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Case Report

Pediatric jugular vein aneurysm (phlebectasia): report of two cases and review of the literature

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ABSTRACT

Jugular vein aneurysms are rare vascular abnormalities that are most commonly encountered in the pediatric population. We report two separate cases in infants, both of whom presented with enlarging neck masses and were found to have jugular vein aneurysms. Diagnosis was established with duplex ultrasonography, computed tomography angiography, digitally subtracted catheter venography, and magnetic resonance imaging in one case and magnetic resonance imaging with magnetic resonance angiography/magnetic resonance venography, gray scale ultrasonography, and digital subtraction catheter venography in the other case. Both aneurysms were treated by surgical resection.

Introduction

Arterial aneurysms are commonly encountered in adult clinical practice, arising from multifactorial vascular degeneration and typically presenting in sixth to eighth decades of life. Comparatively rare entities, venous aneurysms frequently result from congenital deficiencies of the vessel wall rather than degenerative processes, and more often come to attention in the pediatric population [1–3]. Anatomically, venous aneurysms most frequently occur in the upper extremity but are seldom reported since they are typically asymptomatic [4]. Furthermore, upper extremity and cervical venous aneurysms typically pose no significant risk to a patient's health with no serious sequelae reported. Incidental lower extremity venous aneurysms have been described in adults found during workup for pulmonary embolism (PE) [4], with the aneurysm most commonly involving the popliteal vein [4,5]. Venous aneurysms of the deep system tend to have higher morbidity than those involving the superficial system related to thromboembolic events and recurrent pulmonary emboli [4].

When encountered in children, neck masses can pose a diagnostic dilemma due to the relatively broad differential diagnosis in this age group. However, the differential diagnosis is limited when a neck mass enlarges when performing

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a Valsalva maneuver. Diagnostic considerations for a neck mass that enlarges with Valsalva maneuver include superior mediastinal tumor or cyst, laryngocele, or jugular phlebectasia [6].

Duplex ultrasonography scanning is usually sufficient for the diagnosis of a venous aneurysm, but three-dimensional ultrasonography, computed tomography, magnetic resonance imaging (MRI) with magnetic resonance angiography/magnetic resonance venography, and catheter-directed venography can also be performed to improve morphologic evaluation [7,8].

Case 1

A 3-month-old term male infant with no significant prenatal or perinatal complications presented with a left-sided neck mass. The mass was first noted at birth and continued to increase in size during the first 3 months of life. Physical examination revealed a firm, mobile mass deep to the left sternocleidomastoid muscle without tenderness or overlying skin changes. There was no history of trauma.

Ultrasound revealed a complex left cervical mass with a lamellated appearance centrally and no internal vascularity (Fig. 1). Contrast-enhanced MRI (Fig. 2) and computed tomography imaging (Figs. 3A and 3B) confirmed an ovoid lesion with peripheral enhancement and intralesional pooling of contrast. The differential diagnosis based on cross-sectional imaging favored low-flow vascular malformation, although the imaging characteristics of the central portion of the lesion made it difficult to exclude the possibility of a solid tumor such as teratoma or complex branchial cleft cyst. Catheter venography using a direct puncture technique helped clarify the diagnosis, revealing a large, irregular left internal jugular vein aneurysm with slow flow into collateral vessels, ultimately draining into the left brachiocephalic vein (Fig. 4). Outflow compression maneuvers failed to elicit reflux of contrast into the intracranial dural venous sinuses.

Due to concerns regarding growth of the aneurysm and intraluminal thrombus, the patient underwent surgical resection. A partially thrombosed venous aneurysm within the carotid sheath was confirmed intraoperatively. The abnormal vein and a small amount of normal adjacent internal jugular vein were excised from skull base superiorly to the facial vein confluence inferiorly, preserving the facial vein and normal caliber inferior aspects of the internal jugular vein. Intraoperative cranial nerve monitoring was performed due to the close proximity of the aneurysm to cranial nerves VII, XI, and XII.

The postoperative recovery period was uneventful, and the patient was discharged on the second postoperative day. Contrast-enhanced MRI performed 8 months after resection revealed patency of the inferior internal jugular vein below the facial vein confluence and no recurrence of the venous aneurysm. Ultrasound performed 18 months postresection revealed no recurrent venous aneurysm. At the 18-month follow-up clinic visit, the patient was developing normally with no symptoms or physical examination evidence of lesion recurrence.

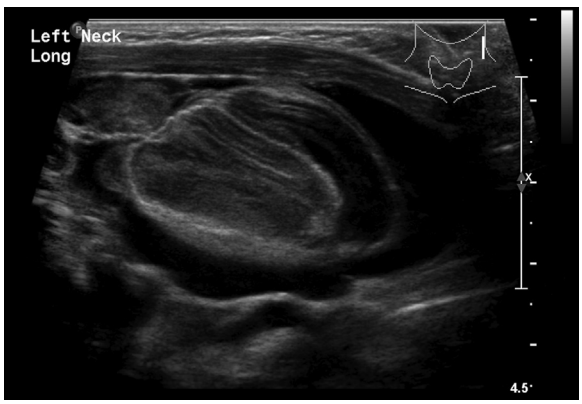

Fig. 1 – Longitudinal grayscale ultrasound image reveals a complex lamellated mass in the left neck with some peripheral flow but no internal vascularity on color Doppler (not shown).

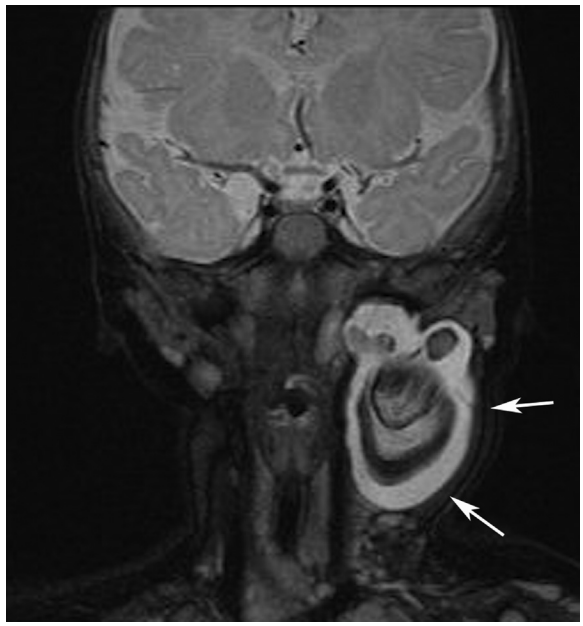

Fig. 2 – Coronal T2 MRI reveals an ovoid, lamellated mass with surrounding high T2 signal in the region of the left internal jugular vein (arrows). MRI, magnetic resonance imaging.

Case 2

A 6-month-old term female infant product of a twin gestation with no significant prenatal or perinatal complications presented with swelling in her left neck that increased with Valsalva. The mass was otherwise asymptomatic but continued to grow with the child. Initial physical examination revealed a compressible mass in the left supraclavicular fossa only appearing when the patient cried. The mass was nontender, and there was mild discoloration of the overlying skin. There was no history of trauma.

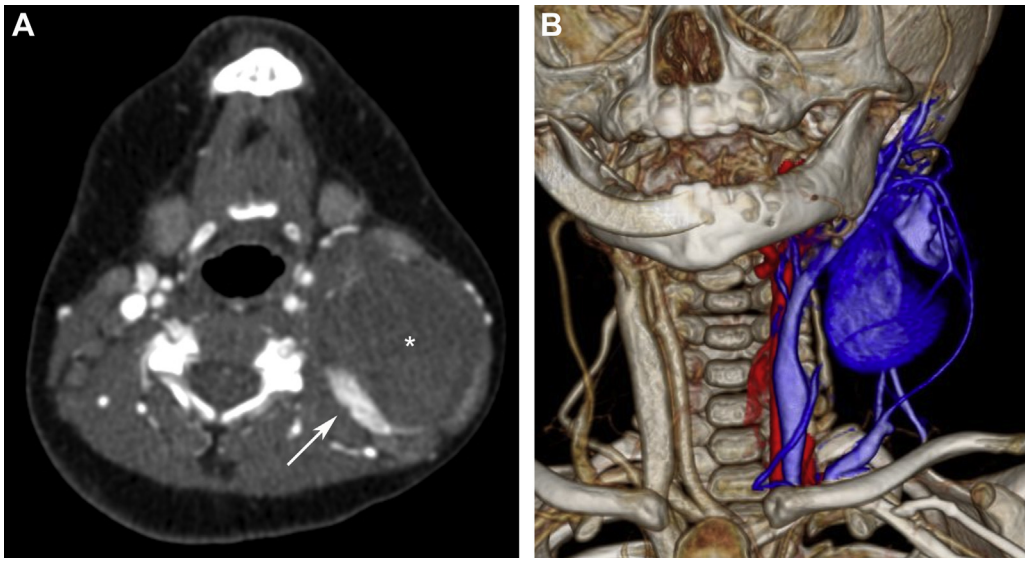

Fig. 3 – (A) Axial contrast-enhanced CT reveals a solid, nonenhancing mass (*) with surrounding enhancement as well as some contrast pooling along the posteroinferior periphery of the lesion (arrow). (B) Three-dimensional color multiplanar CT reformation shows the relationship of the lesion to the adjacent arterial and venous structures with suggestion of involvement of the left internal jugular vein. CT, computed tomography.

Contrast-enhanced MRI with magnetic resonance angiography/magnetic resonance venography (Fig. 5) revealed a large tubular structure in the left supraclavicular fossa that enhanced during the venous phase with drainage into the left subclavian vein. The differential diagnosis based on cross-sectional imaging favored low-flow vascular malformation. At this point, conservative management was recommended with a plan to readdress the lesion when the patient was 2 years of age. However, the patient returned to clinic approximately 6 months later at 12 months of age with parental concerns about lesion enlargement. Due to cosmetic issues as well as concerns about the risk of life-threatening hemorrhage, definitive treatment was requested by the patient's family. Prior to surgical intervention, catheter venography was performed to elucidate the nature of the lesion and map venous communications. Venography using a direct puncture technique revealed a large saccular aneurysm of the left external jugular vein with sluggish antegrade drainage into

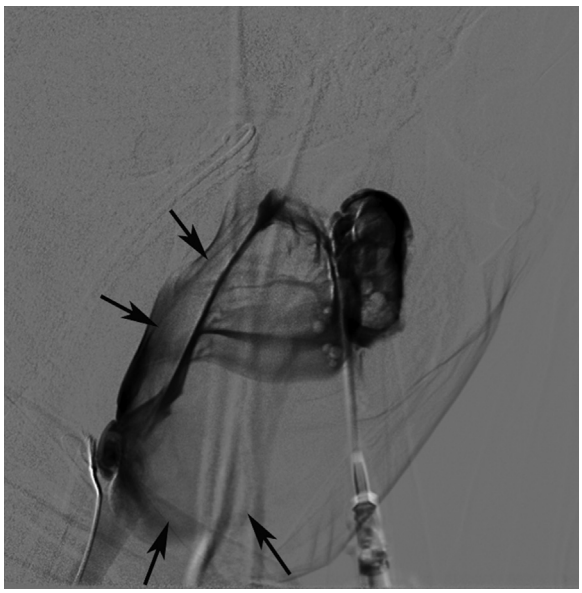

Fig. 4 – Representative image from direct puncture digital subtraction venography shows a large filling defect (arrows) within the left internal jugular vein aneurysm.

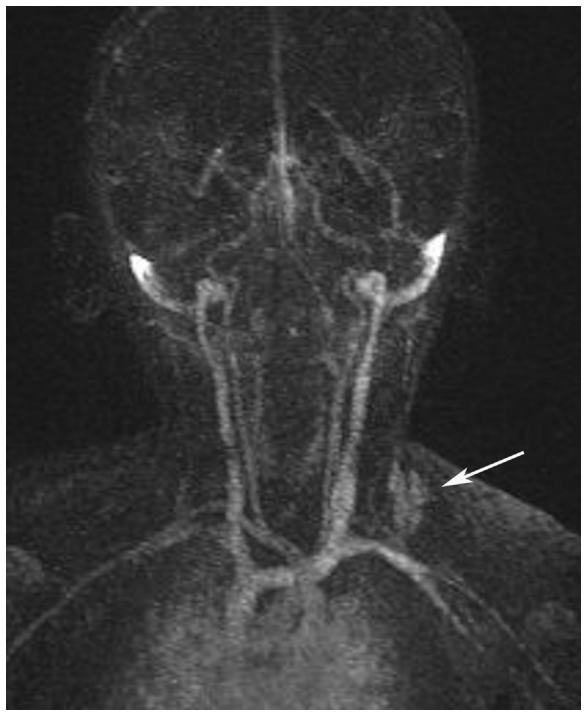

Fig. 5 – Dynamic contrast-enhanced MR angiogram reveals venous phase enhancement of a tubular structure in the left supraclavicular fossa (arrow). MR, magnetic resonance.

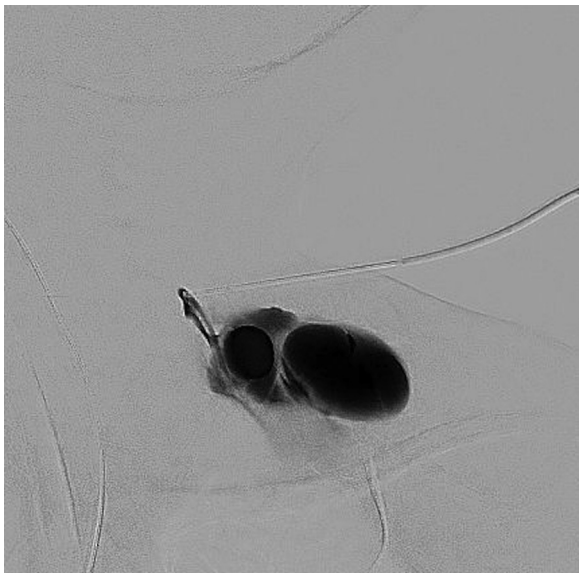

Fig. 6 – Representative image from direct-puncture digital subtraction venography shows a large saccular aneurysm of the left external jugular vein with drainage into the left subclavian vein.

the left subclavian vein (Fig. 6). Preoperative embolization was not performed due to concerns about central embolization.

The patient proceeded to surgical resection immediately following venography. A large vascular mass was encountered just deep to the platysma muscle. Dissection was carried out proximally to a normal-appearing external jugular vein feeding vessel, which was ligated. Careful dissection was performed circumferentially with ligation of several small feeding veins. Dissection was then carried inferiorly with identification and ligation of the tributary draining into the left subclavian vein. Due to the close proximity to cranial nerve XI, continuous trapezius muscle monitoring was performed during the case and the nerve was preserved. The postoperative recovery period was uneventful, and the patient was discharged on the first postoperative day.

Discussion

Primary venous aneurysm is a rare entity. First reported in 1928 [9], only isolated case reports and small case series describing venous aneurysms have been reported in the literature. There are conflicting data regarding the most common location of venous aneurysms. The most common site reported in older literature is the superior vena cava and its tributaries [10], although venous aneurysms were found most frequently in the lower extremity in the largest case series to date [4]. Terminology used to describe abnormal dilation of veins is similarly ambiguous. Some describe true venous aneurysm as a saccular outpouching, while phlebectasia is considered a fusiform dilatation. The terms are occasionally used interchangeably. Histopathologically, phlebectasia typically shows thinning of the muscular layer of the vein wall, while aneurysms show degenerative changes [11]. Regardless of the terminology, the natural history, prognosis, and management strategy depend more on the location of the venous abnormality than the morphology.

Thromboembolic complications including spontaneous thrombosis in cases of jugular phlebectasia and jugular venous aneurysm are reportedly rare [12]. Thromboembolic complications in lower extremity venous aneurysms are far more common, occurring in up to 71% of cases [1]. Venous aneurysm-associated deep venous thrombosis with subsequent PE has been almost exclusively reported in cases of lower extremity venous aneurysm [4]. To the best of our knowledge, there are no cases of deep venous thrombosis/PE related to jugular venous aneurysm thrombosis in the English literature.

There is no clear consensus on the appropriate treatment of venous aneurysms, and recommendations remain equivocal. Some surgeons advocate conservative management, when possible, since venous aneurysms are rarely symptomatic. Others advocate resection due to the potential risk of venous thromboembolism as well as for cosmetic reasons. Authors reporting the largest case series to date (39 venous aneurysms in 30 patients) advocate surgical management of the majority of venous aneurysms with the exception of jugular venous aneurysms, which they report can be treated conservatively [4]. Calligaro et al [1] report that prophylactic surgery is cautiously recommended for low-risk patients with venous aneurysms of the abdomen and strongly recommended for most patients with lower extremity deep venous aneurysms, but that other venous aneurysms should be excised only if they are symptomatic, enlarging, or disfiguring. More recently, endovascular treatment with stent-assisted coiling has been described as a treatment for subclavian venous aneurysm complicated by PE [13].

The first case is unique from a clinical standpoint because the aneurysm was encountered very early in life, had been steadily growing, and was found to have a large intraluminal thrombus on imaging evaluation. Given the lesion characteristics and small degree of diagnostic uncertainty on cross-sectional imaging, the decision was made to resect the aneurysm for tissue diagnosis rather than pursue a conservative management strategy. It is important to note that immediate preoperative catheter venography helped to elucidate the final diagnosis, but the patient proceeded to surgery due to the size of the lesion and the concern for thromboembolic complications given the presence of intraluminal thrombus.

The second case is also unique because the lesion was encountered early in life and grew steadily with the child. The decision to pursue surgical resection was made based on parental concerns about life-threatening hemorrhage and cosmetic issues later in life. As with the first case, preoperative catheter venography helped elucidate the final diagnosis and aided in operative decision-making.

In both of the presented cases, the interventional radiology service was consulted for preoperative catheter-directed venography to assist in operative planning and for consideration of preoperative embolization to minimize intraoperative bleeding. Neither of the lesions was amenable to embolization

due to direct communication with the central veins and the risk of central embolization.

Conclusions

Pediatric jugular venous aneurysm/phlebectasia is a rare entity that can be reliably diagnosed on the basis of clinical history and noninvasive imaging. Early misdiagnosis is often related to lack of familiarity with the pathologic entity. Treatment recommendations vary, with some groups advocating surgical intervention in nearly all cases and others recommending a conservative approach, when possible. There is no clear consensus in the literature regarding the differentiation between venous aneurysm and phlebectasia. However, current treatment strategies rely more on lesion location and symptomatology rather than on histopathologic characteristics.

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