The association between thymoma and MG is known to be related to the intratumorous generation of mature T-cells. These cells play an immunological stimulator role in MG in that they can synthesize anti-ACh receptor antibodies.[2] Thus, thymectomy is considered to be the most effective treatment for achieving sustained improvement, as well as remission in patients with MG. Most neurologists favor the use of this procedure.[8] The presence of ectopic thymic tissue could have prognostic value in determining the outcome of the operation.[9] However, the majority of reported ectopic cervical thymomas have a benign clinical course with no recurrence of the tumor, metastasis or myasthenic symptoms.[3,5] In cases of multiple remnant thymoma, intractable myasthenic symptoms and poor prognosis have been reported.[9] In contrast to previous cases, our patient had only cervical thymoma, but myasthenic symptoms were not well controlled until the mass was excised.

This case is the first report of MG associated with ectopic cervical thymoma, and suggests that inspection and removal of ectopic cervical thymoma is very important for the treatment of MG.

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Synchronous moyamoya syndrome and ruptured cerebral aneurysm in Alagille syndrome

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Abstract

Moyamoya syndrome and cerebral aneurysm formation are rare cerebrovascular manifestations of Alagille syndrome. Although previously reported in isolation, occurrence of these complications in a single patient has not been described. We report clinical and imaging features of synchronous moyamoya syndrome and ruptured cerebral aneurysm in a patient with Alagille syndrome.

Keywords: Alagille syndrome; Moyamoya syndrome; Cerebral aneurysm

1. Introduction

Alagille syndrome, or arteriohepatic dysplasia, is an uncommon autosomal dominant congenital multisystem disorder consisting of bile duct hypoplasia associated with a variety of cardiovascular, skeletal, ophthalmologic, and neurologic abnormalities.[1] Rare cerebrovascular manifestations of this condition include moyamoya syndrome, an idiopathic vascular occlusive disease, and aneurysm formation, both of which have previously been reported in isolation.[2–6] To the best of our knowledge, concurrent

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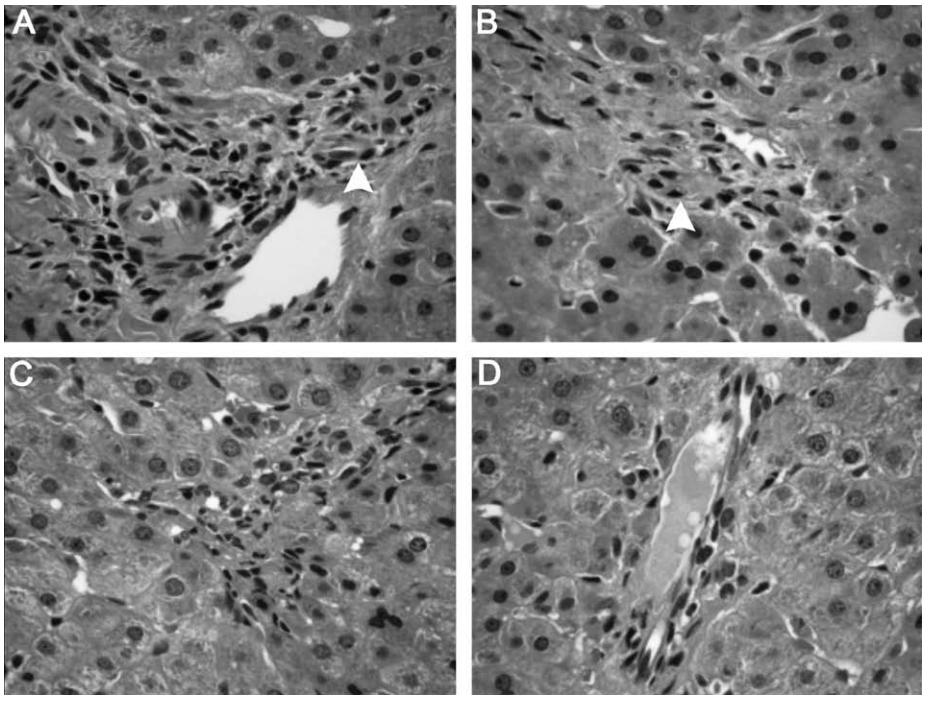

Fig. 1. Histologic sections (hematoxylin and eosin, magnification×60) reveal typical biliary ductal changes seen in Alagille syndrome. Panels A and B show portal regions with bile duct remnants (arrowheads). In panels C and D, portal tracts display absent bile ducts.

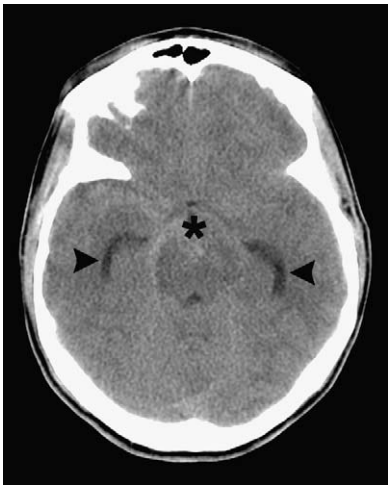

Fig. 2. Non-contrast axial head CT scan demonstrates subarachnoid hemorrhage, with blood occupying the suprasellar cistern (asterisk). Evidence of hydrocephalus includes diffuse cortical sulcal effacement and enlargement of temporal horns of lateral ventricles (arrowheads).

development of these abnormalities in a single patient with Alagille syndrome has not been described. Herein, we present and discuss clinical and radiologic findings of synchronous moyamoya syndrome and cerebral aneurysm formation in a patient with Alagille syndrome.

2. Case report

A 28-year-old woman with Alagille syndrome was brought to the emergency department after developing sudden onset of emesis and malaise followed by a syncopal episode. The patient had previously been in her usual state of good health, and was maintained on cholestyramine for pruritis, the only symptomatic manifestation of her Alagille syndrome at the time. Her liver chemistries on presentation demonstrated mild cholestasis (total bilirubin of 1.3 mg/dL [normal 0–1.2 mg/dL] and alkaline phosphatase of 444 units/L [normal 40–125 units/L]). The patient's diagnosis of Alagille syndrome had been established by liver biopsy (Fig. 1) obtained during infancy, in combination with characteristic clinical features (cholestasis, heart murmur and typical facial features).

Neurological work-up was initiated, and CT scan of the head (Fig. 2) revealed subarachnoid hemorrhage. Following placement of an external ventricular drainage (EVD) catheter, initiation of mannitol and phenytoin for treatment of cerebral edema and seizure prophylaxis, and intubation, cerebral angiography was performed for further evaluation of the subarachnoid hemorrhage. Posterior circulation angiography (Fig. 3A) demonstrated a small, wide-neck vertebrobasilar junction aneurysm as the presumed source of bleeding. Assessment of the anterior cerebral circulation at that time showed evidence of chronic occlusive disease of both internal carotid arteries compatible with angiographic moyamoya syndrome (Fig. 3B).

Because of the patient's poor neurological status, as well as aneurysm location and morphology, endovascular management of the aneurysm was elected over surgical therapy. Attempts at coil embolization were unsuccessful given the wide aneurysm neck. Therefore, treatment using stent exclusion was pursued and successfully achieved utilizing