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RESEARCH ARTICLE

AORTIC COARCATATION IN ADULTS : STILL A LATE DIAGNOSIS

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Abstract

Aortic coarctation is the most common congenital cardiac malformation due to a narrowing of the aorta, most commonly occurring just beyond the left subclavian artery.The diagnosis is mostly neonatal by the systemic palpation of the femoral pulses but can be later discovered at the big child's even at the adult. This fact is illustrated through our observation of a 36-year-old man admitted after a head injury, during his hospitalization the diagnosis of aortic coarctation was made following hypertensive peaks resistant to antihypertensive drugs.

Introduction:-

Aortic coarctation is a common congenital heart disease. It accounts for 6–8% of congenital heart defect cases and ranks as the fifth most common abnormality in this patient group [1]. Although often discovered and corrected during childhood, some patients may be diagnosed in late adolescence or adulthood, As it is in our case report, during evaluation for early onset systemic hypertension and/or when a significant gradient between upper and lower extremity blood pressures is identified.[2]

We illustrate this fact by the case of a young patient with arterial hypertension secondary to an unrecognized coarctation of the aorta diagnosed during complications.

Case report:

We report the case of a 36-year-old patient, hospitalized in intensive care following a cranial traumatism.

Initial clinical examination reveals an unconscious patient with a Galscow score of 11/15. Hemodynamically and respiratory stable with a blood pressure of 12/8 and a heart rate of 75 beats per minute.

The initial brain computed tomography without contrast done at H3, revealed a left deep parietal intraparenchymal hematoma.

The rest of traumatic assessmentdid not reveal any abdominal or thoracic traumatic injuries.

The patient was intubated due to neurological deterioration.

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CT brain of control founds an increase in size of the intra parenchymal hematoma with cingulate and left uncale herniation with supratentorial ischemic strokes of semioval centers and the right thalamus of similar density to the CSF. (fig.1)

During his hospitalization in intensive care, the patient presented unexplained hypertensive peaks, resistant to antihypertensive drugs.

Transthoracic echocardiography founds an aneurysmal dilatation of the aortic sinus measuring 40 mm and bicuspid aortic valve type 1.

CT angiography (CTA) shows a segmentary stenosis of aortic isthmus below the origin of the left subclavian artery, reduction in luminal caliber of the descending aorta measuring 18 mm with axillary, peri scapular, intercostal, and thoracic-abdominal collateral vessels whichare an indirect clue to the severity of the disease. (fig.2 and 3)

We have noted a left ventricular hypertrophy measuring 21 mm in maximum thickness without aneurysmal dilatation of the ascending thoracic aorta.

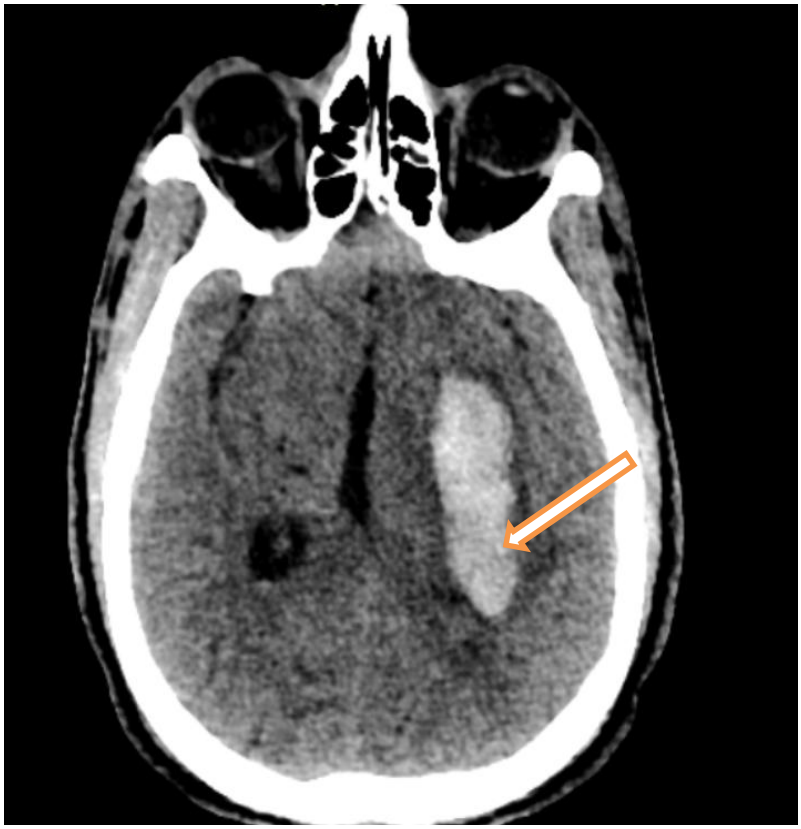

Fig.1:- Axial CT-Brain showing a left parieto-temporal hematoma.

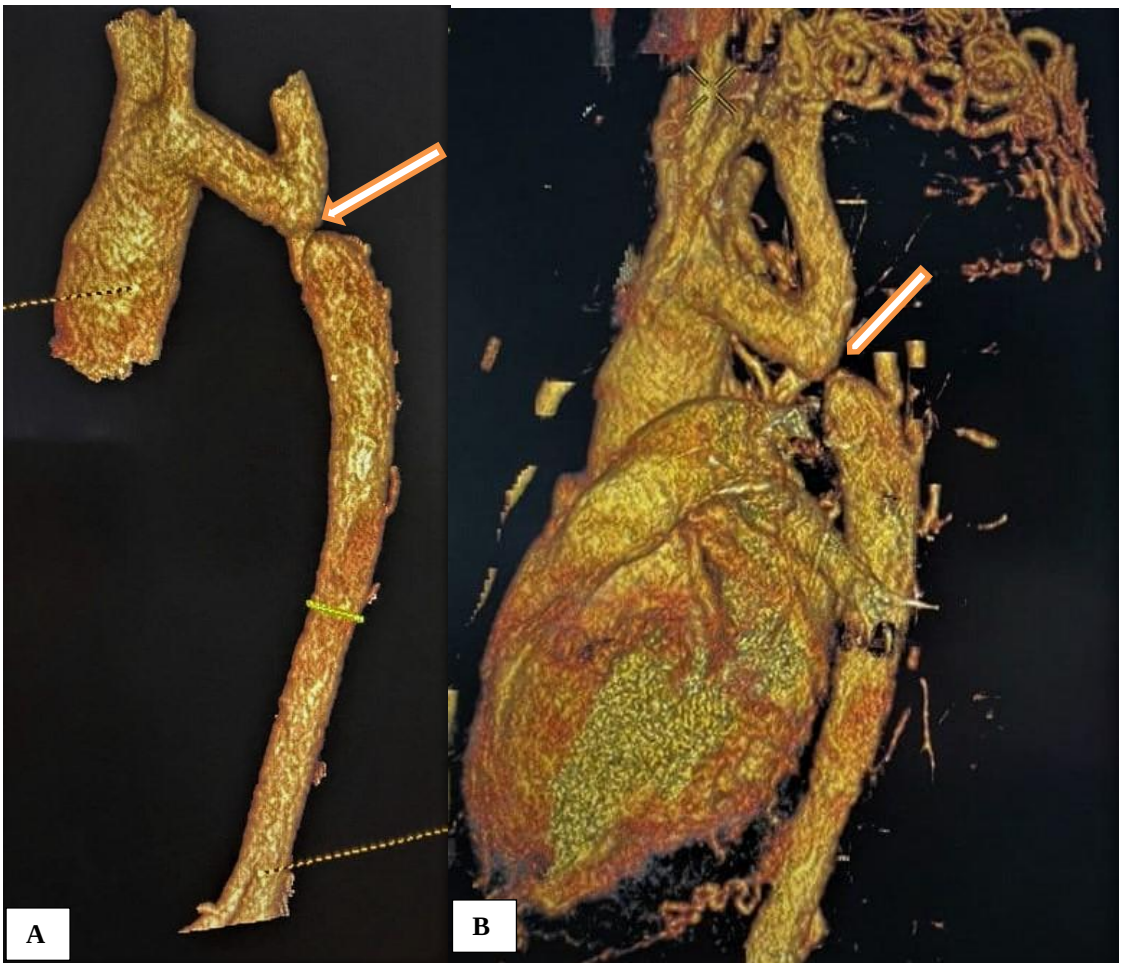

Fig.2:

A:three-dimensional volume-rendered CT angiography shows segmentary stenosis of aortic isthmus below the origin of the left subclavian artery with collateral vessels, without aneurysmal dilatation of the ascending thoracic aorta.

B: image showing an important collateral vesseldue to segmentary stenosis of aortic isthmus.

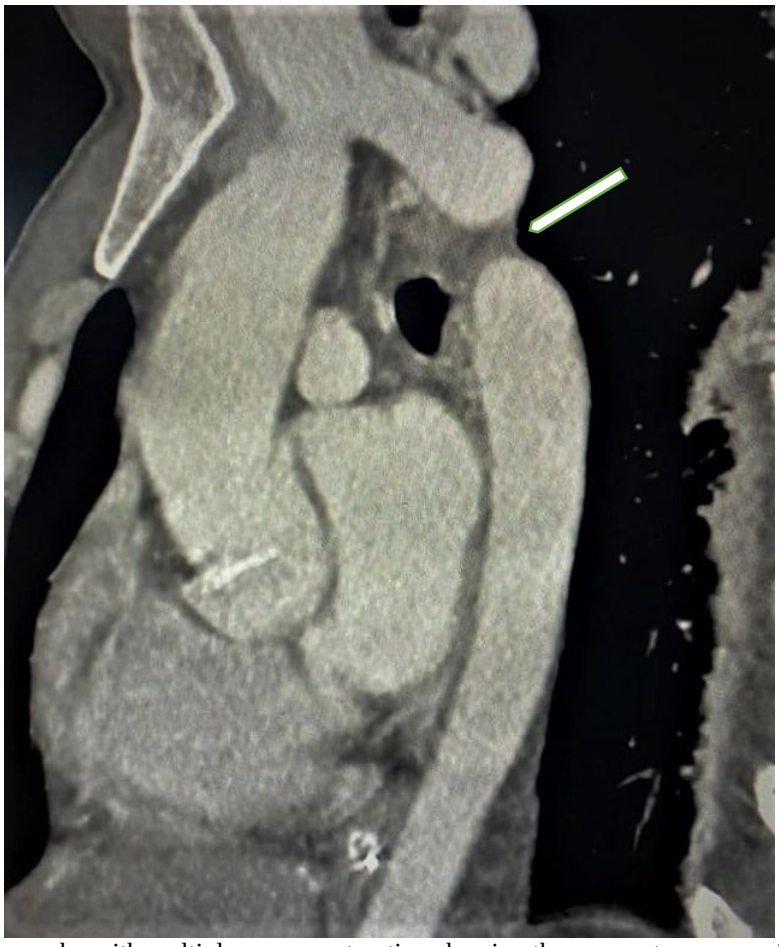

Fig.3:- CT angiography with multiplanar reconstruction showing the segmentary narrowing of the aorta.

Discussion:-

Coarctation of the aorta is a narrowing of the aorta, most commonly occurring just beyond the left subclavian artery. However, it can occur in various other locations of the aortic arch (proximal transverse) or even in the thoracic or abdominal aorta.

The narrowing of the aorta raises the upper body blood pressure, causing upper extremity hypertension. Unrepaired coarctation leads to premature coronary artery disease, ventricular dysfunction, aortic aneurysm/dissection, and cerebral vascular disease by the third or fourth decade of life [3,4,5].

It's often discovered and corrected during childhood, however 10 to 15% of aortic coarctation are diagnosed in adolescence or adulthood.

Non-operated patients usually die before the age of 50 years due to complications of hypertension as it is in our observation,premature coronary artery disease and the formation of aneurysms upstream of the coarctation [6,7,8].

The clinical picture may be variable depending on the presence of other associated congenital heart defects. Premature hypertension is among the most important clinical complication, especially in adult patients, hypertensive heart failure may complicate the clinical picture. Abnormal differences in the upper and lower extremity arterial pulses are hallmark findings in aortic coarctation patients with a difference of 20 mmHg,which can be minimized in case of collateral circulation. Therefore, femoral pulses should be palpated, and blood pressure measurements should be performed separately for each arm. Also systolic, diastolic, or continuous murmurs can be detected during physical examination [9].

The primary goal of imaging in the evaluation of aortic coarctation is oriented toward diagnosis witch includes : Identify the location and hemodynamic significance of the coarctation; determine arch sidedness, branching patterns, and vessel length involved; diagnose other associated congenital anomalies such as bicuspid aortic valve.[10]

CHEST X-RAY :

chest radiography findings may be normal in patients with aortic coarctation. However, there is three conventional signs on the chest X-ray that may be subtle but may provide clues about the presence of an aortic coarctation [11] :

1. Pre- and post-stenotic aortic dilatation creates a notch at the site of the coarctation resembling the shape of the number '3'
2. Notching on the inferior aspect of the third to ninth ribs because of the increased collateral flow through the intercostal arteries.
3. The ascending aorta may be dilated forming a curvilinear shadow along the right sternal border.

Tranthoracic Echocardiography:

Echocardiography is the primary imaging modality for evaluation of the heart. One of the major advantages of echocardiography is the ability to quantify the gradient across the coarctation segment.

The analysis includes different modalities such as two-dimensional ultra Sound, Doppler (continuous wave spectral or color) that improved diagnostic [12], pulsed wave Doppler has also been found to be useful in the diagnosis and evaluation of coarctations [13, 14]

The trans-stenotic pressure gradient can be evaluated from the maximum values of these velocity peaks using the Bernoulli formula. In color Doppler the mosaic flow of the coarctation takes on an hourglass aspect by resolving in the descending aorta where it is very turbulent.

In addition, the presence of associated congenital heart and valve abnormalities and cardiac function can also be evaluated effectively in a noninvasive manner.

However, the main downside of this modality is the poor imaging window in some patients and the long distance between the probe and the isthmic region [15].

Computed Tomography

Computed tomography angiography (CTA) has also become an indispensable tool in the diagnosis and management of this disease.

It offers several advantages over MRI-based techniques: high spatial resolution while imaging the aorta, shorter acquisition times, presence of metallic implants is not a contraindication, and a prior intervention with stent placement does not lead to a signal void artefact [16,17].

Several techniques are now available to evaluate the aorta before and after an intervention. Dual source CTA has been found to have 100% accuracy in evaluating the diameter and length of aortic coarctations compared to findings on surgery in a case series [18]

Excellent anatomic detail of the aorta and coarctation segment are disponible with isotropic reformatted images in different plans, as well as other associating aortic and cardiac abnormalities using ECG gatting.

Three-dimensional volume rendering techniques provide important spatial information not evident on two-dimensional imaging and have made it easier to understand anatomic orientation of the aorta and coarctation segment.[19]

The collateral circulation can also be evaluated which can be an indirect sign of the severity of the stenosis. The report must also include the dimensions of the aortic root and ascending aorta for further follow-up imaging.

CT-based techniques are also useful in the evaluation of postoperative complications: stent fractures, displacement of the stent, aneurysm formation or dissection.

Finally, we can use also the three-dimensional data for intraprocedural guidance during transcatheter interventions using cone-beam CTA (or three-dimensional rotational angiography)[20].

CARDIAC MRI :
Cardiac MRI and MR angiography became a robust technique for the evaluation of the aorta, and they provide excellent anatomic details.

Black blood double inversion recovery T1-weighted images are especially helpful for anatomic detail of the coarctation segment and other neighboring anatomic structures.

MR angiography with white blood unenhanced cine gradient-echo sequences can be helpful for patients who present with contrast allergy and chronic kidney disease.

In addition to anatomic imaging, functional information can also be acquired with phase-contrast imaging. This combination of morphological and functional data has excellent sensitivity (95%) and specificity (92%) for evaluation of aortic coarctation [21]. But there is some limitations on CMR: certain metallic implants may not be MRI safe, claustrophobia, gadolinium contrast may predispose to nephrogenic systemic fibrosis in patient with renal insufficiency, also stents deployed across coarctations create signal void artifact which interfering with diagnosis of complications at the repair site.

Conclusion:-

Aortic coarctation is one of the most common congenital heart diseases that has benefited from the evolution of imaging modalities basedespeciallyon CT-techniques and cardiac MR , which are so helpful for the assessment of this disease and enables in-depth understanding of each patient's unique anatomy and physiology such that we may provide patient specific management option.

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