Clinics in Diagnostic Imaging (49)

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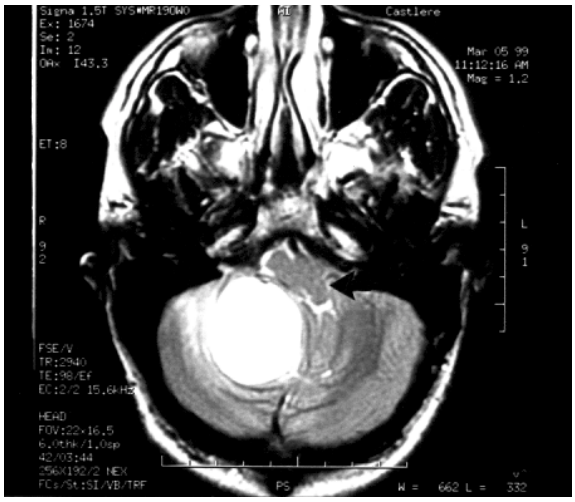

Fig. 1a Axial T2-weighted (T2W) MR image of the posterior fossa.

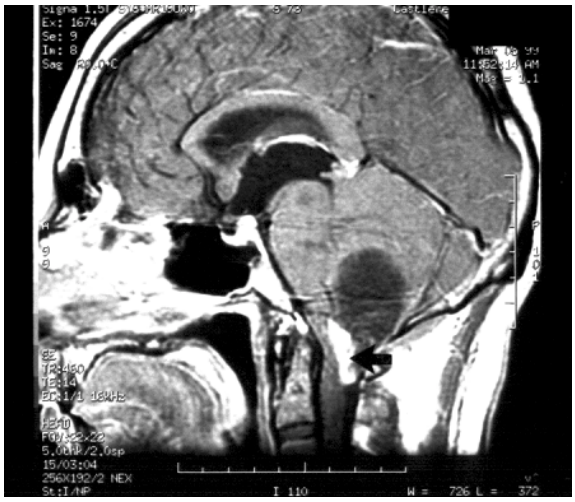

Fig. 1b Enhanced sagittal T1 W MR image of the brain taken in the midline.

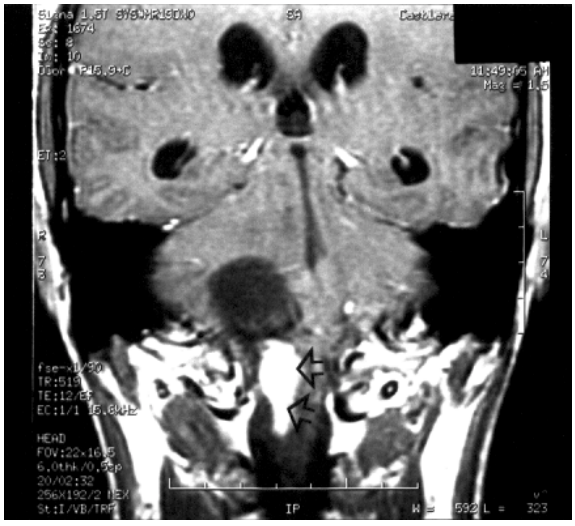

Fig. 1c Enhanced coronal T1 W MR image of the posterior fossa.

CASE PRESENTATION

A 39-year-old Caucasian man was investigated after sustaining minor injuries following a motor vehicle accident. Cranial computed tomography (CT) showed a cystic mass in the posterior cranial fossa. The patient recalled that prior to his accident, he had progressive occipital headaches radiating to the upper neck. He also noticed recent diplopia and blurring of vision. Physical examination showed bilateral papilloedema. All other cranial nerves were noted to be intact. Motor power and tone were normal in all four extremities. Cranial magnetic resonance (MR) imaging was performed (Figs. 1a-c). What do the MR images show? What is the diagnosis?

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IMAGING INTERPRETATION

MR scans of the brain showed a large cystic intrinsic mass, measuring 5 cm x 4.2 cm x 2 cm, arising from the right inferior cerebellar hemisphere and extending into the right cerebellar tonsil. Moderate surrounding white matter oedema, with shift of the medulla oblongata and upper cervical cord to the left, was noted (Fig. 1a). The contents of the cystic lesion were of low signal on T1-weighted and high signal on T2-weighted images, consistent with fluid. The margins of the lesion were better defined following intravenous injection of Gadolinium-DTPA. A moderate-sized oval-shaped strongly-enhancing nodule (arrowed) was seen at the antero-inferior aspect of the lesion, with its base resting against the cervicomedullary junction (Fig. 1b). The fourth ventricle was compressed and deviated to the left. The lateral and third ventricles were moderately dilated as a result of the obstruction by tumour (arrowed) at the fourth ventricular level (Fig. 1c).

Bilateral vertebral digital subtraction angiography (DSA) showed the primary blood supply to be arising mainly from slightly hypertrophied branches of the right vertebral and posterior inferior cerebellar arteries. There was also minor secondary supply from the lateral spinal branches of the right vertebral artery at the level of the first cervical vertebra (Figs. 2a-b). A dense uniform blush, corresponding to the site of the strongly-enhancing nodule seen on MR imaging, was seen. These findings were maximal in the mid- to late arterial phases.

DIAGNOSIS

Cerebello-tonsillar haemangioblastoma

CLINICAL COURSE

Twelve hours after the vertebral angiogram, the patient started to lose consciousness and became unresponsive for 2-3 minutes. Breathing was noted to be heavy and deep. Signs of posterior fossa compression were evident. He was taken to the operating theatre for immediate posterior fossa craniotomy and upper cervical laminectomy. Surgery revealed the right cerebellar tonsil to be herniated through the foramen magnum. A tumour nodule was visible within the right cerebellar tonsil. The cystic component of the lesion was medial and superficial to the inferior cerebellum, near to the 4th ventricle. The entire tumour nodule and cyst were completely resected. The histopathological diagnosis was that of a cerebello-tonsillar haemangioblastoma. The patient made an excellent recovery and was discharged from hospital a week later.

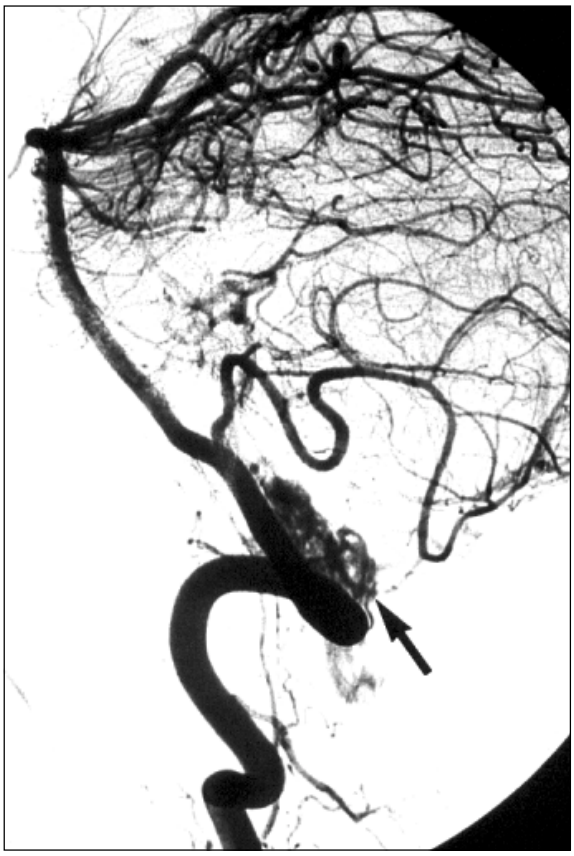

Fig. 2a DSA of the right vertebral artery (lateral projection) of the same patient as in Fig. 1. Mid-arterial phase image shows the mural nodule of the tumour (arrows) being supplied by numerous hypertrophied branches of the posterior inferior cerebellar and right vertebral arteries.

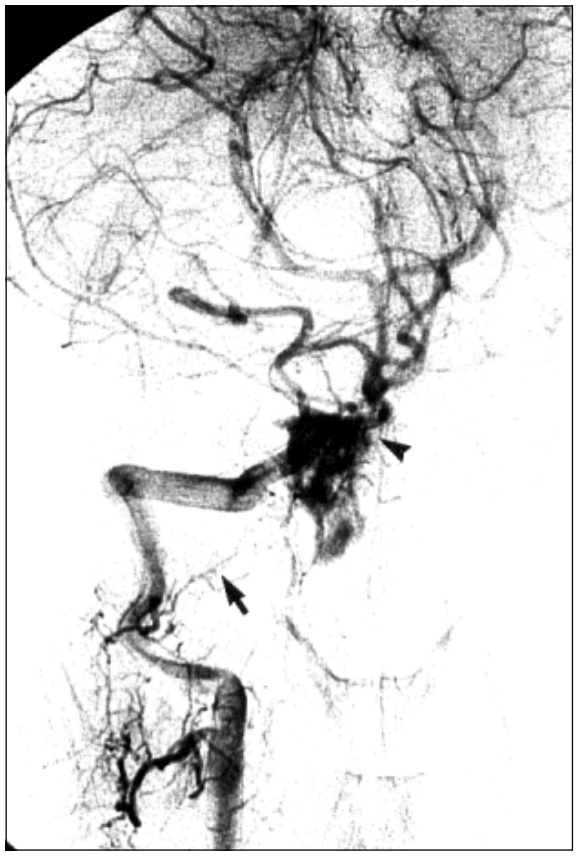

Fig. 2b Right vertebral angiogram (frontal projection) of the same patient as Fig. 2A. Late arterial phase image shows the tumour nodule (arrowhead) being partly supplied by a lateral spinal branch (arrow) of the right vertebral artery.

DISCUSSION

Haemangioblastomas are benign tumours which occur predominantly in the posterior fossa and account for about 1%-2% of all intracranial tumours[1]. Between 80% and 85% of tumours arise from the cerebellum where they comprise up to 7.3% of primary posterior fossa tumours in adults[1]. Characteristically, the lesions present in the third through the fifth decade of life, with a slight male predilection[1]. Affected patients usually have a longstanding clinical history of vague neurological symptoms. Acute symptoms are usually related to sudden raised intracranial pressure, forcing the patient to seek medical advice. Common symptoms include worsening headaches, vomiting, dizziness/vertigo and visual disturbance. The frequently encountered neurological signs are nystagmus, unsteady gait, and papilloedema[2].

Haemangioblastomas may cause polycythemia in some cases[2]. There is also a strong association between medullo-spinal haemangioblastoma and syringomyelia[2]. Between 10% and 20% of solitary haemangioblastomas are associated with the von Hippel-Lindau (VHL) syndrome[3]. The presence of other haemangioblastomas in the central nervous system (CNS) or additional visceral tumours are mandatory for establishing a diagnosis of the VHL syndrome. Pancreatic cysts (70%), renal cell carcinoma (51%) and renal cysts (49%) are the commonest associated visceral tumours, while retinal angiomatosis are the most frequently encountered CNS lesion[4].

Morphologically, haemangioblastomas are seen as well-circumscribed masses. The lesions are characteristically cystic and contain a solid mural nodule situated along the cyst wall. The tumour nodule usually abuts against the pial surface. These morphological features are faithfully reflected in the imaging findings. The cyst contains mucoproteins, amino acids, erythroprotein and haemorrhagic products which usually give its contents a xanthochromic/rusty brown appearance. Up to 30% of haemangioblastomas are reported to be entirely solid[4,5]. Microscopically, haemangioblastomas consist of multiple dilated thin-walled vascular channels which are lined by plump endothelial cells situated in a background of abundant sheets of large pale stromal cells. The cyst wall comprises compressed brain parenchyma and occasionally, reactive neuroglial tissues. Necrosis and haemorrhage may rarely occur but mitotic figures are not usually present[1].

The salient imaging feature of haemangioblastoma is a strongly enhancing mural nodule. This is characteristically demonstrated on both CT and MR imaging following intravenous contrast, although the mural nodule is usually isodense relative to brain on unenhanced CT and is therefore not distinctive (Figs. 3a-b). Sometimes, the nodule is too small to be defined by CT but DSA, with its excellent contrast resolution, is able to delineate it during the mid-arterial/capillary phase. The angiographical features are quite unique and characteristic, showing maximum contrast

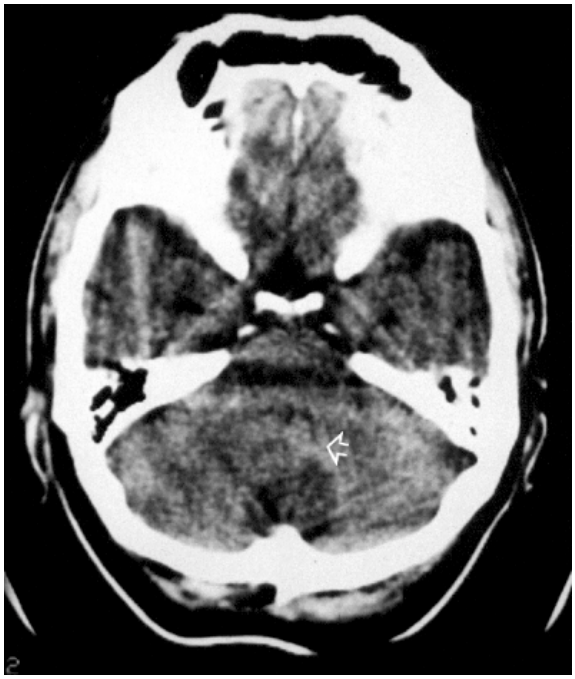

Fig. 3a Unenhanced axial CT scan through the posterior cranial fossa of another patient shows an ill-defined soft tissue density mass (arrow) anterior to a small cystic lesion.

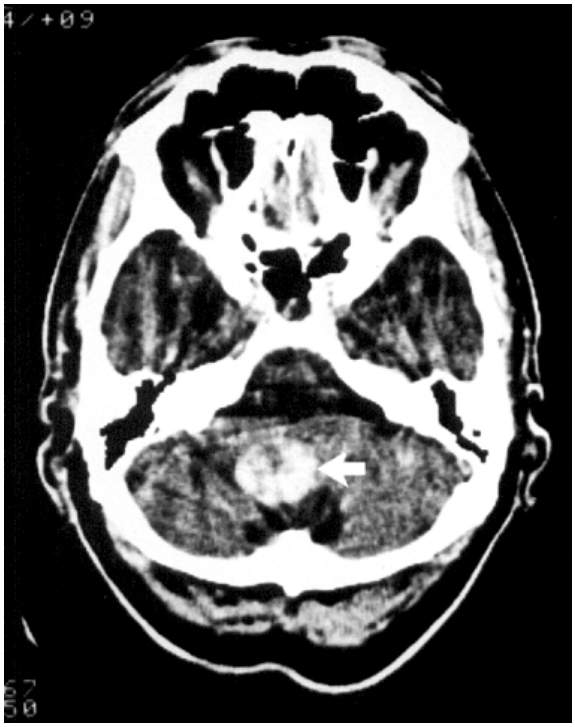

Fig. 3b Enhanced axial CT scan of the same patient as Fig. 3A shows a markedly- enhancing well-demarcated mass representing the solid component of the lesion (arrow). A haemangioblastoma was found at surgery.

uptake by the tumour nodule in the arterial phase which is occasionally associated with an early draining vein. Prior to the advent of MR imaging, the angiographical features were considered virtually pathognomonic, showing hypervascularity, focal nodules and prominent capsular venous channels[6].

Currently, MR imaging is the imaging modality of choice for depicting the signal characteristics, size, texture and extent of haemangioblastomas[5]. Enhanced CT is also useful in showing the cystic and nodular components of the tumour but it lacks the superior soft tissue contrast of MR imaging. Typically, a solid enhancing mural nodule is situated along the wall of a non-enhancing cyst. Sometimes, a solid mass with multiple small internal cysts is the main feature. The typical haemangioblastoma is hypo- to isointense on T1-weighted, and hyperintense on proton density and T2-weighted MR images. In other instances, foci of high signal may be seen within the solid parts of the haemangioblastoma. This T1 shortening may be caused by blood breakdown products and the presence of the lipid-containing stromal cells which are the predominant components of the tumour[7]. Serpiginous signal voids (SSV) indicating its increased vascularity are occasionally visible within or along the surface of the lesion[5,7].

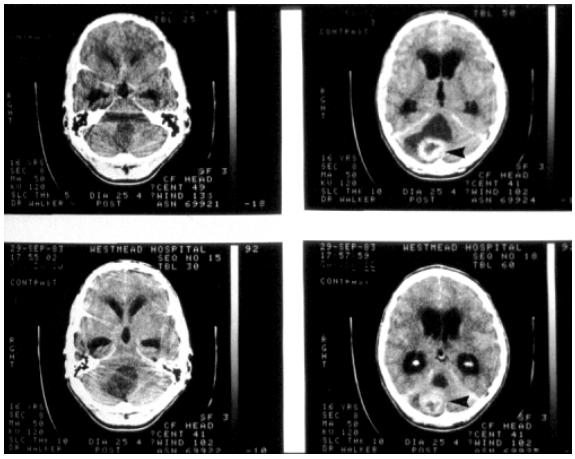

Fig. 4 A series of enhanced axial CT scans through the posterior fossa show a doughnut shaped enhancing mass within a partly- cystic lesion (arrowheads) located in the midline of the cerebellar vermis. Obstructive hydrocephalus is evident. At surgery, a low grade part cystic and part-solid astrocytoma was confirmed.

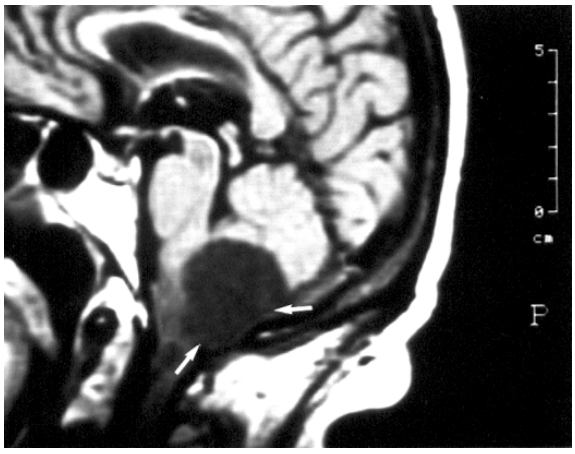

Fig. 5 Unenhanced mid-sagittal T1W MR image through the inferior vermis shows a 4 cm diameter well-defined low signal intensity lesion extending to involve the tonsil (arrows). No intramural nodule is noted in this cystic astrocytoma.

As illustrated in this case, the mural nodule may be situated next to the pial aural surface, sometimes making differentiation from cystic astrocytoma associated with a mural nodule difficult (Fig. 4). Very occasionally, a mural nodule is not discernible even on MR imaging. The cystic component of the haemangioblastoma is then indistinguishable from a cystic astrocytoma (Fig. 5). In these rare instances, however, cerebral angiography can highlight the differentiating feature since the mural nodule in haemangioblastoma will show marked contrast pooling, whereas the structures within a cystic astrocytoma will not. The other main conditions to be considered are a solitary secondary deposit and a foramen magnum meningioma (Fig. 6). Occasionally, a solid lesion with marked SSV and moderate white matter oedema, mimicking a glioblastoma, is encountered. Where haemangioblastomas are multiple, widespread secondary deposits and cavernous haemangiomas must be considered in the differential diagnosis[4]. Cavernous haemangiomas, however, do not show the intense enhancement that characterise haemangioblastomas[7].

Histopathologically, controversy still exists in distinguishing true haemangioblastomas from angioblastic meningiomas. Russell and Rubenstein favoured the diagnosis of meningioma if a lesion with a dural attachment is seen[1]. Surgery may be complicated by bleeding, particularly where the lesions are vascular. Pre-operative embolisation of the tumour using a coaxial catheter system is particularly helpful as selective ablation of the tumour nidus can be accomplished without obliterating the adjacent vessels[8]. It has been stressed that at the upper cervical spinal level, haemangioblastomas characteristically receive their blood supply predominantly from a single pedicle, making preoperative particulate embolisation reasonably safe, effective and rewarding[9]. Surgical resection is considered curative, provided the nodule representing the vascular tumour is completely excised. The cystic portion of the tumour is merely an expression of its secretory products and the reactive gliotic change of the surrounding brain parenchyma[1,4,5]. The prognosis for solitary cerebellar haemangioblastoma is good, with over 80% of patients surviving for 5-20 years following surgical removal[4].

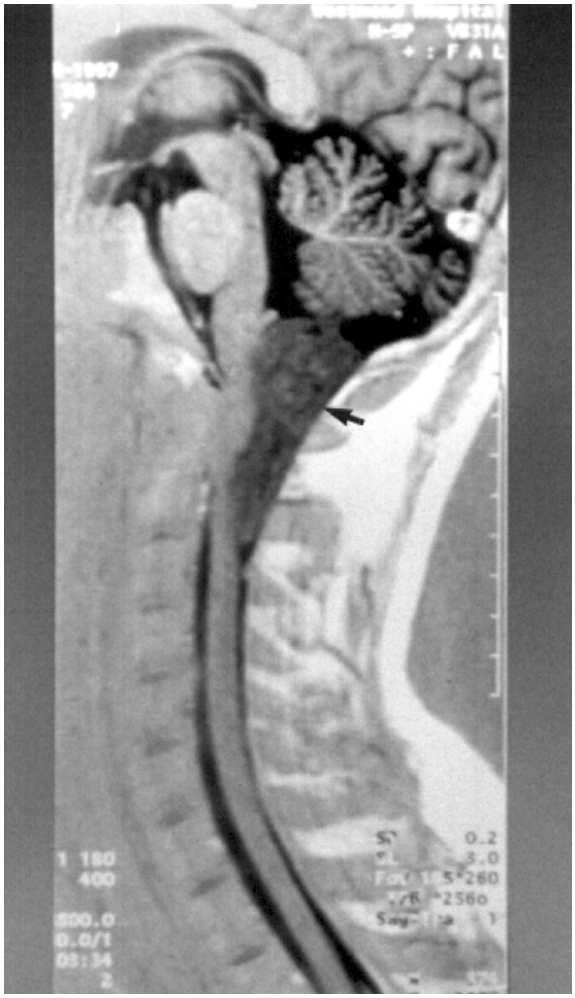

Fig. 6a Unenhanced mid-sagittal T1W MR image taken through the posterior fossa/ foramen magnum region shows a 5 cm long solid extrinsic mass situated posterior to the cervicomedullary junction. The lesion is located below and separate from the inferior vermis.

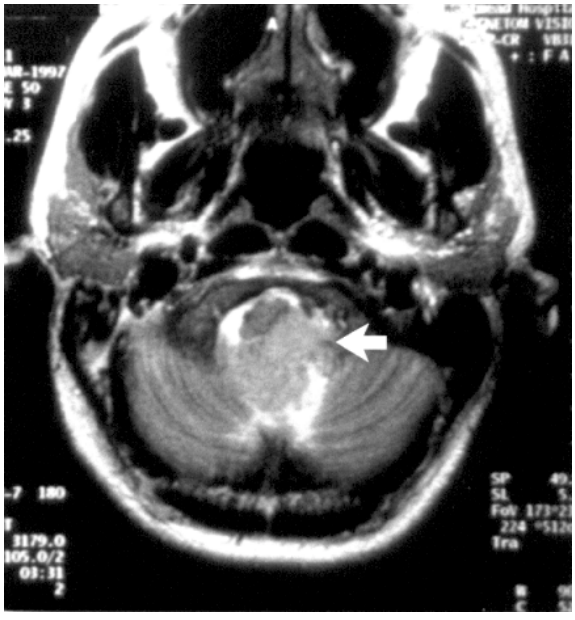

Fig. 6b Axial T2 W MR image taken through the foramen magnum of the same patient as Fig. 6a shows that the lesion is of intermediate signal intensity, suggesting its cellular nature (arrow). A meningioma was found at surgery.

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ABSTRACT

A 39-year-old man with cerebello-tonsillar haemangioblastoma is presented, with emphasis on its morphological and imaging features. Although both computed tomography and cerebral angiography are very useful imaging modalities in helping to establish a radiological diagnosis, magnetic resonance (MR) imaging is more accurate and sensitive in depicting the lesion nature and extent. MR imaging is especially reliable in detecting multiple and other associated lesions within the central nervous system, such that a conclusive diagnosis of the von Hippel-Lindau syndrome can be made in selected cases. It is important to identify isolated haemangioblastomas as they constitute a relatively high percentage of benign resectable tumours within the posterior fossa. The differential diagnosis and imaging features of other posterior fossa tumours are also discussed.

Keywords: Haemangioblastoma, Cerebellar tonsil, Cystic astrocytoma, Digital subtraction angiography, Magnetic resonance imaging

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