Case Report **Open Access**

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Prenatal Diagnosis, 3-D Virtual Rendering and Lung Sparing Surgery by ligasure device in a baby with "CCAM and intralobar pulmonary sequestration"

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Abstract: Congenital cystic lung lesions are a rare but clinically significant group of anomalies, including congenital cystic adenomatoid malformation (CCAM), pulmonary sequestration, congenital lobar emphysema (CLE) and bronchogenic cysts. Despite the knowledge of these lesions increasing in the last years, some aspects are still debated and controversial. The diagnosis is certainly one aspect which underwent many changes in the last 15 years due to the improvement of antenatal scan and the introduction of 3-D reconstruction techniques. As it is known, a prompt diagnosis has an essential role in the management of these children. The new imaging studies as 3D Volume rendering system are the focus of this paper. We describe our preliminary experience in a case of hybrid lung lesion, which we approached by thoracoscopy after a preoperative study with 3D VR reconstruction. Our final balance is absolutely positive.

Keywords: Lung Sparing Surgery, intralobar pulmonary sequestration, pediatric thoracic surgery

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1 Introduction

The term congenital thoracic malformations (CTMs) is a generic term which includes a large spectrum of lesions of the airways and lung. The most common forms of CTMS are: congenital cystic adenomatoid malformations, bronchopulmonary sequestration, bronchogenic cysts and lobar and segmental emphysema. The incidence of CTMs is 3.52 per 10.000 live births [1]. Despite the increasing knowledge about these lesions, many aspects related to pre and postnatal management remain unclear. Many authors suggest that a surgical approach is mandatory but the timing is still discussed. The development of new imaging studies has improved postnatal diagnosis and management of CTMs replacing bi dimensional studies, especially in the evaluation of vascularitation. We have focussed on the use of 3D Volume rendering system as an instrument to improve the surgical approach of these lesions and we have described our preliminary experience in a case of hybrid lesion.

2 Case report

A 28-year-old woman was seen in Antenatal Clinic for the diagnosis of a cystic lesion of the right lower lung. There were no other problems. The Caam Volumetric Ratio was < 1.6 for all pregnancy. A full-term female, 3.120 g, was delivered vaginally. A chest X-ray was performed on the 1st day of life and showed the known cystic lesion in the absence of a mediastinal shift. A chest CT at 6 days of age showed a mass in the right lower lobe, measuring 38 x 15 x 16 mm, with multiple cystis which measured between 3 and 11 mm. Echocardiography was normal. The patient was discharged. Magnetic resonance imaging (MRI) was

performed at 1 month of age confirming the unilateral lesion but with an aberrant sub-diaphragmatic artery supply. The child remained asymptomatic until 5 months when she had an episode of bronchitis. Radiological images were converted with VR render (Figure 1). Surgical excision of CTM was performed at 8 months of age. A thoracoscopic approach was performed. Three 5 mm trocars were placed (1 – V intercostal space in the middle axillary line, 1 – VII intercostal space on the anterior axillary line and 1 VII intercostal space in the posterior axillary line). A 0° mm camera was used. An accessory artery was ligated by 3/0 vycril and excised by ligasure. A lung sparing surgery was performed by ligasure device. The mass was extracted with an enlargement of lowest port (Figure 2). A chest drainage was placed at the end of procedure and it was removed after 24 hours. There were no intraoperative or postoperative complications. The in-patients stay was 8 days. Histological examination was performed as described elsewhere [2-9] and confirmed the preoperative diagnosis of hybrid lesion (CCAM and intralobar bronchopulmunary sequestration). At follow up (6 months) the patient was completely asymptomatic. Scars were well.

Ethical approval: The research related to human use has been complied with all the relevant national regulations, institutional policies and in accordance the tenets of the Helsinki Declaration, and has been approved by the authors' institutional review board or equivalent committee.

Informed consent: Informed consent has been obtained from all individuals included in this study.

3 VR Render

VR render is a well-known visualization method for 3 D reconstruction of medical images. It's an IRCAD image view software that allows the user to visualize bidimensional images in 3D thanks to volume rendering. It works with CT-scan and MRI images. It's also possible to manipulate the volume rendering with multiple clipping planes in order to focus the lesions.

4 Discussion

Even if these malformations are rare, they may lead to morbidity and mortality. Congenital cystic adenomatoid malformation (CCAM) is the most common congenital thoracic malformations (CTMs), representing about 30% of them, and has an incidence of 1:25.000-35.000 live births [10]. It is a hamartomatous lesion, which results from an abnormal development of fetal terminal bronchioles and it is characterized by a multiple cystis with various size. It is connected with the bronchial tree and it is supplied from the pulmonary artery. The original classification is that of Stocker et al. which categorised the lesion into one of three groups on the basis of the size of the cysts and other histological criteria (3). rew Type 1 CCAM contain one or more large cysts (2±10 cm) and make up approximately 50% of cases. Type 2 malformations make up 40% and contain multiple smaller cysts (0.5±2 cm) more uniform in size. Type 3 abnormalities (10%) appear solid on gross exam- ination but contain multiple 0.3±0.5 cm cysts on microscopic examination. The last classification, instead, added other 2 rare types of CCAM, 0 and 4. Type 0 is an essentially tracheoesophageal defect and type 4 an entirely alveolar defect. Due to this new classification, Stocker himself proposed a new name of the esion: congenital pulmonary airway malformations CPAM [11-12]. Bronchopulmonary sequestration (BPS) represents 0.15-6.45% of all CTM [9] and it is a non functioning lung mass which is not connected with the bronchial tree and which is supplied from systemic vessels. Based on its visceral pleura BPS is divided in intralobar (ILS-75%), which shares the pleura with lung and extralobar sequestration (ELS-25%) which has an independent pleura. Fifty percent of ELS are associated with CCAM II type, forming an hybrid lesion [13-16]. A hybrid of CCAM and BPS is a newly recognized congenital lung lesion that demonstrates clinicopatho- logic characteristics of both lesion types. The occurrence of CCAM and BPS in the same patient likely reflects similar embryological origins of these lesions. Both CCAM and BPS are thought to arise early in development. The prenatal diagnosis of this hybrid lesion is difficult, but to date, the preoperative diagnosis is possible with new technologies. Despite the knowledge of CTMs increasing in the last year, some aspects are still debated and controversial. The diagnosis is certainly one aspect which underwent many changes in the last 15 years due to the improvement of antenatal scan and the introduction of three dimensional reconstruction techniques. As it known, a prompt diagnosis has an essential role in the management of these children. Antenatal ultrasonography has increased the awareness and diagnosis of congenital cystic lung lesions, resulting in a better understanding of the natural history of many of these lesions and also allowing provision to be made for delivery and postnatal management [13]. However, postnatal imaging studies are

the focus of this paper. One of the last 3D system is Volume rendering (VR) which has largely replaced other 3D reformatting techniques in evaluations of airway and vascular pathologic processes [16]. It is a free IRCAD Image Viewer Software, based on volume rendering system which allows the user to visualize in 3D the medical images from CT-scan or MRI.

Preoperatively, the use of 3D reconstruction provides unique insights into the anatomical architecture of malformation and allows the user to define the relationship with the adjacent structures. Lee et al. [17] said that the volume rendering, permits to well identify the aspects of vascularization especially the venous drainage resulting in the differentiation of intra and extra sequestration. In our opinion VR render permits to well identify the borders of pleura thus the two forms of sequestration.

Our final balance is absolutely positive. The creation of volume-rendered images allowed the visualization of different tissue types with varying color and opacity (transparency) levels and provides valuable information regarding the spatial relationship of anatomic structures. When we opened thorax of patient we knew perfectly the anatomic profile of the lesion and its relationship with mediastinic structures. The interactive images helped us in the detection of trocars placement. This 3D reconstruction also permitted us to identify the presence of hybrid lesion. We followed the route of systemic artery that supplied the mass and saw the pleural profile [18-30]. Due to all of the preoperative information the operating time was 40 minutes.

5 Conclusion

In conclusion, even if this is our first experience of preoperative 3D VR reconstruction for a lung lesion, we think that three-D reconstruction techniques are very useful in evaluations of airway and lung lesions. The creation of volume-rendered images is a simply process and requires less than 30 minutes. A preoperative 3D study, allows the user to diagnose the lesion and plan the surgery accordingly. Thus making the surgery safer and faster.

Conflict of interest statement: Authors state no conflict of interest

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