Lymphangiome cervico-médiastinal géant chez un enfant avec une détresse respiratoire
Giant cervico-mediastinal lymphangioma in a child with acute respiratory distress

Borgi. A [1], Kebaier. S [1], Hajji. A [1], Ayari. A [1], Louati. A [1], Menif. Kh [1], Bouziri. A [1], Ben jaballah. N [1]

[1] Service de réanimation pédiatrique polyvalente de l'hôpital d'enfants Béchir Hamza de Tunis

RÉSUMÉ

Nous rapportons l'observation d'une fille âgée de 12 mois admise pour une insuffisance respiratoire aigue secondaire à un lymphangiome cervico-médiastinal géant découvert à l'âge de 1 mois. Les difficultés de l'intubation ainsi que de la prise en charge immédiate ont été décrites. La sclérothérapie a échoué à réduire la masse. La chirurgie a été réalisée avec succès. La taille géante ainsi que la double localisation cervico-médiastinale du lymphangiome font l'originalité de notre cas. Les difficultés thérapeutiques sont discutées.

ABSTRACT

We report a 12-months-old girl presented with acute respiratory distress due to a giant cervico-mediastinal cystic lymphangioma diagnosed at the age of one month. We stressed on the difficulties of management and intubation. Sclerotherapy failed to reduce the mass. A successful surgery was performed. This case is atypical because of the size and the location of the mass. Management strategy and the difficulties of conventional methods in such patients are discussed.

Mots clés : Lymphangiome kystique, insuffisance respiratoire aiguë, sclérothérapie, chirurgie.

Keywords : cystic lymphangioma, respiratory distress, sclerotherapy; surgery.

INTRODUCTION

Cystic lymphangioma (CL), also known as cystic hygroma, is a congenital malformation of the lymphatics resulting in a lymphatic flow interruption. It occurs commonly in the neck and axillae. However, 10 % of lymphangiomas in the neck extend into the mediastinum. Treatment consists of surgery and many intralesional injections of sclerosing agents. We reported a failure of sclerotherapy followed by a successful surgery in a case of giant cervico- mediastinal CL in a young child with acute respiratory distress.

CASE REPORT

A 12-months-old girl was referred to our department for acute asphyxia with upper airway obstruction. She had a giant cervico-mediastinal CL diagnosed since the age of 1month and was managed by sclerotherapy. In admission, Prompt resuscitation was started, however multiple attempts at intubation with endotracheal tubes size 4 failed. Finally, she was intubated successfully with a 3,5 sized tube and ventilated in intensive care unit. Pulmonary examination revealed sibilants and a diminished breath sounds at auscultation. A chest radiograph showed a large homogeneous opacity in the left lung field. Chest Computed Tomography (CT) showed a well-defined voluminous 9 x 6.5 cm multilocular cervical cystic lesion and 8 x 6 x9 cm extent cystic lesion in the anterior mediastinum (fig.1).

Auteur correspondant :
Dr Borgi Aida
Address : Pediatric Intensive Care Unit - Children's Hospital of Tunis Bab Saadoun-Jabbary PC: 1007 –Tunis-Tunisia.
Faculty of Medecine University of Tunis El Manar.
Fax : 00 216 71 572 572
Phone : 00 216 20 975 689
Mail : aidabdoc@yahoo.fr

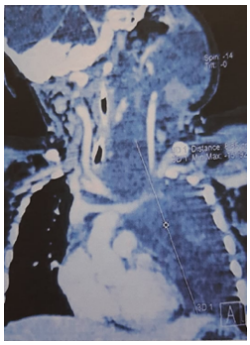

Figure 1 : Computed tomography shows well-defined voluminous cervico-mediastinal cystic lesion.

This lesion extended to the mediastinal vessels, but no invasion was observed. The left main bronchus was compressed by this lesion. There was intracystic hemorrhage. The child has a severe airway distress due to double compression on the trachea and the left bronchus and lung by the giant mass. The trachea was deviated. Tracheostomy could not be performed because of mass localization. Corticotherapy and sclerotherapy were performed for 4 times with ethybloc. After 15 days of conservative approach, the cervical mass enlarged in size and airways were still compressed. Then, surgery was performed via median sternotomy (Fig 2) and left cervicotomy.

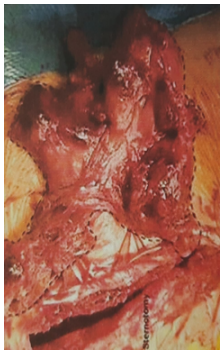

Figure 2 : Intraoperative view shows a large cystic lymphangioma.

For the thoracic part of the tumour, complete excision leading to a best expansion of the left lung was performed. In the cervical region, there was sclerotic tissue around the tumour including vagus and phrenic nerves. Laborious macroscopic complete excision was achieved and complicated by a left recurrent nerve injury. The diagnosis of lymphangioma was confirmed by histopathological study. The post-operative course was complicated by a left phrenic nerve stretch with diaphragmatic paresis and pulmonary infections. Tracheostomy was performed after endotracheal intubation. Diaphragmatic paresis was improved 8 weeks after surgery and the child was discharged with cannula tracheostomy on the 80th post-operative day. She was decannulated 4 years later.

DISCUSSION

Lymphangiomas are rare, benign congenital malformations consisting of focal proliferations of well-differentiated lymphatic tissue comprising complex lymph channels or cystic spaces containing clear or straw-coloured fluid. Most lymphangiomas are present at birth and are detected in the first 2 years of life. Lymphangiomas are most common in the neck and axilla. Ten per cent of lymphangiomas in the neck extend into the mediastinum [1]. Our patient had a CL located in the neck and the mediastinum diagnosed at the age of 1 month. Only a few cases of Cervico-mediastinal CL were reported in child. Karakas et al [2] reported a giant cervicomediastinal cystic hygroma in a 2-year-old child without significant airways compression. Lymphangiomas rarely produce symptoms caused by their soft consistency. However, compression of mediastinal structures can result in chest pain, cough, and dyspnoea. Our patient had a double localization causing severe airway obstruction. The diagnosis is usually easy; however, the management remains controversial. The treatment of choice is surgical excision to avoid superinfection, rapid growth, risk of rupture or emergency laparotomy. Although surgery is the treatment of choice, in many cases complete removal of the CL is not achieved, especially when large vessels and/or nerves are surrounded, due to the high risk for injuring these structures. Postoperative complication rates range from 12 to 33% [3]. One therapeutic option is sclerosis with an injection of Picibanil ® (OK-432), after which several studies have reported total tumor remission, both cervical as well as retroperitoneal [4]. Ogita et al. [5] published the first study of lymphangiomas treated with Picibanil ®, demonstrating the sclerosing effect and reduction in lesion size. In our case, a progressive increase in size due to intra cystic hemorrhage occurred with an acute respiratory distress. Upper airway was obstructed by extrinsic compression. Sclerotherapy was performed again, in intensive care unit, without success. Sclerotherapy is a safer and cosmetically acceptable option of management for such lesions. It's recommended as the first-line treatment modality as an alternative to surgery with the good response [6]. One prospective study was performed to evaluate the role of bleomycin sclerotherapy in the management of different radiological variants of LM. A total of 142 patients were included in this study. The lesions were classified as macro cystic, microcystic, or mixed lymphangiomas based on ultrasonography. All patients were managed by intralesional injection of bleomycin and were recalled after 4 weeks for evaluation. Colour photographs of the patients were taken before the onset of treatment and at each monthly visit and were utilized to assess the response. Following the second, third, and fourth doses, the response was better in patients with the macro cystic variant than in those with the other two variants [7]. Oedema, erythema, and local induration with fever were the most common adverse effects and were more common in younger children. Surgery excision can be technically difficult because of the mass nature. Cases of local recurrence have been reported if the resection is incomplete. In our case, sclerotherapy failed. A successful surgery was performed. Surgery excision should be reserved for those lesions where sclerotherapy failed or at first line when

the mass compromises vital functions. Guidelines for reporting lymphatic malformation case series do not currently exist, making difficult to choose the appropriate treatment [8]. The post operative course in our patient was complicated by a phrenic nerve lesion. The complications after surgical treatment reported in the literature were chylothorax, infection, facial or phrenic nerve palsy, airway obstruction and hoarseness [9].

CONCLUSION

The involvement of the upper airway seems to be the determining prognostic factor in extensive lymphangioma. Surgical excision seems to be the treatment of choice in a severe extrinsic compression giant lymphangioma.

Conflits d'intérêts : Pas de conflits d'intérêt.

REFERENCES

[1] Parkar, Nadeem; Javidan-Nejad, Cylen; Bhalla, Sanjeev. Lymphangiomas (cystic hygromas). Grainger & Allison's Diagnostic Radiology. January 2021.

[2] Karakas O, Krakas E, Boyaci FN et al. Cervicomediastinal cystic hygroma: a case report. J Clin Med Res.2013;5(1):61-3.

[3] Devki C. Shukla and al. Airway management in a neonatal cystic hygroma. Otolaryngology Case Reports 15 (2020) 100157.

[4] Delia María Luján Martínez and al. Utility of conservative treatment in cystic lymphangioma. Spanish Surgery (Cirugía Española, English Edition) 2016; volume 94, Numéro 8, Pages 485-487.

[5] Ogita S., Tsuto T., Tokiwa K., Takahashi T.: Intracystic injection of OK-432: a new sclerosing therapy for cystic hygroma in children. Br J Surg 1987; 74: pp. 690-691.

[6] Bossert T, Gummert JF, Mohr FW. Giant cystic lymphangioma of the mediastinum. Eur J Cardiothorac Surg.2002; 21: 340.

[7] P. Tiwari, V. and al. Bleomycin sclerotherapy in lymphangiomas of the head and neck region: a prospective study. Int. J. Oral Maxillofac. Surg. 2019.

[8] Adams MT, Saltzman B, Perkins JA. Head and neck lymphatic malformation treatment: a systematic review. Otolaryngol Head Neck Surg.2012;147(4):627-39.

[9] Okazaki T, Iwatani S, Yanait et al. Treatment of lymphangioma in children: our experience of 128 cases. J Pediatr Surg.2007; 42(2):386-9.