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Case Report

Internal jugular vein fistula mimicking dural arteriovenous fistula after cardiac pacemaker placement ☆

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ABSTRACT

Carotid jugular arteriovenous fistulas are a documented complication of cannulation of the internal jugular vein. They may present with neck pain, headache, and cardiovascular aberrations. However, carotid jugular fistula secondary thrombus formation after jugular cannulation with radiographic presentation similar to dural arteriovenous fistula has not yet been reported in the literature. Below, we report the case of a 68-year-old male with an incidentally found carotid-jugular fistula secondary to pacemaker placement who had intracranial reflux on imaging, which was ultimately treated successfully through an endovascular approach.

Introduction

Carotid-jugular arteriovenous fistulas (CJ-AVF) are abnormal connections between the carotid artery and the jugular vein. Typical presenting signs and symptoms are dizziness, headache, arterial and venous hypertension, tinnitus, neck swelling, and a pulsatile mass. However, the presenting symptoms of CJ-AVFs may vary based on the location and size of the fistula or be asymptomatic in nature. Etiology is mostly associated with penetrating trauma to the cervical region, iatrogenic trauma, or congenital abnormalities. Overall, CJ-AVFs are rare in occurrence. Catheter angiography is the gold standard for diagnosis and endovascular repair is the most common treatment modality [1–3]. Below, we report a case of acquired CJAVF in a 68-year-old male secondary to internal jugular vein thrombosis, after automatic implantable defibrillator (AICD) placement. The pathophysiology of this fistula was

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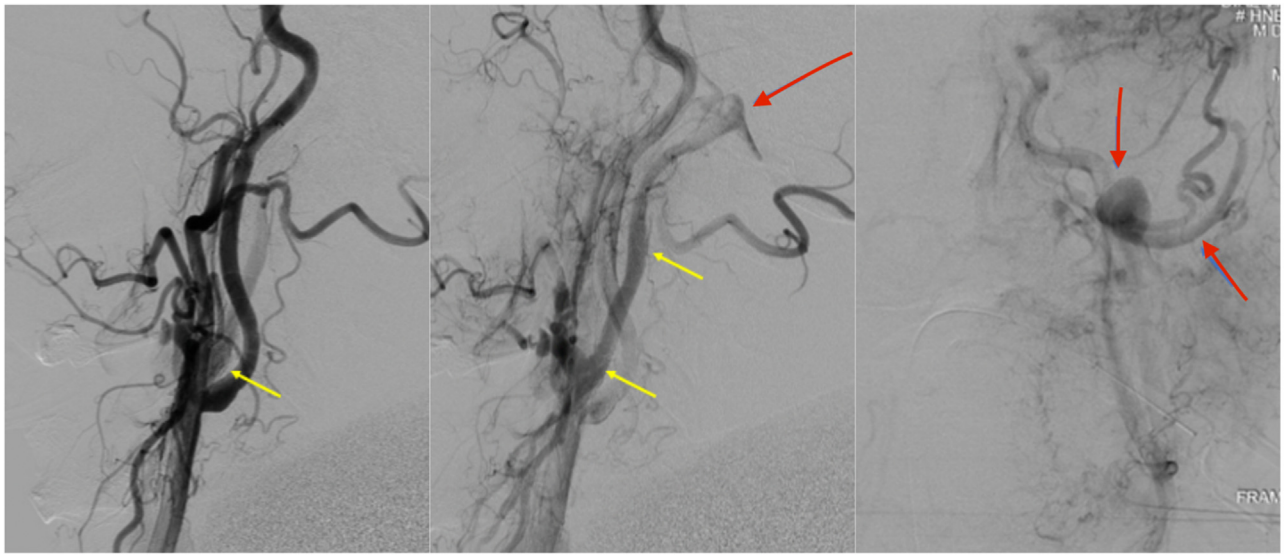

Fig. 1 – (A) Left CCA injection, lateral view. While in the early arterial phase of the injection, there is early venous opacification of the left IJV near the carotid bifurcation (yellow arrow). Numerous small arterial branches supplied the fistula (red arrow) (B) Late arterial phase of the same injection demonstrates opacification of the left IJV (yellow arrow) extending up to the jugular bulb (red arrow) and (C) Frontal view reveals further retrograde drainage with jugular bulb and sigmoid sinus opacification (red arrow).

similar to that of a dural arteriovenous fistula (DAVF) which we discuss below.

Case presentation

A 68-year-old male with a known anterior communicating artery aneurysm followed by surveillance imaging with CTA since 2014 presented for diagnostic cerebral angiography. His past medical history is significant for nonischemic dilated cardiomyopathy status post AICD implantation in 2019. At the time of diagnostic cerebral angiography unexpected arteriovenous shunting from the left external carotid artery into the left internal jugular vein (IJV) was encountered (Fig. 1). Arterial inflow was via numerous branches of the left external carotid anastomosing with left IJV. Venous flow was retrograde draining intracranially via the jugular bulb into the sigmoid and transverse sinuses as well as into the ipsilateral inferior petrosal sinus. No anterograde drainage was observed in the left internal jugular vein beyond the level of C5 suggesting possible obstruction.

Additional noninvasive imaging was obtained to further characterize this incidental lesion. CT venography confirmed suspected occlusion of the left brachiocephalic and distal subclavian veins with the presence of AICD wires while the left IJV was occluded below C5 as well (Fig. 2). Venous collaterals in the neck and thoracic inlet were noted draining into the superior vena cava. Magnetic resonance angiography (MRA) revealed arterialization of the left IJV with retrograde flow extending into ipsilateral cavernous sinus and flow related enhancement of the left superior and inferior ophthalmic veins (Fig. 3). Additionally, there was a cephalad direction of flow in the infe-

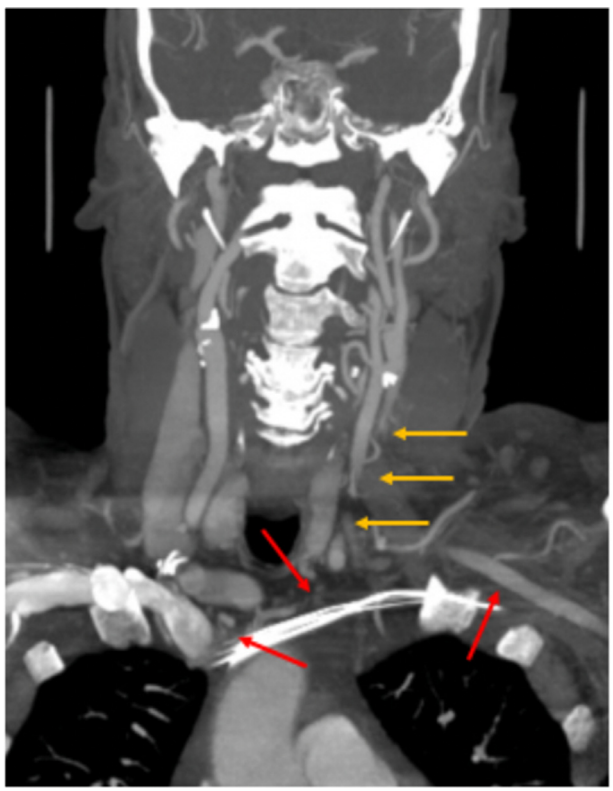

Fig. 2 – CT venography, coronal MIP view, demonstrates left sided AICD leads but absent opacification of the left brachiocephalic and subclavian veins (red arrows). Absence of the left IJV is noted as well (yellow arrows).

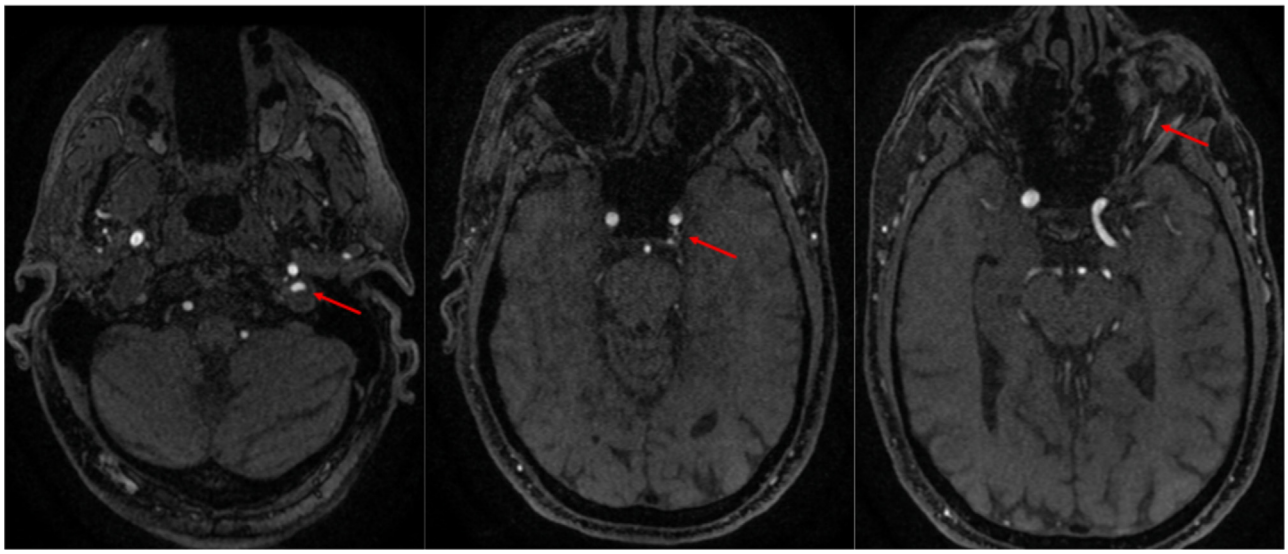

Fig. 3 – (A) 3D TOF MRA demonstrates flow-related enhancement within the left jugular bulb indicative of arterial flow (red arrow). (B and C) Arterialization noted within the left cavernous sinus and ophthalmic vein (red arrow).

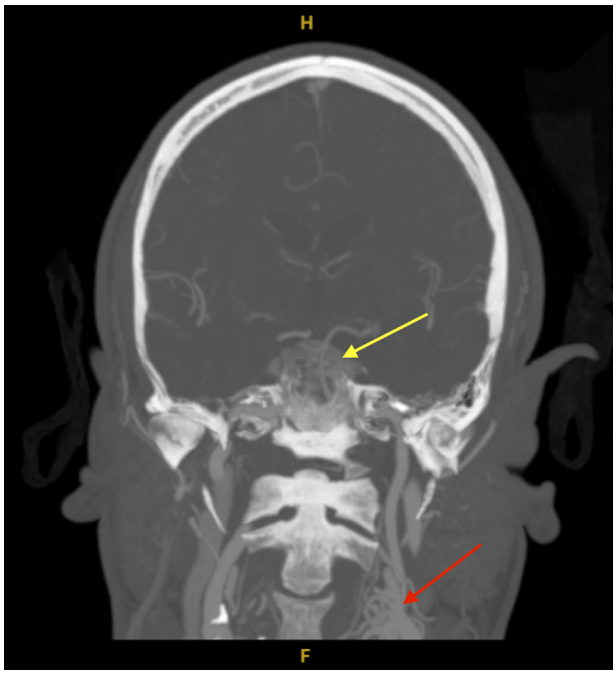

Fig. 4 – CTA, coronal MIP view, performed for surveillance of anterior communicating artery aneurysm (not here visualized), with partially visualized arterial feeders (red arrow), and arterialization of the cavernous sinus (yellow arrow).

rior petrosal sinus into the cavernous sinus, nonetheless the diameter of the left superior opthalmic vein was normal. In retrospect, CTA head demonstrated presence of the fistula in 2021 and its absence on the preceding CTA in 2017 (Fig. 4). It was concluded that the EC-IJ fistula developed secondary to an occlusion of the left IJV sometime after placement of the patient's central venous pacemaker leads in 2019.

Clinically he was neurologically intact with no complaints of headache, pulsatile tinnitus, visual disturbance, or retro-orbital pain. Although the malformation was asymptomatic in nature, the lesion was behaving similar to a Cognard IIa + b DAVF therefore treatment was recommended. However, his aneurysm grew to 6 mm from 2 mm over the course of 4 years, therefore the aneurysm was treated first. He underwent successful intrasaccular flow disruption with Woven Endovascular Bridge embolization via a transradial approach. Following a 6-week period to recover from his aneurysm embolization and observe for no delayed complications, he then underwent endovascular treatment of the fistula. Due to the numerous arterial feeders, it was not possible to occlude the fistula via primary arterial embolization and this required a venous approach. Therefore, the procedure was performed utilizing the right basilic vein and right radial artery for access. With a sheath positioned in the right IJV, a catheter construct was advanced in a retrograde fashion through the right sigmoid and transverse sinuses, across the torcula, then across the left transverse sinus and positioned within the left jugular bulb (Fig. 5). Next, the left IJV was embolized using Onyx liquid embolic and platinum microcoils via a dual microcatheter approach. Ultimately the left IJV was occluded in a retrograde fashion from the stump at the C4-5 level until the C2 level. At the completion of the case, arterial angiography confirmed successful occlusion of the fistula and restoration of antegrade flow in the left transverse and sigmoid sinuses (Fig. 6). The left IJV remained patent at the skull base with venous drainage through left paravertebral venous collaterals. In order to prevent propagation of thrombus intracranially from the newly occluded left IJV, he was started on rivaroxaban in addition to his preoperative aspirin.

No clinical complications were encountered, however, he did complain of mild postoperative facial pain and throat swelling that had no correlate on clinical evaluation and no radiographic evidence of swelling or airway compromise. The symptoms were self-limited and resolved by the second week.

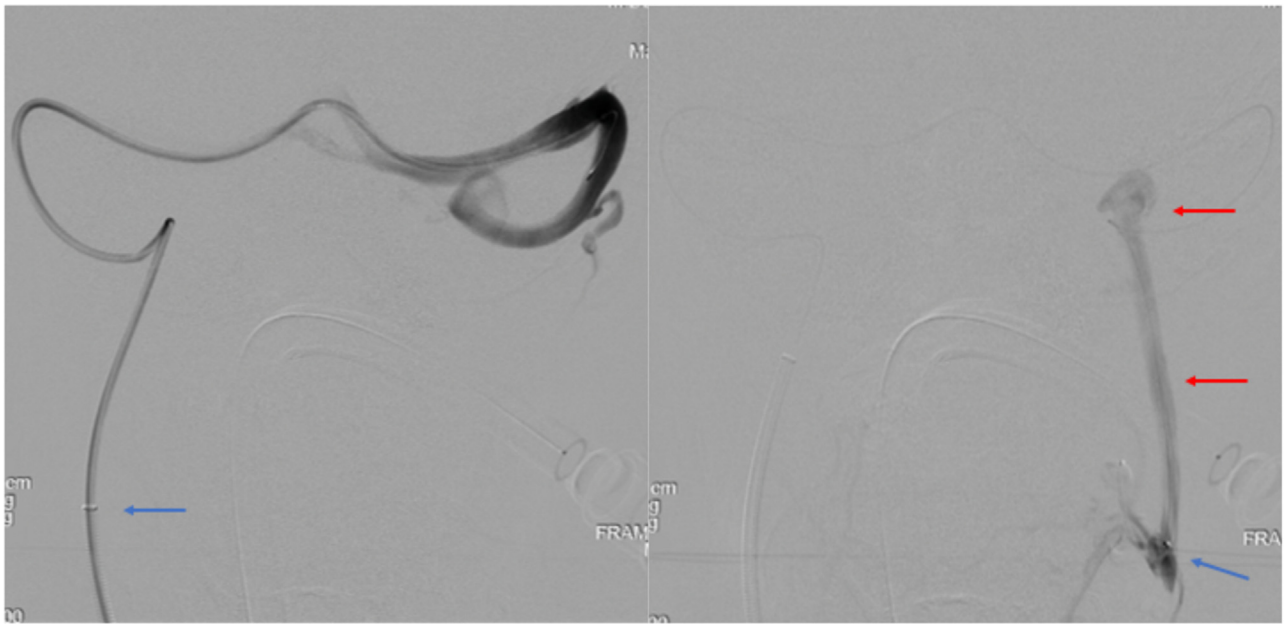

Fig. 5 – (A) 6F long sheath tip positioned in the right IJV (blue arrow). Injection via 6F intermediate catheter with tip in the proximal left sigmoid sinus demonstrates opacification up to the level of the jugular bulb and significant opacification of the left transverse sinus and (B) Injection via 0.017″ microcatheter with tip positioned at the stump of the left IJV (blue arrow) demonstrates retrograde drainage of contrast into the jugular bulb (red arrow).

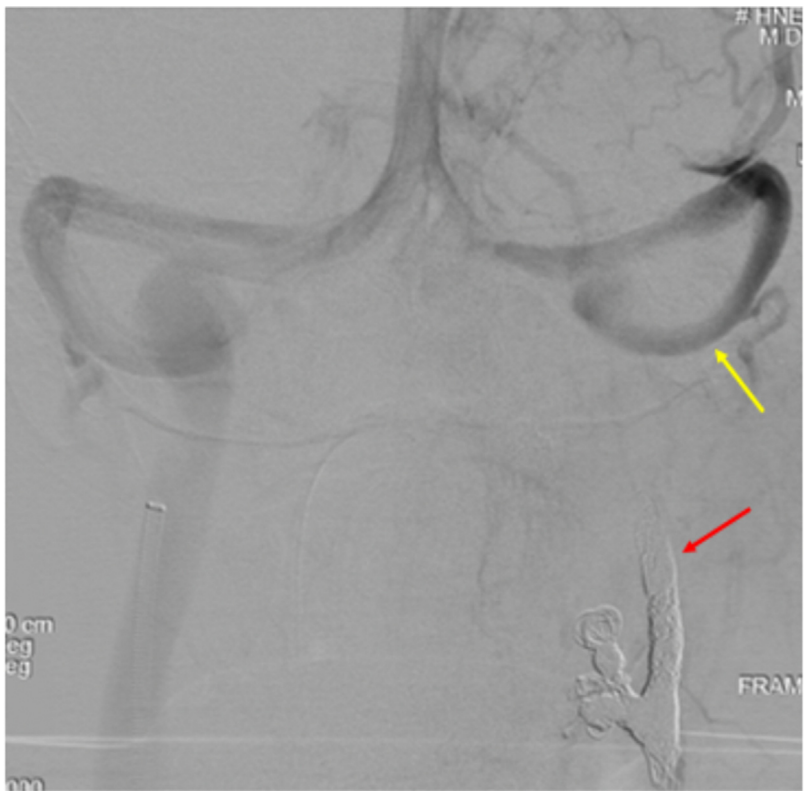

Fig. 6 – Left CCA injection frontal view. Post embolization angiogram while in the late venous phase demonstrates resolution of shunting and new antegrade drainage of the left sigmoid sinus (yellow arrow). The left IJV is occluded beyond the jugular bulb with embolic material noted distally (red arrow).

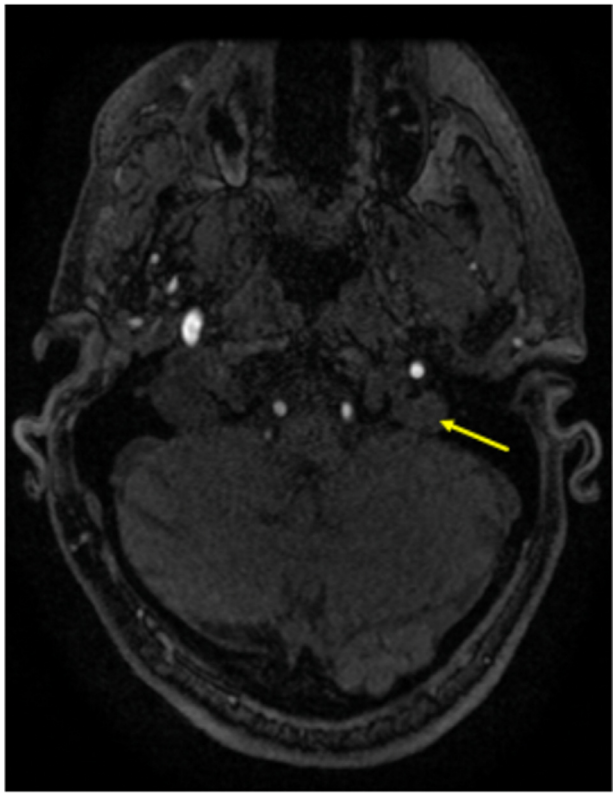

Fig. 7 – 3D TOF MRA after embolization reveals resolution of flow related enhancement in the left IJ bulb (yellow arrow) when compared to Fig. 3A.

At the 6 month postoperative visit, he was neurologically intact and a MRA of the head and neck demonstrated stable occlusion of the fistula (Fig. 7).

Discussion

A few cases of arteriovenous fistulae have been reported after AICD placement, and arteriovenous fistulae arising from central line placement, procedures requiring cannulation of the internal jugular vein, and trauma are also documented [4–7]. Some authors have posited that this may arise in iatrogenic settings as a consequence of inadvertent "double puncture" involving both the internal jugular vein and carotid artery [2]. These patients can present in a variety of ways, including high cardiac output states, severe epistaxis, peripheral edema, focal and nonfocal neurological deficits, elevated intracranial pressure, and jugular venous distention. These fistulae can often be visualized through ultrasonography or CT angiography, and treatment is generally indicated, as when left untreated, they may lead to intractable heart failure and embolus formation. Treatment involves ligation of the fistula, and can be performed with either endovascular approaches, including liquid embolization and stent grafting, as well as open approaches to ligate the fistula [1–7].

Importantly, these were direct fistulae and not associated with thrombus formation or reflux into the intracranial sinuses, as was the case in our patient. Given the occlusion of the IJV in conjunction with numerous arterial feeders into the wall of the occluded vessel and subsequent arterialization with intracranial retrograde sinus and cortical vein flow, this fistula was radiographically similar in appearance to a Cognard IIa + b DAVF [8]. To our knowledge, this is the first time that a lesion of this type has been described. Ostensibly, the occlusion of the IJV produced local conditions of angiogenesis, ultimately leading to the formation of multiple feeder vessels connecting the 2 vessels. Thus, it resembles the pathophysiology of DAVF.

DAVF are classified by the Borden and Cognard systems, which both broadly categorize DAVF into direct, and indirect forms [8,9]. Direct DAVF drains directly into the cortical veins, whereas indirect DAVF drains into dural venous sinuses. Direct DAVF are associated with greater morbidity and mortality. The classification systems utilize the direction of flow to further characterize DAVF to guide management and prognostication. The etiology and pathogenesis of DAVF remains poorly understood, however, the greatest association is with venous sinus thrombosis which leads to subsequent fistula formation through local angiogenesis mechanisms. A three stage model has been proposed by Kultulk et al., with an initial thrombus stage, followed by microscopic angiogenesis, and finally recanalization of the thrombosed sinus by arterial feeders. This process is associated with the angiogenic hypoxia factor 1, vascular endothelial growth factor, stroma derived factor alpha, and matrix metalloprotease 9. Other theories involve the awakening of dormant embryonic arteriovenous connections from venous sinus occlusion, and *de novo* formation of the DAVF with secondary thrombus formation [10,11].

Presenting symptoms of dizziness, and headache, along with signs of increased intracranial pressure are common features of both CJ-AVF and DAVF [1–3,7–9,12]. Although this patient was asymptomatic, the retrograde sinus flow with arterialization of the inferior petrosal sinus and ophthalmic veins raised concern for future hemorrhage and vision loss, and thus the patient was treated [12].Treatment approach can vary, like symptomatology, by location and size of the fistula Endovascular treatment is the most common modality of treatment. Open surgical ligation and stereotactic radiosurgery have also been explored as treatment options. Endovascular approaches are minimally invasive, require a shorter hospital stay, and are better suited for patients with underlying comorbidities or increased surgical risk. An open surgical approach may be selected when the angioarchitecture is less amenable to endovascular approaches. Surgical options include ligation of the fistula and arterial repair utilizing grafts, suture, or end-to-end anastomosis. Open surgical approach results in a greater operating time and carries higher mortality risk as well as a chance of postoperative neurological deficits [13–15]. However, due to the rarity of CJ-AVFs, most reports are case studies and there is a lack of long term follow up studies. Thus, a definitive superiority of an endovascular approach compared to an open surgical approach cannot be made. In our patient, the location of the fistula, intracranial involvement and its numerous arterial feeders, informed our decision to treat endovascularly.

The insidious onset and asymptomatic presentation of CJ-AVFs increase the risk of potentially life-threatening complications, especially if these malformations are not discovered and treated appropriately. CJ-AVFs can arise secondary to a variety of different causes, but there are trace reports of their occurrence as a side effect of pacemaker lead occlusion. This report presents a unique case of an incidentally found ECA-IJV AVF, secondary to an exceedingly rare etiology, and demonstrates endovascular embolization as an efficacious treatment option.

Patient consent

Written informed consent was obtained from the patient.

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