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ASYMPTOMATIC DOUBLE AORTIC ARCH ACCIDENTALLY DIAGNOSED FOR A CHILD WITH T -CELL LYMPHOMA - CASE REPORT

Hălmaciu Ioana[1], Suciu Bogdan Andrei[1], Roșca Sorin[1], Nagy Bota Monica Cristina[1], Trâmbițaș Cristian[1], Brînzaniuc Klara[1], Ivănescu Adrian[1]

[1] University of Medicine, Pharmacy, Science and Technology Tîrgu Mureș

Suciu Bogdan Andrei

University of Medicine, Pharmacy, Science and Technology Tîrgu Mureș, Romania, Gheorghe Marinescu str. No 38
email: suciubogdanandrei@yahoo.com

ABSTRACT

Introduction. Double aortic arch (DAA) represents a vascular malformation generated by the persistence of the right dorsal aorta from the intrauterine life. An aortic ring is formed, that surrounds the trachea and esophagus, resulting in difficulty in breathing and swallowing.

Case report. We report the case of a 13 years old male child who was admitted to the pediatric surgery department accusing the presence of a paravertebral subcutaneous lesions. Histopathological result of the excised lesion revealed the presence of peripheral T-cell lymphoma. Examination of computer tomography angiography (Angio-CT) revealed the presence of a complete arterial chain (aortic double arch - DAA) around the trachea and esophagus, without signs of compression.

Most cases are diagnosed in the first year of life, the literature reports a few cases of DAA diagnosed late, to the adolescent or adult. Clinically most of the anatomical variants are usually symptomatic especially that are associated with congenital heart defects, including also Fallot tetralogy.

Conclusions. Angio-CT is a very useful method in diagnosing arterial or venous malformations, symptomatic or asymptomatic.

Keywords : double aortic arch, CT angiography

Introduction

In embryological development process, cardiovascular system is the first system that achieve a functional state. The development process of the blood vessels which come from the thoracic aorta is extremely complex and a number of genetic factors are incriminated in producing abnormalities on aortic arches segments. (1) From embryological point of view the aortic arches (AA) develops in close connection with the development of the pharyngeal arches. (1,2)

Each pharyngeal arch contains one arterial AA (symmetrical system). The I, II and V AA, will disappear and the IIIth, IVth and partialy VIth AA will remain to form the permanent, asymmetric superior aortic system. (3,4,5)

The large blood vessels of the body will be formed from the IIIth, IVth and VIth AA, so if there is any abnormal regression or the persistence of any part from the AA it may produce a number of abnormalities. (1)

In normal embryological development the AA is formed from the fourth left AA. Double aortic arch (DAA) occurs due to the persistence of the right and the left fourth AA along with the right and the left dorsal aortas. Once formed the two AA fuse giving birth to a vascular ring that includes the trachea and esophagus. (6)

In case of patients with DAA, the intra-

pericardial part of ascending aorta shows no abnormalities, but after the thoracic aorta is crossing the pericardium it divides into a left arch located anterior (paratracheal) and a right arch located posterior. The two AA once formed will fuse in posterior and left side forming the thoracic aorta. (7)

The most frequent type of DAA is the functional type in which the left common carotid artery (CCA) with the left subclavian artery (left SA) are coming from the left AA and the right CCA with the right SA are coming from the right AA. (8)

Three subtypes of the DAA have been described:

- left dominant (20-25%), in which the left AA is larger,
- right dominant (70-75 %), in which the right AA is larger,
- symmetrical (5 %), in which the two AA are equal. (9,10)

Case report

We report the case of a 13 years old boy, who was admitted to the pediatric department of surgery accusing the presence of a subcutaneous paravertebral lesion. Medical history does not revealed any history of trauma or other associated pathological conditions. On the body system examination there were no clinical signs to incriminate the presence of an acute or chronic pathology, with one exception, the tumor located paravertebral in the dorsal region, with gradual increasing. The tumor was surgically removed. Morphological result revealed the presence of peripheral T-cell lymphoma, diagnostic confirmed by the Pathology Department of Tg. Mureş.

A computer-tomography (CT) examination was performed, besides clinical investigations and laboratory (peripheral smear, blood count, biochemistry, immunogram, serology, medulograma). A thoracic CT scan with contrast substance (Angio-CT) was performed for detecting the stage of the disease and determining the therapeutic protocol. The CT revealed the presence of a complete arterial chain (DAA) around the trachea and the esophagus but without signs of compression at this level. (Figure1)

The left AA had a diameter of 115 mm and the right one 190 mm. (Figure 2)

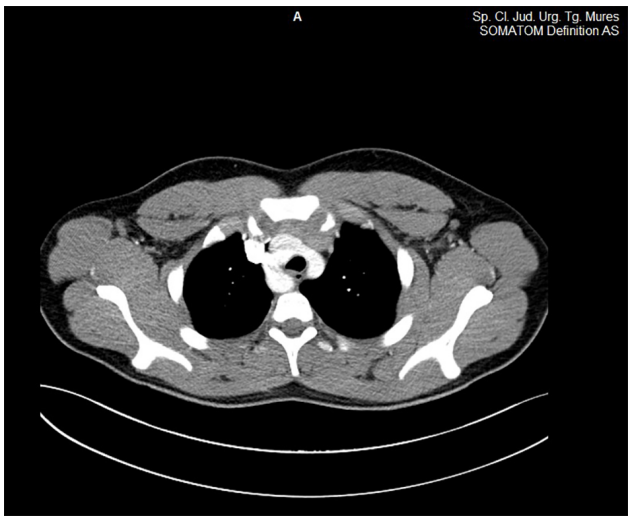

Figure 1 Thoracic CT angiography, axial section. DAA image.

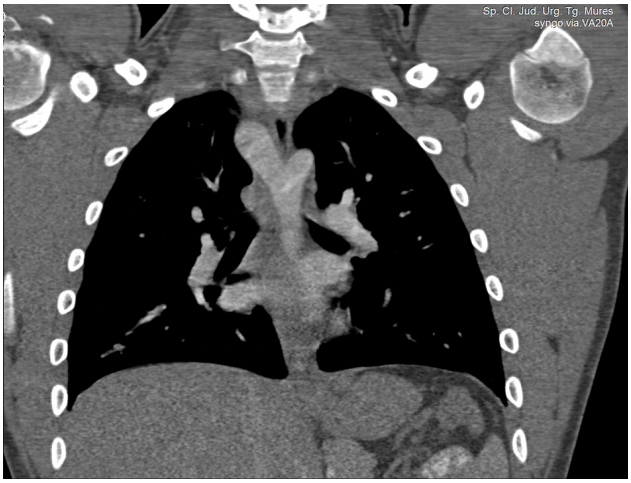

Figure 2 Thoracic CT angiography, coronal section . The the two AA are coming from the ascending aorta.

Also, the left SA and the left CCA were coming from the left AA and the brachiocephalic trunk from the right AA. (Fig. 3). Thoracic and abdominal aorta was normal. (Fig.4)

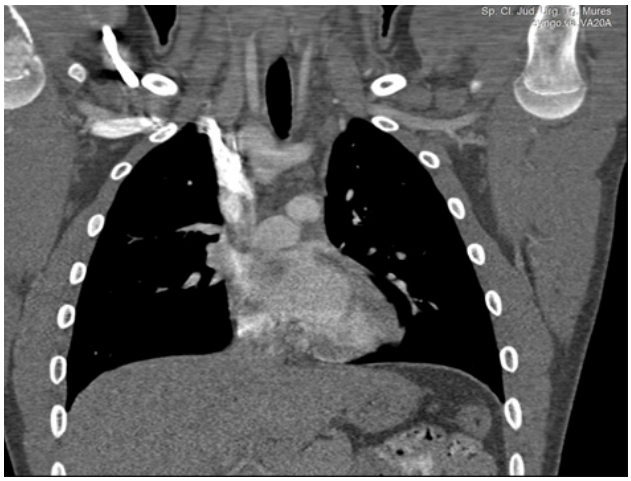

Figure 3 Thoracic CT angiography, coronal section .The origin of emerging arteries from the two AA

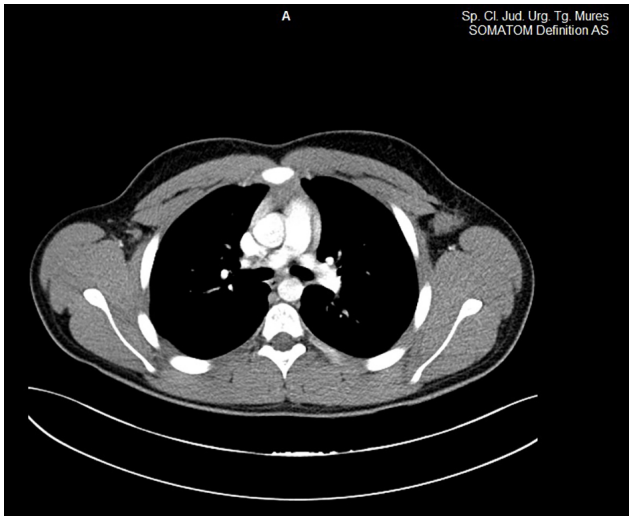

Figure 4 Thoracic CT angiography, axial section. Normal image of the thoracic aorta.

A group of lymph nodes of about 22 mm was observed at left axillary area. No cervical or mediastinal lymph nodes were presents.

Discussions

DAA represents the forming of a completely closed arterial arch around the trachea and the esophagus. The compression phenomena of these organs occurred most often in the first month of life with the presence of respiratory symptoms. (11,12) The literature reported very few cases diagnosed late or during adolescence or adulthood. (10,13-18) Clinically most of the anatomical variants are usually symptomatic especially that are associated with congenital heart defects, including also Fallot tetralogy. (12)

Among symptomatic variants, without associated diseases, the most common is the constrictive traheo-esofagian vasculo-ligamentar ring. In these cases, the clinical picture is noisy by the presence of digestive symptoms (dysphasia lusoria, cyanosis) respiratory symptoms (stridor, cough, apnea) and circulatory symptoms (murmur, cyanosis, chest pain).(3,19,20)

In our case the patient was investigated for others pathologies and the discovery of the arterial malformation was purely accidental. It revealed the presence of a dominant right DAA. The incidence of this form is 70-75 % as reported in the literature. The patient was completely asymptomatic and the CT angiography examination has not found any compression exerted by the arterial ring.

The surgery is the treatment in most cases after the diagnosis of this malformation. In this case the patient was asymptomatic and without signs of compression and the surgery was not recommended. The recommendations on this case were surveillance and periodic evaluation till the first symptoms of vascular disease will appear.

In the literature were cited very few cases of silent DAA diagnosed in the sixth and seventh decade of life. (21,22)

Conclusions

Angio-CT is a very useful method in diagnosing arterial or venous malformations, symptomatic or asymptomatic.

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