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Dissecting Aneurysm of the Recurrent Artery of Heubner in a Patient with Ssteogenesis Imperfecta Kevin Mansfield MD; Ralph Rahme MD

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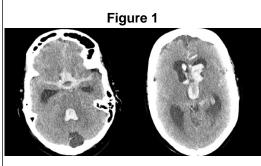


Introduction

Dissecting aneurysms of the recurrent artery of Heubner (RAH) have not been previously observed. Likewise, intracranial dissections in the setting of osteogenesis imperfecta (OI) are extremely rare.

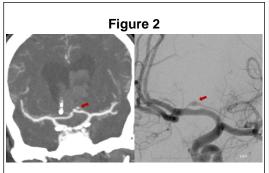
Case Report

A 50-year old woman with OI type I (autosomal dominant, COL1A2 gene mutation) presented with massive spontaneous subarachnoid (SAH) and intraventricular hemorrhage (IVH), resulting in a state of coma with decorticate posturing (GCS 5, WFNS grade 5), Following CSF diversion, her neurological exam improved slightly (GCS 6T).

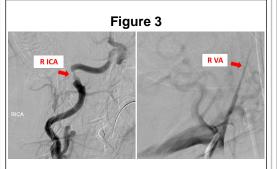


Head CT: diffuse cisternal SAH predominating in the anterior interhemispheric fissure, tetraventricular hemorrhage with hydrocephalus

The patient exhibited clinical signs of OI, including blue-tinted sclerae, joint hyperlaxity, and finger deformities secondary to multiple prior healed fractures. Interestingly, her mother had died of aneurysmal SAH and one of her maternal aunts had undergone surgical cliping of an intracranial aneurysm.

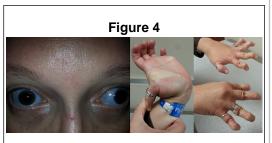


CTA and DSA: 4 mm x 1.5 mm "pearl-onstring" fusiform aneurysm of L recurrent artery of Heubner, probable mural thrombus impinging on superior aspect of A1 segment



DSA: probable prior extracranial dissections of R ICA and R VA

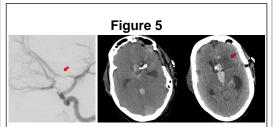
Given the location and configuration of the aneurysm, decision was made to surgically explore and attempt clip reconstruction of the RAH.



Photographs of patient's daughter demonstrate typical features of OI: bluetinted sclerae, joint hyperlaxity Under continuous intraoperative MEP monitoring, the aneurysm was exposed via a standard left pterional transsylvian approach.

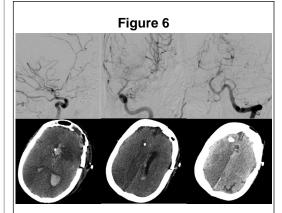


Although patency of the RAH could not be preserved, intraoperative MEPs remained normal. Therefore, aneurysm clips were not repositioned.



Intraoperative DSA: both aneurysm and RAH are completely obliterated by the clip

Postoperatively, her neurological condition was unchanged from baseline, with a persistently symmetric motor exam. Head CT showed expected small infarct in head of L caudate nucleus head without involvement of internal capsule.



DSA and CT on SAH day 6: severe diffuse vasospasm, bihemispheric infarcts (neurological deterioration to GCS 3T)

Given her poor prognosis, medical care was withdrawn at the family's request and the patient ultimately died.

Conclusions

Although rare, intracranial dissections can occur in the setting of OI and can lead to devastating intracranial hemorrhage. Dissecting aneurysms in patients with OI may occur in atypical locations and have a predilection for smaller and more distal arteries, such as the RAH. Management of such aneurysms is particularly challenging and preservation of the parent vessel is often impossible. Intraoperative angiography and neurophysiological monitoring can provide very useful feedback in this setting.

References

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