CASE REPORT

Pseudomyxoma pleuri as a Surgical Emergency: a Rare Case Report

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Received: 30 January 2017 / Accepted: 18 October 2017 / Published online: 27 October 2017
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Abstract Pseudomyxoma pleuri is a rare condition and it mostly results from secondary involvement of an abdominal pathology. Massive pleural disease impedes cardiopulmonary functions and threatens life. We are reporting our experience about managing a middle-age lady with a known case of Pseudomyxoma peritonei, who presented to causality with progressive breathing difficulty in acute exacerbation and cardiopulmonary compromise.

Keywords Pseudomyxoma pleuri · Emergency thoracotomy · Decortication · Pseudomyxoma peritonei

Introduction

Pseudomyxoma pleuri is a rare disease and almost always results as a secondary involvement from pseudomyxoma peritonei, most notable being appendicular, ovarian, or colonic origin. Their prognosis depends on the organ of origin, histology, grade, and disease extent. The condition is very uncommon and largely mentioned as case reports [1–3]. We are presenting our experience of managing one such case with particular mention on its emergency management.

Case Report

A 40-year-old lady with no comorbidity presented to causality with complains of progressive respiratory difficulty. At presentation, her vitals were as follows: P—120/m, BP—100/70 mmHg, afebrile, SPO₂—88% and RR—40/m. No air entry on the left-side lung field with dull note on percussion. Chest X-ray showed homogenous opacity in the left lung field with mediastinal shift to the right side (Fig. 1a). A left intercostal chest tube was placed onsite but fluid was highly viscous and could not be drained via a 32 Fr chest tube. She was admitted in the ICU and developed further respiratory distress with metabolic acidosis and hemodynamic instability requiring mechanical ventilation with inotropic support.

On further inquiry, we noted history of mucinous appendiceal tumor post incomplete cytoreductive surgery, followed by adjuvant FOLFOX-based chemotherapy with advice for completion of abdominal cytoreductive surgery with HIPEC in June 2014. She refused surgery and was lost to follow-up till present admission. Contrast CT imaging of the thorax and abdomen showed gross left pleural effusion with evidence of loculations and mediastinal shift with gross intraabdominal mucinous ascites/deposits (Fig. 1b, c). In view of a collapsed and trapped left lung with mediastinal shift and hemodynamic instability requiring mechanical ventilation and progressive deterioration, she was advised for emergency thoracotomy and decortications as a life-saving procedure. On exploration, there was thick gelatinous fluid-filled left pleural cavity with dense fibrous loculations (Fig. 2a). There was dense fibrous peel trapping whole of the left lung with no obvious lung or mediastinum infiltration; however, there were dense deposits on the diaphragm. Post surgery, the lung fully expanded on table and the optimum cytoreduction including complete pleural decortication was achieved leaving behind no measurable disease (Fig. 2b). Postoperatively, she was

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Fig. 1 Chest x-ray (a) and CT chest topogram (b) showing homogenous opacification of the left hemithorax with mediastinal shift to the right (b); CT chest showing gross left pleural effusion with evidence of loculations

and midline shift (c); chest X-ray showing fully expanded lung with no evidence of effusion/opacity on the 6th post-operative day (d), Incidental note made about calcified opacity likely benign calcification

electively ventilated and on POD1 was weaned of inotropic support and mechanical ventilation with discharge on the POD 6 (Fig. 1d). Later after recovery, she underwent abdominal cytoreductive surgery with complete cytoreduction and hyperthermic intraperitoneal chemotherapy. The peritoneal

cancer index (PCI) was 19 on exploration and we did complete cytoreduction to achieve a PCI of 0. The histological diagnosis was high-grade mucinous appendiceal tumor hence put on chemotherapy after tumor board discussion. Currently she is 2 months post-surgery and on chemotherapy.

Fig. 2 Intraoperative picture showing mucinous material (a), expanded lung after complete decortication (b). CW, chest wall; L, lung

Discussion

Cystadenoma of the appendix is the most frequent cause of Pseudomyxoma peritonei, but it can also occur in mucus-producing adenocarcinomas of the appendix, lung, large and small bowel, breast, stomach, pancreas, and fallopian tubes/ovary [1–3]. Pseudomyxoma with pleural involvement, which is also described as Pseudomyxoma pleuri, is an uncommon presentation. The literature is sparse and largely case report based [2–4]. Majority of the patients develop pleural involvement after primary disease and limited cases has been reported with simultaneous presentation. Their pathway of spread is largely speculative [2, 4]. In our patient, spontaneous transdiaphragmatic spread is the likely mechanism, as abundance of mucinous material with obvious infiltration of the diaphragm which was evident on exploration.

Pleural extension itself carries a poor prognosis. Cytoreductive surgery with intrapleural chemotherapy is a valuable treatment option and it is considered when preoperative or intraoperative pleural involvement is confirmed [4, 5].

Extensive pleural disease may present as an emergency and become a difficult case scenario where patient's general condition does not permit any surgical intervention. It is always a clinical dilemma about whether to proceed with high-risk surgery or wait for some recovery with conservative management. To the best of our knowledge, this is the only case report presenting as emergency management of this scenario. Emergency thoracotomy served the basic purpose of life saving.

The optimum curative treatment includes complete cytoreduction with hyperthermic intrathoracic chemotherapy (HITOC) [2], but in our case, owing to the surgical emergency, we did not offer her HIT. On later date, we have planned

for abdominal cytoreduction and hyperthermic intrapleural chemotherapy, but patient declined for surgery.

Conclusion

Pseudomyxoma pleuri is a rare condition and should always be considered as differential for breathing difficulty and pleural effusion in a diagnosed case of Pseudomyxoma peritonei. Even this rare case occasionally presents as emergency and one should be proactive and prompt.

Compliance with Ethical Standards

Conflicts of Interest The authors declare that they have no conflict of interest.

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