CASE REPORT

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Cavernous Angioma of the Oculomotor Nerve

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Cavernous angiomas of the cranial nerves are rarely reported. We report a case of a 33-year-old man affected by a cavernous angioma originated in the oculomotor nerve with it's palsy. Preoperative radiological findings are difficult to differentiate it from meningioma or neurinoma. Postopertive pathological report discloses it as cavernous angioma. We discuss radiological, pathological features and management of this vascular lesion of the cranial nerve.

KEY WORDS : Cavernous angioma · Oculomotor nerve · Cranial nerves · Vascular malformation · Hemangioma.

Introduction

Cavernous angiomas are vascular lesions which occasionally involve the central nervous system. Some reports represent these lesions are account for about 8~15% of CNS vascular malformations[12]; in 75~85% of cases they have a supratentorial hemispheric localization[5]. Pathologically, cavernous angiomas are composed of large, sinusoidal vascular channels whose walls are tightly opposed to each other without any intervening neural parenchyma[17]. Cavernous angiomas arising from the cranial nerves are rarely reported. Furthermore there were only four cases of cavernous angiomas arising from the oculomotor nerve have been reported[16,18,22,23]. In this report, we describe the clinical, radiological and neuropathological findings in a patient with cavernous angioma of the third cranial nerve. And we discuss the differential diagnosis and management of this vascular lesion of the cranial nerve.

Case Report

A 33-year-old man experienced progressing pupil dilatation and ptosis of the left eye, followed by double vision during 6months. Initial symptoms of his left eye were eye dryness and pain. Ophthalmologist recognized patient's third nerve palsy about 6months later after his intial symptoms. There was the left oculomotor nerve palsy including ptosis, unreactive dilated pupil and upward and downward gaze palsy with partially impaired medial gaze in neurological examination. The MRI scan showed a small, about 7mm in diameter, extra-axial mass abutting on the left uncus and cavernous sinus. The lesion showed isointensity on both the T1 & T2-Weighted images and dense homogeneous enhancement after gadolinium administration which suggesting the meningioma(Fig. 1).

We performed a left frontotemporal craniotomy under the preoperative impression of meningioma. But we found a reddish-brown nodular mass consisted of small tortuous red vessels on it's surface. That was arising from the left oculomotor nerve just proximal to the it's entry of the cavernous sinus(Fig. 2). The outer vascular surface of the nodular mass was well dissected from neighboring tissues, but the lesion was not completely demarcated from the third nerve at the site of it's origin. Attempts to dissect the lesion from the nerve without any nerve injury were seemed to be impossible because the lesion appeared to be originated from the nerve itself. We tried to excise the lesion as much as possible with gentle manipulation of the nerve and performed near total removal of the lesion with preserving it's continuity of the oculomotor nerve. We could not find any visible feeding or draining vessels which were shown in the case of the arteriovenous malformations.

Histopathology revealed a vascular lesion composed of multiple, large, relatively thick and hyalinized channels lined by flat endothelium and separated by intervening fibroconnective tissue stroma. The vascular structures had more or less thick walls with pinkish hyalinization and their inner surface was lined by endothelium without elastic lamina

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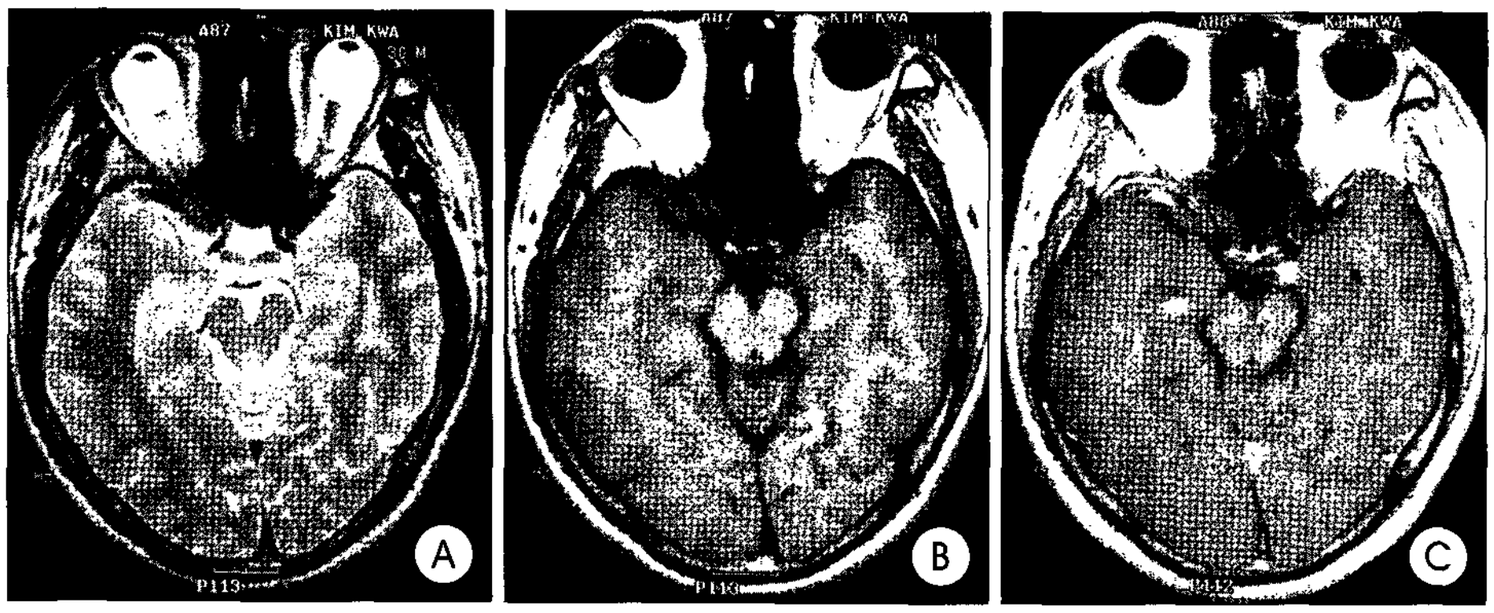

Fig. 1. A, B : T2-and T1-weighted axial magnetic resonance(MR) images showing the iso-signal intensities of the lesion. C : Gadolinium-enhanced axial T1-weighted MR images depicting a well enhanced tumor, about 7mm in diameter, abutting the uncus & cavernous sinus on the left side.

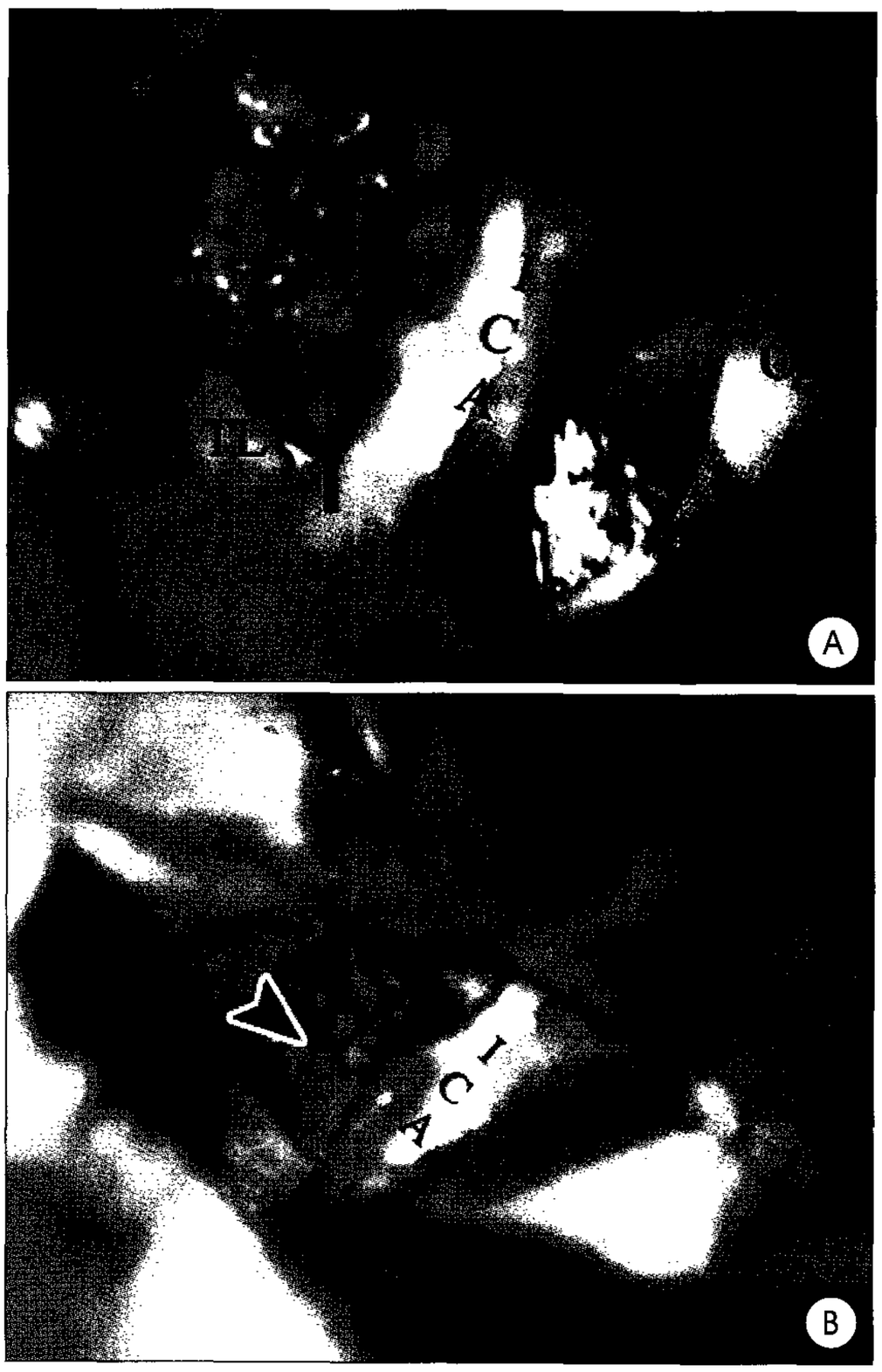

Fig. 2. Intraoperative view A : A reddish-brown nodular mass arising from the left oculomotor nerve just proximal to the it's entry to the cavernous sinus. (arrow=oculomotor nerve ; arrow head=lesion ; TL=temporal lobe ; ICA=internal carotid artery ; ON=optic nerve) B : The lesion is excised near-totally from the oculomotor nerve(arrow head) with preserving it's continuity.

(Fig. 3A). The organized thrombi were packed in some vessels and a small amount of the hemosiderin was deposited there. There was a little portion of positive finding with immune staining for neurofilament, considered some neural substance of the oculomotor nerve(Fig. 3B) which was excised together with vascular lesion. There was no staining tissue with synaptophysin. On immunohistochemical stain with smooth muscle actin, some vascular walls contained scanty attenuated smooth muscle fibers (Fig. 3C).

The patient's oculomotor nerve palsy worsened after surgery, but it became improved 2months later. The patient feels diplopia in right lateral gaze but no diplopia in primary position of the eyes at six months after surgery. Slight narrowing of the palpebral fissure and dilated pupil of the left eye did not improved but there was no evidence of recurrence in the brain MRI findings during one year and a half follow up period. He works in his previous job as an operator of crane with help of sun glasses.

Discussion

Cavernous angiomas originating from cranial nerves are rarely reported. Cranial nerves such as the optic nerve and chiasm[1,7], oculomotor, trigeminal[15], intracranial portion of the seventh and eighth nerve[4,8,9,20] and hypoglossal nerve[3] have been affected. There were four reported cases of cavernous angiomas arising from the oculomotor nerve[16,18,22,23]. These vascular lesions of the cranial nerves are presented as cavernoma, cavernous angioma, cavernous hemangioma or cavernous malformation. All these different terminology for the cranial nerve angiomas make us to be confused in understanding the true pathologic nature of the lesions. As far as we know, the difference between cavernous (hem)angiomas which belongs to the vascular malformations and cavernous type hemangiomas (cavernous hemangiomas) which belongs to the vasoformative neoplasms is not clearly defined pathologically in the literatures. It was very hard for us to make a pathologic diagnosis of our case.

According to the four reported cases of cavernous angiomas arising from the oculomotor neve there was no comments about terminological problems of the cavernous angioma in their reports. M.C.Petit-Lacour et al.[19] who reported hemangioma of the porus acusticus mentioned that cavernous hemangiomas are true tumors whereas cavernomas, often improperly called cavernous hemangiomas, are vascular malformations. But they did not describe the difference between the two vascular lesions pathologically. A. Biondi et al.[2] who

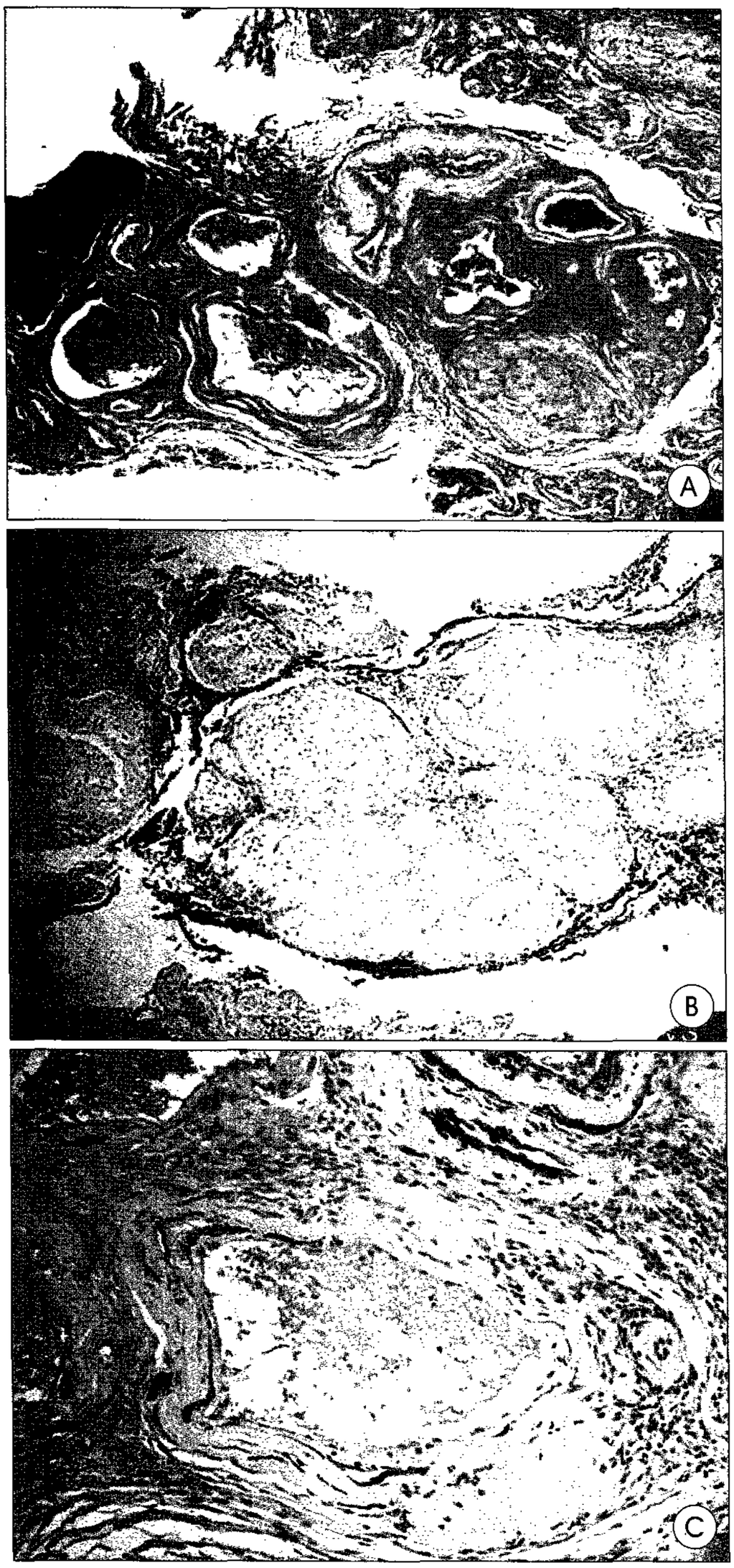

Fig. 3. A : A cluster of distended venous structures containing red blood cells. The walls are irregularly thickened and hyalinized. Thin endothelial cells are lined. The organizing thrombosis is noted(H&E, X10). B : Scanty neural tissue(brown in color) is present around the blood vessel cluster. (Neurofilament immunostain. X10) C : Some blood vessels have attenuated smooth muscle fibers (brown in color) in their walls. (Smooth muscle actin immunostain, X20).

reported five cases of intracranial extra-axial cavernous angiomas studied differences between extra-axial cavernous angioma and hemangioma of infancy. They concluded that adult intracranial extra-axial cavernous angiomas are vascular malformative lesions rather than involuted hemangiomas of infancy and recommend that the term 'hemangioma' should be reserved for the common tumoral lesion of infancy although there are some debate on it. But it was difficult for us to make a definitive diagnosis of this case with the pathologic findings. Is it cavernous (hem)angioma (vascular malformation) or cavernous hemangioma (vascular tumor)? All previously reported four cases of the oculomotor nerve angiomas are presented as cavernous angiomas and two of them are very similar to our case in the intraoperative and pathologic descriptions. But they didn't mentioned about pathological difference between vascular malformative cavernous (hem)angioma and vasoformative neoplastic cavernous hemangioma. And they did not mentioned about terminological problems of the lesion. We decided our case of the vascular lesion of the oculomotor nerve as cavernous angioma (vascular malformation) according to A. Biondi's point of view. But what is this lesion actually? These intracranial extraaxial vascular lesions to be determined whether these are vascular malformation or vasoformative neoplasm.

According to previously reported three cases of cavernous angiomas of the oculomotor nerve they transected the oculomotor nerve to achieve complete removal of the lesion and resulted in complete third nerve palsy. Upon their experience of surgical management of cranial nerve cavernous malformations, Vivek R. Deshmukh et al.[6] mentioned that these lesions pose a significant threat to cranial nerve function because of the eloquence of it's origin. They recommended surgical excision of all lesions with cranial nerve symptoms because of their aggressive clinical behavior damaging involved cranial nerves. And they emphasized that complete resection of the lesion with sharp microdissection, minimal retraction of the cranial nerve and no attempt to remove the hemosiderin-stained tissue are standard of care for recovery of nerve function and no recurrence. We tried to remove the lesion maximally with minimal oculomotor nerve injury and it was possible to preserve it's continuity of the oculomotor nerve but failed to achieve meaningful functional recovery in this case.

With differential diagnosis of this vascular lesion of the third cranial nerve, primary tumors in the intracranial portion of the oculomotor nerve should be considered. Only nine cases of schwannoma or neurofibroma have been reported[23]. Primary eosinophilic granuloma[10], primary glioblastoma multiforme[21], malignant meningioma[11] and rhabdomyoma[14] have also been found to arise from the oculomotor nerve. Schwannoma may contain angiomatous parts, which are marked in some cases[13]. However, there was no schwannoma component in our case.

The common symptoms of patients with oculomotor nerve tumors are headache, diplopia, and dysfunction of other cranial nerves. Enlargement of the tumor resultant of repeated hemorrhage supposed to be the reason of progressing the

third nerve palsy in this patient .

In most cases, radiological preoperative diagnosis may be elusive and a definitive diagnosis can only be achieved upon microscopic study of the surgical samples.

The patient has developed no additional symptoms until 18 months after surgery, but recurrence should be considered because of incomplete removal of the lesion.

Conclusion

Based on the present and other reported data, we suggest that cavernous angiomas of the cranial nerves must be considered in the differential diagnosis of intracranial extra-axial lesions presented with cranial nerves dysfunctions, together with neurinoma and meningioma. In the management of these cranial nerve cavernous angiomas, gross total removal with best efforts to preserve nerve function is the standard of care even though it is not always possible.

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