Non-atherosclerotic supra-aortic trunks pathology: imaging findings and correlation between US, CT and MRI.

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Learning objectives

To review the non atherosclerotic supra-aortic trunks entities, its pathophysiology and differential diagnosis.

"Not to be missed" imaging findings, review and correlation between different techniques.

Background

Evaluation of supra-aortic trunks (SAT) is challenging. On the one hand, the SAT are not directly accessible for clinical evaluation and on the other hand, radiological studies are excellent in demonstrating a SAT lesion and its extent.

1. VARIANT ANATOMY OF THE AORTIC BRANCHES VESSELS:

1.1 Bovine arch
1.2 Aberrant right and left subclavian artery
1.3 Variant origin of vertebral arteries

2. DEVELOPMENT ANOMALIES

2.1 Congenital absence or hypoplasia of the ICA
2.2 Cervical ICA duplication
2.3 Fenestration of the ICA
2.4 Marfan syndrome

3. THORACIC OUTLET SYNDROME

4. CRANIOCERVICAL ARTERY DISSECTION

4.1 ICA dissection
4.1.1 ICA dissection complications: Pseudoaneurysm and pseudofenestration
4.2 Vertebral artery dissection

5. DURAL ARTERIOVENOUS FISTULA (Supraaortic trunks haemodynamic repercussion)

6. RADIATION TREATMENT

7. CATHETER-INDUCED VASOSPASM

7. FIBROMUSCULAR DYSPLASIA (FMD)

8. ARTERITIS

8.1 Takayasu arteritis

8.2 Behçet disease

8.3 Temporal arteritis

**Findings and procedure details**

1. VARIANT ANATOMY OF THE AORTIC BRANCHES VESSELS (Fig. 1 on page 9):

Abnormal formation of the 1\textsuperscript{st}, 2\textsuperscript{nd} and 3\textsuperscript{rd} arch vessels results in abnormal branch vessels:

1.1 **Bovine arch** : The most common variant of the aortic arch. Occurs when the brachiocephalic artery shares a common origin with the left common carotid artery.

1.2 **Aberrant right subclavian artery** (arrises from de 4\textsuperscript{rd} arch)

**Aberrant left subclavian artery** (right sided arch).

1.3 **Variant origin of vertebral arteries**.

2 VARIANT ANATOMY OF THE INTERNAL CAROTID ARTERY:
2.1 **Cervical ICA duplication:** Involves the entire cervical segment, from the carotid bifurcation up to the skull base.

2.2 **Fenestration of the ICA:** Extremely rare anatomic variant. Short vascular segments with two parallel arterial lumens.

Differential diagnosis: Pseudofenestration (see 5.1.1 Complications at ICA dissection)

3. **DEVELOPMENT ANOMALIES**

3.1 **CONGENITAL ABSENCE** *(Fig. 2 on page 9) OR HYPOPLASIA OF THE ICA* *(Fig. 3 on page 10):*

- Rare anomaly.
- Asymptomatic until the carotid stenosis is not well compensated by the contralateral carotid artery (normally with atherosclerotic changes) or its collaterals to the circle of Willis.
- Should only be diagnosed if the petrous segment of the ICA is absent or hypoplastic.
- Compensatory hypertrophy of CoAA and PCA.
- Can simulate occlusion or long ICA stenosis.
- Associated with cerebral aneurysms.

Absense or hypoplasia of the vertebral artery are normal variants.

3.2 **MARFAN SYNDROME** *(Fig. 4 on page 11):*

- Multisystemic hereditary connective tissue disease: defect in FBN1 gene.
- Cardiovascular complications tend to be most frequent cause of death: Aortic dissection is the most frequent.
- Dissection at cranioencephalic vessels is less frequent. Also may be seen tortuosity and ectasia of the extracranial carotid arteries and vertebral arteries.

3.3 **NEUROFIBROMATOSIS TYPE 1** *(Fig. 5 on page 12):*

- The most common phakomatosis.
- Common intracranial vasculopathy. Extracranial vasculopathy is rare.
4. **THORACIC OUTLET SYNDROME** *(Fig. 6 on page 13):*

- Congenital or acquired compression of brachial plexus, subclavian artery or subclavian vein as they pass through the thoracic inlet.
- Clinical diagnosis.

Plain radiography: exclude an underlying bone abnormality.

Ultrasound, contrast-enhanced CT, MRI or conventional angiography: detect vascular compression and its cause.

Imaging is performed with the patient's arms both in the raised and neutral position (for comparison): shows vascular stenosis and complications (such as false aneurysm, thrombosis and distal emboli).

5. **CRANIOCERVICAL ARTERY DISSECTION**

5.1 **INTERNAL CAROTID ARTERY (ICA) DISSECTION:**

20-25% of strokes in patients less than 45 years-old.

Causes of dissection:

- Most common cause: traumatic dissection (severe blunt head and neck trauma).
- Spontaneous: secondary to a trivial trauma (as a trigger to dissection), with underlying arteriopathy (fibromuscular dysplasia) or connective tissue disorders (Marfan syndrome, Ehlers-Danlos disease).
- Iatrogenic: antecedent neck manipulation (catheter-induced vasospasm).

Only 1/3 of patients have symptoms. Due to a lack of specific signs, the diagnosis often goes unrecognised for some time:

- Low grade stenosis: neck pain, headache, ipsilateral Horner syndrome, pulsatile tinnitus and cranial nerve palsy.
- High grade stenosis: ischaemic stroke.

SAT ultrasound *(Fig. 7 on page 14):* Most dissecting aneurysms are missed with this technique.

- Thickened hypoechoic vessel wall: mural hematoma and thrombus cannot be differentiated with B-mode imaging.
• Distal parts of the ICA mainly demonstrate the hemodynamic abnormalities (stenosis or occlusion): Increased flow resistance at left ICA and reduced flow velocity. Wall abnormalities are hardly depicted at distal parts of the ICA.

CT without contrast: insensitive for a dissection. May demonstrate ischaemic changes within the brain.

CT-Angiography (Fig. 8 on page 15):
• Irregularity of the vessel wall.
• Narrow lumen distal to a stenotic focus in the ICA.
• Enlargement of the dissected artery: in an occlusive dissection.
• Intramural hematoma appears isoattenuating to the surrounding muscles and cannot be differentiated from atherosclerotic thickening or thrombus.

MR-Angiography:
• Crescent-shaped mural thickening hyperintensity = wall hematoma on subacute dissection (between 7 days and 2 months). Bright on T1-weighted with fat saturation (Fig. 9 on page 16)
• Early and chronic stage: hematoma is usually isointense to surrounding structures.

5.1.1 ICA DISSECTION COMPLICATIONS:
• Pseudoaneurysm formation (Fig. 10 on page 17)
• Pseudofenestration: double barrel lumen. Contour irregularities and size asymmetry of the limbs of the fenestration (Fig. 11 on page 18).

5.2 VERTEBRAL ARTERY DISSECTION (Fig. 12 on page 19):

Is potentially lethal and can be difficult to diagnose clinically and radiologically.

Clinical presentation: Neck pain and headache, posterior fossa ischaemic events (TIA or stroke), spinal cord infarction, subarachnoid haemorrhage (SAH), cervical nerve root impairment.

5.2.1 Extracranial vertebral artery dissection:
• Typically occur at the C1 or C2 level (V3)
• With or without intracranial extension
CT/CT-Angiography

- Demonstrate posterior fossa ischaemia or subarachnoid haemorrhage.
- Identify an occluded vertebral artery or mural thrombus (thickened wall, often with some surrounding stranding).
- CTA (especially with coronal and sagittal reformats): demonstrate irregularity of the lumen. Arterial wall narrowing more easily appreciable.

MRI/MR-Angiography

- Far greater sensitivity to small foci of ischaemia (using DWI).
- Ability to image the vessel lumen (MRA)
- More sensitive at imaging intramural haemorrhage.
- Crescent sign at Fat saturated T1 axial images: sickle shaped hyperintensity in the wall of the affected vessel (Fig. 13 on page 20).

6. DURAL ARTERIOVENOUS FISTULA (Fig. 14 on page 21)

- 10-15% of all AV shunting cerebral vascular malformations.
- Typically have multiple feeders and are usually acquired.
- Frequently as a result of neovascularisation induced by previously thrombosed dural venous sinus (typically transverse sinus).
- Pulsatile tinnitus, cranial nerve palsies, venous hypertension.

SAT ultrasound and hemodynamic changes (Fig. 15 on page 22):

- Annual follow-up to detect possible hemodynamic changes associated with dural AVF: Reduced flow resistance at ECA (ECA RI is correlated with the effectiveness of treatment and its sensitive to the clinical evolution of dural AVF). Increased systolic and diastolic flow velocity.
- Evaluate intracranial arteriovenous shunt surgery.

7. RADIATION TREATMENT:

- Neck irradiation significantly increases the thickness of the carotid artery wall during the first year after radiation therapy.
- Fibrosis and long stenosis at CCA and ICA.
- Also there are some neck surgeries that produce focal fibrosis in adjacent vessels.
8. FIBROMUSCULAR DYSPLASIA (FMD) (Fig. 16 on page 23):

- Alfa1-antitripsina defect.
- 30 to 50 year-old women (3:1).
- Fibromuscular thickening of the arterial wall = stenosis.
- 30% at craniocervical arteries: ICA is affected en ¾ cases.
- Location: at the C2-C3 space, sparing the major extracranial vessels.
- 65% Bilateral involvement of the cervical ICA.
- Vertebral artery involvement is less common (10%) and frequently coexists with carotid disease.
- Symptoms: transitory cerebral stroke, subarachnoid hemorrhage associated to aneurysms or craniocervical artery dissection.
- Complications: Arterial dissection, sacular aneurysms, distal embolisation (of thrombus formed in aneurysm), AV fistulas.

CT-Angiography or MR-Angiography:

- Visualisation of small or peripheral lesions.
- String of beads sign: alternating stenoses and dilatations, causing a string of beads appearance.
- Less commonly the stenosis has a smooth tapered appearance, or dissection may be seen.

CT and MRI: ischaemic damage.

9. ARTERITIS

9.1 TAKAYASU ARTERITIS:

- Granulomatous large-vessel vasculitis.
- Primarily affects the aorta and its main branches.
- Most common location at neck: subclavian artery and CCA. Often affected over long segments on both sides.
- Segmental and patch inflammation = stenosis, thrombosis and aneurysm formation: Ischaemic symptoms.

9.2 BEHÇET DISEASE:

- Multisystemic and recurrent inflammatory vasculitis of unknown aetiology.
- 20-40% of patients develop vascular complications.
- Usually affects large vessels in the form of occlusion or thrombophlebitis of major veins.
- Arterial occlusion and pseudoaneurysms of the ICA may also be seen to a lesser extent.

9.3 TEMPORAL ARTERITIS (Fig. 17 on page 24):

- Older than 50 year-old patients.
- Headache and increased erythrocyte sedimentation rate.
- Any portion of the extradural arterial circulation within the head and neck may be involved, most commonly at the carotid siphon.
- The definitive diagnosis is generally made with temporal biopsy.

Images for this section:

Fig. 1

(A) BOVINE ARCH: The brachiocephalic artery (BCA) shares a common origin with the left common carotid artery (LCCA).  
(B) ABERRENT RIGHT SUBCLAVIAN ARTERY arising on its own from the 4th arch, after the left subclavian artery (instead of arising from the first branch with the right common carotid as the brachiocephalic artery).  
(C) AORTIC ARCH ORIGIN OF THE LEFT VERTEBRAL ARTERY: 5% of left vertebral arteries will have a variable origin with the most common being as a branch of the aortic arch (arrow).
(A) **CONGENITAL ABSENCE OF THE ICA**. Should only be diagnosed if the petrous segment of the ICA is absent (arrow).

(B) **HYPOPLASIC LEFT CCA** (arrowhead) and absense of the left ICA (*).
CONGENITAL HYPOPLASIC OF THE LEFT ICA: filiform flow at cervical (arrows in A) and petrous (arrows in B) segments of the left ICA. CT image shows a hypoplastic osseous carotid canal.
MARFAN SYNDROME:
Tortuosity and ectasia of both extracranial (SAT ultrasound on A) and intracranial carotid arteries (CTA on B).
Incidental finding in a orbit CT perfomed under the suspect of a orbital cellulitis on a 2 years-old patient.

Fig. 4
NEUROFIBROMATOSIS TYPE 1:
(A) Complete left ICA occlusion (*). Vascular calcification at the theoretical distal ICA territory, that suggest previous ICA pseudoaneurym.
(B) Hypoplastic petrous segment of the left ICA.

Fig. 5
(A) **C7 TRANSVERSE MEGA-APOPHYSIS**: anomaly regarding cervicothoracic transition (arrow)
(B) Circumferential thickening of the left subclavian artery (perivascular inflammatory changes) next to the occlusion (B). **Hypertrophy of the subclavian artery branches** (C).
C: **Progressive stenosis**: filiform flow till complete occlusion of the left subclavian artery (arrow).
D: Proximal occlusion of the left subclavian artery (yellow arrow). **C7 transverse mega-apophysis** (blue arrow).

**Fig. 6**
(A) Normal flow at right ICA.
(B) **LEFT ICA DISSECTION**: Increased flow resistance at left ICA. Reduced flow velocity.
(C) Thickened hypoechoic ICA wall (arrowheads): Mural hematoma and thrombus cannot be differentiated.

Fig. 7
ICA acute & chronic dissection

**ACUTE ICA DISSECTION:**
(A) Irregular left ICA contour (arrowheads). Vascular loop at distal cervical segment of the left ICA (*).  
(B) Intimal flap next to the petrous segment of the left ICA (arrow).

**CHRONIC ICA DISSECTION:** MRI performed 16 days later: Progression of the dissection compared to previous CTA.  
(A) Chronic dissection: hematoma is usually isointense to surrounding structures (square).  
(B) String sign, stenosis > 70% (arrow). 1 - pseudoaneurysm. 2 - pos t-stenotic dilatation.

**Fig. 8**
ICA subacute dissection

Axial T1 weighted FSE FatSat

WALL HEMATOMA ON SUBACUTE DISSECTION:
Crescent-shaped mural thickening hyperintensity at T₁-weighted with fat saturation (arrow).

Fig. 9
(A) **PSEUDOANEURYSM** arising from the medial part of the distal cervical segment of the left ICA (arrowhead)

(B) Narrowed lumen (arrow) distal to the pseudoaneurysm till the petrous segment of the left ICA (**DISSECTION**).

*Fig. 10*
Fig. 11

PSEUDOFENESTRATION SECONDARY TO LEFT ICA DISSECTION:
Distal to the cervical segment of the ICA, there is an asymmetric double barrel lumen (true and false ICA limbs of the fenestration).
Vertebral dissection from its origin at the subclavian artery (A), narrowing of its lumen at its middle portion (B y C) and complete occlusion at C2 (D).

Fig. 12
Subacute thrombosis/wall hematoma at right vertebral artery (arrows at A, B and C). No flow at cervical segment (c1-c2) of the right vertebral artery suggestive of dissection (arrowheads on C).

(C-D) Ischaemic event at posterolateral right bulb secondary to the dissection (arrows).

Fig. 13
Patient with pulsatile tinnitus.
(A and B) Chronic thrombosis of right transverse sinus with multiple “flow voids” within it.
(C-E) Enlarged and tortuous arterial feeding branches from right ECA. High flow through draining veins.
Characteristic findings of a long-standing adult-type dural A-V fistula.
Supraaortic trunks Doppler-ultrasound and haemodynamic features of the same patient:
Reduced flow resistance at right ECA (right IR ECA: 0,50. Compared to a left IR ECA: 0,83).
Increased systolic and diastolic flow velocity at right ECA.

Findings that suggest dural fistula.
(A) STRING OF BEADS SIGN: multifocal beaded and tortuous stenosis and dilatations along both ICAs (arrows). Preocclusive stenosis on the right ICA. 75% stenosis at cervical segment of the left ICA (NASCET-style derived percent stenosis).

(B) Subacute ischaemia secondary to FMD.

Fig. 16
TEMPORAL ARTERITIS:
(A) Prominent, tortuous and beaded right temporal artery. Hypoechoic mural thickening around the perfused lumen (arrows).
(B) The right temporal artery shows a hypoechoic wall thickening with a diameter of up to 1.4mm around the perfused lumen.
Conclusion

The detailed knowledge of SAT pathologies, as well as the proper interpretation of US, CT and MRI findings, allows a proper diagnose, contributing to a better therapeutic management.

Personal information

References