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Thymoma with Histological Heterogeneity- A Case Report

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Abstract: Thymomas are tumours of thymic epithelial cells, and typically have a background of immature T-cells (thymocytes). An elderly female came with progressive drooping of both eyelids and difficulty in breathing. She was diagnosed as having Myasthenia gravis. A CT-thorax was done which revealed a thymoma. On histological examination it revealed mixed histological features of Type B2 and Type B3 thymomas which are relatively rare and have a poor prognosis after extended thymectomy.
Keywords: thymoma, heterogeneity, myasthenia gravis.

INTRODUCTION

Thymomas are tumours of thymic epithelial cells which are uniquely associated with autoimmune diseases such as myasthenia gravis [1]; however the factors associated with the prognosis of thymoma remain poorly understood. The term thymoma is independent of the presence or number of lymphocytes. Nearly all thymomas present in adult life. The usual location of the thymoma is the antero-superior mediastinum, however, this tumor can also occur in other mediastinal compartments (although a posterior location is very rare); in the neck, within the thyroid, within the pericardial cavity, in the pulmonary hilum, within the lung parenchyma or in the pleura itself. Radiographically, thymoma usually results in a lobulated shadow.

CASE REPORT

A 58 year old female was admitted in the Department of Neurology and Movement Disorders at Kannur Medical College. She came with progressive drooping of both eyelids and difficulty in breathing. She was on Ayurvedic treatment for the same. Previously she was on neostigmine with a diagnosis of myasthenia gravis. Examination revealed worsening of weakness on exertion .A neostigmine test was performed which was positive. She also has easy fatigability of lids and limbs, thus confirming the diagnosis of myasthenia gravis. There was worsening of symptoms and she was admitted in the ICU where she was treated with IV Neostigmine. She made a gradual improvement on oral neostigmine and steroid tablets. We did a CT thorax which revealed a thymoma.

A plain and contrast axial CT sections of the thorax revealed a thymoma (contrast enhancing small mass lesion of soft tissue density in the anterior mediastinum- left side). She underwent thymectomy. Grossly, the thymectomy specimen measured 10.0 x 9.0x 2.0 cms with a growth measuring 7.0x4.0cms.The growth had a variegated appearance, having grey-white areas admixed with haemorrhagic areas. The growth was surrounded by adipose tissue.(Fig.1).

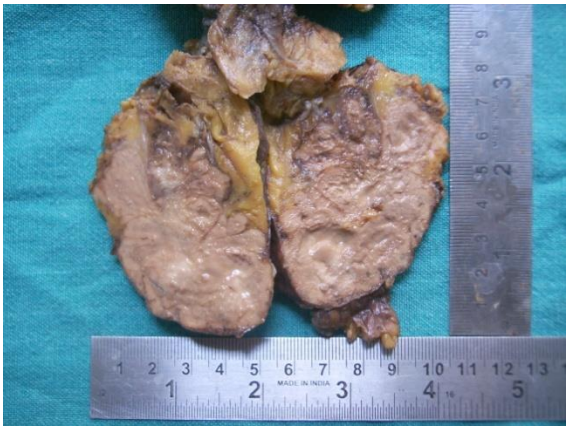

Fig.1.Thymectomy specimen showing thymoma with capsular penetration.

HISTOPATHOLOGY AND DISCUSSION

HPE showed scattered plump cells with vesicular nuclei and distinct nucleoli (neoplastic thymic epithelial cells) among a heavy population of non-neoplastic lymphocytes. Perivascular spaces with lymphocytic infiltration, proteinaceous fluid and perivascular arrangement of tumour cells resulting in a

palisading effect are seen .These features are suggestive of type B2 thymoma (Fig.2). Other foci in the same tumour showed epithelial cells having a round or polygonal shape and exhibiting no atypia. They are admixed with a minor component of lymphocytes, resulting in a sheet- like growth of the neoplastic epithelial cells(Fig 3).These features are suggestive of type B3 thymoma. Hence the tumour was diagnosed as thymoma type B2/B3 [1]. Collections of foamy macrophages and areas of necrosis are also seen. The tumour showsorganotypic features like lobulation, medullary differentiation, perivascular spaces and immature T-lymphocytes and is also seen invading the capsule and adipose tissue around the thymus (i.e. invasive thymoma) unlike thymic carcinomas which show overt cytologic features of malignancy lacking organotypic features [2]. The present case is reported because of the relative rarity of the tumour and the poor prognosis after extended thymectomy.

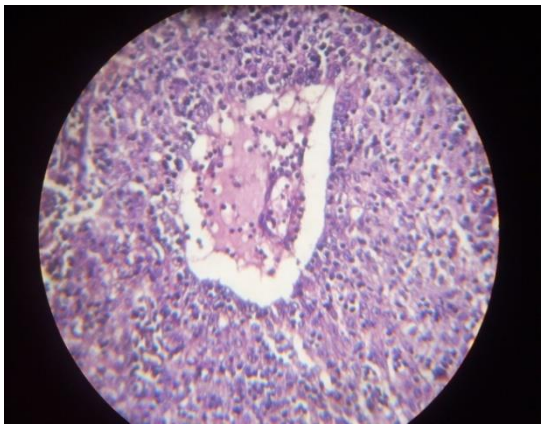

Fig.2.Thymoma type b2 showing plump cells with vesicular nuclei and distinct nucleoli among a heavy population of lymphocytes, perivascular spaces and perivascular palisading of tumour cells are seen.

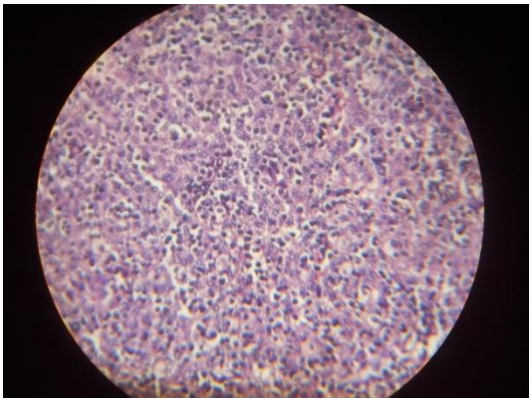

Fig.3. B3 thymoma-sheet-like growth of round to polygonal neoplastic epithelial cells with minor component of lymphocytes.

Immunohistochemistry

There is a close histologic relationship between the microscopic subtype of thymoma and the likelihood of invsion, as follows A<AB<B1<B2<B3<C [1].The epithelial cells of thymoma exhibit reactivity for keratin, Epithelial Membrane Antigen (EMA) and Carcino Embryonic Antigen(CEA).The T-lymphocytes are reactive for CD99 and CD1a [3]. Philipp Strobel *et al*. Also found in their study that B2 and B3 are potentially aggressive tumours with a potential for distant metastasis (approximately 15% of the cases), usually to lung,liver, bone and soft tissues [4]. The microscopic type of thymoma shows a close correlation with prognosis, according to this scheme of increasing clinical aggressiveness: A<AB<B1<B2<B3<C [1].This is reflected in the study by Gang C et al. in which type B2, B3 and C thymomas had significantly worse prognosis than type A, AB and B1 thymomas [5]. Thymoma Type B3 is a poor prognostic factor for survival after extended thymectomy. Age is an independent prognostic factor, suggesting favourable prognosis of thymoma after surgical treatment [6, 7].

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