PICTORIAL ESSAY

Mimics of Pituitary and Pineal Germ Cell Tumours on Imaging: A Pictorial Essay

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INTRODUCTION

Intracranial germ cell tumours (GCTs) comprise 0.4% to 9.4% of primary intracranial neoplasms and intracranial germinoma accounts for 50% to 70% of all intracranial GCTs.^{1,2} They are characteristically located in the suprasellar and pineal regions. Apart from GCTs, other diseases are also found in the suprasellar and pineal regions.

In this pictorial essay, mimickers of intracranial GCTs are illustrated. These GCT mimics are based on a retrospective analysis of 313 consecutive cases collected over 29 years at a single hospital with a tentative or initial diagnosis of GCT but proven to be otherwise on histology.

GERM CELL TUMOURS

The most common locations of GCTs are the pineal

gland and suprasellar regions. The levels of some oncoproteins may elevate in the serum or cerebrospinal fluid (CSF), including alpha-fetoprotein, beta-hCG, and placental alkaline phosphatase, depending on the tumour types.³ Germinoma and teratoma are the most common and the second most common types of intracranial GCTs, respectively. Approximately 90% of intracranial germinoma patients are diagnosed before the age of 20 years.² The two most common locations of intracranial germinoma are the pineal region (37%-65%) and the suprasellar region (25%-49%), with approximately 8% of cases showing bifocal involvement in these two locations.^{3,4} The male-to-female ratio is 1.88:1 in the suprasellar region; the ratio is even higher for pineal germinomas.1 Tumour location, size, and resultant endocrine dysfunction are the main causes of clinical symptoms and signs in germinoma. The prodrome in suprasellar lesions can last from months to years, longer

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than that in pineal lesions. In suprasellar germinomas, symptoms related to diabetes insipidus often occur first. followed by other endocrine dysfunctions. As the tumour grows, visual impairment may occur due to compression of the optic chiasm; obstructive hydrocephalus is also possible if the drainage of CSF is affected. In pineal germinomas, the aqueduct and dorsal midbrain can be affected, resulting in obstructive hydrocephalus, diplopia, and Parinaud's syndrome. Other symptoms due to tumour dissemination or metastasis can develop.⁴ Computed tomography (CT) of the head seldom shows calcification of the germinoma, but when located in the pineal region, it can enlarge and engulf the pineal calcification. In magnetic resonance imaging (MRI), germinoma usually presents as iso- to hyperintense to grey matter on T1-weighted and T2-weighted images, with marked enhancement and cyst formation, and hydrocephalus, as well as water restriction on diffusionweighted imaging (DWI). Dissemination via the CSF and invasion of adjacent brain parenchyma also commonly occur.^{3,4} Because of the risk of CSF seeding, imaging evaluation should include the entire neuroaxis; however, even with extensive involvement, the prognosis of germinomas is good because of the radiosensitive nature of these tumours.³ The prognosis of teratoma varies, depending on the histological findings. Some clues to the diagnosis of germinoma have been proposed, such as engulfment of the pineal calcification and bifocal involvement with normal alpha-fetoprotein level in the serum and CSF. Nonetheless, surgical confirmation is usually required.^{3,4} In our cases, there are typical instances of engulfment of the pineal calcification, doublet lesions, and also infiltrative lesions involving both frontal lateral ventricular walls (Figure 1). However, nongerminomatous GCTs can also demonstrate bifocal involvement.1,2



Figure 1. Germinoma. Case 1 (a) showing engulfment of the pineal calcification by a pineal germinoma (arrow) on axial nonenhanced computed tomography image. Case 2 (b) showing doublet germinoma lesions with a larger pineal component (arrow) and a smaller suprasellar component (arrowhead) on enhanced sagittal T1-weighted fat-suppressed image. Case 3 showing a rare presentation of a germinoma with bilateral enhancing frontal horn wall infiltration (arrows) on coronal (c) and axial (d) T1-weighted fatsuppressed images.

MIMICS OF SUPRASELLAR GERM CELL TUMOURS

Craniopharyngioma

Approximately 1.2% to 4.0% of paediatric brain tumours are craniopharyngiomas, which are commonly located in the sellar and suprasellar regions.⁵ The

adamantinomatous type is more common in childhood. Imaging features include calcifications and cystic components, which are found in our case (Figure 2).

Glial Cell Tumour

When located in the suprasellar region, the glial cell



Figure 2. Adamantinomatous craniopharyngioma. Calcification is identified in the suprasellar tumour (arrow) on axial nonenhanced computed tomography (a) with strongly enhancing soft tissue and cystic portions of the tumour (arrow) on sagittal T1-weighted fat-suppressed image (b).



Figure 3. Glial cell tumour. The case displays suprasellar oedematous infiltration along both optic tracts (arrows) on nonenhanced axial T2weighted fluid-attenuated inversion recovery image (a). The main tumour portion (arrows) is located in the suprasellar and retrochiasmatic with heterogeneous region, iso- to hypointensity on sagittal unenhanced T1-weighted image (b) and strong heterogeneous enhancement on the axial (c) and sagittal (d) T1-weighted enhanced images.

tumour is commonly referred to as an optic pathway glioma. These cases are usually diagnosed at 4 to 5 years of age.⁶ Imaging findings include involvement along the course of optic pathway with variable enhancement and cystic components as well as calcifications. Sometimes, oedema in the optic pathway near the tumour is a diagnostic clue (Figure 3).⁷

Lymphocytic Hypophysitis

Lymphocytic hypophysitis is an autoimmune inflammatory disease with lymphocytic infiltration involving the pituitary gland, stalk, and hypothalamus, which can affect adults and children of both sexes.⁸ Possible common symptoms include headache, hyper or hypofunction of pituitary gland, and diabetes insipidus.⁸ Depending on disease involvement, typical imaging findings are strong enhancement and enlargement of the pituitary gland, stalk, and hypothalamus. Sometimes, loss of the neurohypophyseal bright spot is noted (Figure 4).⁸ Delayed enhancement of the whole pituitary gland due to this disease is also described in the literature.⁸ Repeated imaging studies are sometimes necessary because lesions might be found months after an initial normal imaging study.⁸



Figure 4. Lymphocytic hypophysitis. The case demonstrates the lesion presenting as thickening of the pituitary stalk with tapering in the sella; there is loss of the normal bright signal (arrow) in the neurohypophysis on sagittal T1-weighted image (a). Marked enhancement of the lesion (arrow) is found on enhanced sagittal T1-weighted image (b).



Figure 5. Pituicytoma. This case shows a well-defined iso- to hypointense suprasellar lesion (arrow) on sagittal T1-weighted image before contrast injection (a) and the lesion (arrow) is strongly enhanced after contrast injection (b). The lesion is iso- to hyperintense (arrow) on axial T2-weighted fluid-attenuated inversion recovery image (c).

Pituicytoma

Pituicytomas are rare benign tumours originating from a type of glial cell, the pituicyte, in the neurohypophysis and pituitary stalk. They usually affect adults with a slight male predominance.⁹ The imaging presentation of pituicytoma is nonspecific but it usually presents as a welldefined homogeneously or heterogeneously enhancing solid mass in the sellar or suprasellar region (Figure 5). Calcification, adjacent bony changes, or necrosis are absent, but cystic portions sometimes can be identified.⁹

Pituitary adenoma

Pituitary adenoma is more common in adults but can be found in children.⁷ It may display signal intensities similar to those of the adjacent normal pituitary gland on pre-contrast MRI and relatively less enhancement on



Figure 6. Pituitary adenoma. The tumour (arrow) involves the sellar and suprasellar regions and has marginal calcification and diffuse hyperdensity on nonenhanced axial computed tomography image (a). On axial T2-weighted image (b), the tumour (arrow) is mainly iso- to hyperintense, with focal hypointensity, possibly due to haemorrhage. The tumour (arrows) enhances heterogeneously in sagittal (c) and coronal (d) contrast-enhanced T1-weighted fat-suppressed images.

Pituitary/Pineal Germ Cell Tumour Mimics



Figure 7. Primitive neuroectodermal tumour. The tumour is hyperdense on nonenhanced axial computed tomography image with displacement of the pineal calcification (arrow) from the midline by the tumour (a). Water restriction is noted on diffusion-weighted imaging (b) and apparent diffusion coefficient (c) images. The tumour (arrows) is iso- to hyperintense on axial T2-weighted fluid-attenuated inversion recovery image (d) and heterogeneously hypointense with heterogeneous enhancement on sagittal T1-weighted images (e and f).

enhanced MRI. Cystic components, calcification, and haemorrhage may be visible (Figure 6). The tumours can display suprasellar or parasellar extension. Dynamic contrast-enhanced MRI is useful to detect a relatively hypointense microadenoma in its early enhancement phase.¹⁰

MIMICS OF PINEAL GERM CELL TUMOURS

Primitive Neuroectodermal Tumour

The primitive neuroectodermal tumour of the pineal

gland is also known as a pineoblastoma, which is a highly malignant tumour and accounts for 40% of pineal parenchymal tumours. The diagnosis of pineoblastoma peaks before the age of 20 but can be at any age.³ The tumour is usually > 3 cm and the pineal calcification, if present, is displaced from the midline by the tumour. Because of its high cellularity, pineoblastomas are hyperdense on CT and show water restriction on DWI (Figure 7). Heterogeneous enhancement and obstructive hydrocephalus are present and CSF dissemination is common.³



Figure 8. Meningioma. There is a well-defined hyperintense tumour (arrow) with minimal hypointensity on T2-weighted image in the pineal region (a). Strong enhancement with a small dural tail (arrow) is found on enhanced axial T1-weighted image (b). Mild water restriction is noted on diffusion-weighted imaging (c) and apparent diffusion coefficient image (d).

Meningioma

Pineal region meningiomas are uncommon, accounting for 6.2% of all pineal tumours and 0.3% of all intracranial meningiomas.¹¹ Because of the highly cellular nature, meningioma shows hyperdensity on CT and water restriction on DWI (Figure 8). Strong enhancement is noted. Calcifications and a dural tail are sometimes present.³

Pineal Cyst

Pineal cysts can be found radiographically in 23% of patients, with a female predominance. Although they can be found at all ages, they are most commonly identified in adults. Typically, the lesion is < 15 mm; larger lesions may show intracystic haemorrhage.^{3,12}

The imaging findings are typically well-defined cystic lesions with water signal intensity inside. Sometimes the cystic portion shows hyperintensity on fluid-attenuated inversion recovery images because of the proteinaceous contents. Wall enhancement can be found and rarely, and enhancement of the suspected cystic part has been reported and was present in our case (Figure 9). The likely mechanism is passive diffusion of the contrast into the cyst.^{3,13}

Papillary Tumour of the Pineal Region

Papillary tumour of the pineal region is a rare neuroepithelial tumour located in the pineal region. It can affect children and adults without sex difference.³ On imaging studies, this tumour is described as a wellPituitary/Pineal Germ Cell Tumour Mimics



Figure 9. Pineal cyst. The case shows a pineal lesion (arrows) with heterogeneous hypointensity on sagittal T1-weighted image (a) and hyperintensity on axial T2-weighted fluid-attenuated inversion recovery image (b). About two-thirds of the lesion (arrows) enhances on sagittal (c) and axial (d) T1-weighted images.

defined enhancing lesion with possible cystic portions. Usually, there is absence of fat, haemorrhage, melanin or calcification. Sometimes, there is T1 signal hyperintensity in the lesion (Figure 10), which is considered to be due to proteinaceous content.³ One of our cases shows calcification at the lesion site (Figure 10), but it could be just due to normal pineal calcification considering the old age of the patient.

Pineal Parenchymal Tumours

Pineal parenchymal tumour of intermediate differentiation accounts for about 20% of all pineal parenchymal tumours and can be found at any age with a slight female predominance.³ Imaging findings are variable and may be indistinguishable from pineocytoma or pineoblastoma. Usually, this tumour presents as a

lobulated heterogeneously enhancing lesion, sometimes with a cystic component. Pineal calcification can be displaced to the periphery by the tumour. Because of its high cellularity, it can show hyperdensity on CT and water restriction on DWI (Figure 11). Local invasion is reported in approximately 80% of cases.^{3,14}

CONCLUSION

This pictorial essay suggests that the presence of doublet lesions in both the suprasellar and pineal regions, although less common, might be a useful clue for intracranial germinomas. However, imaging diagnosis for single germinomas in either the suprasellar or pineal region remains challenging because of the overlapping imaging presentations of intracranial germinomas and their mimics. When the mimics are also frequently observed



Figure 10. Papillary tumour of the pineal region. Case 1 shows a well-defined bilobulated hyperintense lesion (arrows) in the posterior commissure on sagittal T1-weighted image (a), which is hyperintense on axial T2-weighted fluid-attenuated inversion recovery image (b) and homogeneously enhancing on enhanced axial (c) and coronal (d) T1-weighted images. Case 2 shows a hypodense pineal tumour with probable displacement of the pineal calcification (arrows) to the margin of the gland on the nonenhanced axial computed tomography image (e). The tumour (arrows) is heterogeneously hyperintense on T2-weighted fluid-attenuated inversion recovery image (f) and heterogeneously hypointense on unenhanced (g) and enhancing on contrast-enhanced (h) sagittal T1-weighted images.

in children or young adults, such as craniopharyngioma and glial cell tumour in the suprasellar region and primitive neuroectodermal tumour in the pineal region, the diagnosis becomes even more difficult.

Imaging diagnosis of intracranial GCTs is challenging due to their diverse presentation and overlapping appearance with other diseases. The phenomenon of doublet lesions in the suprasellar and pineal regions may be a clue to diagnose germinoma but is uncommon and might also happen in other tumours.

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Figure 11. Large pineal parenchymal tumour of intermediate differentiation. Case 1 shows a large lobulated tumour (arrows in [a] and [b]) in the pineal region with extension into the left lateral ventricle and invasion of the adjacent corpus callosum. It is iso- to hypointense on sagittal T1-weighted image (a), with heterogeneous signals on axial T2-weighted image (b). Diffusion-weighted imaging (c) shows water restriction and apparent diffusion coefficient image (d). Case 2 displays a heterogeneously hyperdense tumour (arrows in [e] to [h]) in the pineal region on nonenhanced axial computed tomography image (e), with hypointense and hyperintense parts on the axial T2-weighted fluid-attenuated inversion recovery image (f). There are hypo-, iso-, and hyperintense portions on nonenhanced sagittal T1-weighted image (g) and some enhancement on axial T1-weighted image (h).

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