Gigantic enlargement of the thymus gland

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ABSTRACT

True massive thymic hyperplasia is a very rare entity, characterized by an increase in the size and weight of the thymus gland, without an apparent cause. Surgery has been required in patients with severe respiratory distress. We present an idiopathic true massive thymic hyperplasia in a 5-month-old boy.

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True massive thymic hyperplasia is a very rare entity, characterized by an increase in the size and weight of the thymus gland, without an apparent cause. The gland is enlarged without disruption of the normal architecture or any pattern of abnormal cellular proliferation. The majority of the cases occur between the ages of 1-15 years, and the next common age group is one year of age or younger. Most of the patients are symptomatic, and can mimic other important diseases, including lymphofollicular hyperplasia, thymoma, lymphoma, and germ cell tumors. Separating these entities has required fine needle aspiration or biopsy. Surgery has been required in patients with severe respiratory distress. We are presenting this case to draw the attention of the physicians to this rare presentation of the true thymic hyperplasia, which is frequently seen in the pediatric age group, and to focus on the role of surgery.

Case Report. A 5-month-old boy presented with respiratory distress progressive in course, and not responding to medications. Initial radiographic imaging showed a huge anterior mediastinal mass extending into, and occupying most of the right hemithorax (**Figure 1**), with marked contralateral mediastinal shift. Percutaneous needle biopsy was carried out to exclude lymphoma, and revealed normal thymic tissue. Within a week, the mass has increased its size significantly, and he became more distressed. A computed tomography was repeated and showed that the mass had already filled both thoracic cavities with bilateral marked lung collapse (**Figures 2a & 2b**). Thymectomy was performed through a median sternotomy, and the thymus was found compressing the surrounding structures, but without any infiltrations. The thymus gland was soft, lobulated, well-encapsulated, homogenous, and weighed 380 grams (gms). The operation and postoperative course were uneventful. He was extubated at the end of the procedure, shifted to the ward after 24 hours, and was discharged from the hospital on the 5th postoperative day. Histopathologic examination confirmed true thymic hyperplasia. Two years later, he is doing well without recurrence (**Figure 3**).

Discussion. Levine and Rosai,[1] in 1978, separated thymic hyperplasia into 2 categories based on gross and histologic criteria. The first is lymphoid or follicular hyperplasia, characterized by the presence of lymphoid follicles with activated germinal centers. The second is true hyperplasia, characterized by an increase in the size and weight of the gland, which retains normal morphology and microscopic architecture for the patient's age.[1,2] Hyperplasia of this order is not known to occur in any other organ.[3] True hyperplasia occurs in 3 different clinicopathologic forms, a true thymic hyperplasia without any other disease is extremely rare. Enlargement of the thymus gland is reported as a form of rebound phenomenon in a number of conditions like recovery from severe stress situations, after administration of steroids, and after treatment of malignant tumors. Finally, thymic hyperplasia has been described in association with endocrine abnormalities, sarcoidosis, and Beckwith-Wiedman syndrome.[2,4-6]

Idiopathic true thymic hyperplasia is a very rare condition, characterized by marked to massive enlargement of the thymus gland, with no apparent cause; fewer than 50 cases have been recorded. The majority of

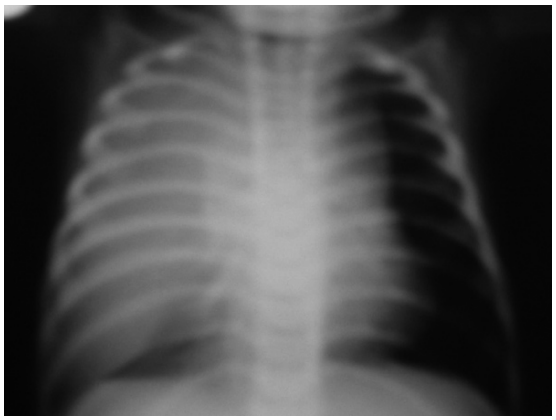

Figure 1 - Preoperative chest x-ray showing huge anterior mediastinal mass extending into, and occupying the right hemithorax.

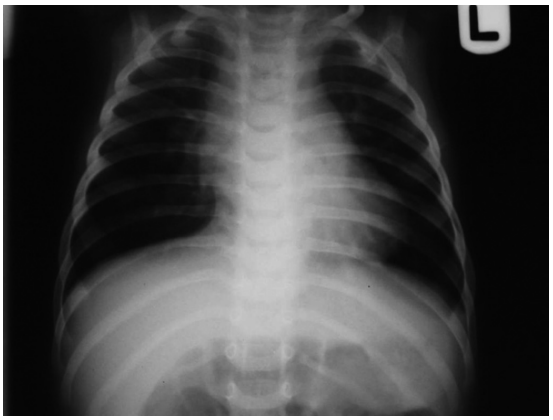

Figure 3 - Repeat chest x-ray 24 months after thymectomy.

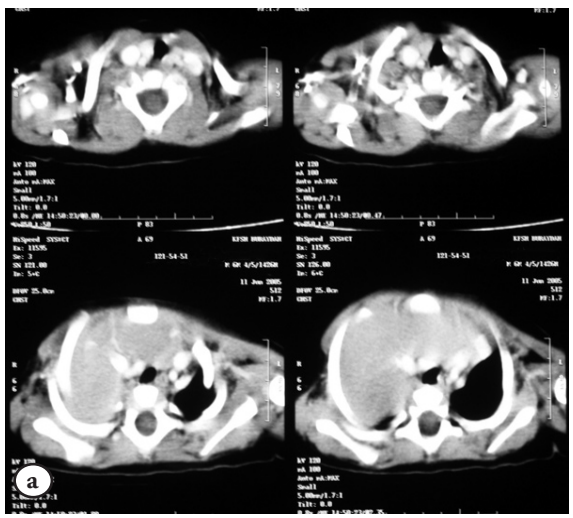

Figure 2 - Preoperative computed tomography of chest showing **a**) mass-filled thoracic cavities, and **b**) bilateral lung collapse.

the cases occur between the ages of 1-15 years. The next common age group is one year of age or younger, and is very rare after the age of 15 years. Most of the cases are symptomatic (85%); cough, dyspnea, respiratory distress, and respiratory tract infections are the most common symptoms.[4] This clinical presentation can mimic other important diseases, including thymomas, lymphomas, and germ cell tumors. Separating these entities has required fine needle aspiration or biopsy.[7]

By reviewing our own experience in the last 15 years, we have seen many cases of asymptomatic or mildly symptomatic thymic hyperplasia in newborns, infants, and older children. They all required symptomatic treatment, re-assurance, and follow-up. This was the first case to have a stormy presentation and a very rapid rate of growth of the thymus, with no response to medications including steroids. Fine needle aspiration was carried out and confirmed the diagnosis, followed by rapid surgical removal of the gland. Surgery is necessary to relieve symptoms of mediastinal compression in rapidly growing tumors and for histologic analysis. It could be accomplished via a median sternotomy, a clamshell incision or posterolateral thoracotomy.[3,4,8] In the new born, the gland reaches a mean weight of 15 gms. In the early neonatal period, the gland reaches its largest relative size and continues to grow until puberty to a mean weight of 30 - 40 gms. It has been reported that the gland weighed 8 - 10 times, or more of the normal weight.[8] In our patient, the removed thymus weighed 380 gms, had a smooth surface with no infiltrations of the surrounding structures. Microscopically, there was no infiltration with any type of cells. The operation is curative, and no subsequent recurrence has been recorded after complete excision.

In conclusion, idiopathic true thymic hyperplasia is a very rare disease occurring without any cause, and affects children between 1-15 years of age. It commonly

presents with manifestations of respiratory distress, and needs symptomatic treatment and reassurance. Rarely, the mediastinal compression symptoms are so severe that they require surgical excision.

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