Case Report

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Surgery Section

Giant Thymoma from Ectopic Thymic Tissue in Children: A Case Report and Review of Literature

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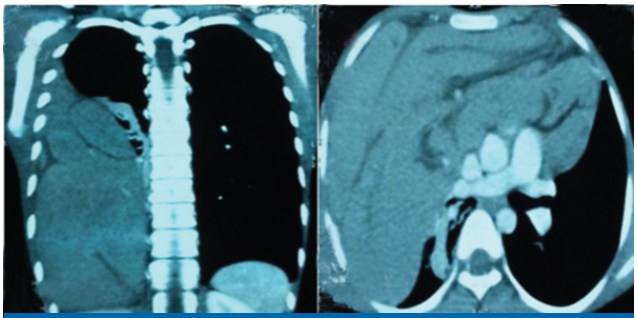
ABSTRACT

Thymoma is a rare tumor in paediatric age group. It is a locally invasive slow growing tumor with occasional occurrence of extra thoracic metastasis. The report of incidence of thymoma from ectopic thymic tissue is extremely rare and to the best of our knowledge, no case yet has been reported on a giant thymoma arising from ectopic thymic tissue in paediatric age group. We present a case of giant thymoma originating from ectopic thymic tissue in a young girl. A 13 years old female child had complaints of failure to thrive and she was found to have a large mass in the right hemithorax with encasement of mediastinal vessels in investigations. Intraoperatively, well encapsulated mass with size of 20 × 30× 15 cm was resected completely through a lower right fourth space "trap door" thoracotomy incision. Histology confirmed the tumor as World Health Organization type B2 thymoma with heavy proliferation of lymphocytes and in Masaoka stage I. The post operative course of the patient was uneventful. The rarity of it among paediatric patient makes the management challenging.

Keywords: Mediastinal mass, Thymectomy, Thymic Carcinoma

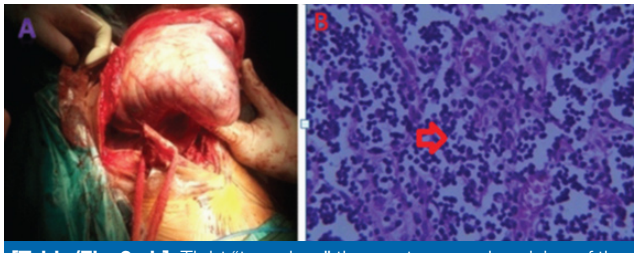
CASE REPORT

A 13-year-old girl was evaluated for failure to gain weight and a mediastinal mass was detected on routine chest X-ray. There was no history of cough, chest pain, fever or hemoptysis. There was decreased in breath sounds on right chest. Her body weight was 20 kg. Computerized tomography (CT) of the thorax [Table/Fig-1a,1b] showed a large well demarcated lesion occupying lower two third of the right hemithorax, anterior mediastinum with encasement of mediastinal vessels and compression obliteration of the right lower and middle lobe bronchus. There was no evidence of tumour infiltration to the surrounding structures. The CT-guided percutaneous biopsy was inconclusive.



[Table/Fig-1a,b]: CT thorax shows a welldefined massin right hemithorax withmultiple septations (a). Extension of the mass to contralateral hemithorax with encasement of aorta andpulmonary arteries (b).

A neurogenic tumour, ectopic thymoma or thymic cyst kept in mind as differential diagnosis. Due consent was taken and surgical excision of the mass was proceeded with median sternotomy. The tumour was massive [Table/Fig-2a] and for complete extracapsular excision it required extension of the incision in toright chest through 4th intercostal space. The mass was well circumscribed andbased towards right mediastinal pleura. It was encasing partly the innominate vein and a portion of the superior vena cava. It did not invade any of the mediastinal structure. The feeding vessels to the tumor were from right internal mammary artery. There was no mediastinal lymphadenopathy, pleural effusion or pleural



[Table/Fig-2a,b]: Tight "trap door" thoracotomy and excision of the mass (a). Scattered plump cells, vesicular nuclei and distinct nucleoli with proliferation of lymphocytes (b).

based nodule. The tumour was resected in toto. It weighted 2200 grams and 20 x 30 x 15 cm in size. Gross examination revealed multiple nodular mass with intact capsule. The cut section had fish flesh appearance. Microscopy revealed a neoplastic epithelial component with proliferation of scattered plump cells with vesicular nuclei and distinct nucleoli along with heavy proliferation of lymphocytes ([Table/Fig-2b] red arrow). The feature was consistent with type B2 thymoma according to WHO Schema for the Classification of Thymic Epithelial Neoplasms (1999, 2004). There was no transcapsular invasion of tumour cell suggesting Masaoka stage-I thymoma. The patient had uneventful recovery. Adjuvant treatment was not considered as complete resection was done and histology was favorable. She is asymptomatic after one year of follow- up.

DISCUSSION

Thymoma is an epithelial neoplasm of thymic gland [1]. It is exceedingly uncommon in children and accounts for approximately 1.5 to 4% of pediatric mediastinal tumors [2,3]. Thymomas are slow-growing tumors and usual local extension is confined to the pleura, pericardium, or diaphragm. The occurrence of extrathoracic metastases is uncommon [4].The incidence of thymoma arising from ectopic thymic tissue is about 4% [5]. Complete surgical resection gives best prognosis and late recurrence is rare [6].

Most mediastinal neoplasm in paediatric patients are lymphomas (40%) or neurogenic (33%) [7]. Thymomas are extremely rare in children. Furman and colleague identified only 1 thymoma from 4712 cases of childhood neoplasm [8]. On review of 30 years (year 1979 to 2008) of English literature by Liang et al., only 30 cases were reported of thymoma in patients less than 18 years of age [7]. The incidence among males and female were similar. The aberrant thymic nodules present in neck, skull, mediastinum and at the bronchus. There are only a few reports about lung and pleural based giant thymic tumors [5]. To the best of our knowledge, no case yet has been reported on a giant thymoma arising from ectopic thymic tissue in paediatric age group. Thymoma patients are usually asymptomatic. Symptoms include cough, dyspnoea, dysphagia, hoarseness or para neoplastic syndrome [7,2]. Approximately 15% of patients with myasthenia gravis (MG) have thymoma, while approximately 35% of patients with thymoma have MG [1,8]. However, this association is about 13% in young population [7]. These tumors are typically aggressive with poor outcomes. As many as 80% of thymomas are identified by routine chest radiographs [5]. Thymomas present on the CT scan as a spherical or ovoid, smooth, anterior mediastinal mass [9]. The tumor enhances homogenously. It may be heterogeneous or even cystic because of areas of hemorrhage and necrosis. The tumor may contain coarse or curvilinear calcifications. Intravenous contrast is necessary for staging of thymoma. The features of vascular involvement include an irregular luminal contour, soft tissue and vascular encasement [5,9]. Ipsilateral pleural nodules are suggestive of regional metastasis.

The accuracies of fine-needle aspiration (FNA) is varies from 77 to 100%. Immunohistochemical stains can aid in the identification of the epithelial and lymphocytic components of thymoma [10,11]. The identification of the epithelial and lymphocytic is aided by immunohistochemical stains. Resection of an anterior mediastinal mass can be planned without a preoperative biopsy when peripheral lymph nodes are not found on a thorough clinical and radiographical evaluation.

Various attempts have been made to classify the histological subtypes of thymoma [1,4]. The tumors with true malignant cytologic characteristics are considered thymic carcinomas and similarly an invasive thymoma is refers to a malignant thymoma. The cortical type tumors tend to have a less favorable prognosis than the medullary type. Bergh et al., did propose a 3-tiered staging system [1,11]. The original 4-tiered Masaoka staging system was proposed in 1981, and a modification of this classification was done by Koga [1,11,12].

It is a postoperative pathologic staging. The stage I tumors are completely encapsulated. Stage II tumors violate the capsule, either grossly or histologically. In stage IIA the thymoma is growing into the capsule and in IIB the tumor has grown through the capsule into surrounding tissue. Stage III tumors have obvious invasion into contiguous structures. Stage IV tumors have regional or distant metastases.TNM system was proposed in 1991 in which the original Masaoka system for the T descriptor and grouped any N or M-positive tumor into stage IVb (pleural or pericardial nodules alone are T4, stage IVa) [13]. Modifications of this ere proposed by Tsuchiya in 1994 and the WHO in 2004. In WHO classification, the thymic epithelial neoplasms divides into 3 categories, types A, B and C.1 Basically two major types of thymoma were identified, depending on whether the neoplastic epithelial cells and their nuclei showed a spindle or oval shape (type A) or a round epithelioid appearance (type B). Presence of these 2 cell types were designated type AB tumor.

B1, B2, and B3 subdivision of type B thymoma was made on the basis of the proportional increase (in relation to the lymphocytes) and emergence of atypia of the neoplastic epithelial cells. Tumor showing overt cytologic features of malignancy is regarded as type C thymoma. Complete surgical resection is the current recommendation for stage I thymoma. Postoperative adjuvant radiation therapy is recommended for incompletely resected stage II and III thymoma [1,7]. Thymoma of stage III-IVa should have surgery when possible and postoperative adjuvant radiation therapy is recommended, perioperative chemotherapy may be given to cases of incomplete resection. Non resectable stage III-

IVa tumors should be treated with chemotherapy, followed by surgery or radiation therapy. The stage IVb thymoma should receive chemotherapy only. Completeness of resection is the most significant prognostic factor in thymoma [14,7]. Those of stage III-IVa, which is not resectable at presentation, should be treated with multi model therapy including chemotherapy, followed by surgery or radiation therapy. The stage IVb thymoma should receive chemotherapy. Completeness of resection is the most significant prognostic factor in thymoma [14,7].

Interestingly, resected giant thymomas tend to be of low-grade thymomas. The average five and ten-year overall survival rates of 92 and 88 percent, respectively, in stage I thymic neoplasms when they undergo complete resection. The recurrence rate is 1% [14,15].

CONCLUSION

Thymoma from ectopic thymic tissue is entirely rare in children and the diagnosis is challenging. Male sex and younger children presents in advanced tumour stage. Complete surgical resection gives best prognosis and must be considered regardless of tumour size if curative resection is possible.

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