CASE REPORT

Dural arteriovenous fistula associated with a glomus jugulare tumour presenting with only pulsatile tinnitus.

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SUMMARY

We present the second known case of a dural arteriovenous fistula (DAVF) associated with a glomus jugulare tumour in a 66-year-old man and the first with a presenting symptom of pulsatile tinnitus. The tumour occluded the left internal jugular vein at the bulb. Our patient opted for monitoring, but the tinnitus progressed and became debilitating, prompting him to proceed with embolisation of the tumour. Angiography revealed a DAVF of the left transverse sinus with retrograde flow. Embolisation of 80% of the tumour did not relieve symptoms. The patient returned for embolisation of the DAVF. Occlusion of the DAVF achieved symptomatic relief. A quandary develops during a procedure when the surgeon discovers that another intervention could satisfy the patient, while the patient is under anaesthesia. The higher flow in the DAVF likely causes the tinnitus in those with a patent sigmoid sinus, and embolisation of the DAVF alone could achieve relief.

BACKGROUND

Intracranial dural arteriovenous fistulas (DAVFs) or dural arteriovenous malformations (AVMs) typically occur in the cavernous and sigmoid venous sinuses; these account for approximately 10% to 15% of intracranial AVMs.¹ One mechanism for the pathogenesis suggests that venous outflow obstruction produces hypertension that causes ischaemia. This in turn is believed to lead to irregular angiogenesis resulting in DAVFs.² The second proposed mechanism suggests that physiological arteriovenous shunts are opened by increased sinus pressure.³ The following report presents the second published case of a patient who developed a DAVF in association with an ipsilateral glomus jugulare mass at the jugular bulb.

CASE PRESENTATION

Our patient presented with symptoms of headache and pulsatile tinnitus at 66 years old. The workup revealed an enhancing mass involving the left jugular bulb. Our patient had no cranial nerve deficits at that time and opted for only monitoring the lesion. Over the following 6 years, he developed progressive worsening of the pulsatile tinnitus. Additionally, our patient developed hoarseness and difficulty swallowing, which was attributed to an anterior cervical spine surgery that occurred during the intervening period. Our patient noted no other

neurological deficits. Over the 6 years, the tumour showed progressive growth (figure 1A–D). Surgical resection was discussed at length with our patient who was opposed. He elected to undergo endovascular embolisation in order to decrease or cure the pulsatile tinnitus.

TREATMENT

Initial embolisation

A six-vessel cerebral angiogram was performed prior to embolisation. In addition to the large vascular mass at the jugular bulb (figure 1B), a Cognard type IIa DAVF involving the left transverse sinus was identified (figure 2C). The angiogram revealed that the left jugular bulb was occluded by the tumour and provided no cerebral outflow (figure 2D); however, the left jugular vein did fill with drainage from the tumour. We proceeded with Onyx (ev3, Plymouth, Minnesota, USA) embolisation of the mass to determine if the tinnitus would resolve. We were able to obtain approximately 80% embolisation of the tumour (figure 2A,B). Postoperatively, our patient did not note any change in the pulsatile tinnitus and complained of some expected discomfort in the area of embolisation.

Second embolisation

Our patient was followed for 1 month without change in the pulsatile tinnitus symptoms. He was brought back for embolisation of the DAVF, and total occlusion was obtained of all feeding branches including a portion of the tumour (figure 2C) with some residual tumour opacification at the end of the procedure (figure 2D). Postoperatively, our patient noted complete resolution of pulsatile tinnitus and was discharged the following day with mild left-sided facial discomfort.

OUTCOME AND FOLLOW-UP

Three months postembolisation

The patient returned to the clinic 3 months postembolisation and stated that he had minor pulsatile tinnitus. He noted that the tinnitus was more severe prior to surgery. The patient also reported light-headedness, which is exacerbated with activity. He was referred to psychiatry for anhedonia. His cranial nerve deficits have been stable.

Six-months postembolisation

Via phone interview, the patient described resolution of light-headedness and anhedonia. The

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Figure 1 (A) Pre-embolisation axial MRI showing an enhancing tumour (asterisk) at the left jugular bulb. (B) Left external carotid artery injection showing a dural arteriovenous fistula (DAVF) (white arrow) and tumour blush (black arrow). (C) Left vertebral artery injection showing the DAVF (white arrow) and tumour (black arrow). (D) Left external carotid artery injection anteroposterior view showing tumour occluding left jugular bulb (black arrow) with retrograde flow (white arrow) in the left transverse sinus from the DAVF.

patient states that the pulsatile tinnitus is minimal and does not affect his daily life.

DISCUSSION

The aetiology of the turbulent flow in our case presents a unique problem as to which one or if the combination of the aetiologies caused the pulsatile tinnitus. Pulsatile tinnitus is caused by

Figure 2 (A) Left external carotid artery injection anteroposterior (AP) view after 80% embolisation of tumour (black arrow) with continued retrograde flow during the arterial phase (white arrow). (B) Left external carotid artery injection lateral view after 80% embolisation of tumour (black arrow) with remaining dural arteriovenous fistulas (DAVFs) (white arrows). (C) Left external carotid artery injection AP view after complete embolisation of DAVF (white arrow) with stable residual tumour (black arrow). (D) Left external carotid artery injection lateral view after complete embolisation of DAVF (white arrow) with stable residual tumour (black arrow).

venous hypertension and turbulent flow in the cochlea which is drained by the vein of the cochlear aqueduct to the inferior petrosal sinus and finally the jugular bulb. Therefore, flow in the sigmoid sinus is required for pulsatile tinnitus. Matsuyama published a case where it was hypothesised that a DAVF causing pulsatile tinnitus developed after sigmoid sinus thrombosis. The thrombosis was likely caused by hormonal therapy for infertility due to endocrine dysfunction from a hypothalamic hamartoma.⁴ Angiogram showed patent transverse sigmoid junction with DAVF and total relief of symptoms was achieved after embolisation.

A previously published case with a C2 schwannoma experienced bilateral pulsatile tinnitus (the contralateral inferior petrosal sinus opacified with injection). The C2 schwannoma had only distal jugular occlusion due to tumour with retrograde flow through the sinuses as ours did. They chose to treat in two stages, embolisation and resection, leading to complete resolution of the tinnitus.⁵ The DAVF recurred at 6 months and was treated with gamma knife surgery total occlusion and no recurrence at 33 months with total resolution of tinnitus.⁵ A second patient with a meningioma was treated first with embolisation of the feeding arteries, followed by excision of the meningioma, the DAVF and the left tentorium leading to complete resolution of the tinnitus.⁶ A third patient with a meningioma was treated with partial resection of the meningioma with resolution of the DAVF and tinnitus.⁷ The fourth patient with a meningioma was treated with excision of the meningioma and the invaded transverse sinus to the transverse-sigmoid junction.⁸ This case was the only one in which complete resolution of the tinnitus was not achieved; however, it was decreased postoperatively.⁸ Angiography showed unchanged DAVF at 2 months postresection and occlusion at 9 months postresection with no comment on change in tinnitus.⁸ Tinnitus response to therapy appears to depend on the resolution of the DAVF.

We present the second reported case in the literature of a glomus jugulare tumour accompanied by a DAVF, and the first presenting with only pulsatile tinnitus. The first reported case (case two from paper) was by Arnautović *et al* that presented with significantly more intracranial extension than ours and led to the predominant symptoms of cranial nerve palsies and hemiparesis with dizziness and hearing loss. Arnautović *et al* concluded that the leading risk factor for development of a transverse-sigmoid sinus DAVF was occlusion of the dominant dural venous sinus due to tumour invasion.⁶ Our case, however, revealed symmetric patent transverse dural sinuses, but was also occluded at the jugular bulb. The cause of the DAVF was more likely due to the high retrograde flow within the sigmoid sinus due to the DAVF. We showed that the highly vascular tumour was likely not the cause of the symptoms, and that one should be vigilant to rule out DAVF.

This also brings an ethical dilemma when a patient is under anaesthesia and the cause of the patient's symptoms is discovered during the procedure and is unrelated or different from the initial diagnosis. If the risk versus benefits of treatment versus undergoing repeated anaesthesia are different, then the physician must take time with added anaesthesia time to discuss with family members to obtain a new informed consent. In our case, due to the patient's age, we felt obliged to proceed with good chance of successful treatment. However, with further insight, we believe that initial embolising of the DAVF could have adequately provided relief of symptoms and he could have forgone the embolisation of the glomus tumour.

CONCLUSION

The importance of this case is to acknowledge the goals of the treatment. Due to the patients age and comorbidities, surgical resection was a poor option and embolisation was chosen. A good outcome can be achieved especially when we remember to take the anatomy into consideration for embolisation of feeding vessels to cut off flow to the inferior petrosal sinus. Therefore, for patients presenting with jugular bulb tumour and tinnitus, angiography and embolisation of DAVF are reasonable courses of action.

Learning points

- Symptomatic relief from pulsatile tinnitus with embolisation of arteriovenous (AV) fistula.
- Dural AV fistula with neoplastic occlusion of jugular bulb with pulsatile tinnitus.
- Pulsatile tinnitus requires patency of sigmoid sinus to jugular bulb.

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