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Case report

Long-term results of sclerotherapy for cervical-thoracic-mediastinal lymphatic and venous malformations: A case report

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ABSTRACT

Cervical-thoracic-mediastinal LVMs in infants is rare, and very difficult to be cured because of life-threatening risk and recurrence. An infant with cervical-thoracic-mediastinal LVMs was treated in our department. Left neck was thick at birth, usually depressed, lazy and inactive. He showed dyspnea after about 3 min of automatic activity, with increased wheezing and open-mouth breathing, squatting after walking no more than 6 steps. There were masses and bulges in the left submandibular area and the left neck. The left cervical root and left clavicle were full. LVMs was diagnosed, and treated by sclerotherapy with bleomycin and triamcinolone acetonidein in a phased and step-by-step manner. The thoracic LVMs and mediastinal LVMs were cannulated under general anesthesia B-ultrasound guidance with an indwelling catheter; drugs were administered via the catheter. After several sessions, the submandibular LVMs and cervical LVMs were completely regressed, the thoracic LVMs and mediastinal LVMs were approximately 95 % regressed. Follow-up of 11 years, there was no recurrence; the lungs, thoraxes, and spines were well developed and free to move. Surgical resection of thoracic-mediastinal LVMs has high life-threatening risk, especially in children, and can't remove all of the LVMs. Comparing with surgical resection, sclerotherapy has the advantages of minimally invasive, low risk, conveniently repeated treatment, obvious curative effect. Therefore, cervical-thoracic-mediastinal LVM in infants can be effectively cured by sclerotherapy; the sclerotherapy has no obvious side effect on the development.

1. Introduction

The neck-chest-mediastinal lymphatic and venous malformations (LVMs) are rare and can bring fatal danger to infants and young children. The most common serious complications are respiratory failure caused by respiratory obstruction, followed by internal bleeding, thrombosis, etc. Even adults are seriously threatened in this situation [1–3].

Many cases of thoracic-mediastinal LVMs have been reported. According to the reports, some literatures only introduced the diagnosing process, but patients weren't treated [4–7]; some patients were treated, recent outcomes and complications were described, but long-term results were missing.

11 years ago, we treated an 18-month-old child with cervical-thoracic-mediastinal LVMs. Based on our many years of experience with bleomycin sclerotherapy in thousands of LVM patients, we adopted a careful phased, step-by-step, low-dose, and combined medication to treat the child through sclerotherapy, and the tumor steadily shrank and regressed smoothly. Subsequently, we conducted a long-term follow-up to observe whether there was any relapse and the impact of the treatment on lung, thorax and spine development.

2. Case

At the time of consultation, the patient was 18 months old, and male. His parents reported that they found that the left neck was thick at birth, and he was usually depressed, lazy and inactive. He showed dyspnea after about 3 min of automatic activity, with increased wheezing and open-mouth breathing. He squatted after walking no more than 6 steps. Recently, the above symptoms have worsened. He was referred to several hospitals and was diagnosed with lymphatic and venous malformation. In view of the seriousness of the disease and the risk of treatment, no treatment was given in these hospitals. Physical examination: There were masses and bulges with soft texture, no tenderness and blurred boundaries in the left submandibular area and the left neck. The left cervical root and left clavicle were full, and the mass was soft

Abbreviations: LVMs, lymphatic and venous malformations.

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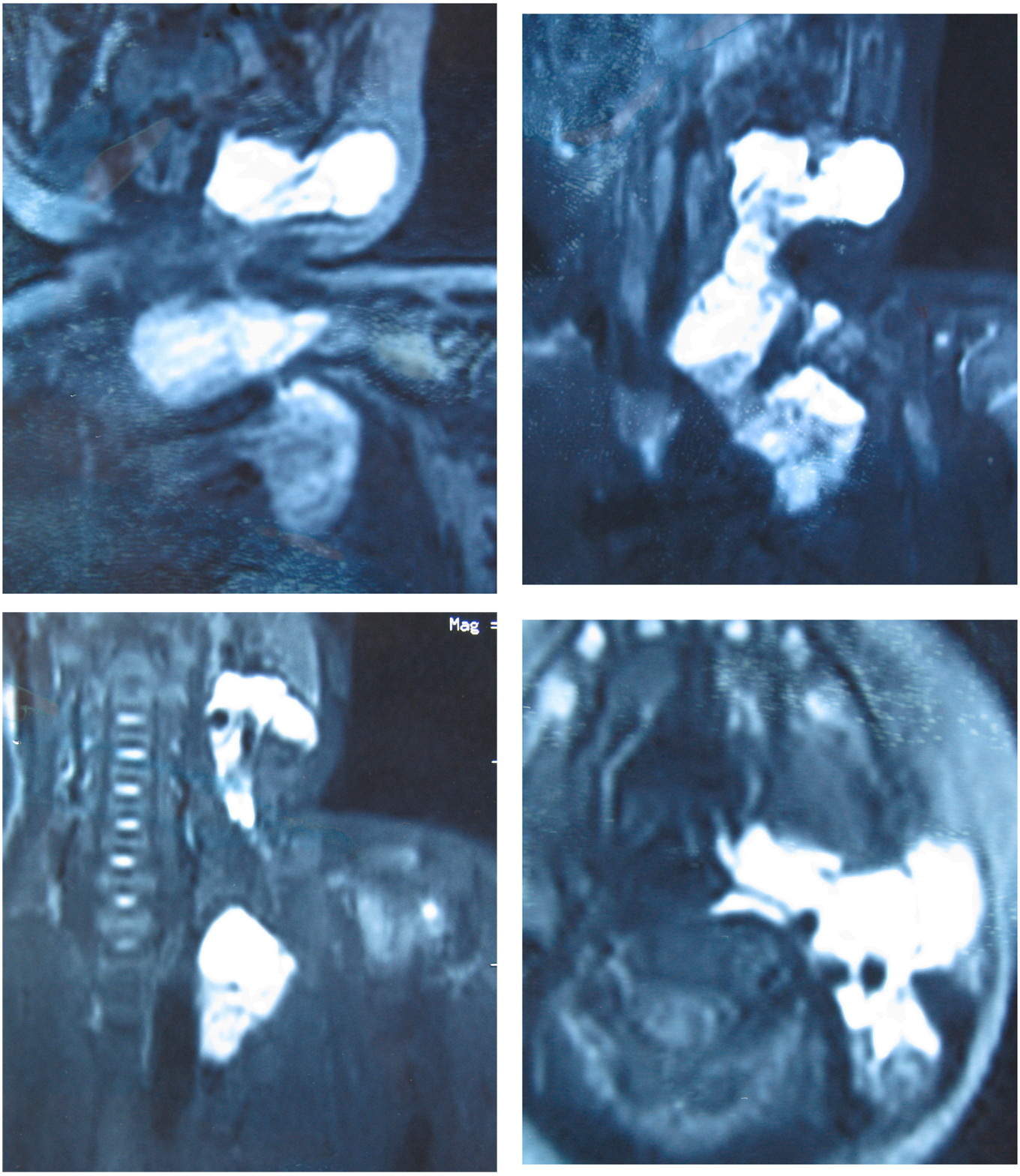
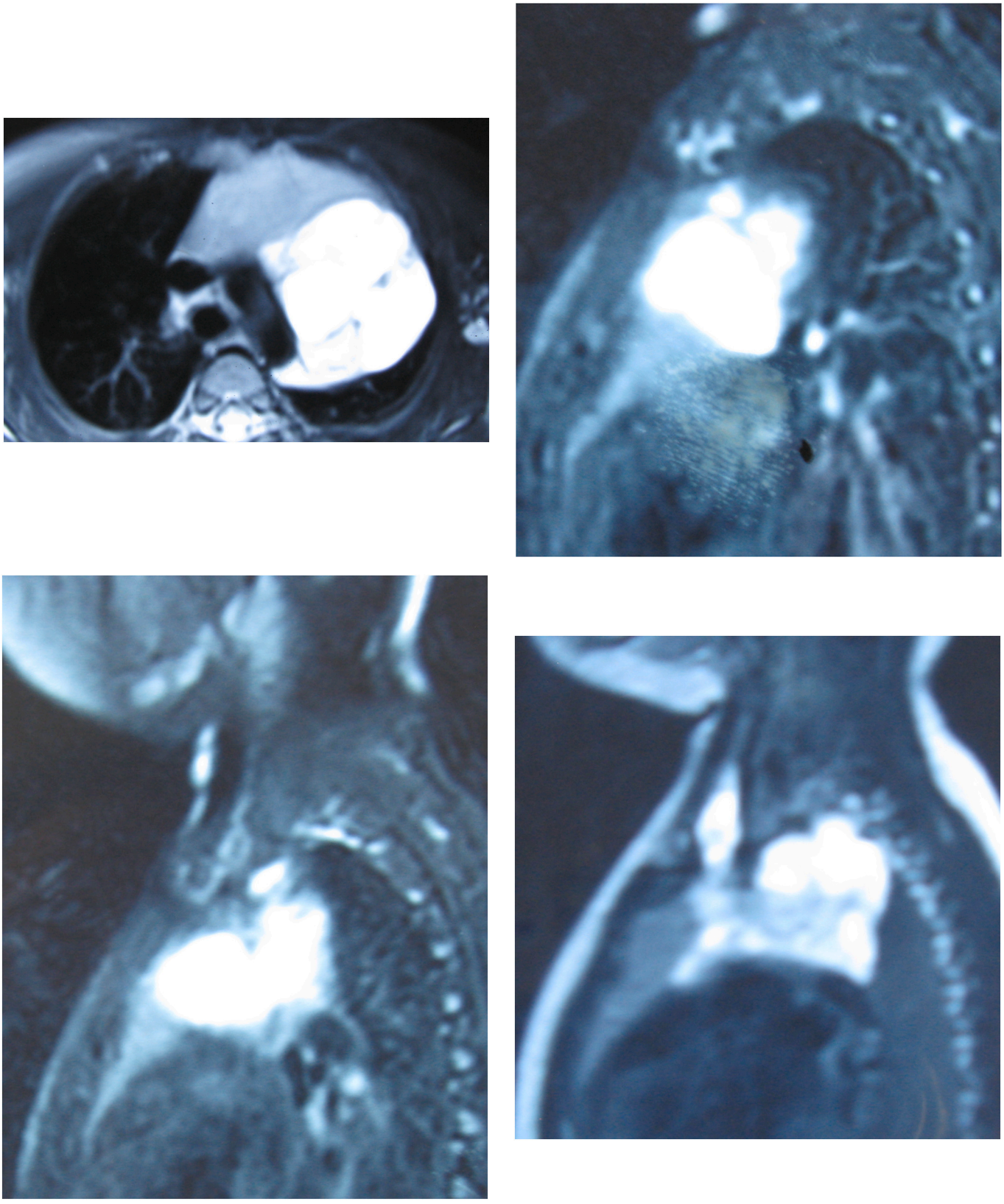


Figs. 1–8. Before treatment, 18 months old, enhanced magnetic resonance imaging. Low density and polycystic masses in the left submandibular area, left cervical area, upper left thoracic cavity and upper mediastinum. The mass wound wrapped around the aortic arch and subclavian artery and vein.

without tenderness. The breath sounds on the upper side of the left chest were disappeared, and the breath sounds on the lower side were weakened. Percussion on the upper side of the left side of the chest showed solid and turbid sounds.

Pale yellow liquid was aspirated from the left submandibular area and dark red liquid was aspirated from the left supraclavicular area. The contrast-enhanced MRI showed low density and polycystic masses in the left submandibular area, left cervical area, upper left thoracic cavity and upper mediastinum. The mass wound wrapped around the aortic arch and subclavian artery and vein (Figs. 1–8).



Figs. 1–8. (*continued*).

According to the symptoms, physical examination and enhanced magnetic resonance examination, the diagnosis of lymphatic and venous malformations were confirmed.

Treatment regimen: The partitioned and phased treatment was adopted.

First, the sclerotherapy of left submandibular area and left cervical vascular lymphatic malformation was performed first; after the massed in the submandibular area and neck regressed, the sclerotherapy of the

Table 1
Drug use in different parts.

	Bleomycin	Triamcinolone acetonide	Number of treatments	Effect
Submandibular and cervical LVM	1000–1500 units/time	6–8 mg/time	9	100 % regression
Thoracic and mediastinal LVM	1000–2000 units/time	8–10 mg/time	15	95 % regression

left side of the chest was performed; when the cysts in the above areas regressed and alleviated, the sclerotherapy for the mediastinal angiolymphatic malformation was performed.

Mode of administration: The LVMs in the submandibular area and neck were treated with sclerotherapy in outpatient department. Under the guidance of B-ultrasound, they were punctured into the capsule cavity, and the liquid was aspirated. The liquid was aspirated as clean as possible, and then the drug was injected into the cysts.

The patient was hospitalized when the treatment of the thoracic and mediastinal LVMs would be performed. General anesthesia was given, and catheters were placed in the cysts under the guidance of B ultrasound, and the sclerotherapy was adopted for 3 consecutive days. Only moderate amount of fluid was aspirated from the cystic cavity in each treatment to avoid large location moving of the mediastinum and lung which could cause accidents.

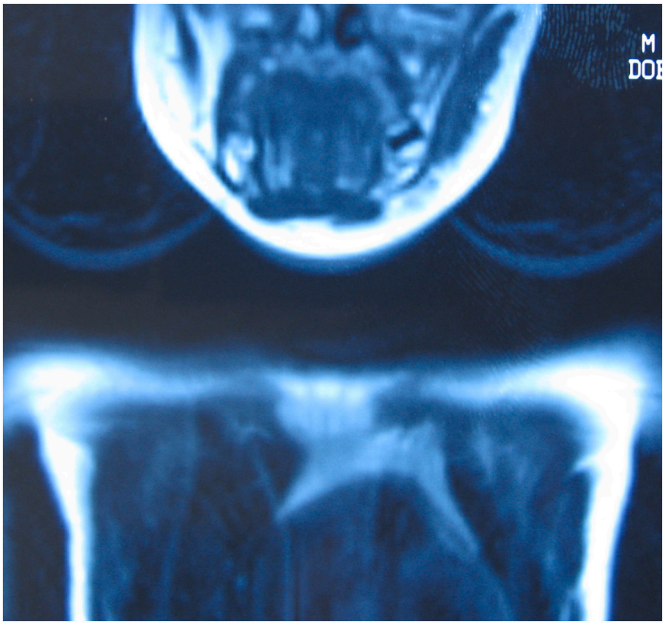
Sclerotherapy drug: Bleomycin combined with triamcinolone acetonide. Drug use in different parts was listed in Table 1.

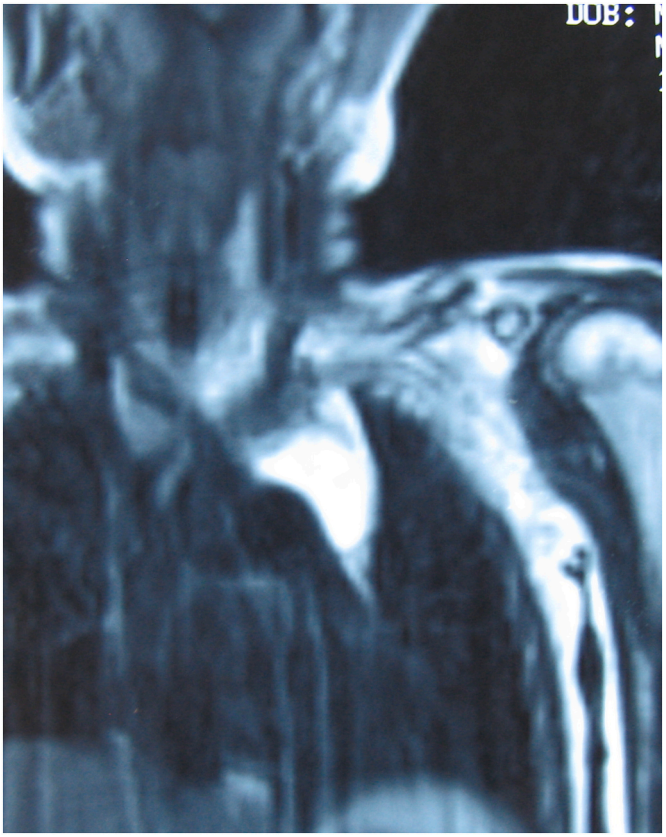
Reexamination: After nearly 2 years of treatment, the cervical LVM and mediastinal LVM were almost regressed, with a small amount of residual LVM in the chest, mainly located at the aortic arch. The further sclerotherapy was waived in view of the high risk of puncture. There was no abnormality in the thorax of the patient. The respiratory sound appeared with auscultation on the left upper chest, but it was weaker than that of the contralateral side (Figs. 9–14). There were no abnormalities in the spine. The moving ability was enhanced, dyspnea after movements was relieved.

Recent examination: After treatment 11 years, the patient was 14 years old, 178 cm height (his father 169 cm height, his mother 162 cm height), 70 kg weight, grew up without obvious scoliosis, the spine activity is normal. Breathing smoothly, the respiratory sound clear, bilateral chest movements were symmetric, bilateral thoraxes were symmetrical. Computed tomography showed that there were no abnormalities in the neck, both of the lungs were clear, original focal site of left superior lung was almost normal. There was a small amount of cysts around aortic arch (Figs. 15–22).

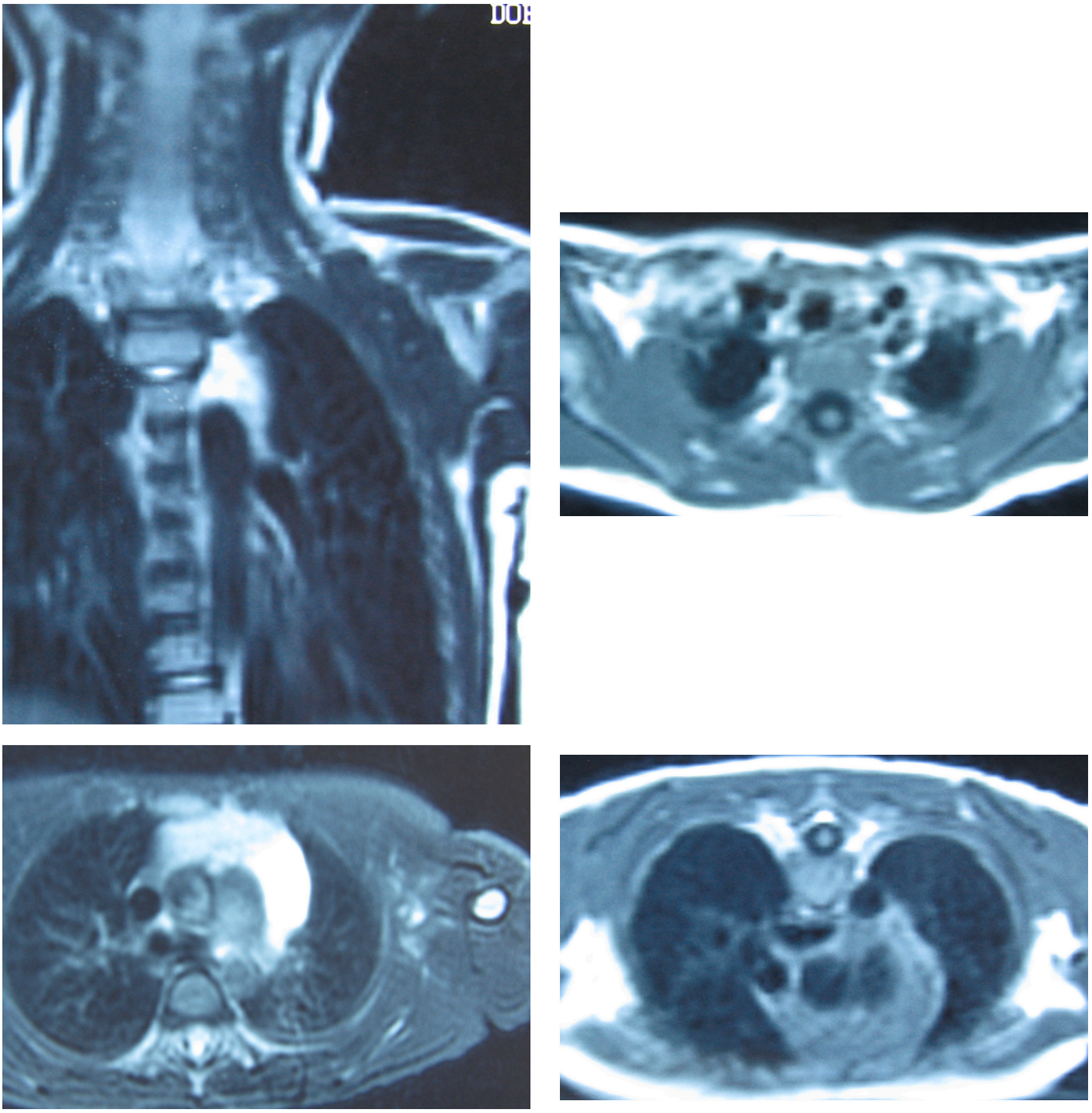
3. Discussion

The treatment of vascular malformations of the thoracic mediastinum includes surgical resection and sclerotherapy. Marc Riquet et al. [8] reported the treatment of 3 cases of thoracic mediastinal lymphatic malformation, including 2 adults and 1 juvenile case. The injury caused during thoracotomy was relatively large, the intraoperative bleeding was relatively heavy, and there were complications such as nerve damage. So-Hyun Nam et al. [9] performed surgical resection of LM in 3 infants and young children, but it was difficult to completely remove the lesions, and one of the infants died of postoperative infection. Amy W. Cheng et al. [10] performed surgical resection of LM in 3 children with thoracoscopy, which is less invasive, but the intraoperative risks are still relatively high, such as bleeding. Shigeru Ueno et al. [11] analyzed previous literatures in mediastinal LMs in Japan. Direct surgical resection of the mediastinal LMs accounted for a relatively low proportion, the potential risks associated with surgery were higher and may be





Figs. 9–14. After treatment 2 years later, 3 years old, enhanced magnetic resonance imaging. Only a small amount of residual LVM in the chest, mainly located at the aortic arch.



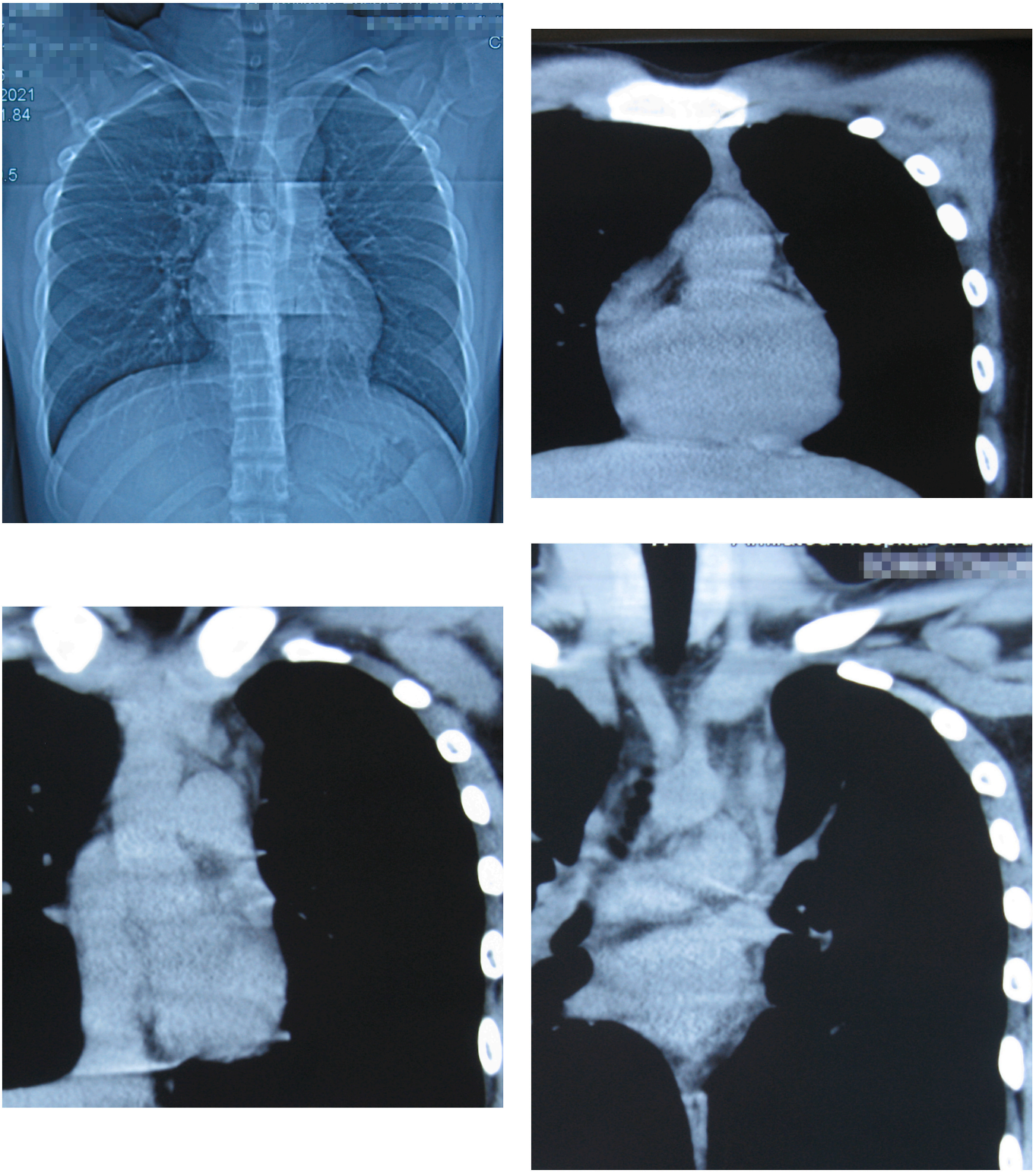
Figs. 9–14. (*continued*).

serious. Therefore, direct surgical resection of LM is generally not recommended unless it is an emergency.

Sclerotherapy is a common approach to treat hemangiomas and vascular malformations. Shou-jiang Tang et al. [12] performed sclerotherapy for thoracic mediastinal LVMs by endoscopy, Omer Karakas et al. [13] treated cervical-mediastinal LVMs with bleomycin and the lesion almost regressed. Compared with thoracotomy, sclerotherapy has many advantages, such as being minimally invasive, less operation impact on the patient, and low risk. But Nader Ghaffarpour et al. [14] have different opinion. They summarized and analyzed the complications of their previous sclerotherapy for mediastinal LMs. They found that sclerotherapy could cause fever, scar contracture, necrosis, possible blockage of the trachea, and swelling. Given the high incidence of complications associated with sclerotherapy, they concluded that surgery could be an option for the treatment of mediastinal LMs.

As a matter of fact, the complications of sclerotherapy are associated with many factors, for example, sclerosing agents, methods and mode of administration etc. Some of the sclerosing agents have strong collapsing force, such as absolute ethanol, complications occur in all probability; others destroy cyst gently, such as bleomycin, complications occur rarely. If the agents were administrated in large dosage and given repeatedly in short period, complications occur easily.

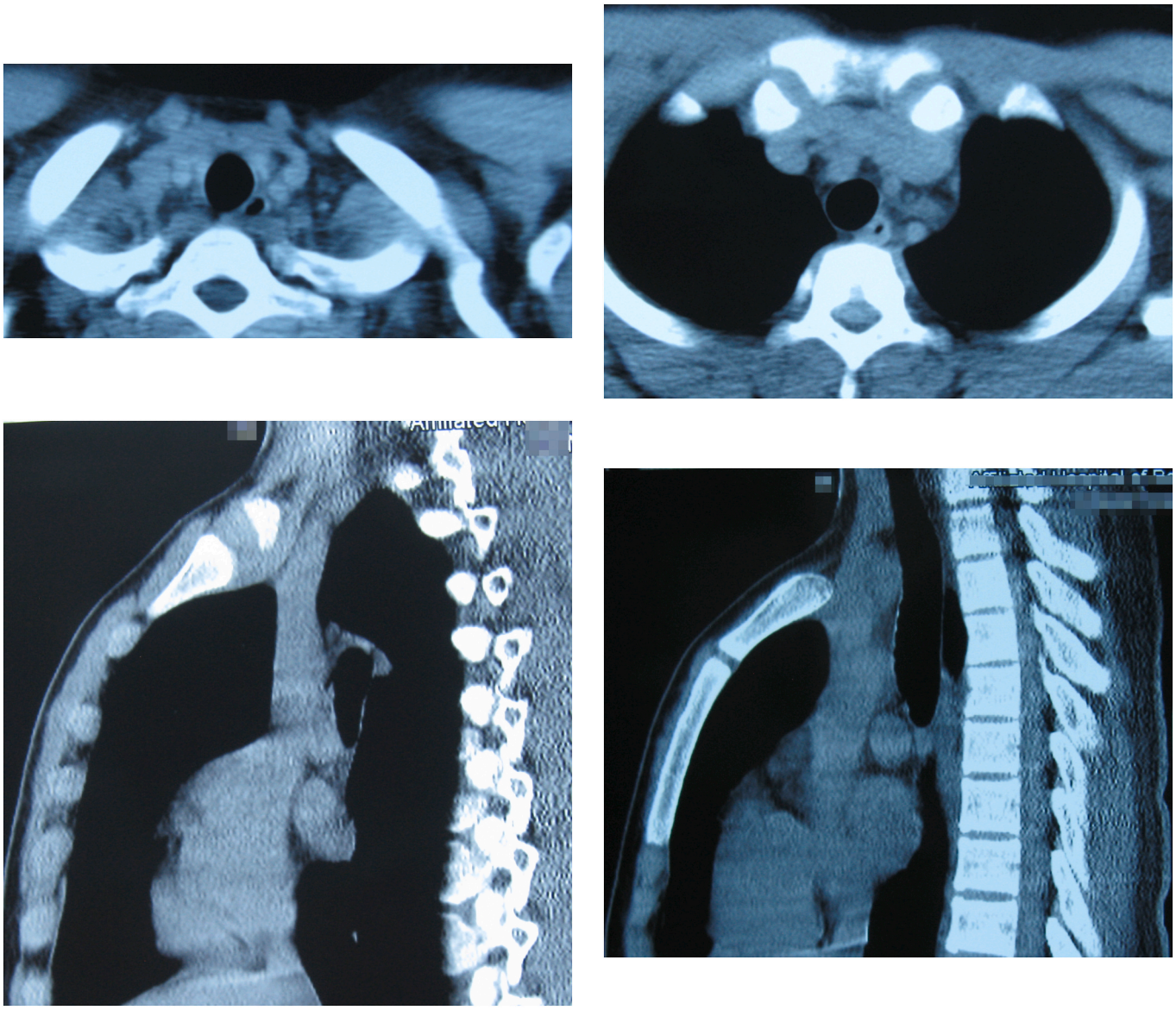
We paid full attention to the dose and concentration of the drug when using bleomycin, thus avoiding complications. One thing worth noting with bleomycin is that the tissue became stiff after multiple intralesion applications. In order to avoid the adverse effect, we used a small



Figs. 15–22. After treatment 11 years later, 14 years old, computerized tomography, grew up without scoliosis, bilateral thoraxes were symmetrical. There were no abnormalities in the neck, both of the lungs were clear, original focal site of left superior lung was normal. There was a small amount of cysts around aortic arch.

amount of triamcinolone acetonide in combination, which not only avoided the sclerosis of the tissue, but also reduced the swelling and fever that could be caused by bleomycin; in addition, triamcinolone acetonide itself also has a therapeutic effect on LMs. The combination of bleomycin and triamcinolone acetonide provided therapeutic synergy while reducing the side effects of bleomycin.

During the treatment, indwelling catheter in the cystic cavity can not only avoid the risk of repeated puncture, but also enable the drug to repeatedly act on the cystic wall and destroy it more thoroughly. The treatment of this patient indicated that continuous multiple



Figs. 15–22. (*continued*).

administrations of small doses by an indwelling catheter effectively avoided the complications of the sclerosis process.

We were also very concerned about the long-term effects of sclerotherapy of cervical-thoracic-mediastinal LVMs on the development of children's lungs, thorax and spine. We followed up the developmental process and the recurrence of the lesions in the child. After 11 years of follow-up observation, no recurrence was found of the patient's cervical-thoracic-mediastinal LVMs, indicating that the treatment was relatively thorough, and the treatment method was safe and feasible. Further observation showed that the patient's overall development was good with normal development of the lungs, thorax, and spine, which indicated that triamcinolone acetonide effectively prevented the fibrotic adhesion and adverse impact on the surrounding tissues and organs during sclerotherapy.

4. Conclusion

Sclerotherapy should be the preferred treatment for cervical-thoracic-mediastinal LVMs. Appropriate procedures and methods with reasonable combined medications guaranteed a good therapeutic response.

This case reports were in line with the SCARE 2020 criteria [15].

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Ethical approval

N/A.

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Guarantor

Quan-Feng Luo.

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CRediT authorship contribution statement

Quan-Feng Luo performed the initial surgery operation and all sessions of treatment, reviewed the literatures, and wrote the manuscript. Ye-Hua Gan reviewed the case report and manuscript.

Declaration of competing interest

There are no conflicts of interests.

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