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Case Report

Pituitary enlargement in a carotid-cavernous fistula: An atypical imaging manifestation ☆

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ABSTRACT

A carotid-cavernous fistula is a rare abnormal communication between carotid vasculature and the cavernous sinus. Development of a carotid-cavernous fistula often results from trauma, but may be spontaneous in the setting of predisposing risk factors. Suspicion for a spontaneous fistula is understandably low on routine non-contrast imaging. In this article, we present a case of a carotid-cavernous fistula initially presenting with the potentially underrecognized imaging manifestation of diffuse pituitary enlargement identified on a non-contrast CT, later revealed to be due to the presence of the fistula.

Introduction

A carotid-cavernous fistula (CCF) is an abnormal communication between carotid vasculature and the venous plexus of the cavernous sinus. Presenting symptoms may include pulsatile exophthalmos, proptosis, progressive visual loss, pulsatile tinnitus, and cranial nerve palsies. CCFs can be categorized as direct (communication between intracavernous internal carotid artery and cavernous sinus) or indirect (communication via branches of the internal/external carotid circulation) which have different etiologies. Aneurysm rupture, trauma, and vasculopathies, such as collagen deficiency syndromes and fibromuscular dysplasia, can result in or increase the risk of developing a direct CCF. Noninvasive imaging with CT angiography is the initial modality of choice for evaluation of a suspected CCF. Imaging features supporting the diagnosis of a CCF include an enlarged superior ophthalmic vein, bulging cavernous sinus, and abnormally early enhancement of the cavernous sinus [1].

We report a case of a CCF initially presenting with the atypical imaging manifestation of rapid pituitary enlargement due to vascular congestion from the CCF, demonstrate the more typical imaging features also present, discuss the vascular relationship between the pituitary and cavernous sinus, and discuss the association of fibromuscular dysplasia and CCF.

Case report

A 62-year-old female presented with the gradual onset of a severe left-sided headache and constant pulsatile tinnitus. A non-contrast CT scan of the head was performed which was

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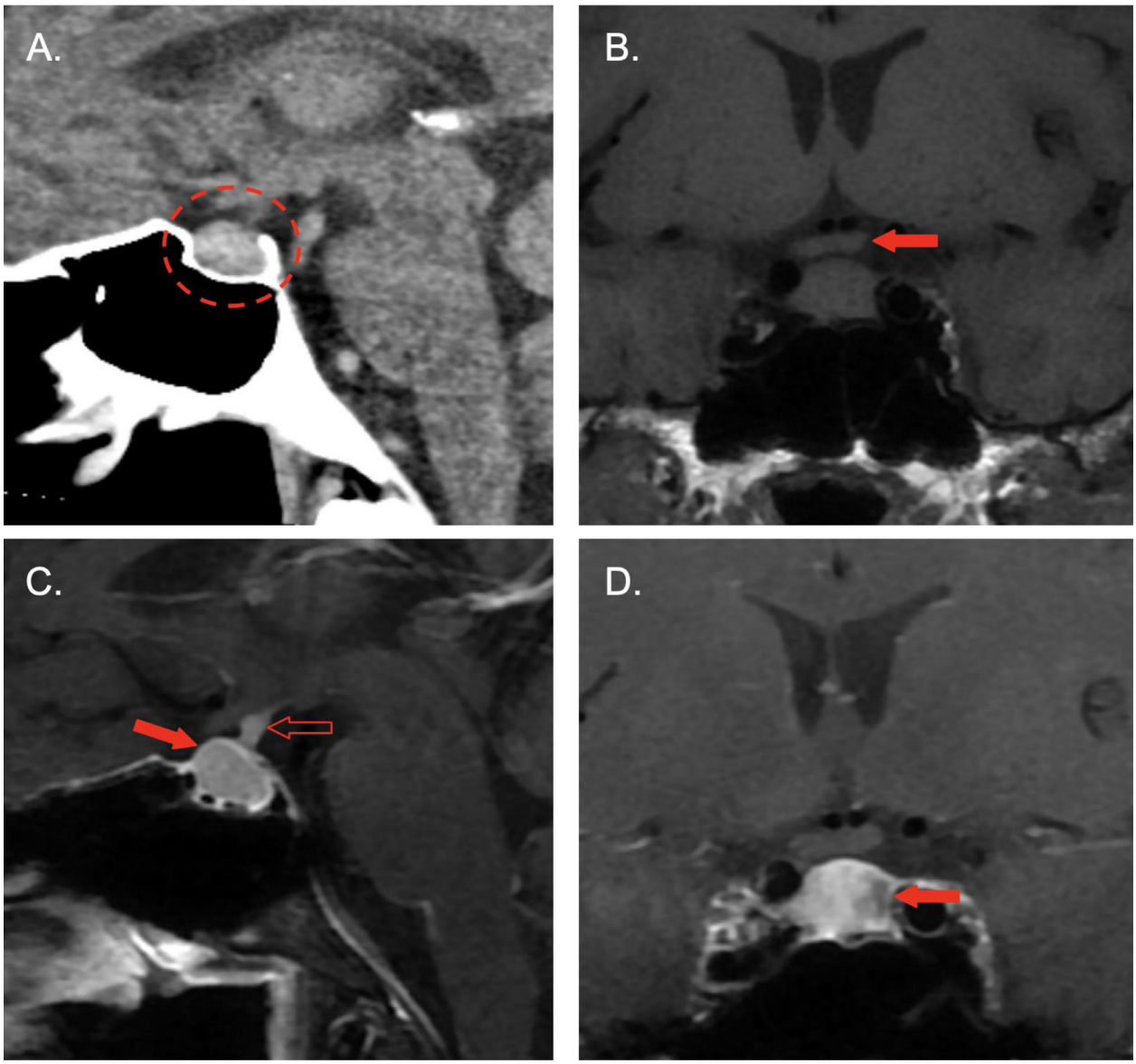

Fig. 1 – (A) Sagittal noncontrast CT of the head demonstrates a hyperattenuating pituitary gland with a superior convex margin (dashed circle). (B) Coronal T1 MRI sequence shows no mass effect of the enlarged pituitary gland on the optic chiasm (solid arrow). (C) Sagittal fat-suppressed T1 postcontrast MRI sequence demonstrates an enlarged anterior pituitary gland (solid arrow) and normal enhancement of the infundibulum (open arrow). (D) Coronal fat-suppressed T1 postcontrast MRI sequence reveals a region of heterogeneous enhancement in the left aspect of the anterior pituitary gland (solid arrow).

negative for intracranial hemorrhage, but remarkable for a hyperattenuating pituitary gland with a convex superior margin (Fig. 1A). The symptoms and CT appearance of the pituitary gland prompted further investigation with a pituitary protocol MRI. On the initial MRI, the enlarged pituitary gland approached, but did not exert mass effect upon the optic chiasm (Fig. 1B). The anterior pituitary gland demonstrated regional heterogeneous enhancement without a discrete hypoenhancing lesion (Fig. 1D). The infundibulum was normal (Fig. 1C). The patient was referred to endocrinology for workup of a suspected pituitary macroadenoma. Hormone laboratories were unrevealing.

One month later, the patient developed right-sided ptosis, diplopia, and acute onset of a right-sided headache. A noncontrast head CT and CT angiogram of the head and neck were obtained which demonstrated further enlargement of the pituitary gland (Fig. 2A). The left superior and inferior ophthalmic veins were enlarged (Figs. 2B and C) and there was abnormal bulging and early enhancement of the cavernous sinuses (Fig. 2C). Angiography of the neck was also remarkable for a beaded appearance of both cervical internal carotid arteries (Fig. 2D). Findings were interpreted as being highly suspicious of a CCF and underlying fibromuscular dysplasia. MRI brain confirmed progressive enlargement of the anterior

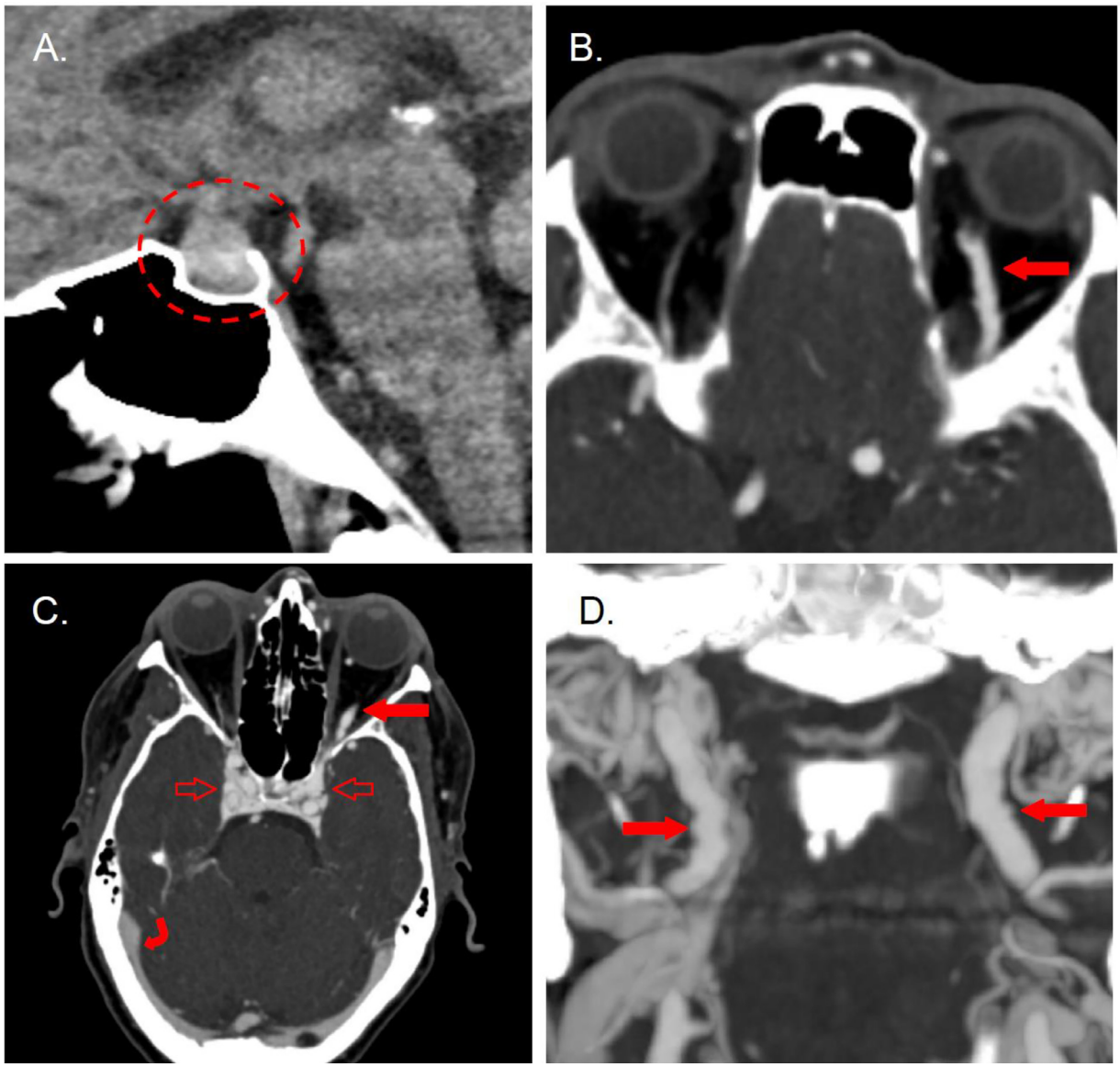

Fig. 2 – (A) Sagittal noncontrast CT of the head reveals further enlargement of the pituitary gland with cranial extension into the suprasellar cistern (dashed circle). (B) Axial CT angiogram of the head shows an enlarged left superior ophthalmic vein (solid arrow). (C) Axial CT angiogram of the head demonstrates enlargement of the left inferior ophthalmic vein (solid straight arrow), enlargement and early enhancement of the cavernous sinuses (open arrows). Expected venous enhancement is demonstrated in the dural venous sinuses (curved arrow). (D) Coronal maximum intensity projection (MIP) of the CT angiogram of the neck reveals a beaded appearance of both cervical internal carotid arteries (solid arrows).

pituitary gland which now contacted the undersurface of the optic chiasm (Fig. 3A) and the proximal aspect of the optic nerves. The pattern of enhancement was also noted to have changed compared with the prior MRI (Fig. 3B), but no discrete hypoenhancing lesion was identified.

A conventional cerebral angiogram was performed revealing a high flow left CCF with venous drainage from both cavernous sinuses to the superior and inferior ophthalmic veins, left inferior petrosal sinus, bilateral pterygoid venous plexi, and the left superficial middle cerebral vein (Figs. 4 and 5). Venous coiling of the left cavernous sinus was performed.

The patient developed complete palsy of the left 6th nerve and near complete palsy of the left 3rd nerve after coil embolization. These symptoms have gradually improved and right-sided ptosis has resolved.

Discussion

Carotid-cavernous fistulas are rare arteriovenous shunts that may be spontaneous or traumatic. Contents of the cavernous

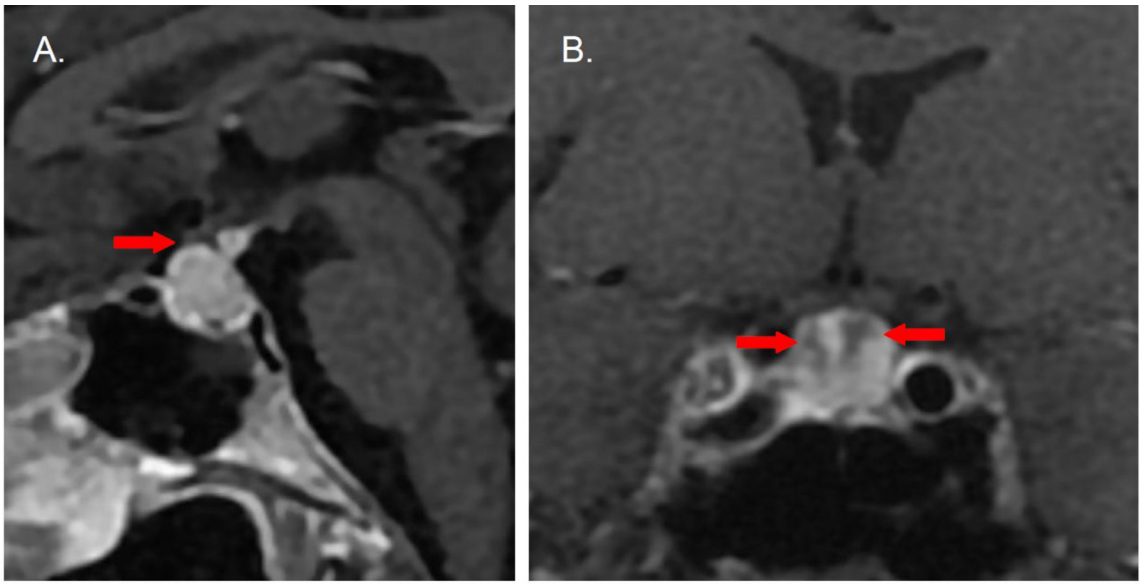

Fig. 3 – (A) 3D Sagittal CUBE T1 post-contrast MRI sequence shows enlargement of the pituitary gland which contacts the undersurface of the optic chiasm (solid arrow) and proximal aspect of the optic nerves (not shown). (B) Coronal fat-suppressed T1 postcontrast MRI sequence demonstrates a changed enhancement pattern compared with the prior MRI with linear regions of hypoenhancement (solid arrows), but no discrete lesion.

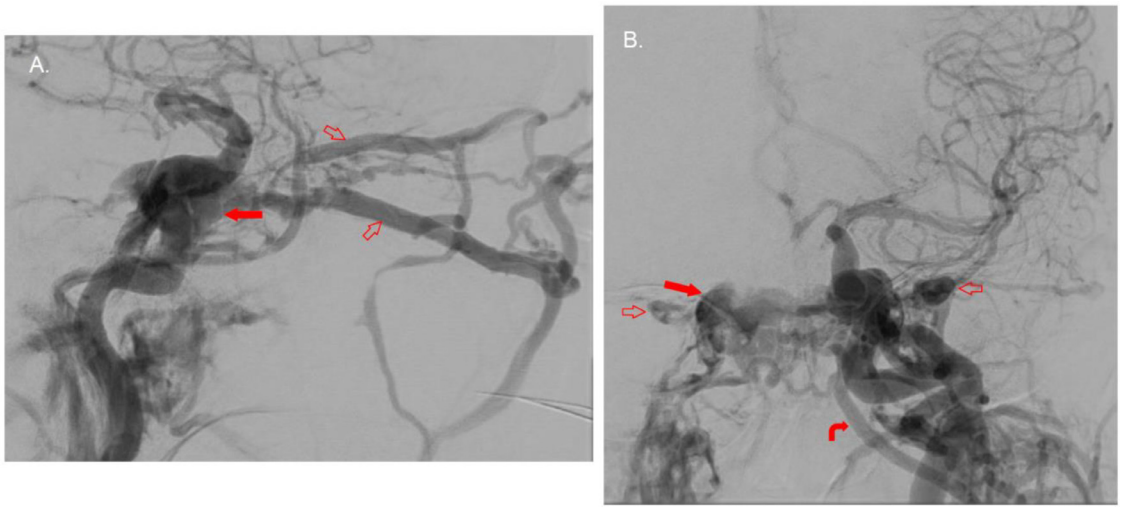

Fig. 4 – (A) Sagittal digital subtraction cerebral angiogram of the left internal carotid artery demonstrating early opacification of the cavernous sinus (solid arrow) and the superior and inferior ophthalmic veins (open arrows). (B) Coronal digital subtraction cerebral angiogram of the left internal carotid artery shows early filling of both cavernous sinuses (solid straight arrow), both superior ophthalmic veins (open arrows), and the left inferior petrosal sinus (curved arrow).

sinus include the cavernous internal carotid artery, cranial nerves III, IV, V1, V2, and VI. The presence of a fistula within this anatomically rich space allows for a relatively predictable pattern of clinical symptoms and imaging findings. Symptoms can include cranial nerve (III, IV, V1, V2, VI) palsies, pulsatile exophthalmos, proptosis, progressive visual loss, and pulsatile tinnitus. Common imaging manifestations of a CCF can include an enlarged superior ophthalmic vein, bulging cavernous sinus, and abnormally early enhancement of the cavernous sinus [1]. While several of these typical clinical and imaging features of a CCF were present in this case, the initial presentation highlights a potentially underrecognized imaging finding of diffuse pituitary enlargement in the setting of a CCF.

Detection of subtle diffuse pituitary enlargement can be challenging given its variable physiologic appearance. Imaging evaluation of the pituitary gland should include correlation with patient demographics as normal pituitary size varies with age, gender, and pregnancy status [2,3]. On average, pituitary height in females is greater than that of age-matched

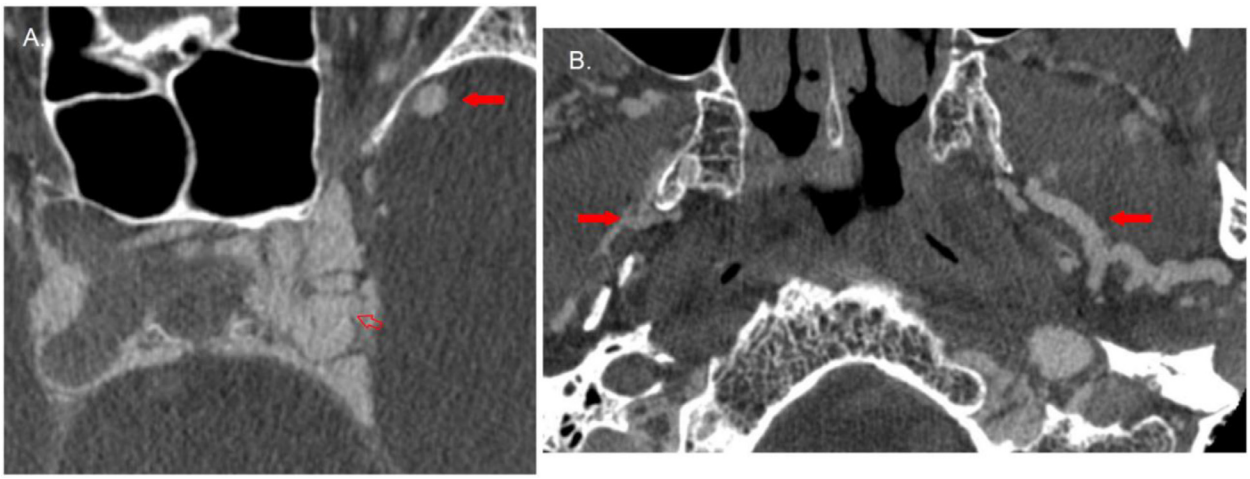

Fig. 5 – (A) Axial selective CT angiogram of the left internal carotid artery demonstrates early opacification of the left superficial middle cerebral vein (solid arrow). The presumed location of the CCF fistula is also found on this image (open arrow). (B) Axial selective CTA of the left internal carotid artery demonstrates early opacification of both pterygoid venous plexi (solid arrows).

males with pituitary height peaking in the third decade [2]. The prevalence of an upward convex margin is also greater in patients younger than age fifty [4]. Deviations from the expected pituitary morphology for age and gender, as in our case, should therefore merit further investigation if detected on routine CT or MRI. Beyond physiologic etiologies, the differential diagnosis for diffuse pituitary enlargement is broad and includes hypothyroidism [5], Addison disease [6], and neuroendocrine tumors [7]. Medications such as gonadotropin releasing hormone (GnRH) analogs and antipsychotics have also been implicated in pituitary hyperplasia [7].

This unique case demonstrated not only atypical pituitary morphology for the patient's demographics, but more importantly, rapid enlargement over 1-month period without a discrete lesion detected by MRI. The unexplained diffuse pituitary enlargement was only elucidated after the CTA of the head was obtained which demonstrated several imaging findings suspicious for a CCF. Pituitary enlargement in the setting of a CCF may occur as the cavernous sinus contains a venous plexus draining the anterior and posterior pituitary gland [8]. Abnormally high flow and pressure through the cavernous sinus may then impair normal venous drainage from the pituitary gland leading to enlargement [9]. Understanding this vascular relationship is important to prevent misdiagnosing pituitary pathology.

The CTA neck was also remarkable for a "string of beads" appearance of the cervical internal carotid arteries suggesting underlying fibromuscular dysplasia [10], an established risk factor for the development of a spontaneous CCF. This vasculopathy, affecting small and medium-sized arteries, can be complicated by dissection, aneurysms, thromboembolism relating to aneurysms, and arteriovenous fistulas [11]. Intracranial involvement of fibromuscular dysplasia can lead to subarachnoid hemorrhage from aneurysm rupture or dissection. If intracavernous, aneurysm rupture can result in a CCF [12,13] and the culprit microaneurysm may be angiographically occult [13].

As demonstrated by this case, nonspecific symptoms such as headache and tinnitus can pose a diagnostic challenge. The atypical, potentially underrecognized, imaging manifestation of rapid diffuse pituitary enlargement may occur in the setting of a CCF and should prompt further diagnostic workup if identified on routine CT or MRI. Using all imaging clues and recall of the vascular relationship of the pituitary with the cavernous sinus was of utmost importance in determining a unifying diagnosis in this case rather than considering the rapid enlargement of the pituitary as separate pathology.

Patient consent

Informed consent was obtained from the patient for publication of their case.

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