Lymphoblastic Leukaemia Presenting as a Carotid-Cavernous Fistula

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SUMMARY – *A carotid-cavernous fistula is a life-threatening condition characterised by an abnormal communication between the carotid arterial vessels and the cavernous venous system. Although these fistulae can arise spontaneously, they mainly occur after trauma, especially road traffic accidents, falls and penetrating cranial or orbital injuries. The mainstay of treatment involves endovascular embolization, but in those patients where this is not possible or where embolization fails, direct surgical intervention and ligation of the artery may be necessary. Here we describe an interesting case of a suspected carotid-cavernous fistula which turned out to be cavernous sinus syndrome secondary to lymphoma.*

Case Report

An 18-year-old male attended the Emergency Department after suffering a head injury whilst playing rugby. A non-contrast CT head was reported normal and the patient was subsequently discharged home. Nine days later, he returned to the Emergency Department complaining of persistent left frontal headache associated with one day history of vomiting, bloody discharge from the left ear and left-sided ptosis. A repeat non-contrast CT head showed hyperdensity of the left cavernous sinus and he was referred to our regional neuroscience institution for further management.

On admission, the patient was alert (GCS 15) and haemodynamically stable. Examination was unremarkable except for left-sided facial numbness in the V2 distribution of the trigeminal nerve and left ptosis. A CT head scan showed significant widening and hyperdensity of the left cavernous sinus when compared to the original CT head scan. Findings were highly suspicious of a carotid-cavernous fistula therefore a cerebral angiogram was performed on the following day. This did not demonstrate a fistula, but delayed venography revealed retrograde flow from the late-filling cavernous sinus into the superior ophthalmic vein (Figure 1). An MRI was requested to ex-

clude haematoma formation and subsequent compression of the cavernous sinus. MRI imaging demonstrated an enhancing parasellar mass extending out through the left foramen ovale into the masticator space closely associated with the mandibular division of the left trigeminal nerve (Figure 2). A smaller mass was located in the right parasellar region. These findings raised the suspicion of lymphoma and subsequent thorough examination of the patient revealed a left submandibular mass. On direct questioning the patient admitted to a non productive cough and feeling generally unwell over the past three weeks. A subsequent chest X-ray demonstrated an extensive superior mediastinal mass with superior vena cava obstruction and tracheal compression. Biopsy of the submandibular mass was inconclusive, however biopsy of a surrounding lymph node under ultrasound guidance revealed lymphoblastic leukaemia/lymphoma.

Discussion

Carotid-cavernous fistulae can be classified into four distinct categories according to the Barrow classification¹: type A is a direct shunt between the cavernous portion of internal carotid artery (ICA) and the cavernous sinus,

Figure 1 A-D) Catheter angiography. Sagittal (A,B) and coronal (C,D) images following injections into the left (B,D) and right (A,C) internal carotid arteries. No evidence of a carotid-cavernous fistula was demonstrated but during late venous imaging there was late filling of the cavernous sinus on the left (red arrow in B and D) and retrograde flow in the superior ophthalmic vein (black arrow in B and D).

whereas types B, C and D are indirect dural arteriovenous fistulae supplied by the ICA, external carotid artery or both internal and external carotid arteries respectively. Type A fistulae are high-flow and usually associated with trauma¹. In addition, they occur more commonly in males due to their increased susceptibility to traumatic incidents^{2,3}. The clinical presentation varies depending on the size and location of the fistula but common symptoms include proptosis, conjunctivitis, chemosis, corneal ulceration, pulsating exophthalmos and visual impairment^{4,5}. In addition, the diversion of arterial blood can result in ischaemia of cranial nerves III-VI causing abnormal ocular movements and altered sensation to the face in V1

and V2 distribution⁴. As our patient presented with traumatic head injury, a focal neurological deficit and vascular abnormalities on primary CT head, a carotid-cavernous fistula seemed a likely diagnosis. However, the negative finding after cerebral angiography and the appearance of a suspicious mass on MRI led us to consider other possibilities. This case demonstrates the importance of considering multiple causes of cavernous sinus syndromes (despite the convincing history for a single diagnosis). It also highlights the importance of excluding them systematically through thorough history-taking and examination. These steps are essential to provide a rapid accurate diagnosis, thereby avoiding unnecessary invasive investigations.

Figure 2 Unenhanced head CT at initial presentation (A) and at 10 days (B). The left cavernous sinus is expanded and hyperdense (white arrow). C) Coronal T1 weighted MR imaging post gadolinium. The left parasellar homogeneously enhancing mass is demonstrated extending through the foramen ovale into the masticator space (white arrow heads). There is a contralateral mass of similar signal characteristics in the right masticator space (white arrow).

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