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Full Length Case Study

DETECTION OF AN ANTERIOR MEDIASTINAL MASS BY AUTOPSY AND ITS DIAGNOSIS AS THYMIC LYMPHOMA- AN EXTREMELY RARE CASE

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ABSTRACT

The diversity of pathologic processes that may reside in a small anatomic compartment like mediastinum is impressive. The lesions may be neoplastic and non-neoplastic; and may include proliferation of somatic epithelial, lymphoid, mesenchymal and germ cell types. Anterior mediastinum is the preferred site for mediastinal masses, of which 40% or more are of thymic origin. Again approx. 90% of all thymic masses are in the anterior mediastinum. About 40-50% of patients having anterior mediastinal mass are asymptomatic and when symptoms appear, those are mostly due to structural compression and displacement. These neoplasias are most common in the fifth and sixth decades of life and distributed evenly between men and women. In the present case study autopsy was performed in a case of sudden death, which had history of respiratory distress for some time and was being treated elsewhere by quacks. A huge anterior mediastinal growth was found on autopsy and diagnosis was confirmed by histopathology and immunohistochemistry.

INTRODUCTION

Very often natural deaths form the basis of medicolegal investigations, if they have occurred suddenly, in apparently healthy persons. Autopsy of cases of death due to natural causes is a challenging task to an autopsy surgeon, not only due to the fact that they sometimes present with multitude of pathological findings but also due to totally unexpected outcomes that open up to the utter surprise of the autopsy surgeon. Respiratory distress in a young adult is not an uncommon presentation in any emergency health care setup but detection of a huge anterior mediastinal mass by autopsy in such a clinical presentation is a rare finding in Forensic Pathology. In the present case study it has been attempted to present an extremely rare autopsy finding of huge anterior mediastinal mass, leading to unexplained death of a young adult male, and its subsequent diagnosis with the help of ancillary investigations.

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Case History

A 21 years old male patient was brought to the casualty department of NRS Medical College, Kolkata, in the late evening hours of mid- December, 2011 with complaint of severe Respiratory Distress for last few hours. He had history of Respiratory Distress for last 4-5 months and was being treated elsewhere by non-qualified practitioners in a remote village of a neighbouring state. The patient succumbed before any diagnosis could be made. Autopsy Examination was done to ascertain the cause of death in the said case of unexplained, sudden death and a huge Anterior Mediastinal Growth was found. Tissue from the growth was sent for Histopathology and Immunohistochemistry. The final Diagnosis was Thymic Lymphoma.

Salient Autopsy Findings

Naked Eye Findings

1) A greyish white, solid, compact mass measuring 12 inches x 8 inches, inside the Thoracic Cavity, occupying the anterior mediastinum, more towards left side of the

midline, intimately adhered to the Sternum in its posterior aspect, Pericardium and Left Pleura.

2) Heart was normal in size and shape weighing 250 gms and was found encased within the growth
3) Left Lung was collapsed, weighing 200 gms.
4) Right Lung was congested, oedematous, weighing 700 gms.
5) Other viscera were found congested and rest of the cavities was unremarkable.

Fig. 1. The growth as taken out from thoracic cavity

Histopathology

Sections showed a growth comprising of small rounded cells with round nuclei, at places packed in solid trabecular and nesting pattern.

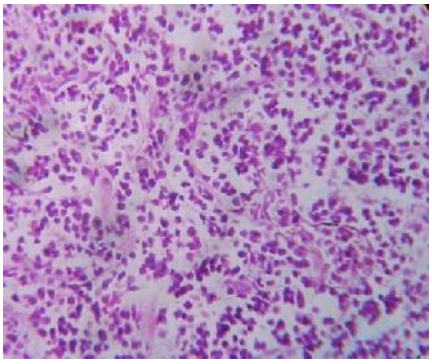

Fig. H/P 40X

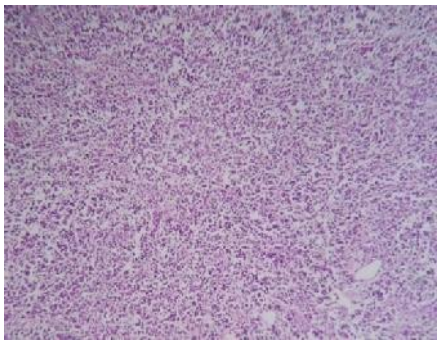

Fig. H/P 10X

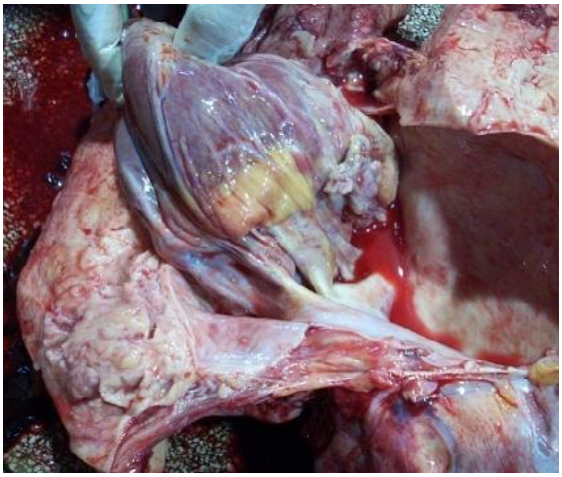

Fig.1a. The groth was adherent to the pericardium

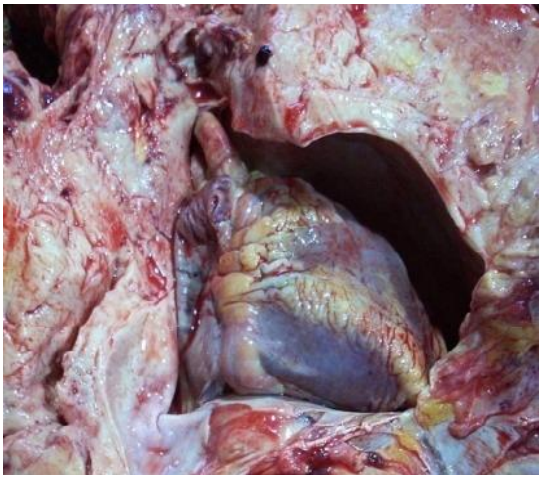

Fig. 2. The heart was found encased within the growth

Immunohistochemistry

Chromogranin -	Negative
NSE -	Negative
CD 45 -	Positive
CD 20 -	Positive
EMA -	Negative
PAN CK -	Negative
CD 5, CD 3, CD 21 -	Negative
BCL 2 -	Negative

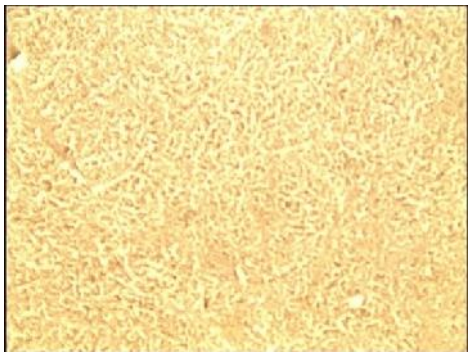

Fig. Immunohistochemistry CD4510X

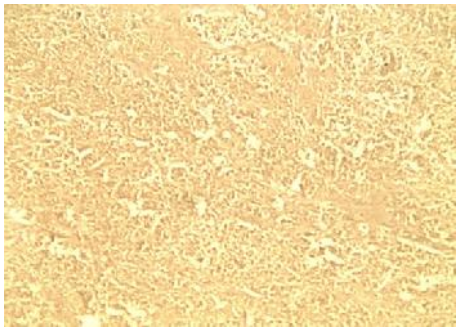

Fig. 7. Immunohistochemistry CD2010X

Diagnosis

Considering the naked eye findings, Histopathological findings and results of Immunohistochemistry all together, it can be opined that the features are suggestive of "THYMIC LYMPHOMA."

DISCUSSION

The mediastinum is the portion of the thoracic cavity located between the pleural cavities, extending anteroposteriorly from the posterior aspect of the sternum to the spine and sagitally from the thoracic inlet to the diaphragm.

The numerous organs and anatomical structures, it contains, make it a "Pandora's Box". An arbitrary division of the mediastinum into Superior, Anterior, Middle and Posterior compartments has proved useful since most cysts and neoplasms have a predilection for one compartment over the other (Rosai J. Rosai, 2012). The anterior mediastinal compartment is that which is ventral to the anterior cardiac border and the aortic root; it most commonly harbours thymic epithelial tumours and cysts, germ cell neoplasms, lymphoproliferative epithelial lesions, retrosternal thyroid glandular proliferations, parathyroid lesions, malignant lymphomas, aorticopulmonary type paragangliomas and non-neurogenic mesenchymal tumours. (Rosai J. Rosai, 2012; Mills, 2010). The Thymus is a lymphoepithelial organ, occupying a small place in the anterior mediastinum, consisting of two lobes that are divided into lobules that form the basis of anatomic units of thymus. The thymus is covered by a fibrous capsule from which fibrous bands (trabeculae) penetrate the parenchyma, dividing it into lobules. Histologically the lobules have two distinct regions. The peripheral region-which is called the cortex, is divided into the outermost or subcapsular cortex and inner or deep cortex. The central region is called the medulla. On H/E stained sections the cortex appears dark blue to purple because of the predominance of lymphocytes (80-85%); whereas the medulla appears eosinophilic because of predominance of epithelial cells (Greer *et al.,* 2009).

The two major cell types of thymus are endodermally derived epithelial cells and bone marrow derived lymphocytes. Thymic lymphocytes have a T-cell phenotype-a whole range of differentiation exists among them; the better defined stages being those of subcapsular thymocyte, medullary thymocyte and mature T-lymphocytes. Other cells normally present in the thymus include B-cells, interdigitating reticulum cells, Langerhan's cells, mast cells, eosinophilic and the usual non specific type of stromal cells (Rosai J. Rosai, 2012; Greer *et al.*, 2009; Levine and Rosai, 1978; Muller-Hermelink *et al.*,1986; Kumar *et al.*, 2010).

The Thymus undergoes normal involution after puberty although it never disappears completely. Islands of thymic tissue are consistently found on microscopic examination of the pericardial fat and sometimes also in the retrosternal fat, following autopsy in different age groups (Dimaio and Dimaio, 2001). Thymoma is the most common cause of an anterior mediastinal mass in adults. The other major causes are lymphomas (2nd most common), germ cell tumours etc. After combination chemotherapy for another malignancy, teenagers and young adults may develop a rebound thymic hyperplasia in the first few months after treatment. About 40-50% of patients of anterior mediastinal mass are asymptomatic; masses are detected incidentally on routine chest radiographs. Symptomatic patients may have cough, chest pain, respiratory distress, fever, wheezing, fatigue, weight loss, night sweats and occasionally features of obstruction of Superior Vena Cava. About 30% of patients with thymoma have Myasthenia Gravis (Longo *et al.,* 2011). Detection of an anterior mediastinal mass by autopsy, either medicolegal or pathological, is not a common happening. The same can be said about Thymic mass also. Though cases wherein anterior mediastinal mass is detected by autopsy, have been reported sporadically in literatures; they are previously diagnosed with the help of radiological investigations (chest radiograph, CT Scan etc.).

Again the incidence of such autopsy finding is unknown (not found in standard text books, journals and scholarly articles). 'Lymphoma' is an umbrella term that loosely refers to several dozen independent categorical types and subtypes of cancers of the lymphatic system. Thymic lymphoma, better known as Primary Mediastinal (B-cell) Lymphoma, is one of the subtypes of lymphoma, develops from the lymphocytes in the thymus. Hodgkin's disease of the thymus is also a well recognized entity and it is known to spread locally to lung parenchyma, mediastinal structures, and the sternum (Steiner, 1943; Marshall and Wood, 1957; Fechner, 1969; Keller and Castleman, 1974). Intrathoracic Hodgkin's disease involving the chest wall has also been reported (Goldman, 1971; Eiser and Samarrai, 1975).

Primary mediastinal lymphoma is an uncommon condition. It constituted 2.4 percent of 1,403 cases reviewed by the Non-Hodgkin's Lymphoma Classification project. Distribution is worldwide, with documented series reported from the United States, Europe, and the Far East. The experienced pathologist can often suspect the diagnosis when presented with an invasive but localized primary neoplasm of the mediastinum in a young individual (15 to 45 years old), with female preponderance (M:F-1:2). The typical location (anterior mediastinum), the huge size, the typical histopathological appearance supplemented with immunophenotyping (these cells uniformly exhibit CD45- the leukocyte common antigen and CD20 and also noteworthy for regular absence of CD21) confirm the diagnosis (Johnson and Davies, 2008).

Conclusion

- Autopsy of a case of Sudden, Unexplained Natural death is basically a challenge for the Autopsy Surgeon.
- In rare cases histopathology and other modern ancillary investigations may give us surprising results.

- In Autopsy cases of death due to diseased condition, histopathological examination may be suggested as compulsory.

Conflict of Interest

This article was not sponsored by anyone and was done exclusively by the authors with their own resource and interest.

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