Acute affection of the thorax in children due to pulmonary blastoma

ABSTRACT

Background: In children, acute affectation of the thorax is presented during pleuropulmonary infection, bronchopulmonary malformation and malignant tumors. We had the opportunity to treat patients with malignant tumors of the thorax and we present our findings.

Clinical cases: We studied three patients with a diagnosis of pleuropulmonary blastoma. All patients were treated by surgical resection of the neoplasm and one patient received pneumonectomy. Diagnosis of blastoma was confirmed histologically and one had positive histochemical markers to vimentin and S-100. Only one patient died.

Conclusions: 1. Different cellular types of pleuropulmonary blastoma were confirmed using histochemical markers. 2. The pediatric surgeon facing a case of a child with acute thorax affection due to pleuropulmonary blastoma must undertake thoracotomy on an urgent basis.

Key words: Blastoma, pleuropulmonary, children.
BACKGROUND

In pediatric patients, an acute condition of the chest is a clinical syndrome characterized by irritability, anxiety, progressive shortness of breath, fever, pain with pressure on the affected side and, in addition, signs of condensation or pleural effusion. This syndrome appears together with bronchopulmonary infection, or malformations and thoracic tumors. The opportunity of having treated some patients with this disease motivated us to report our experience. Our objective is to analyze a series of cases with diagnosis of thoracic neoplasm and compare our observations with recent publications.

METHODS

We performed a retrospective, observational, cross-sectional and descriptive study of the clinical records of patients with a diagnosis of malignant thoracic tumor at the Sonora State Children’s Hospital (Hospital Infantil del Estado de Sonora, HIES) in Mexico.

CLINICAL CASES

Case 1

We present the case of a 15-year-old female who had a left nephrectomy, which was performed at 7 years of age without diagnostic confirmation. The present ailment evolved during a 23-day period with rhinorrhea, fever of 39°C and progressive dyspnea that lead to orthopnea and incapable of tolerating dorsal decubitus position. On physical examination the patient weighed 41 kg, height 1.5 m, temperature 36°C, and blood pressure 110/70 mmHg; Neck examination noted venous engorgement and increased circumference in its inferior portion. Chest showed deformity at the expense of the right side. Intercostal spaces were open with total dullness and inaudible breath sounds. Abdominal examination revealed an old surgical scar, no pain and no organomegaly. The rest of the examination was without significant findings. Chest x-rays demonstrated a total homogeneous opacity on the right side without calcifications or air bronchogram and with left mediastinal deviation. Laboratory exam revealed hemoglobin 10.0 g/dl and leukocytes 16,300/mm³ with predominance of lymphocytes. Bronchoscopy showed hyperemic mucosa with trachea deviated to the left and the right main bronchus with decreased diameter but without modification of the dimensions with sustained ventilation. Pre-operative diagnosis was an acute condition of the chest due to probable malignant tumor and superior vena cava syndrome.

Right thoracotomy showed a reddish-gray friable, hemorrhagic tumor adherent to the pulmonary hilum and that also compressed the superior vena cava and adjacent vessels. Upon attempting to release the vascular compression, the patient suffered cardiac arrest without response to the usual resuscitation maneuvers. Histopathological diagnosis was embryoma of the right lung (Figure 1). Autopsy was not done.

Figure 1. Case 1. Embryoma. Cells with pleomorphism, scanty cytoplasm and loose matrix with some vessels.
Case 2

We present the case of a 5-year-old male without prior clinically significant medical history. The current ailment evolved during the previous 20 days with unquantified fever, cough, signs of respiratory insufficiency, and pleural effusion syndrome. In the Emergency Department a right chest tube was placed and connected to a water seal. A pediatric surgeon was consulted and during physical examination the following was noted: The patient was thin, pale and irritable. Chest showed signs of respiratory insufficiency, a pleural tube on the right draining scant serosanguineous material, dullness in the entire right hemithorax and inaudible respiratory sounds. The remainder of the physical examination was without change. Laboratory findings were hemoglobin 13.4 g/dl and leukocytes 28,000/mm³ with predominant neutrophils. Cytology of the pleural fluid was negative for malignant cells. Chest computed tomography showed total right opacity, no calcifications and no air bronchogram. The mediastinum was found deviated to the contralateral side (Figure 2). On initial bronchoscopy the trachea was observed to be deviated to the left and the carina was not well visualized. Preoperative diagnosis was an acute condition of the chest due to a probable malignant pulmonary tumor. Findings of the right thoracotomy showed a tumor that occupied the entire chest cavity and displaced the entire lung downwards and forward without invasion. The neoplasm was grayish in color and hemorrhagic. In some areas its consistency was increased and very bloody with a weight of 460 g. The site of growth came from the parietal pleura between the 4th and 5th intercostal spaces. There were no intraoperative problems. Histological diagnosis was thoracic leiomyosarcoma. The plan was to administer chemotherapy with vincristine, actinomycin and cyclophosphamide scheduled for 2 years together with radiation therapy.

The patient progressed well for 1 year; however, during a follow-up visit, a round opacity appeared when reviewing the chest x-rays. It was localized in the right superior lobe and part of the superior mediastinum. Shortly thereafter, another thoracotomy was carried out to resect a tumor that did not invade the right superior lobe (Figure 3). Once the child recovered he was referred to the Oncology Service to continue with chemotherapy. Histological diagnosis was recurrence of leiomyosarcoma.

Case 3

We present the case of a female 2 years 5 months of age. Clinical and surgical history included infectious respiratory problems and respiratory insufficiency treated for 8 months with multiple antibiotics without improvement. She had a right chest tube placed and it was...
removed after 1 week after noting improvement. At 2 years 3 months of age and due to a clinical picture similar to the one described, she had a right thoracotomy performed and remained hospitalized for 28 days. The diagnosis was pulmonary bullae. The current symptoms upon her admission to the Emergency Department were irritability, dyspnea, grayish skin, very visible superficial venous network on the right anterior part of the chest and with an increase in volume, total dullness and no ventilation, tachycardia and tachypnea. On physical examination the following were demonstrated: weight 12 kg, height 86 cm, temperature 37.6°C, heart rate 110 beats/min, and blood pressure 100/70 mmHg. The right hemithorax showed signs of respiratory insufficiency, increased voice transmission, inaudible respiratory sounds, and the tip of the heart showed a left deviation. Pain was demonstrated upon pressure of the right rib cage. Dark blood was collected at pleural puncture. Preoperative diagnosis was acute condition of the chest most likely due to lung tumor. Laboratory tests reported hemoglobin 11.9 g/dl, leukocytes 7,800/ mm³ and α-fetoprotein 5 g/l. Plain X-ray of the chest showed total right opacity without calcifications or air bronchogram and with displacement of the mediastinum towards the left side. Ultrasound of the chest showed solid and fragmented images with hemorrhage and perilesional fluid without calcifications. Computed tomography with a cut at the level of the carina in the right main bronchus showed an intrabronchial extension of the tumor (Figure 4). Right thoracotomy findings were firm parieto-visceral fibrous adhesions and compression of the superior and middle lobe. The middle and inferior lobes were released from where the friable tumor emanated. The appearance was cerebroid, dark red in color, very fragmented, and with dark hemorrhagic areas. Right pneumonectomy was performed (Figure 5). Histopathological diagnosis was spindle cell malignancy with numerous abnormal mitoses and abundant thin-walled vessels. Histochemical markers were positive for vimentin and S-100 (Figure 6). The diagnosis was undifferentiated sarcoma with neurogenic tissue of the right lung. Postoperative evolution was favorable and the patient was referred to the Oncology Service for chemotherapy.

RESULTS

There were 12 patients seen with the diagnosis of thoracic neoplasm and only three had pleuropulmonary blastoma.

Figure 3. Case 2. Round tumor with thin pseudocapsule, regular vascularization and without right upper lobe invasion.

Figure 4. Case 3. Computed tomography; cut at the level of the carina. Main right bronchus shows an intrabronchial extension of the tumor.
Pleuropulmonary blastoma is a dysmembrionic blastematous and malignant mesenchymatous type of neoplasm. According to Seballos and Klein, it was recognized for the first time by Barrett and Barnard in 1954 who interpreted its histological characteristics as nephroblastoma. Spencer, in 1961, reported that this type of tumor arises from the mesodermal tissue constituted by immature mesenchymal and epithelial cells. In 1988, Wright wrote that Manivel mentioned that it is a neoplasm almost exclusively of children, but in adults appears at about 30 to 40 years of age with an epithelial component. Dehner, in 1994, proposed classification of the pleuropulmonary blastoma according to its macroscopic characteristics into the following types: Type I cystic, Type II cystic-solid and Type III solid; type I has a better prognosis than types II and III.

Figure 5. Case 3. Tumor invades the three lobes. The tip of the clamp shows no bronchial invasion.

DISCUSSION

The frequency of presentation is rare, but currently there is an international registry for pleuropulmonary blastoma of children’s hospitals and clinics in Minnesota where cases from 1987 to 2008 are concentrated and total > 200 patients. With regard to gender there is no significant difference and in children its prevalence is < 5 years of age. The majority of the patients have a history of recurrent lung infections that have been treated by several physicians without clinical improvement. This results in a diagnostic delay until the child shows signs of an acute thorax and then thoracic tumor is considered. Imaging studies should examine in a sequential manner and preferably with an imaging specialist so as to have a good clinicoradiological correlation and thereby determine the characteristics of the tumor (cystic, solid, with effusion, etc.) and attempt to determine the nature of the neoplasm and its extension. On computed tomography the densities can be measured in Hounsfield units (HU) to observe differences between solid and cystic lesions, with or without effusion and with the coronal and cross-sectional cuts to determine the exact dimensions and characteristics of the neoplasm (Figure 4).

Histological diagnosis is established with H/E stain and some special stains to see fibrosis and morphology of the malignant cells. For some time now, histochemical markers are useful to verify the existence of pleuropulmonary blastoma. Vimentin is a 57-kDa protein that is expressed in all mesenchymal cells and S-100 protein is linked to the calcium transporter and signals glial tissue, chondrocytes and epithelial tumors as in the cases herein reported (Figure 6).

Currently, the etiology of pleuropulmonary blastoma continues to be unknown; however, in some cases it is associated with trisomy 2, 8, and 12 and with neoplasms of the ovary and uterus, leukemia and Hodgkin’s lymphoma. Studies in families with alterations in the DICER 1 (ribonuclease enzyme that helps generate the microRNA of the genes that control cell growth) conclude that its loss alters the synthesis of microRNA and promotes proliferation of mesenchymal cells.
Chemotherapy given to children with pleuropulmonary blastoma has a variety of schemes including vincristine, cyclophosphamide and actinomycin D, etoposide, cisplatin, etc. Some patients also receive radiation therapy without improvement of survival.

Kaneko et al.\textsuperscript{12} reported the case of a 4-year-old female with recurrence of pleuropulmonary blastoma at 29 months postoperatively for which a regimen of both chemo- and radiotherapy was indicated. Autologous stem cell transplant was done with a notable improvement. This latter treatment may help patients with tumor recurrences.

Pleuropulmonary blastoma also presents itself in domestic mammals such as horses, dogs and sheep. Woolford et al.\textsuperscript{13} reported on a horse fetus with a cystic solid pleuropulmonary blastoma and the histopathology was similar to that reported in humans. It was also positive for histochemical markers such as vimentin and cytokeratin, for which the blastematous character of the neoplasm is confirmed.

Participation of the pediatric surgeon in the treatment of patients with pleuropulmonary blastoma is crucial because the neoplasm should be resected entirely, seeking an improved prognosis. Unfortunately, the majority of patients have tumor recurrences; therefore, it is advisable to educate parents about the importance of ongoing chemotherapy treatments and follow-up visits.\textsuperscript{14}

Treatment of a patient with severe neoplastic problems requires the participation of various professionals and with unified criteria in order to take best decisions. In the medical literature reviewed and in the evolution of the reported series, it is evident that the delay in early diagnosis is due to the low frequency of cases with pleuropulmonary blastoma, and inadequate physical examination of children’s chest. The desire to improve the prognosis for these patients should be achieved with all available resources, thereby immediately directed to determine the nature of the neoplasm. For this reason, we recommend the next indications to be followed:

1. During the prenatal stage, methodical monitoring by ultrasound in order to discover any pulmonary lesions and follow its progression until the child is born.
2. During the neonatal period and up to 2 years of age, if the child suffers an infectious pneumonia that does not disappear in 2 weeks, it is necessary to perform computed tomography.
3. If the patient improves for a short time and once again presents a similar clinical picture, it is indicated that all imaging studies be performed to rule out neoplasm.
4. All these characteristics are present in older children and in adolescence; therefore, they should be treated in the same and complete manner.

5. Postoperative medical follow-up should be done with professional interest and with a strong dose of humanism.

In conclusion, the different cell types that form pleuropulmonary blastoma are verified with histochemical markers. The pediatric surgeon, when faced with a patient with an infection of the chest due to pleuropulmonary blastoma, will need to perform emergency thoracotomy with the goal of resecting the primary tumor along with any implants and, if possible, any metastases.

“Clinical diagnosis is a very playful devil that, when we think we have him trapped in our fist, escapes between our fingers laughing at us”…. Marín R.15

REFERENCES