Case Report

Thymoma in a child- A case report

Meena Patil*, Vaishali Aphale**, Sujit Joshi***

Dept. of Pathology, B.V.D.U.Medical college, Pune, Maharashtra, India.

*Assistant Professor. ** Associate Professor, *** Assistant Professor
Correspondance: E-mail meenipatil@yahoo.co.in

Abstract

Thymomas are neoplasms arising from thymic epithelial cell, regardless of the presence or number of lymphocytes. Thymoma is an uncommon & slow growing neoplasm. Thymoma comprise about 20-30% of mediastinal mass in adults but only about 1% in pediatric patients. Patients usually present with mass-associated respiratory symptoms, superior vena cava syndrome, or paraneoplastic syndrome including myasthenia gravis. We report a case of thymoma in a two year old female child presented with breathlessness & upper respiratory tract infection for 10-15 days. Due to the limited no. of cases, knowledge & experience with thymoma in pediatric patients, the diagnosis & treatment are very challenging for this age group.

Key words: Mediastinal, Epithelial cells, Paraneoplastic syndrome, Myasthenia gravis, Pediatric.

Introduction

Primary thymic lesions such as thymic cysts, hyperplasia, carcinoma & thymomas comprise approximately 2-3% of all pediatric mediastinal tumors (Grosefeld 1994). The usual location of thymoma is anteosuperior mediastinum & may be discovered incidently by chest radiography during routine examination. They can produce symptoms as cough, dyspnoea, palpitation, & substernal or interscapular pain. A no. of paraneoplastic syndromes may be associated with thymoma. Classically, thymomas are encapsulated, circumscribed, lobulated neoplasms. Thymomas are comprised of varying proportions of epithelial celss & lymphocytes. However the epithelial cells are the only neoplastic elements in such tumors[2]. Thymoma can generally be categorised as non-invasive(stage I) and invasive (stage II-IV) (Masaoka et al 1981). Treatment of thymoma includes Surgery, irradiation and chemotherapy[3].

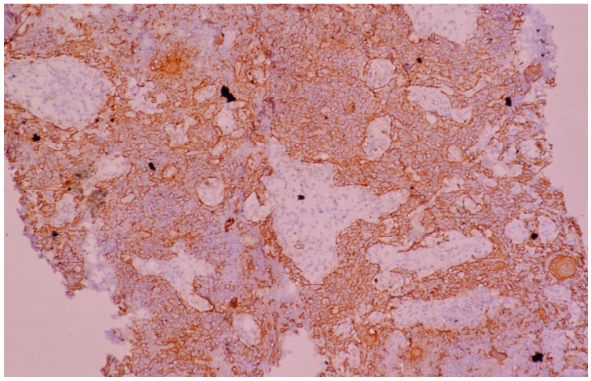
Case Report

We presented a case of one year old female child presented with complains of cough & coryza on & off since one month, breathlessness since 8 days and H/o fever since 2-3 days. Investigations were done. X-ray chest- Showed gross widening of the mediastinum with homogenous opacification predominantly involving superior mediastinum s/o Lymphoma or thymic mass. USG Chest:- Large solid anterior mediastinal mass s/o lymph nodal/ Thymic mass. MSCT Thorax with contrast:- Large, well-defined, isodense, diffusely enhancing anterior mediastinal mass extending into the middle mediastinum s/o neoplastic etiology such as lymphoma/ Thymic mass

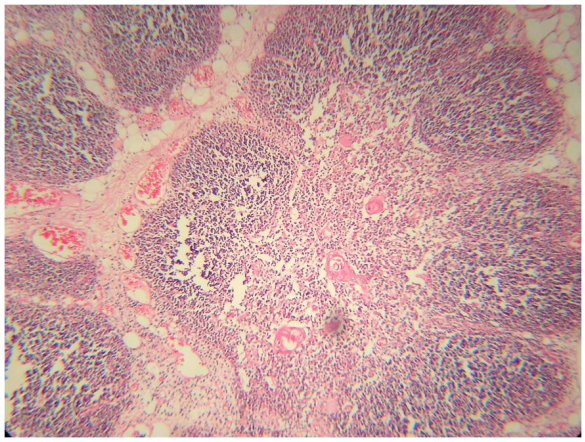
Serum Alfafetoprotein (AFP)- was normal which rule out seminoma. CT- guided biopsy of anterior mediastinal mass was done. On histopathology impression was given as S/o Thymic mass with D/D- 1.Thymoma (cortical type), 2. Lymphoma

IHC was done & it showed positivity for CK, CD3, Tdt, & LCA. Excision of anterior mediastinal mass

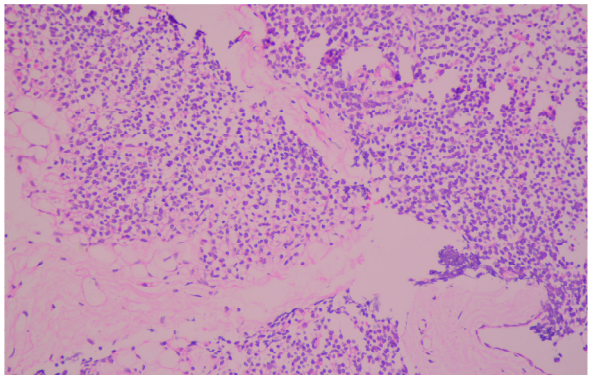
was done by right thoracotomy. We received a single, encapsulated tumor mass measuring 13X7X3 cm. External surface was smooth, lobulated & at places congested. Cut surface was homogenous, grey-white. Consistency was soft to firm. Microscopically an encapsulated thymus tissue comprised of many lobules of variable size separated by thin fibrous septae were seen. Each lobule was rich in lymphocytes surrounded by thin layer of epithelial cells. At places congested, dilated blood vessels & Hassals corpuscles were also noted. No evidence of necrosis/atypia/ or capsular invasion was noted. Diagnosis was offered as Cortical Thymoma(Type B1) with no evidence of malignancy.



CK1

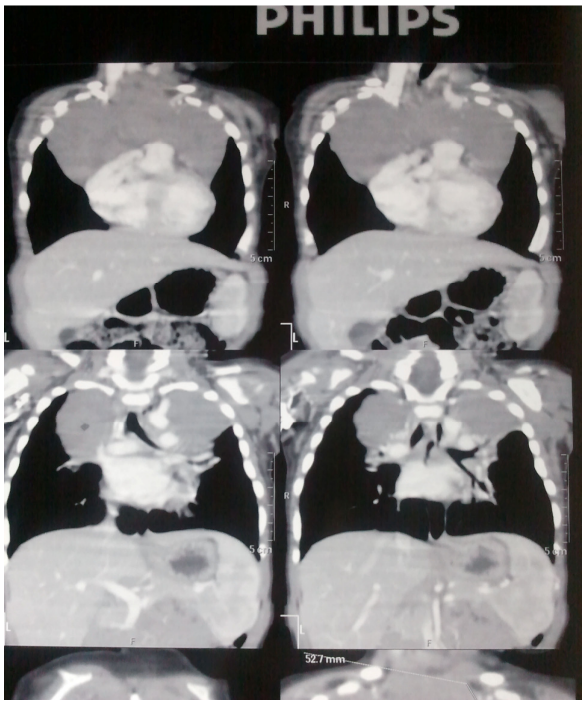


H&EX100 THYMOMA



HE1

www.ijbamr.com P ISSN: 2250-284X E ISSN: 2250-2858



MS CT THORAX THYMOMA

Discussion

Thymomas are neoplasms of thymic epithelial cells, regardless of the presence of relative no. of non-neoplastic lymphocytes[4]. Nearly all thymomas present in adult life. Thymomas in children are exceptional. Therefore, it is important to recognise this tumor as distinct from others like mediastinal teratoma, lymphoma, malignant histiocytoma & ewing sarcoma/PNET. Thymoma and thymic carcinoma are uncommon tumors with an annual incidence of approximately 1-5 per million population. The usual location of thymoma is the anterosuperior mediastinum; however it can also be seen in other mediastinal compartment, in the neck, within the thyroid, pericardial cavity, pulmonary hilum, within lung parenchyma or in pleura itself[5].

The etiology of thymoma is still largely unknown. They have been repeatedly observed in patients with paraneoplastic syndromes as myasthenia gravis, pure red cell aplsia, acquired hypogammaglobulinemia etc. Thymomas are usually discovered incidentally by chest radiography during routine medical examination, as lobulated shadow that may be calcified. They can also produce symptoms such as cough, dyspnoea, palpitation & substernal or interscapular pain[2]. Grossly, the typical thymoma is well – encapsulated, solid, yellowish gray & lobulated. Foci of necrosis, cystic degeneration are common, particularly among larger tumors[5].

Microscopically, the majority of thymomas are composed of a mixture of neoplastic epithelial cells & non-neoplastic lymphocytes. The epithelial cells may have a round-polygonal (plump), stellate or spindal shape with vesicular nuclei. The lymphocytes may appear mature(inactive) or show varying degrees of activation[5].

There has been much controversy concerning the best choice for the classification of thymic tumors. Among the two most influential proposals, the first is descriptive scheme originaly proposed by Lattes et al and adopted by Bernatz et al. The The second scheme, which is based on morphofunctional

www.ijbamr.com P ISSN: 2250-284X E ISSN: 2250-2858

principles, is of Muller-Hermelink & his co-workers[5].

The WHO Committee for the histologic typing of thymic tumors was opted as new scheme. Thymomas are divided into two major types depending on whether the neoplastic spindle/oval shape epithelial cells, designated as type A; or whether these cells have a dendritic or plump (epithelioid) appearance, designated as type B. Type B thymomas are further subdivided on the basis of the proportional increase (in relation to the lymphocytes) & emergence of atypia of the neoplastic epithelial cells into 3 subtypes, designated as B1, B2, B3 [5].

Traditionally thymomas of any type (other than thymic Ca) that appear totally encapsulated have been regarded as benign; whereas evidence of aggressiveness in the form of local invasion, pleural or pericardial implant or distant metastasis have been designated as malignant[5].

Clinical staging system was proposed by Masaoka et al for thymoma in 1981. In turn, this scheme has been incorporated with some modification into TNM staging system proposed by Yamakawa et al[5].

Type B1 thymoma (Lymphocyte rich thymoma) is distinguishable from the normal non-involved thymus based on architectural diff, including large excess of cortical areas compared to small areas resembling the thymic medulla, fewer Hassall corpuscules, less regular lobulations & a thick fibrous capsule or irregular fibrous septa. B1 thymoma with a high predominance of T lymphocytes may simulate T lymphoblastic lymphoma. An infiltration of lymphocytes into septa & capsule would favour lymphoma. The presence of prominent ck meshwork & low cdk expression in T lymphocytes favours B1 thymoma. Lymphocyte-rich thymoma also distinguished from lymphoid hyperplasia. In lymphoid hyperplasia normal architecture of thymus is maintained. In contrast, in type B1 thymoma there is effacement of most or all of the thymic structure[5].

Other differential diagnosis for thymoma are large cell lymphoma, thymic Hodgkin lymphoma, thymic seminoma, thymic carcinoid which are seen in anterior mediastinum. These are distinguished on the basis of histological pattern and immunohistochemistry. Thymoma is positive for keratin. Large cell lymphoma is positive for B-lymphocyte markers, Hodgkin lymphoma positive for CD 15 & CD 30, seminoma positive for PLAP, CD117 & carcinoid for chromogranin & synaptophysin[5].

Prognosis of thymoma is dependent on tumor stage, WHO histologic type & completness of resection. Type A & AB thymoma in stages I & II always follow a favourable clinical source. They are considered benign tumor of low malignant potential. Type B1 thymoma have a very low malignant potential, rare local recurrences or late metastases may occur. Type B2 & B3 thymoma & thymic carcinoma are clear cut malignant tumors. The prognosis of combined thymomas may be determined by the most malignant component. Paraneoplastic pure red cell aplasia, other cytopenias or hypogammaglobinemia have an adverse effect; whereas paraneoplastic myasthenia gravis had no or positive factor on survival[5].

The primary treatment of thymoma is surgical excision. For the entirely encapsulated thymomas that have been removed in toto, no additional therapy is necessary; regardless of their microscopic type. Thymoma associated with gross invasion or implants, excision should be supplemented with radiation therapy[5].

In our case type B1 thymoma was differentiated from lymphoma and seminoma. Immunohistochemistry for epithelial markers was positive which ruled out lymphoma. Serum

www.ijbamr.com P ISSN: 2250-284X E ISSN: 2250-2858

alfafetoprotein level was normal which ruled out seminoma. As it was encapsulated tumor total excision was done in toto. As it is type B1 thymoma it has low malignant potential. No additional therapy was given and patient has advised follow up.

References:

1. David H. Rothstein, Stephen D. Voss, Michael Isakoff, Mark Puder. Thymoma in a child:case report and review of literature. Pediatric Surgery International, July 2005, Volume 21, Issue 7, pp 548-551.
2. Christopher M. Flecher. Tumors of Lymphoreticular System:Thymoma. Diagnostic Histopathology of Tumors, 3rd ed., vol.2, 2007, p.1317-31.
3. Hamidah A, Poulsaeman V, Suria AA, Zarina AL, Zulfiqar MA, Jamal R. Thymoma:A case report and Review of Literature. Med & Health 2010; 5(1):49-54
4. William D. Travis, Elizabeth B., H. Konard Muller-Hermelink, Curtis C. Harris. Thymomas. WHO Classification of Tumors-pathology & Genetics- Tumors of the Lung, Pleura, Thymus & Heart. Lyon, France, 2003.
5. Juon Rosai. Mediastinum-Thymoma. Rosai & Ackerman's Surgical Pathology, Vol.1, 9th ed., 2005, p.466-481

Date of submission: 23 March 2013
Date of Provisional acceptance: 30 March 2013
Date of Final acceptance: 28 April 2013
Date of Publication: 03 June 2013
Source of Support: Nil ; Conflict of Interest: Nil