Diagnosis and Surgical Treatment of Intradiaphragmatic Extralobar Pulmonary Sequestration: a Report of 10 Cases

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Abstract

Background Intradiaphragmatic extralobar pulmonary sequestration(IDEPS) is a rare type of pulmonary sequestration (PS) and has been reported in very few studies. The purpose of this study is to access diagnosis and operative treatment of an IDEPS.

Methods Patients with PS who were diagnosed and treated in our center from January 2015 to December 2020 were retrospectively analyzed, in order to identify patients with IDEPS. Clinical datas regarding diagnosis, surgical procedures and outcomes were documented.

Results From January 2015 to December 2020, 215 patients with PS were surgically treated in our center, including 87 patients with extralobar pulmonary sequestration(EPS). 10 cases were identifed as IDEPS (2 males, 8 females; age, 5 months to 21 months). Prenatal ultrasounds and enhanced computed tomography (CT) showed the presence of IDEPS in 4 cases and 7 cases, respectively, and a three-dimensional (3D) reconstruction software was perfectly performed to identify the location of the lesions in 3 cases._The surgeries were smoothly performed by laparoscopic surgery in 1 case, video-assisted thoracic surgery (VATS) in 5 cases(Group 1) and Da Vinci robot-assisted thoracoscopic surgery(DVRATS) in 4 cases(Group 2). In Group 1 (5 females; the mean age, 8.0 months), the average operative duration, intraoperative blood loss volume, length of stay after operation, and postoperative thoracic catheter indwelling duration were 48 minutes, 3.8ml, 6.4 days and 2.2 days, respectively. That of Group 2(1 male, 3 females; the mean age, 9.5 months) were 80 minutes, 3.5ml, 4.3 days and 1.5 days, respectively. The IDEPS cases who followed up rang from 3 months to 36 months were included in this retrospective study and no side effect had appeared.

Conclusions It's challenging to diagnose an IDEPS through prenatal ultrasound or CT, and a 3D reconstruction software was proved to be capable to assist on the assessment of an IDEPS. We suggested early surgery to removel of an IDEPS, and the best path was accessing the mass from the chest. DVRATS or VATS for the treatment of an IDEPS is safe, feasible and effective. Furthermore, compared with VATS, DVRATS is 3D magnified view, more flexibility and precision.

Background

Pulmonary sequestration (PS) is a rare congenital pulmonary development anomaly in infant and children, defined as pulmonary tissue lack of a tracheobronchial connection and has a systemic arterial supply, occurred in approximately 0.15%~6.4% of all pulmonary malformations[1][1].

According to the presence or absence of visceral pleura, PS is ususlly divided into two types: intralobar pulmonary sequestration (IPS) and extralobar pulmonary sequestration (EPS). IPS usually appears at the lower lobe of both lungs, especially at the left lower lobe. EPS accounting for 0.1 to 1.8% of cases is the second most common lung malformation in children[2], and mainly locates between the left lower lobe of the lung, especially near the mediastinal spine[3], accidentally occurs at the inner part of the diaphragm, abdomen and neck[3–6]. In same cases, IPS and EPS may co-exist[3, 4]. As in EPS, Intra-diaphragmatic extralobar pulmonary sequestration (IDEPS) occurs most often on the left[3, 5], was first reported by Caulet in 1962, and accounting for13%-18% of the PS[5, 7].

It has been reported that the rate of extralobar sequestration infection in the diaphragm was 16.7 % [3]. The infection may cause diaphragmatic edema, adhesion and elevation, making it more difficult to separate the

diaphragm fibrous tissue from the mass. Meanwhile, respiratory function may be affected because of the elevation of diaphragm. Hence, early surgery management to resect the IDEPS is necessary. In this study, we performed a retrospective study of 10 such cases and summarize experiences in the diagnosis and surgical treatment.

Patients And Methods

1.1 Patients

Data on patients with PS who underwent surgery in our institution from January 2015 to December 2020 were retrospectively collected. The diagnosis of patients with IDEPSs were performed by enhanced computed tomography (CT) after birth. Surgeries were successfully performed to manage with IDEPSs, including video-assisted thoracic surgery(VATS), Da Vinci robot assisted thoracic surgery(DVRATS) and laparoscopic surgery. Patients are free to choose VATS or DVRATS according to their preferences. The gender, age, operative duration, intraoperative blood loss volume, length of stay after opertation, postoperative thoracic catheter indwelling duration and postoperative side effect were documented in this condition.

1.2.1 Da Vinci robot assisted thoracoscopic surgery (DVRATS)

Operations were performed using routine tracheal intubation, general anesthesia and single-lung ventilation. Patient was positioned in the lateral decubitus position with the mass side facing up and a pad was positioned to slight widening of the intercostal space. Four incisions were performed(Figure. 1a): An observation port (8 mm) was placed in the fouth intercostal space at the mid-axillary line, two operation ports (8mm) were placed in the sixth intercostal space at the midclavicular line and the seventh intercostal space at the subscapular line, respectively. The remaining 5 mm port (assistant port) was placed in the fifth intercostal space at the anterior axillary line. During the operation, a mass protruding from the diaphragm to the chest cavity could be found(Figure. 1b). The diaphragm was opened alo9ng the edge of the mass, and the mass was completely resected(Figure. 1c,e). The nutrient arteries were ligated with 3-0 silk thread or clipped with vascular clamp(Figure. 1d). Then, the diaphragm was repaired with interrupted 3-0 barbed suture(Figure. 1f). Lastly, a thoracic drainage tube was routinely placed and the operation was completed. If the single lung ventilation is failure, the pressure of $C0_2$ was maintained at 6~8 mmHg to produce an artificial pneumothorax.

1.2.2 Video-assisted thoracoscopic surgery (VATS)

Routine tracheal intubation, general anesthesia and single lung ventilation. Patient was positioned in the lateral decubitus position with the mass side facing up and a pad was positioned to slight widening of the intercostal space. Three incisions were performed: An observation port (5 mm) was placed in the seventh intercostal space at the mid-axillary line. Two operation ports (5 mm) were placed in the fourth intercostal space at the anterior axillary line, and Xth intercostal of subscapular line. The operation method is the same as above.

1.2.3 Laparoscopic surgery

Routine tracheal intubation combined with general anesthesia. The patient was in supine position with a pad to slightly elevate the abdomen. Three incisions were performed: An observation port (5 mm) was placed in navel. Two operation ports (5 mm) were placed in the four centimeters above umbilicus and the right abdomen. The operation method is the same as above. The pressure of $C0_2$ was maintained at 6~8 mmHg to produce an artificial pneumoperitoneum.

Results

A total of 215 patients with PS were enrolled from January 2015 to December 2020 in our hospital, including 128 patients with IPS and 87 patients with EPS. 10 patients(11.5%) with IDEPS were identified(2 males, 8 females; age, 5 months to 21 months, mean age, 10.1 ± 4.6 months)(Table 1), and 1 case complicated with pectus excavatum. All IDEPSs were located in the left diaphragm in our center, no clinical symptom occurred after birth. All cases except one were discovered first in prenatal ultrasounds that showed the presence of a mass above or below the diaphragm, and 4 cases were diagnosed as an IDEPS. One case was incidentally discovered at operation. Enhanced CT was routinely performed after birth and showed the presence of an IDEPS in 7 cases(Figure. 2). Meanwhile, a three-dimensional(3D) reconstruction software(The Hexa3D company, China) was performed to identify the location of the lesions in 3 cases(Fig. 3). Full diaphragm and the mass can be automatically modelled with 3D reconstruction software through enhanced CT imaging of the case. The aberrant nutrient arteries included a blood supply via the thoracic aorta in 1 case, via the splenic artery in 1 case, via the abdominal aorta in 4 cases, via the small artery branches in 4 cases.

Table 1
Characteristics of IDEPS cases

Case	Gender	Operative age(months)	Surgury	Operative time(min)	IOBLV(ml)	Postoperative stay(d)	TCI(d)	Feeding artery
1	female	12	VATS	30	5	7	1	thoracic aorta
2	female	6	VATS	50	5	6	1	abdominal aorta
3	female	5	VATS	30	5	7	2	small artery branche
4	female	8	VATS	40	2	6	1	abdominal aorta
5	female	11	VATS	90	2	6	6	small artery branche
6	female	11	DVRATS	130	5	3	1	splenic artery
7	female	12	DVRATS	60	5	5	2	abdominal aorta
8	male	6	DVRATS	65	2	5	2	small artery branche
9	female	9	DVRATS	65	2	4	1	small artery branche
10	male	21	LS	120	10	7	3	abdominal aorta

VATS, video-assisted thoracic surgery; DVRATS, Da Vinci robot-assisted thoracoscopic surgery; LS, laparoscopic surgery; IOBLV, intraoperative blood loss volume; TCI, thoracic catheter indwelling.

VATS, DVRATS and laparoscopic surgery had been performed in 5 cases, 4 cases and 1 case respectively. One case(male; age, 21 months) who was treated with laparoscopic surgery cost about 120min, and intraoperative blood loss volume was 10ml. VATS and DVRATS were defined as Group 1 and Group 2. In group 1(5 females; the mean age, 8.4 months), the average operative duration, intraoperative blood loss volume, length of stay after operation, and postoperative thoracic catheter indwelling duration were 48 minutes, 3.8ml, 6.4 days and 2.2 days, respectively. In group 2(1 male, 3 females; the mean age, 9.5 months), the average operative duration, intraoperative blood loss volume, length of stay after operation, and postoperative thoracic catheter indwelling duration were 80 minutes, 3.5ml, 4.3 days and 2.2 days, respectively. The diagnosis of IDEPSs confirming in 10 cases were made according to the histopathological results(Figure. 4). IDEPS patients who followed up rang from 3 months to 36 months were included in this retrospective study and all the results from these checkups were normal.

Discussion

PS was first reported by Rokitansky in 1981, and the etiology and pathogenesis of this disease remain unclear[8], which may be related to the imbalance of cell proliferation and apoptosis[9]. Compared with congenital cystic adenomatoid malformation(CCAM), aberrant feeding arteries of PS mainly arise from the thoracic aorta, followed by the abdominal aorta, intercostal artery, and subclavian artery[3, 10]. Studies[3, 11]have showed that PS was associated with many deformities, such as diaphragmatic hernia, congenital heart disease, cyst of bronchial origin, funnel chest(pectus excavactum or pectus carinatum). With the popularization of prenatal examination, PS has increasingly been detected by prenatal ultrasound or incidentally found on postnatal CT imaging. However, it's still challenging to identify an IDEPS. The study found that [12]the arise of PS during the 9th to 12th weeks of embryonic development may have a higher chance of forming within the diaphragm.

The diagnosis of an IDEPS mainly included Prenatal ultrasound, enchanced CT, or magnetic resonance imaging(MRI), play an important role in the surgical planning and safe operative resection. Prenatal ultrasound is a prefer method for the diagnosis of fetal PS [13], However, due to the special location of a sequestered mass in diaphragm, it is difficult to definitely diagnose if the mass is located in the diaphragm using prenatal ultrasound. The detection rate of fetal IDEPS proving the condition was 40% in our study. MRI is of a value method in diagnosing an IDEPS, which can provide multi-level anatomical location information of lesions, and is superior to prenatal ultrasound in assessing the feeding arteries and venous[14]. However, the diaphragm may be affected by respiratory activity, limiting the application of MRI to identify an IDEPS. Given the limitations of MRI, CT scan is less affected by respiratory activity, and has a higher concordance rate for characterising the lung parenchyma, especially the reformatted imaging of CT scan can demonstrate the presence of a mass enveloped by the diaphragm. What's more, CT is the most accurate method for the detection of a systemic vasculature to the mass. Although CT scan has the risk of ionizing radiation, it is the most efficient choice among the compared methods to identify an IDEPS. Hong et al. [5] reported that 11 cases of IDEPSs had been confirmed after birth using enhanced CT, but Olivieri et al.[15] suggested that a diagnosis could only be confirmed at surgery. In our institution, the concordance rate of CT was 70% and the feeding thoracic aorta and abdominal aorta account for 50%, which were lower compaired with previous study[5, 16, 17]. A 3D reconstruction software was used to provide access to more and higher-quality information about patient's 3D anatomy, making it possible to improve the accuracy and reliability of diagnosis and treatment. This seems to be the first report that a 3D reconstructed imaging clearly showed the relationship between a mass and a diaphragm and was helpful for the identify an IDEPS.

Although the appropriate management of PS remains controversial, majority of surgeons suggested PS should be surgically resected [1, 3, 11, 18], and surgical resection should be performed no more than 12 months of the age[3, 18]. Beatrice et al.[1] reported that paediatric patients operated on after 1 year of age developed more respiratory symptoms than those treated before 1 year of age. Stanton et al.[8] found that the rate of postoperative complications of symptomatic PS were significantly higher than that of asymptomatic PS patitents undergoing elective surgery. However, some studies[18–20] demonstrated that an EPS may remain asymptomatic throughout life and even involute over time, suggesting the safety of just following up EPS without surgery management. In currently, VATS has become the main surgical technique with the advantage of minimally invasive, accurate and rapid recovery. Some studies [21–23]had reported the application of transcatheter arterial embolization (TAE) in the treatment of PS. By embolizing the feeding artery, PS occurs ischemia, necrosis and fibrosis. However, patient may suffer from coil deviation, incomplete embolism, recurrence, hemoptysis and infection. In addition, the risk of exposure to ionizing radiation should be concerned, and further longitudinal study is required to validate this new technique. There are very few reports on the management of IDEPS till now. Due to the special location of an IDEPS, it is difficult to differentiate from a neuroblastoma, an adrenal tumor or a teratoma, and has a risk of infection. Once confirmed, we chose surgery treatment and the best path was access the mass from the chest, which were consistent with Hong et al. [5]. During surgery, the aberrant feeding vessels and ruptured diaphragm should be properly managed to avoid massive bleeding and diaphragm expansion. One case who was treated with VATS in our center suffered from infection, making it difficult to identify the feeding vesses, dissociate the tissue and resect the lesion, all of which led to an increse of operative duration and postoperative thoracic catheter indwelling duration. Laparoscopic surgery was successfully performed in one case, however, it was difficult to found the mass during the operation, and resulted in the prolongation of the operative duation and increase of intraoperative blood loss volume.

To our knowledqe, this is the first report of DVRATS for the treatment of an IDEPS. Compared with VATS, the robotic-assisted systems could allow a three-dimensional (3D) magnified view of the surgical field (10–15 times magnification power), eliminate hand tremors and provide a wider range of motion to the surgical performance, all of which make it easier to complete ligation of the feeding vessel, resection of the mass and subtle suture. What's more, we found that the length of stay after operation and postoperative thoracic catheter indwelling duration in Group 2 were significantly shorter than that of Group 1. However, due to the limited experience and long preparation time, the first case managed with DVRATS cost more time than other cases. The limitations of expensive equipment, high cost and complete absence of touch sensation restrict its wide application.

Conclusions

In summary, IDEPS is a rare congenital anomaly, which most commonly located at the left diaphragm. Misdiagnosis should be avoided before surgery as much as possible. Although it's challenging to diagnose an IDEPS through prenatal ultrasound or CT, enhanced CT is essential technique for identify an IDEPS. This study applied a relatively new technique of 3D reconstruction software to appropriately assist on the assessment of an IDEPS. Early surgery to resect the IDEPS was recommended, and the best path was accessing the mass from chest. The treatment of an IDEPS with DVRATS is as safe, feasible and effective as VATS. Furthermore, compared with VATS, DVRATS is 3D magnified view, more flexible and precise. Small sample size may affect results, however, further clinical studies with more cases are needed.

Abbreviations

VATS: video-assisted thoracic surgery; DVRATS: Da Vinci robot-assisted thoracoscopic surgery; LS: laparoscopic surgery; IOBLV: intraoperative blood loss volume; TCI: thoracic catheter indwelling; EPS: extralobar pulmonary sequestration; CT: computed tomography; MRI: magnetic resonance imaging; 3D:three-dimensional; TAE: transcatheter arterial embolization; CCAM: congenital cystic adenomatoid malformation.

Declarations

Acknowledgements

Not applicable.

Authors' contributions

Xu Han, Yue Gao and Zheng Tan designed the study and submitted the manuscript. Xu Han and Yue Gao collected and analyzed data together. Xu Han drafted the article. Yue Gao and Zheng Tan supervised this study. All authors read the final version of this article and approved for publication.

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Availability of data and materials

All data generated or analyzed are included in this published article.

Ethics approval and consent to participate

Not applicable. Institutional review board was not required was waived for the purpose of this study.

Consent for publication

Not applicable.

Competing interests

All authors declare that they have no competing interests.

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Figures

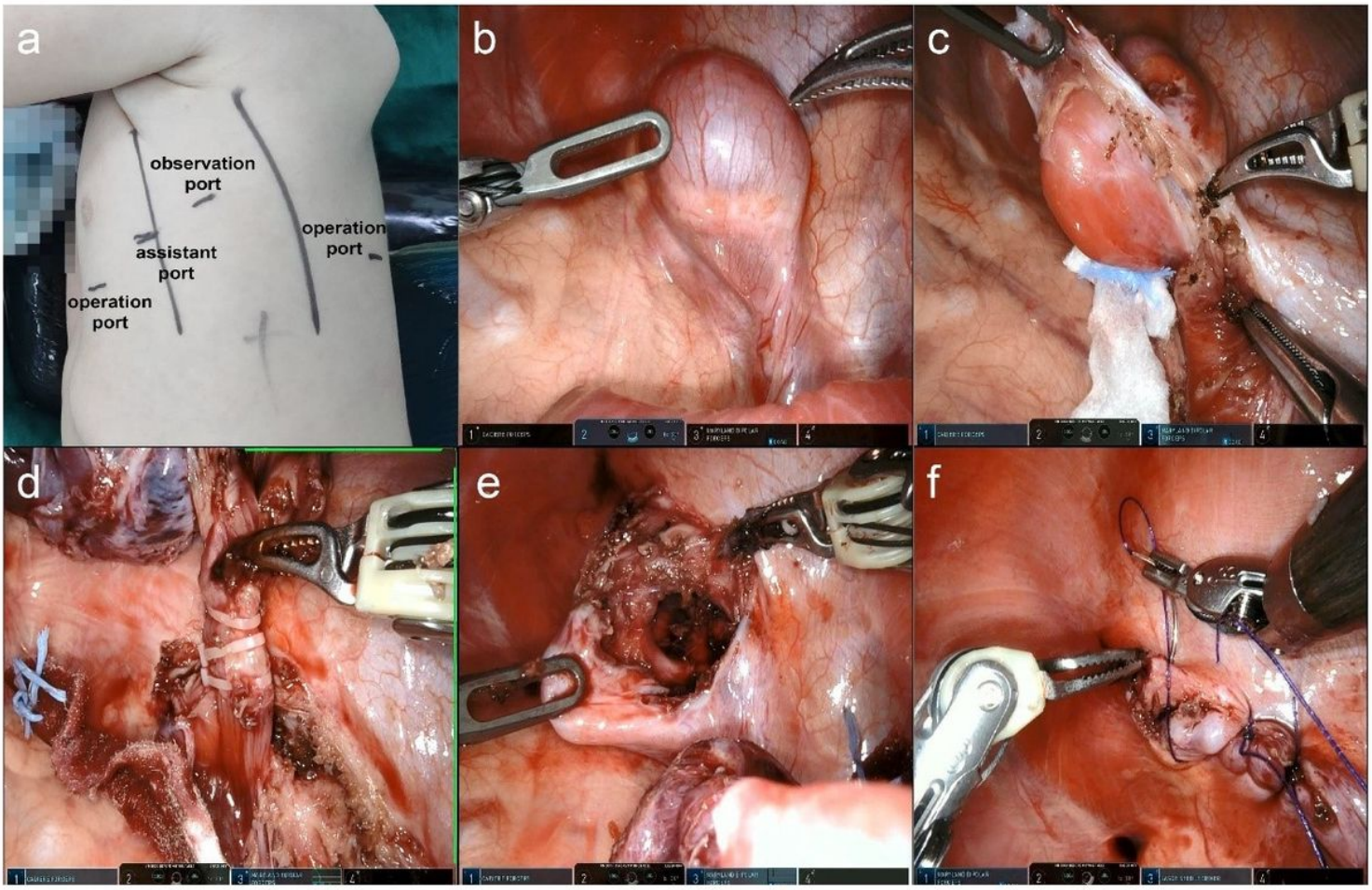

Figure 1

IDEPS treated with Vinci robot-assisted thoracoscopic surgery. Fig.1a, locations of the incisions. Fig.1b, a mass protruding from the diaphragm to the chest cavity. Fig.1c, the diaphragm was opened along the edge of the mass; Fig.1d, the nutrient arter were ligated with vascular clamps. Fig.1e, mass was completely resected; Fig.1f, diaphragm was repaired with interrupted 3-0 barbed suture.

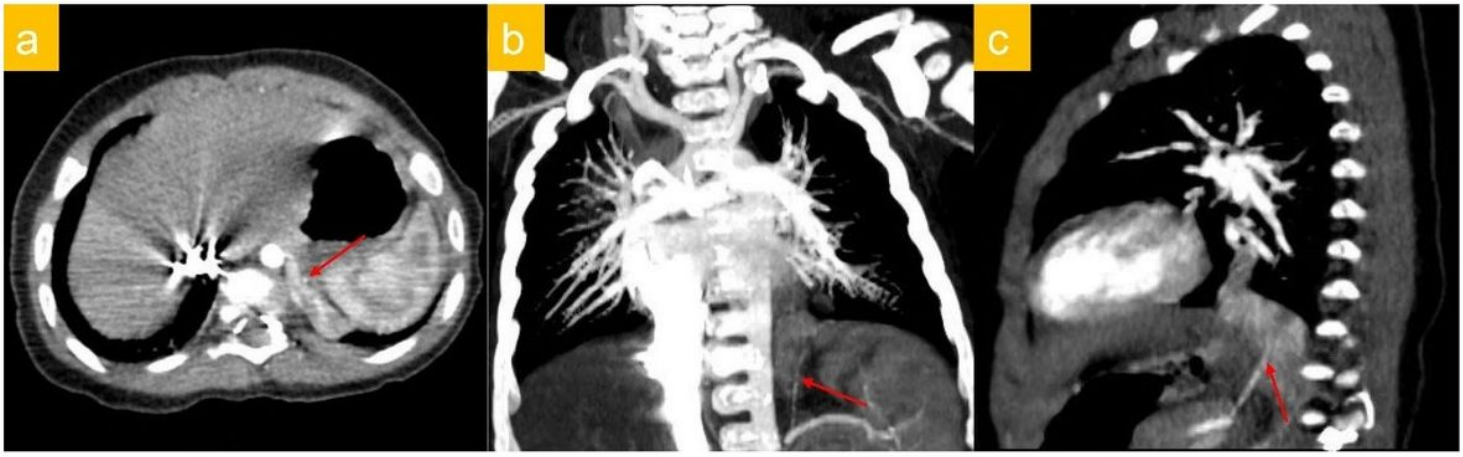

Figure 2

Enhanced CT demonstrated the presence of a solid mass at the left diaphragm area. A single systemic artery (red arrows mark) arising from the abdominal aorta was seen to supply the mass. Fig.2a, horizontal of CT image; Fig.2b, coronal of CT image; Fig.2c, sagittal of CT image.

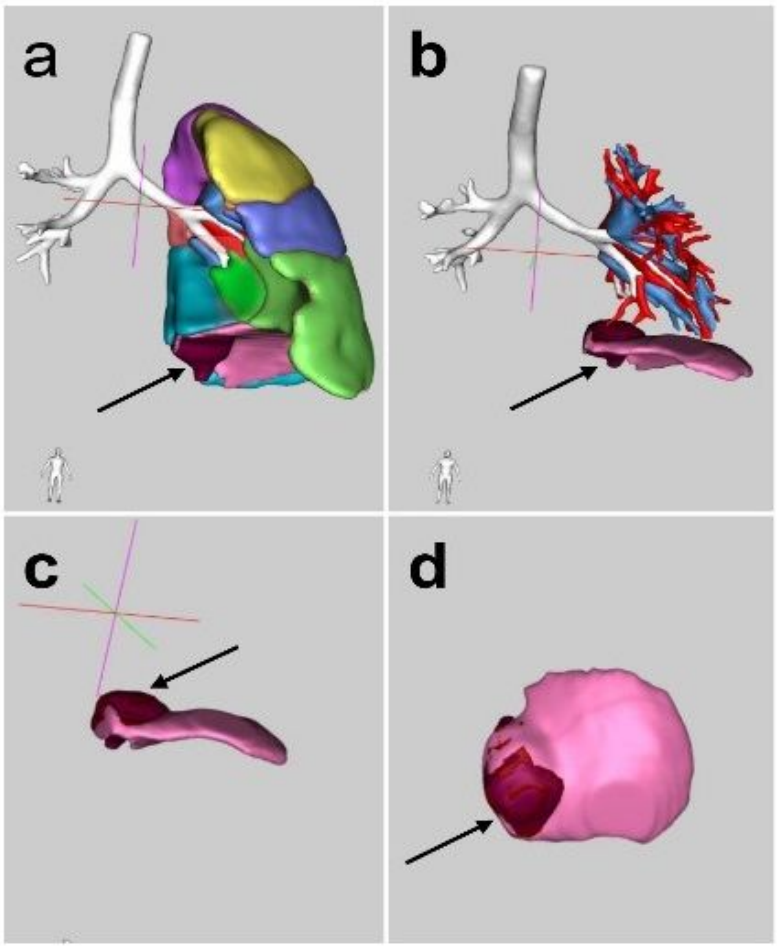

Figure 3

Three-dimensional(3D) reconstruction imaging of an IDEPS patient. The black arrows marked a mass (dark red area) was located in the diaphragm.

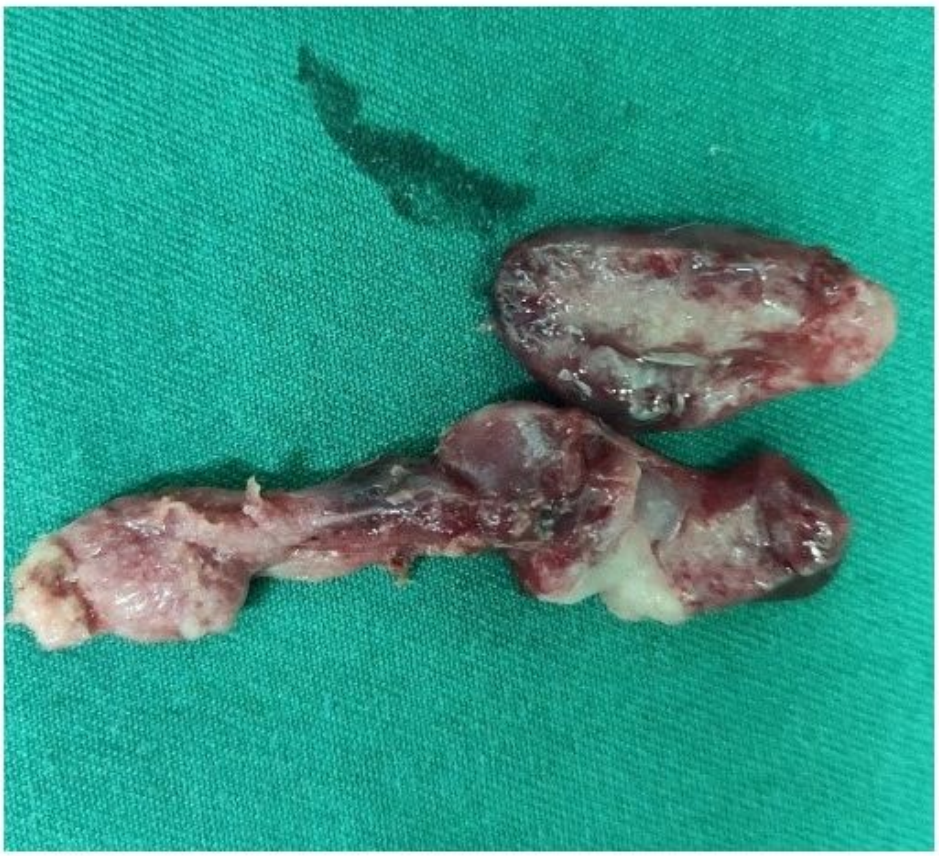

Figure 4

Pathological specimen of IDEPS patient. The section of lesion exhibited a honevcomb-like appearance.