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Multivisceral resection for synchronous inflammatory myofibroblastic tumors of the lung and stomach

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ABSTRACT

Inflammatory myofibroblastic tumor is a mesenchymal tumor which commonly originates from lungs. In this paper we report the successful resection of multiple inflammatory myofibroblastic tumors invading thoracic, mediastinal and abdominal viscera in a 14-year old child. We could achieve an R0 resection with distal esophagectomy, proximal gastrectomy, splenectomy, left hepatectomy, left pneumonectomy, left diaphragm resection and pericardiectomy. To our knowledge, this is the first case reporting synchronous gastric and pulmonary inflammatory myofibroblastic tumors. The patient is alive without disease after 33 months.

1. Introduction

Inflammatory myofibroblastic tumor (IMT) is an uncommon mesenchymal tumor which is comprised of myofibroblastic spindle cells and inflammatory infiltration of plasma cells, lymphocytes and eosinophils. It is more common in childhood and adolescence with a slight predominance in females. Lung, retroperitoneum, abdomen and pelvis are the most frequent locations, besides synchronous IMTs in lung and stomach have not been described before [1]. Surgical resection with clear margins is the main curative treatment. This intermediate-grade, rarely metastasing tumor may act as locally aggressive malignant processes owing to the location and adjacent organ invasion [2]. This paper presents the radical resection of synchronous huge IMTs of lung and stomach which were assumed unresectable previously.

2. Case

Fourteen-year-old female patient had been misdiagnosed with achalasia and underwent multiple endoscopic dilatations in two different institutes. Soon after, surgery was decided for worsening symptoms of the patient. Laparotomy revealed an unresectable thoracoabdominal mass invading gastroesophageal junction and celiac trunk. Incisional biopsy and feeding gastrostomy were performed. Histopathologic examination demonstrated IMT and the patient was considered for palliative chemoradiotheray. After 11 fractions of radiotherapy and 3 cycles of actinomycin-D, vincristine and cyclophosphamide she was referred to our institute with progressive findings. She was cachectic and decayed. Breathing sounds on left hemithorax were decreased and hemoglobin level was 5.9 g/dL. Computed tomography showed total atelectasis on left lung due to a 15-cm hilar mass infiltrating left main bronchus and left pulmonary vein (Fig. 1A). Tumor was invading distal esophagus, stomach, left lobe of the liver and extending along retroperitoneum behind the vena cava (Fig. 1B). Celiac trunk was also invaded (Fig. 1C). Angiographic tomography sections showed replaced right hepatic artery from superior mesenteric artery, thereby potential curative surgery with celiac trunk resection was considered (Fig. 1D). Nutritional support, blood transfusions and respiratory rehabilitation have been provided until the general medical condition of the patient was built up.

Thoracoabdominal incision was performed. The tissues were extremely fibrotic and fragile due to previous radiotherapy and laparotomy, particularly around gastroesophageal junction, that we could not recognize the healthy tissue borders between unattached synchronous tumors. Namely, en-block resection for a single giant tumor was presumed. Left lung, distal esophagus, proximal stomach, left lobe of liver, celiac trunk and spleen were resected (Fig. 2). Partial resection of pericardium and left diaphragm were also performed. Reconstruction of the left diaphragm was carried out by primary sutures. During pericardiectomy, the patient suffered ventricular fibrillation and corrected to normal sinus rhythm after 5 times of defibrillation.

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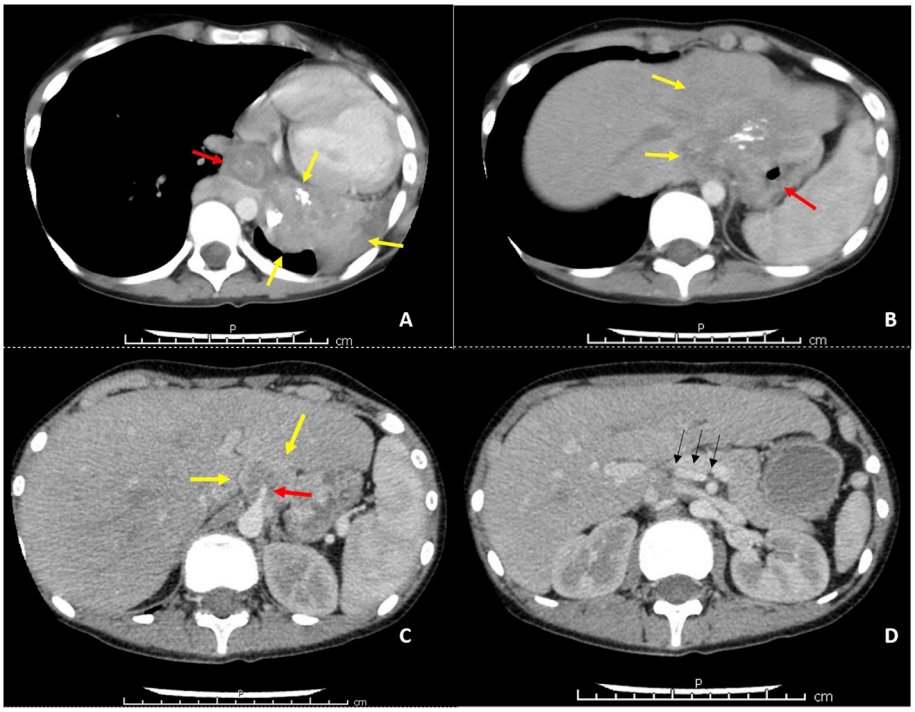

Fig. 1. Preoperative computed tomography sections of the patient (yellow arrows: tumor). A. Tumor originates from lower lobe of the left lung (red arrow: esophagus) B. Gastric tumor invades distal esophagus and left liver lobe (red arrow: stomach) C. Invasion of celiac trunk (red arrow: celiac trunk) D. Replaced hepatic artery from superior mesenteric artery (black arrows: right hepatic artery). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

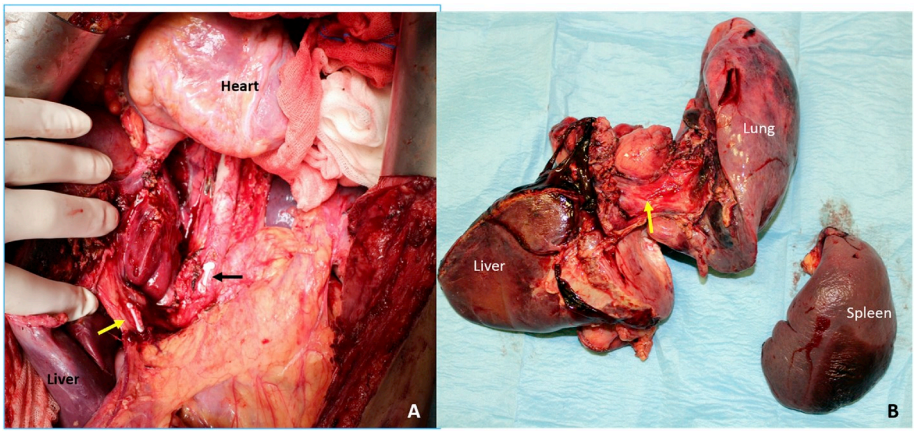

Fig. 2. A. Intraoperative view before esophagogastrostomy (yellow arrow: left portal stump, black arrow: celiac stump) B. Specimen showing en-block resection of lung, pericardium, diaphragm, liver, esophagus, stomach and spleen (yellow arrow: esophagus). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

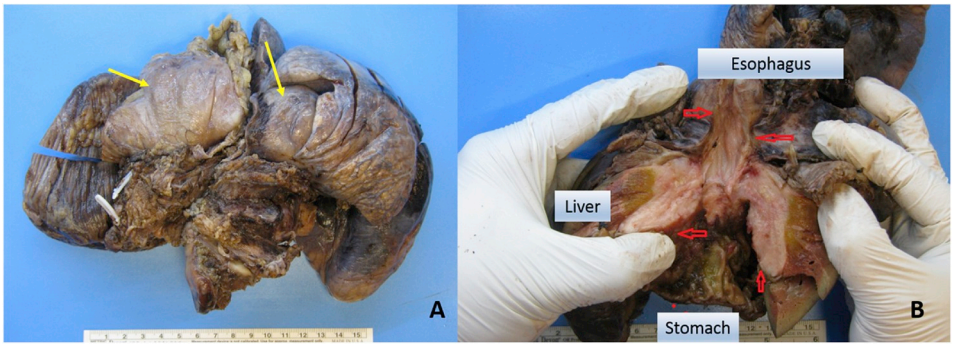

Fig. 3. A. Yellow arrows denote two different tumors originating from lung and stomach B. Destruction of left liver lobe and gastroesophageal junction by the tumor originating from stomach. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

Esophagogastrostomy was performed using 26 mm circular stapler. The operative time was 410 min. Blood loss was 300 ml and 2 packs of erythrocyte suspension, 1 fresh frozen plasma and 200 ml human albumin 20% were administered intraoperatively. The patient was taken to intensive care unit postoperatively. On postoperative day 1 she was extubated, and enteral nutrition was started. She was discharged to clinic on day 6. Four days later, non-invasive mechanic ventilation was needed due to poor saturations and dyspnea. X-ray revealed pneumonia. Wide spectrum antibiotherapy was started and she returned to intensive care unit. Respiratory parameters were still poor and invasive mechanic ventilation was needed, therefore percutaneous tracheostomy was performed. On postoperative day 25, anastomotic leak at esophagogastrostomy was detected. Intraabdominal and thoracic collections were drained by percutaneous pigtail catheters and leak was managed with silicone covered stenting. Mechanic ventilation was stopped on day 45 and the patient was discharged to the clinic. Four weeks later stent and catheters were removed. The patient was discharged from hospital after closure of tracheostomy on postoperative day 69. Histopathological examination revealed three independent IMTs: Two in lung ($7.5 \times 5.7 \times 4.3$ cm and 0.5×2x2 cm) and one in proximal stomach ($6 \times 5.5 \times 4.5$ cm). Surgical margins were clear (Fig. 3). There was no anaplastic lymphoma kinase (ALK) fusion gene rearrangement. No adjuvant treatment including targeted therapy was considered. The patient has been disease-free without any further problem during follow-up of 33 months.

3. Conclusions

Representing less than 1.5% of lung tumors, IMT is a rare and unfamiliar tumor that there are very limited data in the literature to designate the pathogenesis and the optimal treatment [3]. Although it has been considered as a benign or infectious process, current aspect indicates that IMT is a real intermediate-grade neoplasm which carries a non-negligible potential of local recurrence and metastasis [4]. Differential diagnosis is very conflicting due to non-specific symptoms and radiologic findings, besides 30–70% of the patients are asymptomatic. Therefore, a substantial part of the cases is diagnosed postoperatively [1–3]. Given that the most frequent primary lung tumor in pediatric population is IMT, awareness is essential in such cases. Our patient with dysphagia as the major symptom has been treated for achalasia for more than a year. The certain diagnose was obtained after laparotomy and incisional biopsy.

Clinical behavior of IMT varies due to aggressiveness and the location of the tumor. Different approaches in treatment including resection, chemotherapy, radiotherapy, non-steroid anti-inflammatory drugs, and steroids were reported. In a few reports patients had benefit from anti-inflammatory treatment [5,6]. Response to radiotherapy and chemotherapy is anecdotal, even in adjuvant setting [6,7]. In our case, the tumor has progressed in spite of chemoradiotherapy.

Currently, no standard treatment has been admitted. Resection with clear margins, when possible, is the commonly held treatment, however adjacent organ invasions may preclude surgery. Results of radical surgery are satisfying with a 5-year survival rate of 91%. Local recurrence rate has been reported up to 13%, particularly after incomplete resections [8]. We did not observe any sign of recurrence after 33 months follow-up.

Our patient had giant tumors invading essential structures. Thus, surgical risk was extremely high and curative resection possibility was low. Conservative or less invasive treatment options would be cut out for such cases, however other treatment modalities did not provide even palliation and/or regression of IMT in our case. Consequently, we had to push the limits for radical surgery. Nonsurgical treatment modalities may be considered in case of incomplete surgical resection or multifocal/metastatic disease. Until further studies reveal the optimum management to IMT, radical surgery seemed to remain the most effective treatment [8].

Patient consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.epsc.2019.101355.

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