CASE REPORT

Mediastinal mature teratoma in a child- A case report

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SUMMARY

Mediastinal teratoma is an infrequent germ cell tumour and comprises of 1 to 5% of all mediastinal tumours. We report a case of mediastinal mature teratoma in a 12 year old boy who presented to us with persistent non-productive cough, fever and dyspnoea for the past 7 months. Computed tomographic scan of thorax revealed a large anterior mediastinal mass measuring 11.2x9.9x14cm with calcification within. He subsequently underwent a median sternotomy with left subcostal extension (L-incision) and excision of tumour. Histopathology of the tumour revealed a mature cystic teratoma. We would like to report a case of successful surgical management of a large mediastinal mature teratoma in a child.

KEY WORDS:
Mediastinal teratoma, child

INTRODUCTION

Mediastinal teratoma is an infrequent germ cell tumour, comprising of 1 to 5% of all mediastinal tumors.[1] It has an equal distribution in males and females with peak occurrence in the second and third decades of life. Mediastinal teratoma is derived from pluripotent embryonic cells which undergo differentiation into tissues with all three germ cell layers.[2] They can be classified into mature solid or cystic teratoma, immature teratoma and teratoma with malignant transformation.[3] Mediastinal tumour in the paediatric age group is an uncommon occurrence with a wide variety of etiological considerations which determine its management. Herein, we report a case of successful surgical management of a large mediastinal mature teratoma in a child.

CASE REPORT

A twelve year old boy presented to us with persistent non-productive cough, intermittent fever and reduced effort tolerance for the past seven months. There was a fullness over his left anterior chest wall and breath sounds were reduced over the left hemithorax. Chest X-ray showed a mass with calcifications, obliterating the left heart border and trachea was deviated to the right. Computed tomographic scan of thorax revealed a large heterogeneous mass in the anterior mediastinum, measuring 11.2x9.9x14cm in size (Figure 1). The mass has multilocular cystic, solid and fat components and calcification within. Serum beta human chorionic gonadotropin (<0.1IU/L), alpha fetoprotein (0.5IU/mL) and lactate dehydrogenase (236U/L) were tested normal. Ultrasound guided core biopsy of the left anterior mediastinal mass revealed features consistent with mature teratoma. Subsequently, he underwent a median sternotomy with left subcostal extension (L-incision) and excision of tumour. Intraoperatively, a large lobulated anterior mediastinal tumour was seen adherent to the pericardium over the area of left pulmonary artery and left atrial appendage, left chest wall antero-laterally and lingular part of left lung posteriorly. The tumour measured 17x12x7cm and weighed 860gm (Figure 2). A cuff of pericardium was excised en bloc with the tumour and the left pericardial defect was repaired with bovine pericardial patch. A rim of lung parenchyma that was densely adhered to the tumour was resected using stapled wedge resection technique. Histopathology of the tumour revealed a mature cystic teratoma with a mixture of benign mature tissues comprising skin with adnexal structures, mature glial tissue, pancreatic tissue, colonic and respiratory type mucosa, adipose tissue, chondro-osseous elements and fibrovascular proliferations. The patient had an unremarkable post-operative recovery and was discharged well 9 days post-surgery.

DISCUSSION

Mediastinal teratoma occurs in children typically as an anterior mediastinal mass. Other differential diagnosis for an anterior mediastinal mass in children are thymoma, lymphoma, thymic cyst, lymphangiomas, neurogenic tumours, germ cell tumours and mesenchymal tumours. Sixty percent of patients with mediastinal teratoma are asymptomatic and diagnosed incidentally during routine chest X-ray.[3] Compression symptoms are a common presentation, attributed to its mass effect to the mediastinal structures. Our patient presented mainly with respiratory symptoms as a result of the compression effect by the anterior mediastinal teratoma. Trichoptysis is a rare symptom but pathognomonic of mediastinal teratoma due to tumour erosion into the tracheobronchial tree. There have been rare reports of tumour ruptured into the pleural cavity, pericardium, lung parenchyma and great vessels, leading to life threatening complications hence require emergent surgery.[1,3]

Most mediastinal teratomas are benign mature teratoma which have well differentiated germinal derivatives mainly consisting of ectodermal elements. Immature teratoma is

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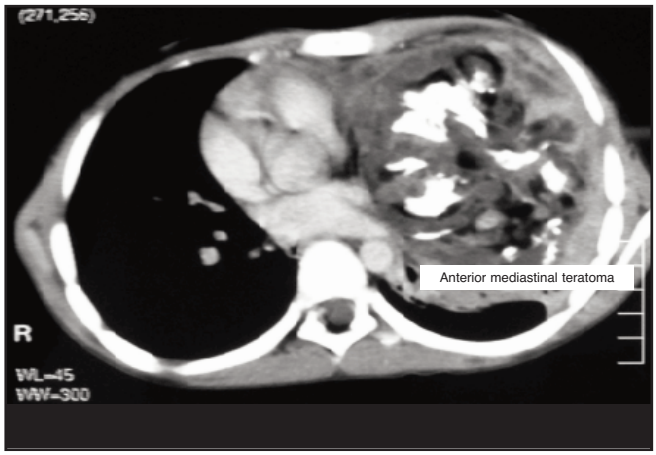

Fig. 1: Computed tomographic scan of thorax revealed a large heterogeneous mass with calcification in the anterior mediastinum extending into left hemithorax.

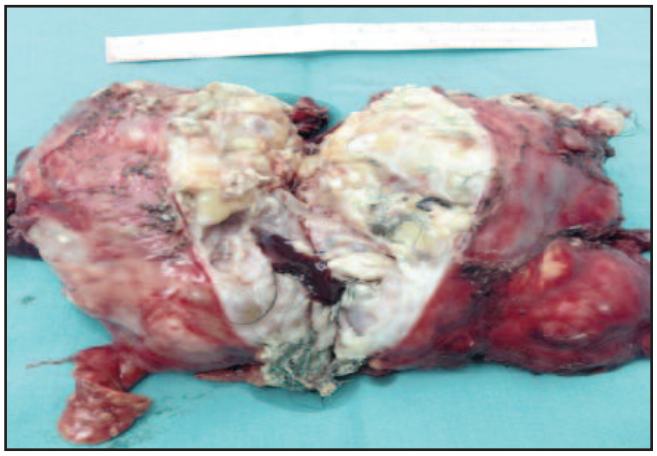

Fig. 2: Cut surface of mediastinal mature teratoma showing presence of hair, fat, cartilage and teeth.

characterized when immature germinal derivatives are seen. Teratoma with malignant germ tumor components has been described in the literature. Teratoma with malignant transformation is rarely encountered and presence of sarcoma, squamous cell carcinoma, adenocarcinoma or carcinoid have been reported.4 Hence surgical treatment is also justified to prevent future malignant transformation.

Computed tomography is the modality of choice to ascertain the diagnosis and to study the extent of mediastinal tumor.1 Mediastinal benign teratomas are typically well-circumscribed heterogeneous mass with radiographic features of multilocular cystic, solid and fatty components with calcifications. These radiographic features may not be present in all cases and only 20-40% of cases show evidence of calcifications.[2] Elevation of tumour markers like alpha fetoprotein and human chorionic gonadotropin may indicate malignancy.

The treatment of choice for mediastinal mature teratoma is complete surgical excision.[1,3] Literature reports have described approaches of mediastinal tumour excision with median sternotomy, anterolateral thoracotomy, posterolateral thoracotomy, clamshell incision or even thoracoscopic surgery for smaller mediastinal tumors.[5] Median sternotomy is the surgical approach of choice for huge anterior mediastinal mass.[1] Median sternotomy was adopted initially in our patient, however tumour extension to the left hemithorax with adherence to the anterolateral chest wall and the left lung required a left subcostal extension for adequate exposure and safe removal. L-incision or inverted-T incision can be extended from median sternotomy for larger anterior mediastinal mass encroaching into the hemithorax. Excision of mediastinal teratoma can be challenging when the tumour is densely adhered to mediastinal structures which may lead to devastating complications. Surgical approach to excise the mass is important. In this case, the mass was huge, compressing the left lung and was also densely adhered to the left pulmonary artery. Double lumen ventilation with left sided endotracheal tube was helpful during lung resection and to achieve good control of the left pulmonary artery during dissection. Large and lateral pericardial defects following en bloc mediastinal tumour resection need to be reconstructed to prevent cardiac herniation and prevent sternal-cardiac adhesion in case a redo surgery is required in the future. In this case, the left pulmonary artery was dissected from the tumour and pericardial repair can prevent kinking of this vessel. Additional procedures like lung wedge resection, lobectomy or prosthetic vascular reconstruction may be required for en bloc resection of mediastinal tumor.[2,5] Kesler *et al* stated that median sternotomy and clamshell incision were commonly adopted for excision of mediastinal tumour and 5% of their patients require cardiopulmonary bypass during excision of mediastinal tumor.[5]

Complete surgical excision is the mainstay of treatment for mature teratoma. Adjuvant therapy has no role in treatment of mediastinal mature teratoma.[2] Platinum-based neo-adjuvant chemotherapy followed by surgical resection or adjuvant chemotherapy for residual disease are considered for management of immature teratoma.[1,5] Radical resection of tumour determines the long term survival and low recurrence rate in patients with immature teratoma.[5] Teratoma with malignant transformation has an aggressive behaviour with rapid progression or metastasis. It has a poor prognosis despite combination of surgery and adjuvant chemo-radiotherapy.

In conclusion, complete removal of mature teratoma poses a challenge when strong adherence to vital mediastinal structures occur. Careful surgical planning and approach must be customized according to the size and extent of mediastinal tumour so as to provide adequate exposure, safe dissection and removal of mediastinal tumour.

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REFERENCES

1. Vohra LS, Talwar R, Mathur M, Mohan CV, Chawla N, Bharathi RS. Giant mediastinal teratoma—bull in a china shop: Management strategies. J Thorac Cardiovasc Surg 2007; 133(5): 1382-3.
2. de Hoyos A, Sundaresan RS. Resection of Mediastinal Teratoma. Operative Techniques in Thoracic and Cardiovascular Surgery. 2001; 6(4):209-20.
3. Jothianandan K, Tibb AS, McLemore M, Keller S, Appel DW. An adult man presenting with hemoptysis caused by mature teratoma with rupture into the bronchus and pericardium and complicated by Haemophilus influenzae infection. J Thorac Cardiovasc Surg 2010; 139(5): e104-7.
4. Shimizu K1, Nakata M, Hirami Y, Akiyama T, Tanemoto K. Teratoma with malignant transformation in the anterior mediastinum. J Thorac Cardiovasc Surg 2008; 136(1): 225-7.
5. Kesler KA. Technique of Mediastinal Germ Cell Tumor Resection. Operative Techniques in Thoracic and Cardiovascular Surgery. 2009; 14(1): 55-65.