J Ped Surg Case Reports 5 (2016) 15–18

Contents lists available at ScienceDirect

Journal of Pediatric Surgery CASE REPORTS

journal homepage: www.jpscasereports.com

Large intra-thoracic desmoid tumor with airway compression: A case report and review of the literature

Tyler D. Robinson [a,*], Matias Bruzoni [b], Peter Than [c], Safwan Halabi [d], James Wall [b]

[a] Boston University School of Medicine, USA
[b] Lucile Packard Children's Hospital at Stanford, Division of Pediatric Surgery, USA
[c] Stanford Hospitals and Clinics, Department of Surgery, USA
[d] Lucile Packard Children's Hospital at Stanford, Department of Radiology, USA

ARTICLE INFO

Article history:
Received 6 October 2015
Received in revised form
17 December 2015
Accepted 18 December 2015

Key words:
Desmoid tumor
Fibromatosis
Intra-thoracic
Dyspnea
Surgery

ABSTRACT

Intra-thoracic desmoid tumors are exceedingly rare tumors in the pediatric population, and can present with compression of cardiopulmonary structures and respiratory compromise. Surgical and anesthetic management of these tumors are challenging. We report the case of a 14-year-old male who presented with dyspnea, found to have a large intra-thoracic mass that was critically compressing his trachea. We provide a review of the literature of desmoid tumors and discuss the management of intra-thoracic masses.

1. Case report

A previously healthy 14-year-old male began experiencing slowly progressive respiratory symptoms two months prior to presentation to care. His previous medical problems included only mild intermittent asthma, for which he occasionally used an inhaler. His family medical history was significant only for diabetes. Initially, he began to notice mild dyspnea on exertion and fatigue. During this time, his parents also noted a 14-pound weight loss. One week prior to presentation, while swimming in a lake, he experienced significant dyspnea.

He presented to an outside hospital's urgent care clinic, where a chest radiograph (Fig. 1) demonstrated a large right chest opacification with mass-effect and leftward shift of the heart and mediastinum, and was immediately admitted to the pediatric intensive care unit (PICU). A contrast enhanced CT scan of the thorax (Fig. 2) revealed a large, homogeneously enhancing right-sided intra-thoracic mass, which crossed 6 cm to the left of midline and compressed the right atrium of the heart. There were no large calcifications or macroscopic fat density identified within the mass. The cross section of his trachea was measured to be 12 mm × 4 mm just superior to the carina. There was cortical irregularity and spiculation noted in the first and second right ribs, suggesting rib invasion by the soft tissue mass or point of origin from the rib of the soft tissue mass. There was aerated lung at the right base. The imaging differential diagnoses included: mediastinal origin lymphoma or malignant germ cell tumor; pulmonary and chest wall origin carcinoid tumor and Askin tumor; and metastatic disease [1].

He had a bedside core needle biopsy under procedural anesthesia of versed, ketamine and dexmedetomidine. He became cyanotic and desaturated, requiring bag-mask ventilation and epinephrine. Subsequently, he was tachycardic with a hyperdynamic precordium. The head of his bed was raised upright, and he was given normal saline and methylprednisolone, with resolution of his tachycardia. An arterial line was placed for continuous monitoring of his blood pressure. A bedside echocardiogram showed significant leftward deviation of the heart and aorta with impaired right atrial filling, but a normal ejection fraction. Preliminary pathology from the core needle biopsy showed low-grade spindle-cell neoplasm.

He was transferred to our hospital in stable condition. He showed no clinical evidence of superior vena cava syndrome. At this time, he did not exhibit any shortness of breath at rest, and was able

* Corresponding author. Tel.: +1 650 815 9807.
E-mail address: tdr@bu.edu (T.D. Robinson).

http://dx.doi.org/10.1016/j.epsc.2015.12.005

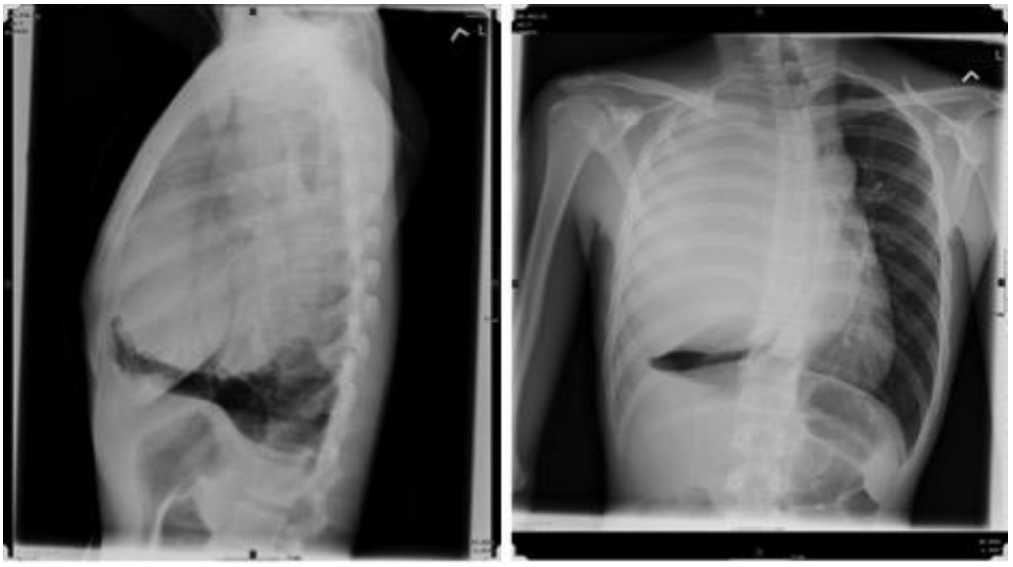

Fig. 1. Radiographs on initial presentation demonstrate a large mass-like opacification of the right chest with leftward shift of the heart and mediastinum including the trachea.

to lie prone comfortably. His father noted that his voice seemed different from one week prior, sounding more raspy and requiring increased effort to speak. Beta human choriogonadotropin, alpha fetoprotein, and urine catecholamines were all within normal limits. Ultrasound examinations of his testes and abdomen were negative for any masses. A second echocardiogram showed significant compression of both atria.

A care coordination conference was held with oncology, general surgery, radiation oncology and intensive care teams. Pre-operative endovascular embolization of the tumor was not planned. Neo-adjuvant use of radiotherapy and chemotherapy were discussed. However, the consensus decision was to proceed with primary surgical resection of the mass, due to the patient's tenuous respiratory status as evidenced by the episode of desaturation and significant dyspnea at the transferring hospital. CT imaging demonstrated a tracheal cross sectional area of 0.48 cm^2, which is 32% of expected for our patient's age and gender. Evidence suggests that for pediatric patients with mediastinal tumors, compression of the trachea to cross sectional areas of less than 50% is extremely high risk for total respiratory collapse upon induction of general anesthesia [2]. The goals of surgery were primary relief of compression of mediastinal structures and pathological definition of the tumor. The most serious risks during surgery were anticipated to be induction of anesthesia because of the compressed proximal airway, and the likelihood for substantial bleeding from the large tumor. The family was counseled about the risks and benefits of surgery, and decided to proceed.

The operative team included two pediatric general surgeons, a pediatric cardiac surgeon, a general surgery resident and the pediatric cardiac anesthesia team. In case of cardiorespiratory collapse on induction of anesthesia, the team planned for rapid decompression of the chest through median sternotomy, and central cannulation for cardiopulmonary bypass. Additionally, femoral venous and arterial catheters were placed preoperatively in preparation for emergent extracorporeal membrane oxygenation.

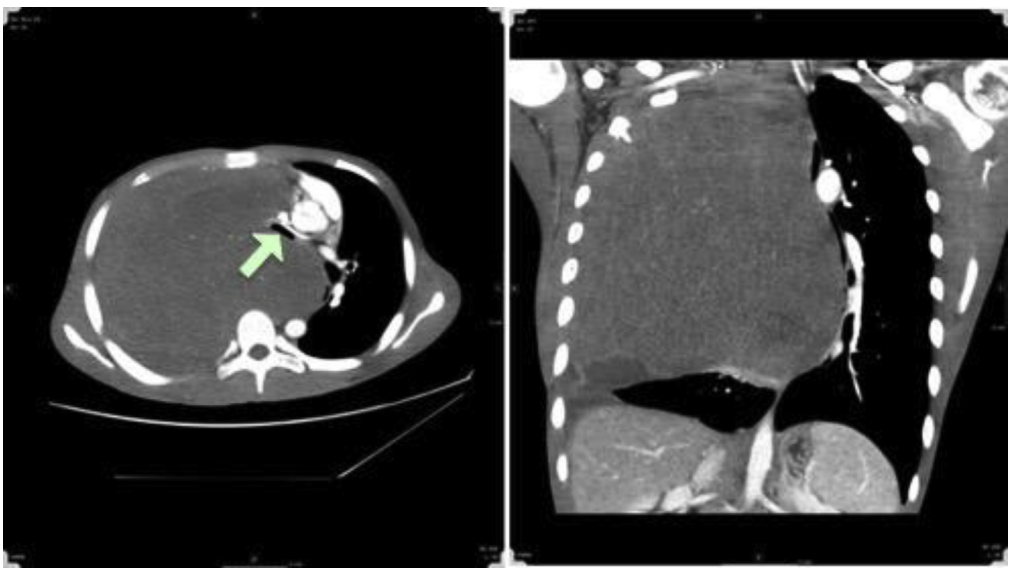

Fig. 2. CT scan demonstrating a large enhancing soft tissue mass occupying the right chest and crossing the midline. There is mass effect and compression on the trachea just superior to the carina (green arrow).