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

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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

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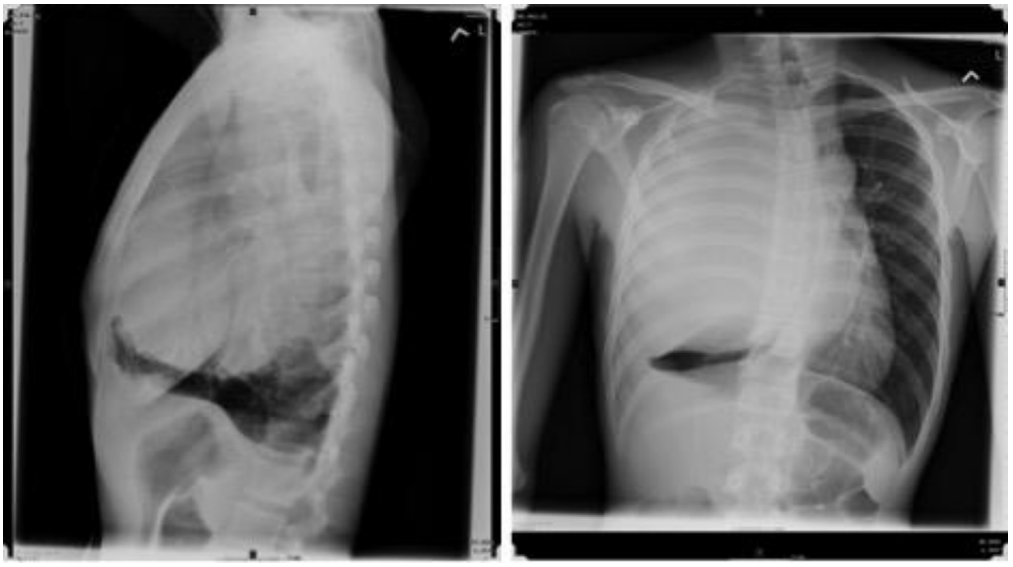

Fig. 1. Radiographs on initial presentation demonstrate a large mass-like opacification of the right chest with leftward shift of the heart and mediastinum including the trachea.

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.

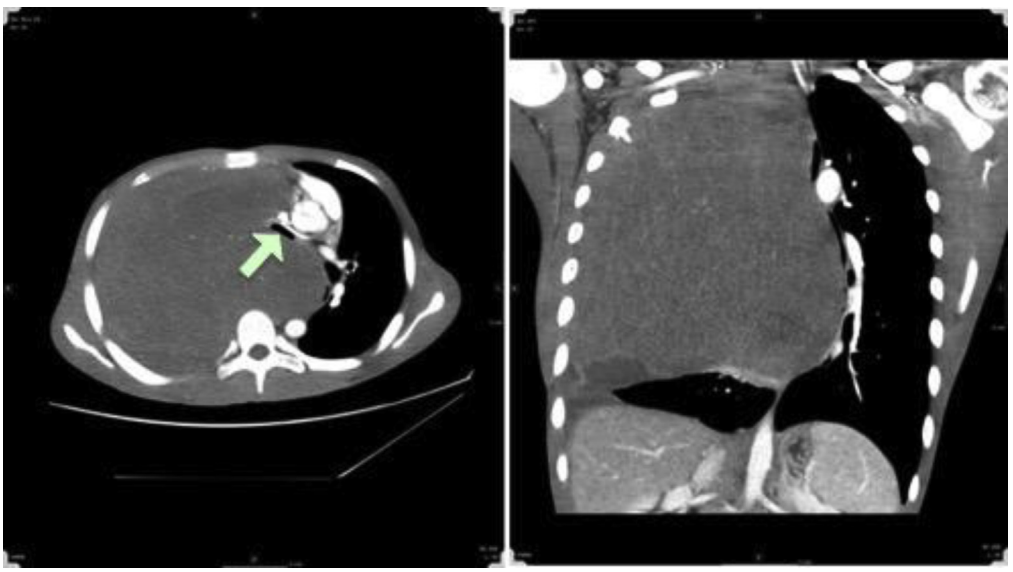

Fig. 2. CT scan demonstrating a large enhancing soft tissue mass occupying the right chest and crossing the midline. There is mass effect and compression on the trachea just superior to the carina (green arrow).

Induction of anesthesia progressed smoothly with ketamine and midazolam sedation. Topical lidocaine was sprayed onto the posterior oropharynx and vocal cords prior to intubation. The patient maintained his oxygen saturation and stroke volume through induction. The incision was a median sternotomy with right thoracotomy for maximum exposure. After the sternotomy, there was an immediate improvement in ventilation and the patient was then fully paralyzed. The right pleura was divided, with immediate visualization of the mass filling the entire right chest. The mass was dissected from the surrounding tissues using a combination of blunt dissection and electrocautery. Effort was made to remove the mass en block, but it proved too difficult to do so because of its large size and the need to control bleeding. Ultimately, the mass was removed in multiple pieces. The mass was safely freed from the pericardium and vena cava medially, the diaphragm inferiorly, and the upper lobe of the right lung medially. Although no major vessels were damaged, the tumor itself was very bloody, requiring many surgical clips and use of an argon beam coagulator. The mass remained adherent to the apex of the right chest and could not be safely separated from the area of the subclavian vessels and the brachial plexus. As much tumor as possible was removed, ultimately leaving a rim of residual tumor approximately 5–10 mm thick. The area was coagulated with the argon beam and absorbable hemostatic cellulose strips. This surface was marked with six large vessel clips.

There were no operative complications, and the patient tolerated the procedure well. Intraoperative blood loss was estimated at 7 L, primarily from the tumor itself. The patient was transfused 21 units packed units of red blood cells, 18 units of fresh frozen plasma, and 4 units of platelets. The patient was transferred to the PICU intubated. He was hypotensive postoperatively, but was resuscitated with crystalloid.

His postoperative course was relatively uncomplicated. He was extubated without issue two days after his operation. He was discharged six days after his operation. Final pathology of the surgical specimen was desmoid type fibromatosis. The tumor measured 31.0 × 23.5 × 11.2 cm and weighed 2.93 kg (Fig. 3).

He continued to recover well at home. He had no residual symptoms from debulking surgery and demonstrated no injury to his brachial plexus. He was followed by pediatric oncology and started on oral sorafenib in an attempt to shrink the residual tumor. An oral agent was chosen over intravenous chemotherapy to facilitate his return to school and normal activities. Radiation therapy was not recommended because of his young age. He was evaluated by genetics and found to be negative for the APC gene mutation.

2. Discussion

Desmoid tumors, also called aggressive fibromatosis, are benign soft tissue tumors arising from musculoaponeurotic or fascial structures. Although they have no potential for metastasis, they are known to be very locally aggressive, and can invade or compress neighboring structures. Additionally, they have high rates of local recurrence, even with complete surgical resection with negative margins. They most commonly affect individuals aged 15–60, and are slightly more common in women than men. Desmoids are rare tumors, comprising approximately 0.03% of all neoplasms and 3% of soft tissue tumors. Approximately two-thirds of all desmoid tumors are intra-abdominal [3]. Tumors affecting the chest wall comprise roughly 20% of all extra-abdominal desmoids, which also occur in the abdominal wall, retroperitoneum, hip or buttock, neck, or extremities [3,4]. Desmoid tumors have a variable clinical course, and include lesions that demonstrate spontaneous regression, long-term stability, slow growth or rapid progression.

The pathogenesis of desmoid tumors is not completely understood, though is likely related to defects in connective tissue.

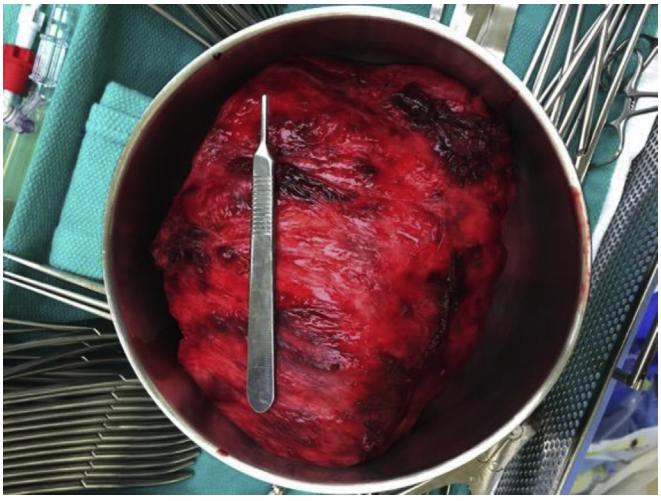

Fig. 3. The first of several large pieces of tumor removed from the chest.

Genetic predisposition, hormonal effects and physical trauma are related to the development of desmoid tumors [4–6]. The majority of desmoid tumors are sporadic. Around 7.5% of all lesions are associated with Familial Adenomatous Polyposis (FAP) [7]. FAP-associated desmoids result from mutations in the adenomatous polyposis coli (APC) gene, whereas sporadic tumors are caused by mutations in either APC or beta-catenin [4,6]. Desmoid tumors have been associated with high estrogen states, especially pregnancy [6,8]. Approximately 30% of patients with desmoid tumors have a history of physical trauma, most commonly prior surgery [6,9].

Intra-thoracic desmoid tumors are exceedingly rare; approximately 40 case reports have been published [10], with around 10 of these found in pediatric patients. Intra-thoracic tumors originate from the chest wall structures such as the intercostal muscles or shoulder girdle and grow inward, or can originate from mediastinal structures. Intra-thoracic desmoid tumors are typically clinically silent until they are discovered incidentally or until they begin to compromise nearby structures. In a recent series of 28 intra-thoracic tumors, 86% presented with palpable lesions, 71% with pain, 25% with loss of motor function, and 7% with hypoesthesia. A total of 11% were asymptomatic and identified incidentally [4]. Very few cases of intra-thoracic desmoid tumor presented with respiratory symptoms [10,11], similar to our case. Even very large intra-thoracic desmoids have presented with no respiratory symptoms [12,13]. The largest tumor removed from the thorax weighed 5.89 kg [10].

The overall mortality from desmoid tumors is roughly 1% or less, due to their inability to metastasize. Fatalities result from mechanical compression or invasion of neighboring vital structures [6]. Consequently, the mortality rate is higher in intra-abdominal and intra-thoracic cases. The goal of care in treating desmoid tumors is reduction of mechanical compression and invasion of neighboring structures. The primary treatment for desmoid tumors is surgical resection, and the measure of treatment success is recurrence-free survival.

There are numerous studies reporting risk factors for desmoid tumor recurrence. In a multivariate analysis 495 patients, the largest cohort of desmoid tumors treated with surgery, only tumor site, tumor size and patient age were associated with recurrence rate [14]. Using a prognostic nomogram published with this study, our patient's 5-year local recurrence-free survival rate is approximately 0.3. The effect of microscopically positive surgical margins is unclear; some series report a higher recurrence rate than in tumors with wide negative margins [6,15,16], while others claim that recurrence rate is independent of microscopic margin status [6,17]. In most studies,

however, macroscopically positive margins have a significantly higher recurrence rate than surgically free margins, with up to 89% of tumors recurring [17,18]. However, even tumors that are surgically resected with negative margins can have recurrence rates of 16–19% [6,15–18]. Other reported risk factors associated with recurrence include anatomic site, size, gender and age [6]. Tumors that affect the extremities have the worst prognosis, followed by intra-abdominal or extra-abdominal tumors. Those affecting the abdominal wall have the lowest rate of recurrence. Tumors greater than 7 cm have higher rates of recurrence. Some studies have reported that tumors affecting women and those affecting younger individuals have higher rates of recurrence [6]. There is preliminary evidence that recurrence rate may be correlated with certain molecular factors, a topic of current research interest.

Historically, desmoids have been thought to be poor candidates for systemic therapy due to their slow growth, low cellularity and typical high differentiation. However, evidence from several retrospective case series suggests that adjuvant treatment modalities may have efficacy in the primary treatment of desmoid tumors and in prevention of recurrence. Radiation therapy has been suggested to be beneficial in preventing recurrence, or for treatment of unresectable tumors [17,19,20], although some have found no added benefit of radiation [21]. There is limited evidence for the use of systemic therapy, though the use of nonsteroidal anti-inflammatory [18], selective estrogen receptor modulation [4], and cytotoxic [17] agents have been proposed. There is a need for a multicenter trial to establish the efficacy of adjuvant therapies [17].

In our case, the tumor was clinically silent until it grew large enough to begin compressing the mediastinum, reducing cardiac output and constricting the trachea. This presented several challenges to the treatment team. The first challenge was the danger of general anesthesia with a greatly compressed mediastinum. There have been several algorithms proposed for the management of mediastinal masses in children [2,22]. An important pre-operative evaluation is tracheal cross sectional area, calculated using a CT image. Evidence suggests that tracheal cross sectional area less than 50% of expected for age and gender have a high risk for total respiratory collapse upon induction of general anesthesia [2,23]. In our case, the distal trachea was reduced to a cross sectional area of 0.48 cm^2 above the carina, just 32% of normal [24]. Thus, prior to the operation, our patient's femoral artery and vein were cannulated for the possible need for extracorporeal membrane oxygenation. Additionally, a pediatric cardiac surgeon was ready to provide central cannulation if necessary. Fortunately, our patient tolerated induction well, and never demonstrated evidence of impending respiratory collapse.

A second surgical challenge was the large size of the mass, which weighed nearly 3 kg and measured over 30 cm long. Even using a full sternotomy and thoracotomy, en bloc extrication of the mass would not have been feasible without the resection of several ribs. Thus, we elected to remove the mass in pieces. Desmoid tumors are not known to be particularly vascular, and bleeding was easy to control by use of surgical clips and the argon beam coagulator. In addition, desmoids do not metastasize, and so could not seed the thorax or disseminate systemically. In biopsy-proven desmoid tumors, we consider resection in multiple pieces to be an appropriate surgical method.

A third surgical challenge was the fact that the origin of the tumor was strongly adherent to the apex of the right chest. Full primary resection of the tumor would have required deep dissection near the subclavian artery and vein and the brachial plexus. Thus, a small cap of residual tumor was left and marked with surgical clips. Adjuvant radiation and systemic therapy may help this tumor to regress. However, further surgical intervention will likely be required.

3. Conclusion

Desmoid tumors are locally aggressive and can become life threatening when they compress neighboring structures. Intrathoracic tumors are very rare, and do not usually present primarily with respiratory symptoms. Special precautions must be taken when managing patients with a tumor compressing the mediastinum. Desmoid tumors have high rates of recurrence even with full surgical resection. Further research is needed to optimize adjuvant treatments of desmoid tumors.

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