Thymic Carcinoma-A Rare Case Report

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ABSTRACT:

Thymic carcinoma is a rare malignant neoplasm of the thymic epithelium. It is differentiated from invasive thymoma by the presence of malignant cytological features and higher incidence of local invasion and embolic metastasis. They constitute a heterogenous group of tumors that present differently in terms of both behaviour and prognosis. Here we present a case of thymic carcinoma in a 24 years old male whose chief complaining were common cold with cough for 3 months.

All the radiological, biochemical examinations along with surgical biopsy with histopathological examination and IHC study was done to reach the definite diagnosis. CECT of the thorax suggest large predominantly cystic anterior mediastinal mass. The final histopathological diagnosis was thymic carcinoma.

KEY WORDS: Thymic carcinoma, mediastinal mass, embolic metastasis, common cold

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I.Introduction

Thymus is an anterior mediastinal lymphoid organ that is mainly composed of epithelial cells in outer cortex ,lymphocytes in the germinal centre(1,2). The thymus is responsible for the processing and maturation of T- lymphocytes(3). Thymic carcinoma is a rare and invasive mediastinal neoplasm that often shows capsular invasion and metastasis. Thymic carcinoma have been reported to account for only 0.06% of all thymic neoplasms(4). The histopathological classification of thymic carcinoma was proposed by Levine and Rosai(5). Histologically different variants of thymic carcinoma described are squamous cell carcinoma, basaloid carcinoma, clear cell carcinoma(6). Clinical symptoms and signs are due to anterior mediastinal mass effects include coughing, chest pain and signs of upper respiratory airway congestion (7).

II.Case report

A 24 years old male was admitted to IPGMER and SSKM Hospital with history of cough and cold for 3 months. He was referred from a district hospital in West Bengal .X ray and CT scan was done. CT thorax suggested fairly large predominantly cystic anterior mediastinal mass lesion compressing left lung (Figure 1). Operation was done and mediastinal mass was sent to Department of Pathology of SSKM for histopathological examination.



Figure 1

(A) X-Ray shows lobulated left cardio-mediastinal contour, lungs are clear, no bony destructive bony lesion.
(B) Computed Tomography (CT) reveals a cystic anterior mediastinal mass compressing left lung

Grossly ,deflated cyst measuring 13x9x1cc. Wall was thickened at places. External surface was smooth. One cystic cavity measuring 3x2x1cc with papillary excrescences noted. (Figure 2)

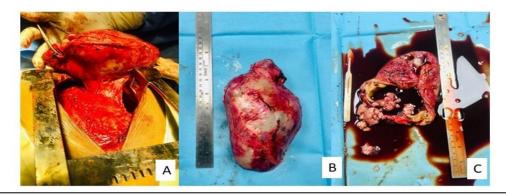


Figure 2.

(A) Intact cystic anterior mediastinal mass during operation.

(B), (C) Intact cystic mass sent to Pathology department. On cut open haemorrhagic fluid came out and deflated cyst measuring 13x9x1cc.

Microscopically sections show neoplastic lesion composed of round to oval cells having salt and pepper chromatin with scanty cytoplasm arranged in sheets and rosettes mixed with reactive lymphoid cells. There is presence of staghorn blood vessels. Extensive areas of cystically dilated blood vessels lined by endothelial cells. Some are filled with blood and some are thrombosed. (Figure 3)

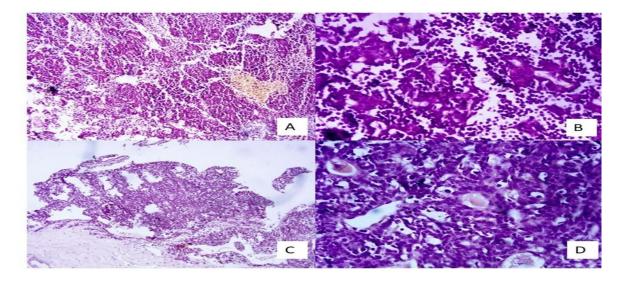


Figure 3.

(A), (C) Thymic carcinoma: Cell are arranged in sheets and rosettes mixed with reactive lymphoid cells (H&E, 10X and 4X respectively)

(B), (D) Sections show presence of cohesive cells with vesicular round nucleus and prominent nucleoli with infiltration of lymphocytes. (H&E, 40X)

IHC was done (Figure 4 and 5), it shows CD117, PAX 8, p63 and Vimentin positivity. P40 was weakly positive. Synaptophysin, WT 1 and CK 7 were negative.

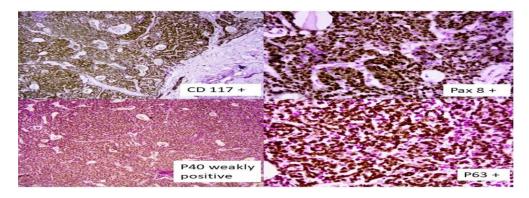
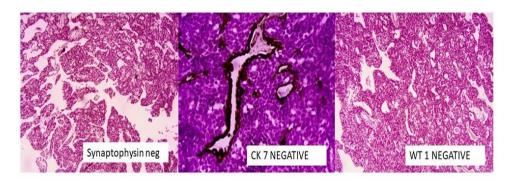


Figure 4. Immunohistochemical stains show positivity for CD 117 (cytoplasm positivity), PAX 8 (nuclear positivity), P40 (nuclear positivity), P63 (nuclear positivity)



Figure~5.~Immunohistochemical~stains~show~negativity~for~Synaptophysin,~CK~7,~WT1.

III.Discussion

Thymic carcinoma is an exceedingly rare tumour. Because of their aggressive nature they are likely to produce hematogenous and lymphatic spread locally and distally. In the present case grossly, the lesion was a multiloculate cyst filled with haemorrhagic fluid. Multiloculated thymic cysts (MTC) are rare and considered to be acquired condition of thymic tissue caused by inflammatory processes[8]. MTCs associated with thymomas or TCs are relatively rare and have seldom described. Till date not more than 50 cases has been described in English literature.[9-14]. Unilocular cysts are considered to be of embryological origin. They generally have clear fluid inside thin walled cyst[8]. As reported by Nakamura and Weissfredt the prevalence of MTCs with thymoma or TC are approximately 10.9 and18.2%,respectively.[12,15]. In the present case the patient showed symptoms of cold for 3 months. On radiological examination a large mass was discovered in the thorax. No other symptoms were there. However, they also reported two of these patients having myasthenia gravis in association with cystic thymic carcinoma[8]. Histologically all of the cases showed prominent cystic changes with malignant changes being confined to small areas. The histological classification of thymic carcinoma has been important because of its prognostic significance. High grade tumors are aggressive and increase incidence of local recurrence and distal metastasis[16]. The two classifications commonly followed and widely accepted are the Masoka staging system and classification by world health organization (WHO) [17, 18].

As described by Shen et al most of the patients in their study were asymptomatic and was discovered having the lesion on radiography. Chung et all reported 305 patients with thymic carcinoma compiled from 1966 to 2000 [19]. Most of these patients were asymptomatic or had developed late symptoms and were treated in the late stages of the disease.

The etiology of MTC is an enigma. Suster postulated that the MTC originates from the medullary duct epithelium of cyst which dilates following inflammatory process.[20].

Nakamura et al. showed immunohistochemical profile of these lesions which supports this hypothesis. The lining epithelium of Hassall corpuscles and MTCs showed similar staining for AE1/AE3, CK13, p63, CK5/6 and D2–40. In the present case along with other markers p63 and p40 were positive. Cystic thymoma is

the most important differential of MTC. Cystic dilatation of perivascular space (PVS) can give rise to cystic thymoma. These lesions do not have lining epithelium. However, in MTC the cyst has a lining epithelium[8]. Other possibilities like lymphoma or germ cell tumor should also be ruled out by extensive sampling, histopathological examination and immunohistochemistry. As per past studies by jeong et al a panel of positive markers, including CD5, CD117, Glut-1, IGF-1R, and MUC1, and negative markers, including beta-5t, CD1a, CD205, and TdT can be used to differentiate thymoma from thymic carcinoma. In our case CD117 was positive along with PAX8 [21]. However other markers were not done. As per past study by Asirvatham et al. PAX-8 was positive in 69.2% of thymic and 5.8% of lung carcinomas. CD117 was positive in 84% of thymic and 26.6% of lung carcinomas [22]. Thus corroborating with our case being PAX8 and CD117 positivity. Surgery is the main stay of treatment. Complete resection represents the most significantly favourable prognosis. In advanced stage, chemotherapy is also used as neoadjuvant therapy [23,24].

IV.Conclusion

Though thymic carcinoma sometimes become very aggressive, but if it is diagnosed in early stage, then the complete surgical debulking is possible and histopathological examination and immunohistochemical markers plays an important role to diagnose the case.

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