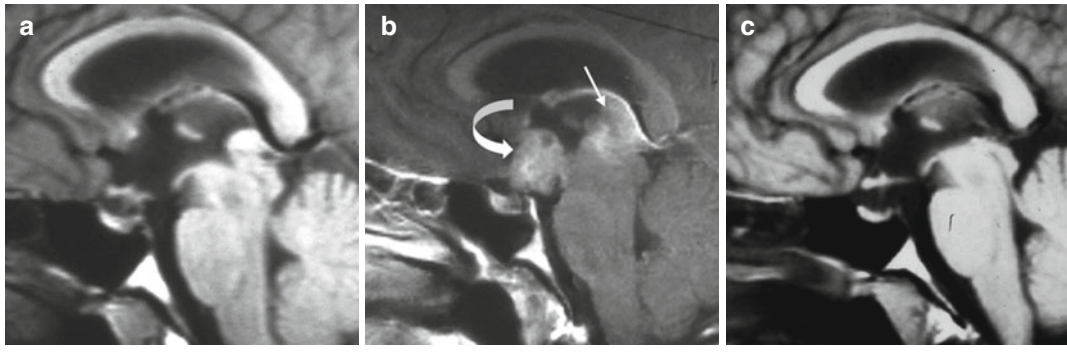

Fig. 31.5 Diabetes insipidus for 1 month. (**a**) Sagittal T1WI showing only a posterior intrasellar hyposignal instead of the physiological T1 hypersignal of the posterior lobe. (**b**) Six months later, on sagittal CE T1WI, demonstration of suprasellar (*curved arrow*) and pineal (*arrow*) lesions. (**c**) After treatment, complete radiological remission. The hypersignal of the posterior lobe is still absent

Further Reading

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Phi JH, Kim SK, Lee J et al (2013) The enigma of bifocal germ cell tumors in the suprasellar and pineal regions: synchronous lesions or metastasis? J Neurosurg Pediatr 11:107–114

Sethi RV, Marino R, Niemierko A, Tarbell NJ, Yock TI, Mac Donald SM (2013) Delayed diagnosis in children with intracranial germ cell tumors. J Pediatr 163:1448–1453