Improved resolution of cardiovascular imaging techniques allows better assessment of the morphology of cardiovascular malformations. Multi-slice computer tomography with three-dimensional reconstruction provides both static and dynamic images with high temporal and spatial resolution. We present a three-dimensional reconstruction of scimitar syndrome using multi-slice computer tomography.

Keywords: Congenital heart disease; Imaging; Tomography

1. Introduction

Right hemi-anomalous pulmonary venous drainage is rare, and consists of a descending right pulmonary vein that enters the inferior vena cava just above or below the diaphragm. The anomalous venous channel produces characteristic X-ray findings of a crescent-like shadow in the right lower lung field, designated as the Scimitar Syndrome. Multi-slice computer tomography (CT) with three-dimensional (3D) reconstruction confers excellent images of congenital malformations for both cardiologist and cardiothoracic surgeons alike [1]. We present a case of Scimitar Syndrome illustrated with 3D reconstruction.

2. Case report

A 33-year-old male was admitted with dyspnea on exertion. His past history included a predisposition to recurrent respiratory tract infections, smoking and hyperlipidemia. His chest radiograph performed on admission (Fig. 1A), revealed a scimitar-shaped shadow in the right lung field. Although the right lung and airway was normal in size, the right hilum appeared hypoplastic. Spirometry revealed moderate airway obstruction pattern, with a FVC of 3.86 l (70% predicted), FEV1 of 2.12 l (52% of predicted) and FEV1/FVC ratio of 81%. On echocardiography, the right atrium was moderately dilated, the left atrium and both ventricles were normal in size and no defect of the atrial septum was identified. The systolic pulmonary artery pressure was estimated at 43 mmHg from echocardiography. Cardiac catheterization detected a left-to-right shunt with a QP/QS ratio of 1.9. The systolic pulmonary pressure was 47 mm Hg. Levophase cineangiography after selective right pulmonary artery injection showed blood from the right pulmonary vein draining abnormally into the inferior vena cava (IVC). Angiography was not sensitive enough to detect any stenosis at the level of the junction between scimitar vein (SV) and IVC (Fig. 1B).

Multi-slice CT with bi-dimensional reconstruction (Fig. 1B) confirmed the abnormal pulmonary venous drainage from the right lung via a large SV that drained infra-diaphragmatically at the IVC, suggesting a partial obstruction of the SV at its junction with the IVC. Other anomalies in arterial supply to the right lung, bronchial arteries, dextrocardia or hypoplastic right lung were not identified. Multislice CT with 3D reconstruction (Fig. 2) showed total anomalous venous drainage of the right lung. The SV was a large collector of the all the right sided pulmonary veins passing anterior to the right hilum. The 3D reconstruction of the confluence between the SV and IVC confirmed obstruction. Three hypoplastic right pulmonary

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veins were detected flowing into the left atrium. Non-arterial supply from the descending aorta to the right lung was identified.

3D reconstruction precisely defined the anatomy of the SV and its relation with other structures for confirmatory diagnosis and facilitation of the planning of operative intervention. This is illustrated by this case report where obstruction of the confluence of the SV with the IVC was accurately delineated.

3. Discussion

An unusual type of partial anomalous pulmonary venous connection is the scimitar syndrome characterized by an anomalous right pulmonary vein that generally drains the entire right lung infradiaphragmatically at the IVC below the right atrial–IVC junction. The anomalous pulmonary trunk, termed SV, usually passes anterior to the hilum of the right lung and entrance into the IVC just superior to the hepatic veins. The atrial septum is usually intact, however, occasionally a defect may be present. In the most instances, anomalies of the right lung may be present, most commonly, right lung hypoplasia with marked mediastinal shift and dextrocardia. In this case, the right lung and airways were normal in size without pulmonary lobar sequestration or hypoplasia of the right pulmonary artery.

Diagnoses and treatment of patients with congenital heart disease demands a complete appreciation of the anomaly and adjacent anatomy in three dimensions. A chest radiograph could only detect the silhouette of the SV and a small right hilum. Two-dimensional echocardiography was unreliable in the detection of anomalous connection of right pulmonary vein to the IVC. The classical diagnosis of Scimitar Syndrome is based on cardiac catheterization and cineangiography. Cardiac catheterization can screen for pulmonary hypertension and detect the presence of a shunt. Calculation of the shunt fraction (QP/QS) is of particular importance in patients with anomalous pulmonary venous drainage without atrial septal defect as the shunt may be greater than expected. Selective angiography is most helpful and showed the anatomic details of the malformation. The identification of the specific anatomic details of the connection of the SV into the IVC are shown by levophase cineangiography after selective right pulmonary artery injection. Angiographically the partial obstruction to the SV can be shown at its confluence with the IVC. Usually, patients are symptomatic if left untreated can result in pulmonary hypertension [2].

The original cardiac imaging systems, such as angiography, produced in two-dimensional images, and necessitates mental reconstruction of the displayed images to formulate a three-dimensional image. Now, 3D imaging systems, namely, magnetic resonance [3–5], volumetrically...
acquired echocardiography [6], and CT, are available and provide three-dimensional data [7,8]. However, 3D images of the heart can only be provided by rapid acquisition, such as ultrafast CT with digital spatial reconstruction [1]. The 3D reconstruction technique, called volume rendering, is used in CT angiography to study the anatomy of blood vessels and provides static and dynamic tomographic images with excellent temporal and spatial resolution. Because of the absence of overlying structures and the 3D nature of ultrafast CT acquisition, anomalous pulmonary venous connections in the Scimitar Syndrome can be accurately assessed.

At this time, the 3D reconstruction process is still labor-intensive and cannot be done routinely. However, improvements in both software and hardware are likely to make this technology increasingly available for medical use in the near future.

References
Appendix A. ICVTS on-line discussion

Author: Sameh Sersar, Cardiothoracic Surgeon, Mansoura University, Cardiothoracic Surgery, Mansoura 123, Egypt

Date: 03-Jun-2003 12:48

Message: Re this new modality, I would like to ask a question. For a developing country, which modality do you prefer MRI, CT or the volumetrically acquired echocardiography? (In regards specifically to the specificity, the experience and the costs?) Does this modality have a postoperative role?

Response

Author: Fernando Hornero, Consultant Surgeon, Hospital General Universitario, Cardiac Surgery, Av Tres Cruces s/n, Valencia 46014, Spain

Date: 20-Jun-2003 14:33

Message: This case is an unusual malformation, and our experience is relatively small to define which could be the best preoperative non-invasive image technique. However for us, based on the experience obtained in other cardiovascular pathologies (aneurysms, aortic dissection, redo-CABG; preoperative studies of the aortic wall calcifications, etc.) MRI and CT 3D imaging reconstruction provide an excellent demonstration of the anatomy and allow a more precise planning of the operation than the conventional angiography study. We do not have experience with the 3-dimensional echocardiography. The CT Scan has the disadvantage, compared with the MRI, of the use of intravenous contrast (allergies, renal failure) and x-ray radiation. However, the new modalities of Multislice CT offer a higher spatial resolution (it allows the performance of sub-millimetric slices) and also a higher temporal resolution (it is possible to scan a large chest area during a single breathhold %bb 15 seconds) than the MRI. Last generation CT have a markedly lower price than MRI-high local magnetic field (1.5 Teslas). Probably, CT scan is simpler and more economic than the conventional angiographic study, as it does not require hospitalization of the patient.

Author: Antonio Corno, CHUV, Cardiovascular Surgery, 46 rue du Bugnon, Lausanne CH-1011, Switzerland

Date: 04-Jun-2003 07:39

Message: Wonderful imaging of the 3D reconstruction of the congenital partial anomalous pulmonary venous return. The technique is a valuable contribution to the decision making process for the surgical treatment of the ‘scimitar syndrome’. The reported case seems quite unusual regarding both the morphology as well as the presentation at an age (33 years) older than generally observed. After the wonderful job performed by the Authors to produce such very clear imaging, the readers, particularly the surgeons, are curious to know what has been the subsequent treatment of the patient. Probably the Authors could add a short comment to provide us with the rest of the clinical history, in order to help the colleagues taking care of these types of patients with more information useful for the decision making process.

Response

Author: Enrico Aidala, Osp. Infantile “Regina Margherita”, Pediatric Cardiac Surgery, P.zza Polonia 94, Torino 10126, Italy

Date: 06-Jun-2003 08:56

Message: Accurate evaluation of congenital anomaly or iatrogenic pathologies of the systemic or pulmonary venous returns is difficult to achieve, particularly in children. Imaging studies include CT, NMR and angiography. The 3D-CT reconstruction of the case by Hornero and coll is really impressive; otherwise it comes after echocardiographic and angiographic studies not useful to define anatomical details to perform the definitive surgical operation (about which nothing is reported!). In children we often have the same diagnostic problems. Recently we evaluated some patients with NMR with excellent results and advantages. Neither contrast media nor X-rays are necessary and, at the moment the new cardiac dedicate software is able to perform a complete anatomic evaluation in a short time (about 10-15 seconds). Moreover with a single exam it is possible to know everything necessary for planning the operation. Although dedicated software and machinery are not as greatly diffused as CT or cath-lab, in the near future I think NMR will be probably the best technique for cardiac study.