CASE REPORT

An uncommon mimic of spontaneous subarachnoid haemorrhage

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We here presented a rare disease entity with a clinical presentation mimicking aneurysmal subarachnoid haemorrhage. A 43-year-old woman presented with a 1-week history of neck pain and dizziness. Computed tomography of brain showed communicating hydrocephalus and subarachnoid hyperintensity suspicious of previous subarachnoid haemorrhage. Investigations revealed no underlying vascular lesion and leptomeningeal biopsy showed diffuse melanocytosis. We go on to discuss the diagnostic features and clinical course of this entity.

Case report

A 43-year-old woman presented with first episode of seizure and a 1-week history of neck pain and dizziness in February 2010. She had a similar attack but without a seizure 1 month earlier. Computed tomography (CT) of the brain showed communicating hydrocephalus and a tinge of subarachnoid hyperintensity suspicious of previous subarachnoid haemorrhage (Fig 1a). Angiography did not reveal any underlying vascular lesion. The hydrocephalus was managed eventually with a ventriculoperitoneal shunt and cerebrospinal fluid was noted to be xanthochromic.

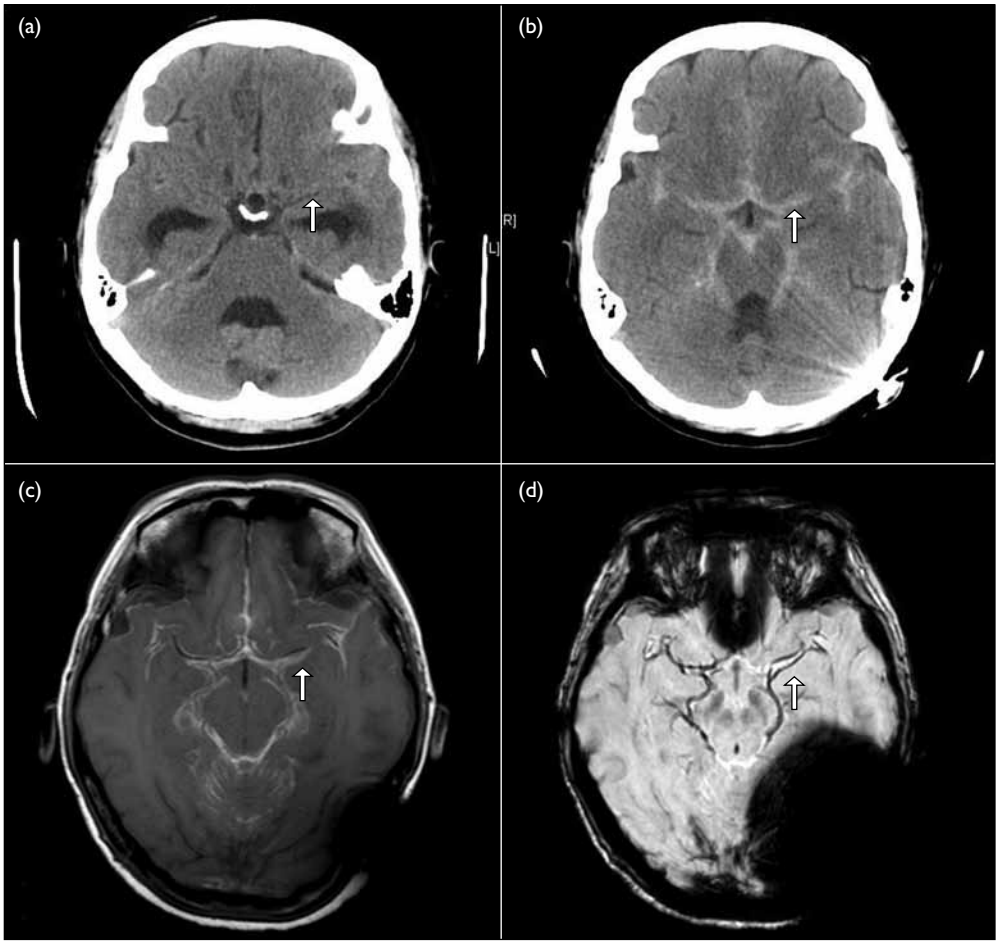

FIG 1. (a) Initial plain computed tomography (CT) shows communicating hydrocephalus and a tinge of subarachnoid hyperintensity suspicious of previous subarachnoid haemorrhage (arrow); (b) plain CT 1 month later shows leptomeningeal hyperintensity mimicking subarachnoid haemorrhage (arrow); (c) non-contrast T1-weighted brain magnetic resonance imaging (MRI) shows basal leptomeningeal hyperintensity (arrow) and artefact from the ventriculoperitoneal shunt; (d) venous blood oxygen level–dependent MRI brain shows no blooming artefact (arrow) [suggestive evidence of blood product]

Key words
Melanoma; Melanosis; Meningeal neoplasms; Subarachnoid hemorrhage

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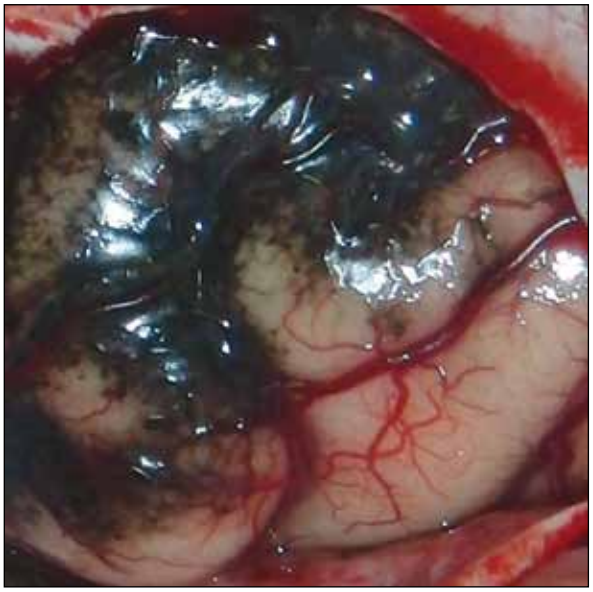

FIG 2. Intra-operative photo shows leptomeningeal dark confluent lesions

A month later she was readmitted with headache, vomiting, seizure, and paraparesis. Brain CT showed basal leptomeningeal hyperdensity (Fig 1b). Magnetic resonance imaging (MRI) revealed leptomeningeal T1 hyperintensity (Fig 1c), negative for blooming artefact suggesting the presence of blood product (Fig 1d), and enhancement in the brain and spinal cord. Craniotomy revealed confluent dark lesions over frontal lobe and basal cisterns with adhesions to the undersurface of the dura (Fig 2). Leptomeningeal biopsy confirmed diffuse melanocytosis. Her neurological condition deteriorated rapidly and she succumbed 1 month later.

Discussion

Diffuse leptomeningeal melanocytosis is a rare condition. It involves the pathological proliferation of melanocytes and production of melanin results in diffuse pigmentation over the surface of the brain and spinal cord.[1] When there is histopathological evidence of pial invasion, it is considered to be malignant (melanomatosis).[2]

The condition of diffuse leptomeningeal melanocytosis or melanomatosis is often associated with a rare phakomatosis called neurocutaneous melanosis, which was absent in our patient. It is a condition of diffuse melanocytosis with giant or disseminated congenital nevi of the skin. About 25% of patients with diffuse melanocytosis have cutaneous manifestation.[3] The CT and MRI appearance of melanocytosis includes diffuse thickening of the leptomeninges with homogeneous contrast enhancement, with or without focal nodule(s). In the MRI, the region of melanocytosis shows T1 shortening (hyperintensity) due to the paramagnetic properties of melanin.[4]

The diagnosis of melanocytosis can be difficult due to the non-specific clinical and radiological findings, especially in patients with a short history. Computed tomography of cistern hyperintensity can be similar to picture of subarachnoid haemorrhage, as in our patient. Alertness to this rare differential diagnosis and MRI are keys to make the correct diagnosis.

Treatment is mainly palliative. Neither chemotherapy nor radiotherapy is effective against melanocytosis.[5] The prognosis is poor, even in the absence of histological malignancy. Most patients deteriorate rapidly after the onset of neurological symptoms and soon die from the disease.

擬似自發性蛛網膜下腔出血的一個罕見病例

本文報告一個臨床特徵與動脈瘤性蛛網膜下腔出血非常相近的罕見病例。一名43歲女性出現頸痛及暈眩已達一星期。腦電腦斷層影像顯示病人有交通性腦積水及蛛網膜下增強，懷疑是蛛網膜下腔出血。仔細檢查後發現病人並無潛在的血管病變，而腦膜活檢則顯示瀰漫性黑色素細胞增多症。本文續討論有關此病的診斷症狀及臨床特徵。

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