Clinics in Diagnostic Imaging (81)

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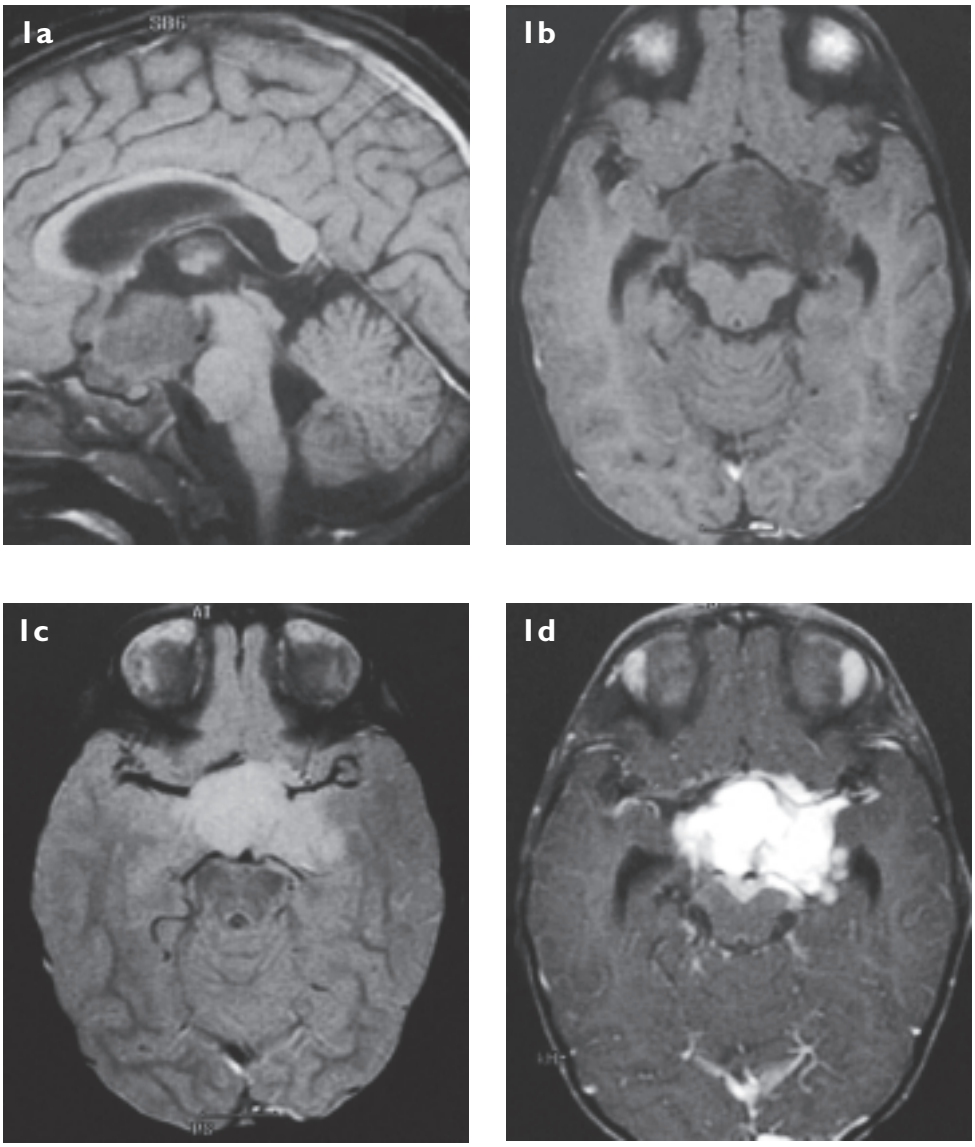

Fig. 1 (a) Unenhanced sagittal T1-weighted MR image. (b) Unenhanced axial T1-weighted (c) Unenhanced axial proton density-weighted and (d) enhanced T1-weighted MR images taken at the level of the suprasellar region.

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CASE PRESENTATION

A 15-month-old boy presented with failure to thrive. Physical examination revealed third degree malnutrition. He was however an alert and hyperactive child. Neurological examination did not reveal any neurological deficit. What do the cranial magnetic resonance (MR) images (Figs. 1a - 1d) show? What is the diagnosis?

IMAGE INTERPRETATION

Unenhanced sagittal T1-weighted MR image (Fig. 1a) and unenhanced axial T1-weighted MR image (Fig. 1b) show a large lobulated hypointense mass in the suprasellar region. The mass is homogeneously hyperintense on the axial proton density-weighted (Fig. 1c) and T2-weighted (not shown) MR images, and enhances homogeneously (Fig. 1d). Mild dilatation of the temporal horns and encasement of the circle of Willis by the mass are seen. Absence of subcutaneous scalp fat is noted on the T1-weighted MR images.

DIAGNOSIS

Hypothalamic glioma with diencephalic syndrome.

CLINICAL COURSE

The patient underwent a craniotomy for a subtotal resection of the tumour. Histopathological examination revealed a juvenile pilocytic astrocytoma. Nineteen days after surgery, the patient was discharged from the hospital. On a follow-up physical examination two months later, his nutritional status had improved. He was subsequently lost to follow-up.

DISCUSSION

Diencephalic syndrome is a complex of signs and symptoms related to hypothalamic dysfunction[1]. This syndrome is a rare cause of failure to thrive in infancy and early childhood. It is characterised by profound emaciation despite normal caloric intake, absence of cutaneous adipose tissue, locomotor hyperactivity, euphoria, and alertness[2]. It occurs almost exclusively in association with space-occupying lesions of the hypothalamic-optic chiasm region, mainly low grade glioma, and more precisely, juvenile pilocytic astrocytoma[1]. It has also been reported in association with other lesions such as midline cerebellar astrocytoma, suprasellar ependymoma, suprasellar spongioblastoma, and thalamic tumour[2]. Classically, this syndrome affects children younger than 12 months, with a mean age of six months for symptomatic onset. The diagnosis may be delayed because brain tumours are often not initially considered.

The astrocytoma associated with this syndrome is larger, occurs at a younger age, and is often more aggressive than other astrocytomas arising in this region[2]. On computed tomography (CT), suprasellar juvenile pilocystic astrocytomas are isodense to slightly hyperdense, and enhance homogeneously. On MR images, they are iso- to hypointense on T1-weighted images, and hyperintense on proton density- and T2-weighted images, with homogeneous enhancement. Dissemination of the tumour throughout the cerebrospinal fluid pathways may be seen as discrete subarachnoidal nodules in the cerebral and spinal compartments, and as areas of diffuse leptomeningeal enhancement on CT and MR images[1,2].

Besides diencephalic syndrome, the neuroendocrine disorders associated with chiasmatic or hypothalamic gliomas are precocious puberty and hypopituitarism[3]. There is also an association of these tumours with neurofibromatosis type 1. Other tumours or diseases that can cause a suprasellar mass in children include craniopharyngioma, germinoma, pituitary adenoma, Langerhans cell histiocytosis, granulomatous diseases, and Rathke cleft cyst.

Craniopharyngioma

Craniopharyngiomas account for 50% of suprasellar tumours in children. Their incidence in children peaks between five to 10 years of age. Craniopharyngiomas are divided into two histologic types, namely, papillary and adamantinomatous. Some tumours have mixed histology[4]. Papillary tumours are found almost exclusively in adults. Adamantinomatous tumours are mixed solid-cystic or mainly cystic lobulated suprasellar or intrasellar/suprasellar tumours occurring in children and adolescents[5]. They often encase the vessels of the circle of Willis.

Children with adamantinomatous craniopharyngiomas present most often with signs and symptoms of increased intracranial pressure. Visual disturbances due to compression of the optic apparatus are also frequently seen. Others present with pituitary hypofunction because of compression of the pituitary gland, pituitary stalk, or hypothalamus[6]. Adamantinomatous craniopharyngiomas have a characteristic appearance on CT. Ninety percent have a cystic component and 90% are at least partially-calcified. The calcification may be seen as a thin circumferential rim (usually around the cyst) or as chunks of calcium within the solid portion of the tumour (Figs. 2, 3). Intravenous administration of contrast medium results in enhancement of 90% of craniopharyngiomas[4].

The most characteristic MR imaging finding is a suprasellar mass that contains a cystic component that is well-defined, internally uniform and hyperintense (Fig. 4) on both T1- and T2-weighted images[6]. The hyperintensity of the cyst on T1-weighted images is due to its high protein content[5,7]. The cystic component may be isointense or hypointense on T1-weighted images. The solid component, which is frequently partially-calcified, is seen as a heterogeneous area. A moderate degree of enhancement of the solid component of the

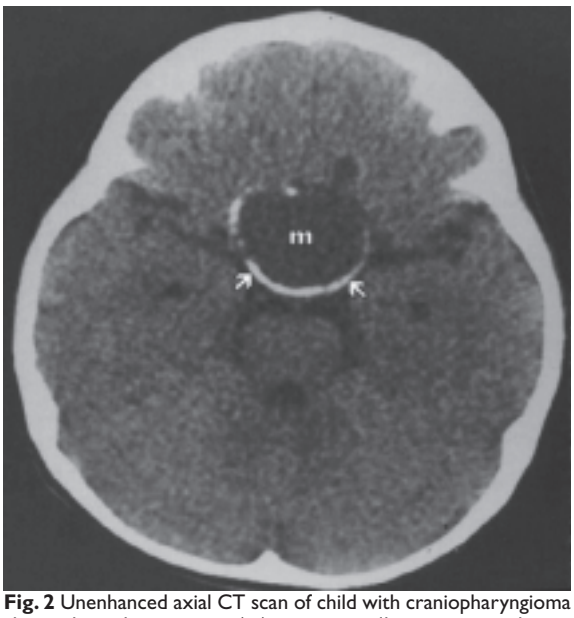

Fig. 2 Unenhanced axial CT scan of child with craniopharyngioma shows hypodense mass (m) at suprasellar region with rim calcification (arrows).

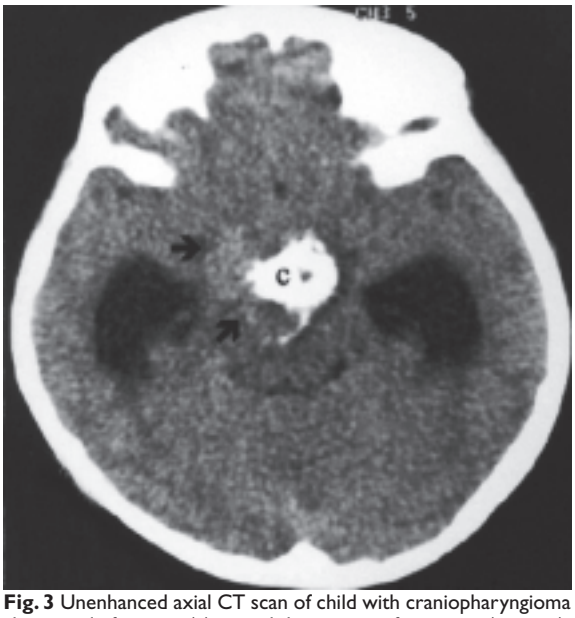

Fig. 3 Unenhanced axial CT scan of child with craniopharyngioma shows calcification (c) in solid portion of tumour (arrows). Note dilatation of temporal horns.

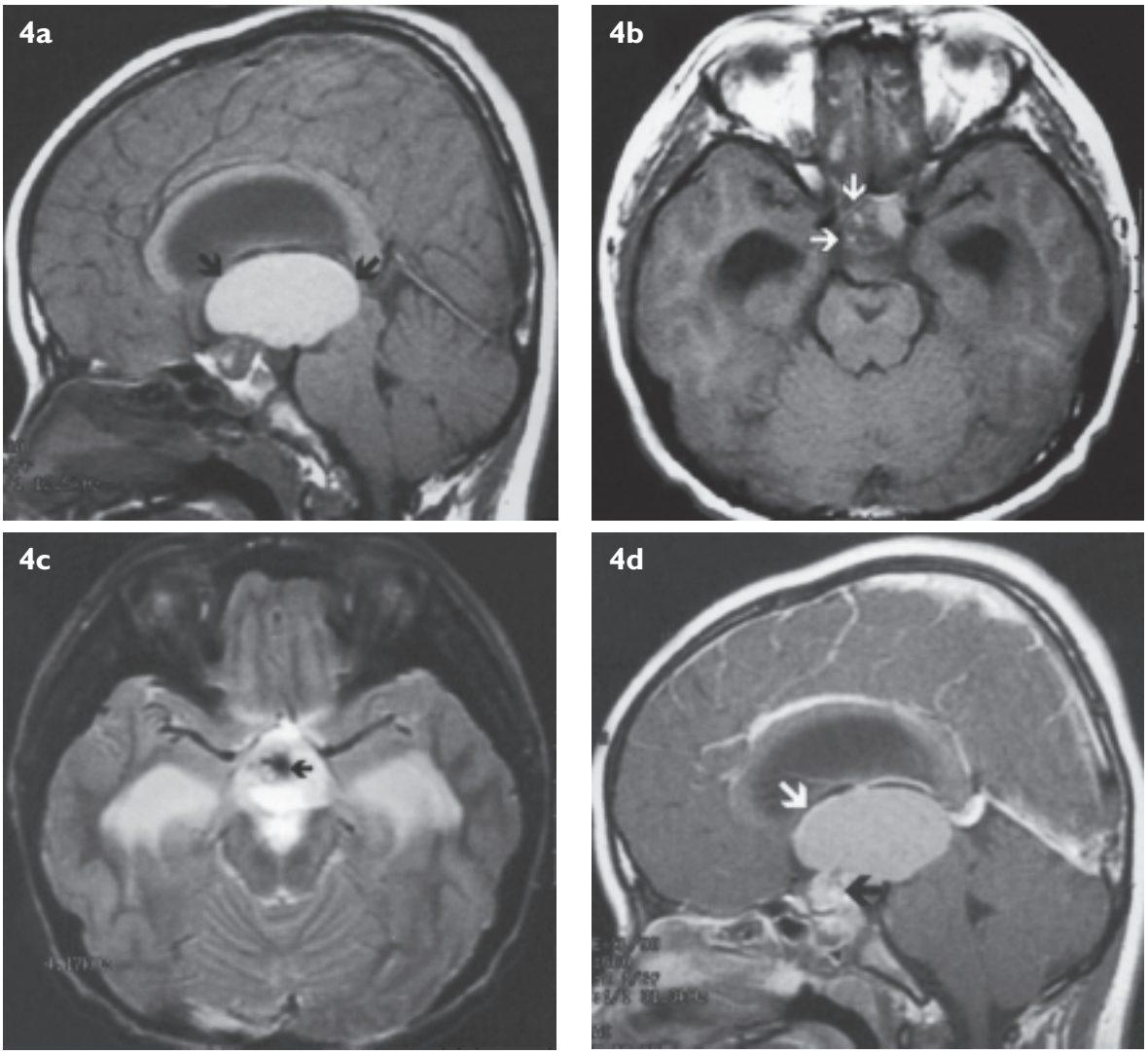

Fig. 4 MR images of child with craniopharyngioma. (a) Unenhanced sagittal T1-weighted MR image shows suprasellar mass with intrasellar extension. The cystic component of the mass (arrows) is hyperintense. (b) Unenhanced axial T1-weighted MR image shows solid component (arrows) of mass, which is heterogenously intense. Note dilatation of temporal horns. (c) Corresponding unenhanced axial T2-weighed image shows slightly hyperdense mass. The small hypointense area (arrow) within mass represents calcification. (d) Enhanced sagittal T1-weighted MR image shows enhancement of solid component of mass (black arrow) and enhancement of wall of the cyst (white arrow).

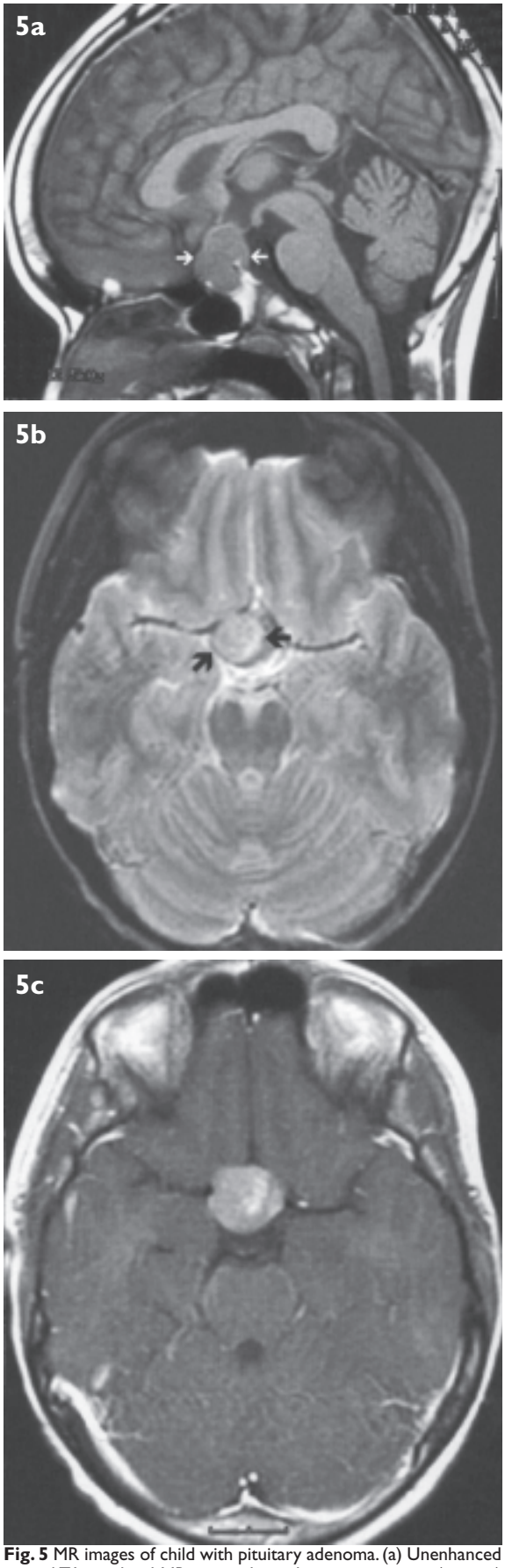

Fig. 5 MR images of child with pituitary adenoma. (a) Unenhanced sagittal T1-weighted MR image shows hypointense mass (arrows) arising from pituitary fossa. (b) Unenhanced axial T2-weighted MR image shows slightly hyperintense mass (arrows). (c) Enhanced axial T1-weighted MR image shows enhancement of mass.

tumour is seen on enhanced T1-weighted MR images. The thin walls of the cyst nearly always enhance.

Germinoma

About 20%-35% of intracranial germinomas occur in the suprasellar region[4,6]. They are usually associated with symptoms indicating hypothalamic involvement such as diabetes insipidus, emaciation, or precocious puberty. They occur in the pituitary stalk and floor of the third ventricle. On CT, they appear as well-marginated iso- to hyperdense masses. Uniform enhancement of the tumour is seen. On MR images, they usually appear as well-marginated tumours that are either round or lobulated. They are mildly hypointense on T1-weighted images, and iso- to hyperintense with grey matter on T2-weighted images, with marked enhancement.

Hypothalamic hamartoma

Hypothalamic hamartomas are rare congenital malformations composed of normal neuronal tissue. Boys are affected more often than girls. The most common presenting symptom is precocious puberty. On CT, hypothalamic hamartomas are isodense with grey matter. On MR images, they are well-defined within or adjacent to the tuber cinereum or mamillary bodies. They are isointense with grey matter on T1-weighted sequences and isointense to slightly hyperintense on T2-weighted sequences[4,6,7], without enhancement.

Pituitary adenoma

Pituitary adenomas are uncommon in children. The clinical manifestations depend on tumour size, hormonal activity, and how far they extend beyond the sella. These manifestations include delayed onset of puberty, galactorrhoea, primary or secondary amenorrhoea, gigantism, and hypercortisolism. Symptoms such as headache or visual symptoms related to mass effect may also occur. Macroadenomas may invade the cavernous sinuses and extend superiorly into the suprasellar cistern, compressing the optic chiasm[3].

MR images (Fig. 5) typically show a mass arising from the pituitary fossa, that is hypointense on T1-weighted images, and compresses normal pituitary tissue. These masses are often hyperintense on T2-weighted images. Enlargement of the sella is almost always seen because of their slow growth and late presentation[6]. They enhance uniformly and intensely.

Langerhans cell histiocytosis

Children with systemic Langerhans cell histiocytosis may have involvement of the pituitary stalk

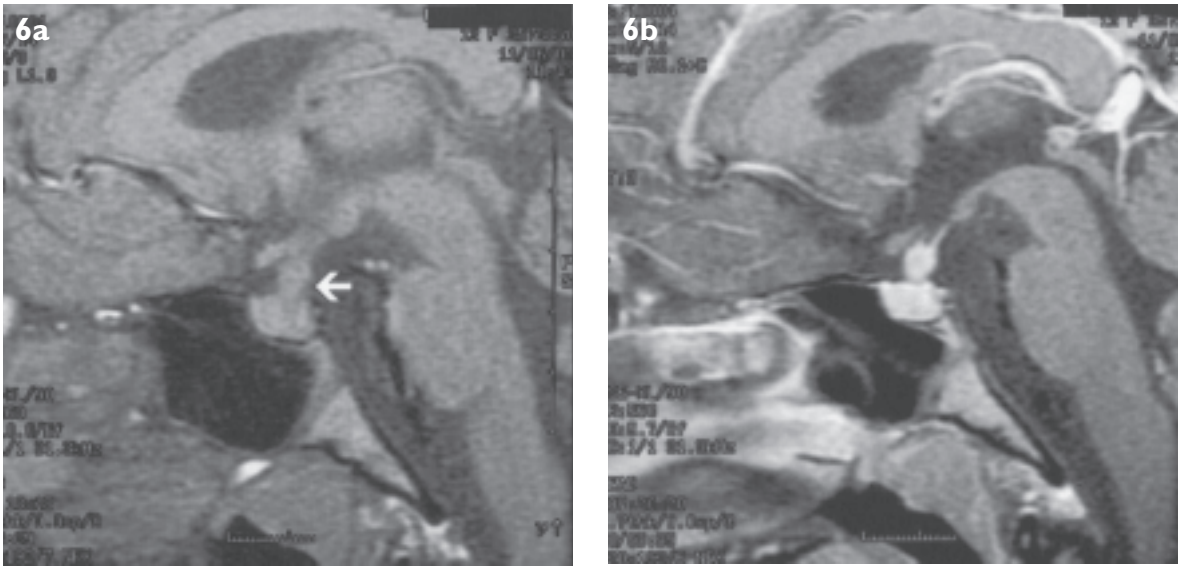

Fig. 6 MR images of child with Langerhans cell histiocytosis. (a) Unenhanced sagittal T1-weighted image shows thickening of pituitary stalk (arrow) and absence of posterior pituitary bright spot. (b) Enhanced sagittal T1-weighted image shows intense enhancement of thickened pituitary stalk.

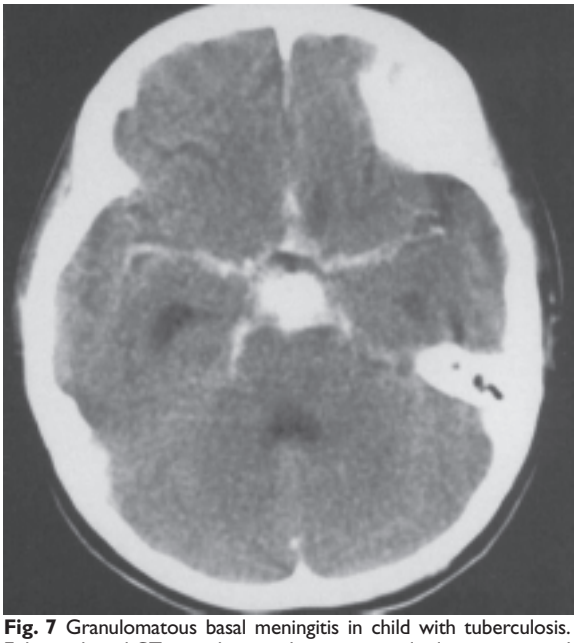

Fig. 7 Granulomatous basal meningitis in child with tuberculosis. Enhanced axial CT scan shows enhancing irregular lumpy mass and abnormal enhancement of the basal cistern.

and hypothalamus. These patients often present with diabetes insipidus. MR imaging shows loss of the normal posterior pituitary bright spot. MR imaging may reveal a solitary mass in the region of the median eminence of the pituitary stalk or thickening of the infundibulum (Fig. 6). The lesion is isointense on T1-weighted images and hyperintense on T2-weighted images, with marked enhancement[3,6].

Granulomatous diseases

Granulomatous basal meningitis is relatively uncommon in patients with intracranial tuberculosis, and is characterised by diffuse or circumscribed granulomatous involvement of the meninges at the base of skull. It often causes visual disturbances, compressing the optic nerve and chiasm. On CT, an irregular lumpy enhancing mass can be seen at the basal cistern as well as dense basal enhancement (Fig. 7). On MR imaging, the granulomatous portion of the basal meninges is hypointense on T2-weighted images. On T1-weighted images, a mass isointense to brain is seen. On enhanced MR images, the basal meninges are heterogeneously intense[8].

Rathke cleft cyst

Most Rathke cleft cysts are small, asymptomatic, and found only at autopsy. Symptoms occur if the cyst has enlarged sufficiently to compress the pituitary gland or optic chiasm. The contents of the cysts are usually mucoid, and are less commonly filled with serous fluid or desquamated cellular debris. On CT, cysts with mucoid content may be hyperdense whereas cysts containing serous fluid are hypodense[7]. On MR imaging, the cysts with mucoid fluid are hyperintense on both T1- and T2-weighted images. The cysts with serous fluid are isointense to cerebrospinal fluid on all pulse sequences[6]. Rathke cleft cysts have no calcification and do not enhance.

ABSTRACT

A 15-month-old boy presented with failure to thrive. Physical examination revealed third degree malnutrition. MR imaging showed a large lobulated mass in the suprasellar region. The mass was hypointense on T1-weighted MR images, hyperintense on proton density-weighted and T2-weighted MR images, and enhanced homogeneously. The patient underwent a craniotomy for a subtotal resection of the tumour. Histopathological examination revealed a juvenile

pilocytic astrocytoma that caused the diencephalic syndrome. Clinical presentation and imaging findings of the various tumours or diseases that can cause suprasellar masses in children are discussed and illustrated via further examples.

Keywords: Suprasellar masses, Magnetic resonance imaging, Computed tomography, hypothalamic glioma, juvenile pilocytic astrocytoma

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ERRATA

The authors of the SMJ article "Adult Community Acquired Bacterial Meningitis in a Singaporean Teaching Hospital. A Seven-Year Overview (1993-2000)" in the Decemeber 2002 SMJ wish to make a correction to the original table (Table II) they submitted. The correct table is as follows:

Table II. NUH meningitis cases.

Case number	Sex	Age	Temp (°C)	Neck stiffness	CSF pressure (CM)	CSF white cells per µl	CSF white cells differentials (%)	CSF Protein (mg/dl)	CSF glucose (Blood glucose) (mmol/l)	CSF Culture	CSF Latex Agglutination	Blood Culture
1	F	14	39.6	yes		4030	P79	1.28	2.8 (6.1)	n	n	S. pneumoniae
2	M	23	38.6	yes	29	540	P61L26	0.78	3.4 (4.6)	n	n	n
3	F	65	36.7	no		2340	P40L56	2.47	2.8 (7.4)	n	n	n
4	F	36	36.8	no	high	3150	P94L4	1.18	3.0 (5.1)	Group B strep	Group B strep	Group B strep
5	M	61	37.5	no		380	P93	1.02	5 (9.7)	n		n
6	M	50	35.2	yes		2088	P86	6	1.2 (9.3)	n	N. meningitidis	n
7	F	46	39.8	yes		300	P60	7.39	0.8 (9.8)	n	S. pneumoniae	S. pneumoniae
8	F	45	36.5	no	38	810	P81	1.08	1.2 (5.4)	n	n	n
9	M	35	38.5	yes		891	P13L85	1.19	3.2 (5.2)	n		Group B strep
10	F	44	40	no	34	433	P38L53	2.63	2.5 (7.3)	n	n	n
11	M	64	38	no	26	873	P40	1.91	3.5 (7)	n		n
12	M	27	39	yes	26	370	mostly P	2.86	4.5 (7)	n	n	n
13	F	64	39.5	yes	42	300	P38L44	3.87	5.3 (16.8)	n	n	L. monocytogenes
14	M	42	40.8	yes		136	P24L69	1.92	<0.6 (6.4)	L. monocytogenes	n	L. monocytogenes
15	M	60	39.3	yes		13000	P98	6	0.5 (6.2)	S. pneumoniae	n	S. pneumoniae
16	M	20	febrile	yes	33	1400	P84	1.97	1.1 (5.7)	N. meningitidis	H. influenzae	n
17	M	32	38.7	yes	31	4050	P85L12	3.99	4.5 (7.1)	n	n	n
18	M	70	36.7	yes	13.8	1035	P84L16	13.65	10 (17.6)	n	n	K. pneumoniae
19	M	62	37.7	yes	18	954	mixed P/L	5.02	0 (9.4)	S. pneumoniae	S. pneumoniae	S. pneumoniae
20	M	55	38.9	yes	23	25	P94L5	1.94	0.1 (5.4)	n		S. suis
21	M	71	37.6	yes	23	216	mostly P	20	4.8 (12.8)	n		n
22	M	40	36	yes	22	28	P88L14	>12	<0.6 (11.2)	Group B strep	Group B strep	Group B strep
23	M	47	39.6	yes		244	P50L46	15.7	0.8 (15)	n	n	n
24	M	59	37.8	yes	19	133	P84L2	14	6.5 (13.3)	n	n	K. pneumoniae
25	M	72	35	yes	low	3645	P82L19	0	5.3 (14)	n		n
26	M	64	38.6	yes		1210	P90L8	8.4	2.1 (7.7)	n	n	N. meningitidis

P: polymorphs, L: lymphocytes, L.: Listeria S: Streptococcus, strep: streptococci, N: Neisseria, H: Haemophilus, K:Klebsiella

The first author of the SMJ article "The Pattern of Utilisation and Accuracy of a Commercial Nucleic Acid Amplification Test for the Rapid Diagnosis of *Mycobacterium Tuberculosis* in Routine Clinical Practice" in the August 2002 issue would also like to make the correction to the name. It should be K C Yee.