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Malignant pleural mesothelioma in a child

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ABSTRACT

Malignant pleural mesothelioma (MPM) is an aggressive malignancy that occurs extremely rarely in the pediatric population. It carries a dismal prognosis. Adult studies are often used to guide therapy in the pediatric population, as a limited number of case reports form the body of pediatric literature. Herein, we document the course and treatment of an 8-year old male diagnosed with MPM. The diagnosis came after he presented to his family physician with dyspnea and was found to have a large right-sided chest mass on subsequent imaging. Through an initial right pneumonectomy and subsequent chest wall excision, followed by chemotherapy with Pemetrexed and Cisplatin he remains virtually disease free today, almost 2 years following surgery.

1. Paragraph setting the train of thought

Malignant pleural mesothelioma (MPM) is an aggressive malignancy that occurs extremely rarely in the pediatric population and carries a dismal prognosis [1]. Less than 5% of presenting cases are in children, and less than 300 cases of such have been published in the literature [1,2]. MPM in children appears to represent a different entity from its adult counterpart, as it is not associated with asbestos or radiation exposure [1,3], and much of the information regarding the disease is derived from the limited number of case reports available. There is no accepted standard of treatment for childhood MPM, and much of our current therapy is guided by the data available from adult studies. We report a case of an 8-year old male diagnosed with MPM who was successfully treated with pneumonectomy, chest wall excision, and subsequent chemotherapy using Pemetrexed and Cisplatin with hopes of adding to the pediatric body of literature and improving the outcomes for pediatric patients with MPM.

2. Case report

An eight year old male who originally presented to his family physician complaining of dyspnea, weight loss and lethargy was referred to the department of Pediatric Surgery at the University of Alberta/Stollery Children's Hospital. There was no known exposure to asbestos or radiation. A chest radiograph and CT scan (Figs. 1 and 2 respectively) demonstrated a large right-sided chest mass that appeared to be originating from the right upper lobe and encroaching on the right middle lobe. The dimensions of the mass measured roughly 10.6 × 7.5 × 7.7 cm.

Needle biopsy was initially preformed on the mass, and suggested the possibility of malignant cells; however, it was non-diagnostic. The differential remained broad, including infectious (including fungal, as the child had recently traveled to Arizona) and inflammatory etiologies in addition to neoplastic possibilities.

With the aim of ascertaining a diagnosis, the child was taken to the operating room for a thoracotomy and open lung biopsy. A few expert opinions from cancer centers indicated that frozen section pathology was consistent with an undifferentiated or poorly differentiated carcinoma, such as a choriocarcinoma. The pathology was then reviewed by Dr. Chris Fletcher, chief of onco-pathology at the Dana-Farber/Harvard Cancer Center and a final diagnosis of malignant mesothelioma—epithelioid tye was reached. Dr. Fletcher specializes in onco-pathology including mesothelioma. Pathological findings of note included electron microscopy of the tumor cells that showed numerous long, narrow microvilli, and hematoxylin-eosin staining of the tumor that showed cords of tumor cells and epithelioid nuclei with large pink eosinophilic cytoplasm (Fig. 3). Immunohistochemical staining of the tumor with an antibody against epithelial membrane antigen was also positive (Fig. 4). Immunohistochemistry also demonstrated multifocal convincing

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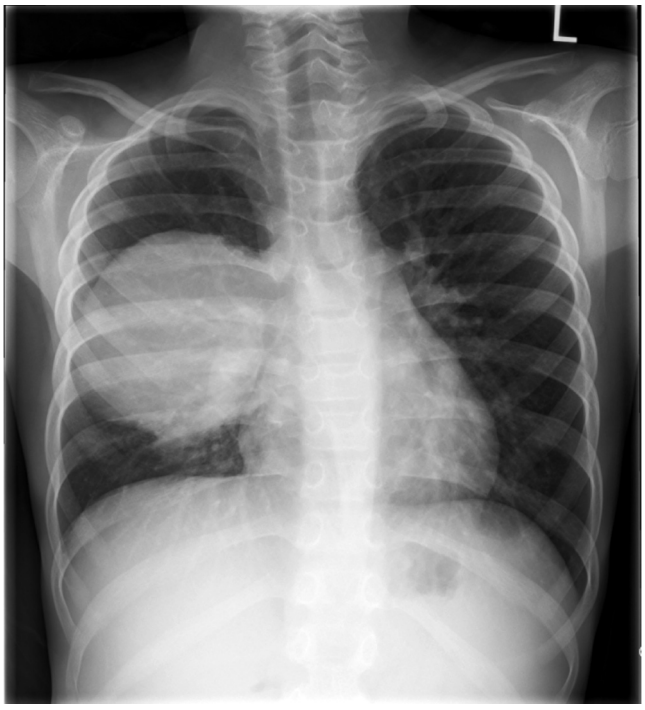

Fig. 1. Chest radiograph with right-sided chest mass.

positivity for calretinin and WT1 to support the diagnosis of mesothelioma. Synovial sarcoma was not ruled out by genetic analysis, as upon expert review in Boston, the diagnosis of synovial sarcoma was not further considered following the immunohistochemistry results. All of these results supported the diagnosis of malignant mesothelioma—epithelioid type.

Bone and PET scanning failed to demonstrate any evidence of metastatic disease, and the lesion appeared to be well circumscribed and localized to the right lung (Fig. 5). As such the child was taken to the operating room for a right thoracotomy and

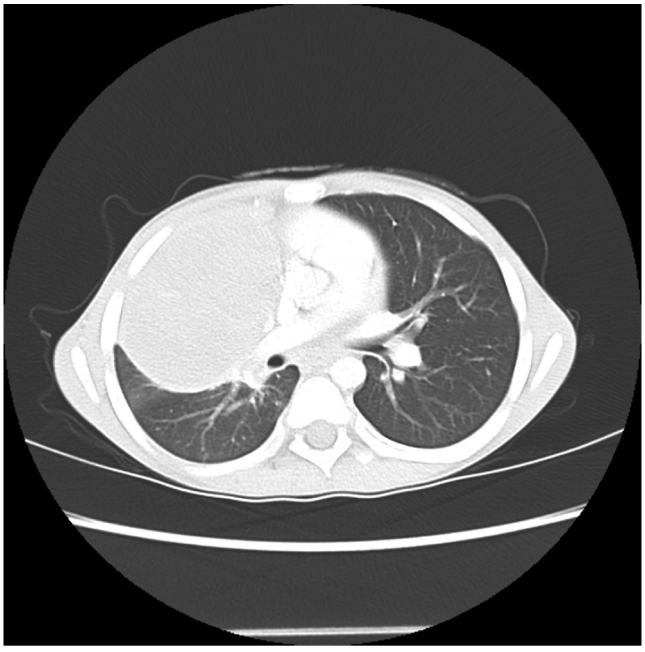

Fig. 2. CT scan showing axonal view of right chest mass.

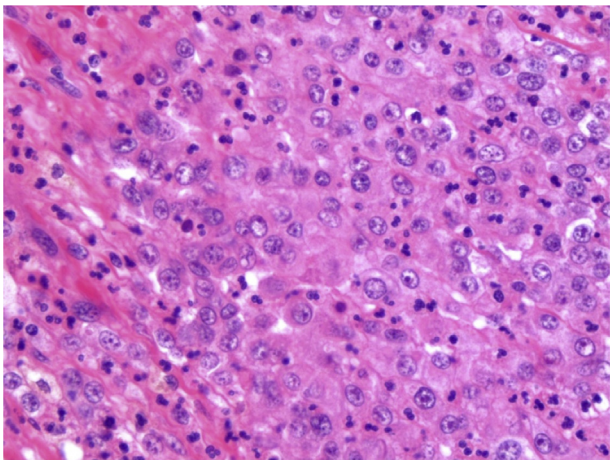

Fig. 3. Hematoxylin-eosin staining of the tumor showing cords of tumor cells and epithelioid nuclei and large pink eosinophilic cytoplasm (400× original magnification).

pneumonectomy. Intraoperatively, the tumor did not have the characteristic appearance of a malignant mesothelioma, and there was no evidence of a pleural rind encasing the lung or invading the chest wall. Instead, the tumor appeared to be well localized within the visceral pleura and extending into the lung parenchyma (Fig. 6a and b). The tumor did not appear to be of hilar origin, and this would be unusual if it were the case. Most commonly the tumor is found in the pleural lining surrounding the periphery of the lung. We believe that the tumor could have possibly originated in the visceral pleura of the horizontal or oblique fissure, and had grown to involve all three lobes of the right lung. A decision was made to perform a right pneumonectomy, as there was no evidence of invasion or spread to the chest wall, diaphragm or pericardium in the setting of a tri lobar tumor.

Post-operatively, the child did well, but unfortunately developed a small metastatic lesion in the thigh that was excised. Additionally, the child was found to have recurrent disease involving the right chest wall on PET scanning roughly two months following pneumonectomy. As a result, he was again taken to the operating room for excision of the right chest wall and removal of ribs four through eight. The pathologic margins were clear, and Prolene mesh was used to replace the chest wall.

The child received chemotherapy post-operatively and has had a very good response. He received eight courses of chemotherapy for a total duration of one year of therapy. Six courses of Pemetrexed

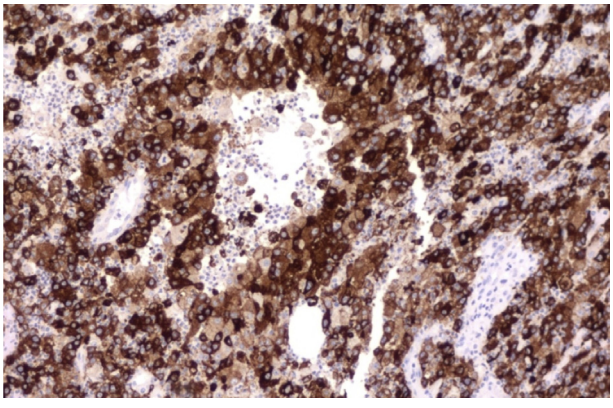

Fig. 4. Immunohistochemical staining of the tumor with an antibody against epithelial membrane antigen (100× original magnification).