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Commentary

The thymoma tale

Thymic epithelial tumours form the most important differential diagnosis for anterior mediastinal masses, among which thymomas constitute nearly 50 per cent of these lesions. These are infrequent and possess certain unique characteristics[1]. The thymus plays a central role in the development of self-immunological tolerance by appropriate conditioning of T-cell progenitors that enter the gland. Consequently, thymic pathology (including thymomas) leads to the generation of a variety of autoantibodies. Hence, a significant proportion of patients with thymomas present with features of paraneoplastic syndromes (PNSs) to the extent of exclusion of chest-related symptoms. The most common manifestation is myasthenia gravis, induced by anti-acetylcholine esterase antibodies, identified in close to 95 per cent of patients with available clinical data in a large series of Indian patients published in this issue[2]. It must be borne in mind that there are also other PNSs (including multiorgan autoimmunity), secondary cancers as well as opportunistic infections, which are explained in part by acquired T-cell immunodeficiency[3]. The presence of PNSs, paradoxically, was associated with favourable features[4].

On histology, the thymomas, in general, are seen to be composed of epithelial cells and lymphocytes in varying proportions. However, this seemingly innocuous admixture has led to several classification systems over decades[5]. The classification used currently and in this article by Guleria *et al*[2] is the WHO 2015[6], which identifies five subtypes (A, AB, B_1, B_2 and B_3) depending on the morphology of the epithelial cells (spindloid cells, epithelioid cells or their combinations) and proportion of lymphocytes. The authors concluded that the thymomas of B_2 and AB subtypes were the most common in the Indian population, in contrast to the studies from the Western world. However, in a recent, multi-institutional study of 1470 patients with thymomas from 11 countries, a similar distribution was found[7]. Furthermore, the Indian studies quoted[2] are either clinical (without a thorough histological review) or have been studies of 'mixed bag' of mediastinal tumours. Hence, to have an idea of the actual distribution, there is a need to have multi-institutional collaborations with thymomas in sufficient numbers. It would also be important to follow a protocol[8] for clinicoradiological, gross and histological assessment of such tumours, as there are some studies indicating that even tumour sizes are useful in predicting prognosis[9].

Another feature of thymomas is that the histotypes do not necessarily correlate with the clinical behaviour, prognosis and overall survival, which explains the use of the modified Masaoka-Koga staging system[10]. With time, there would presumably be more robust criteria for prognosis and therapeutic management such as utility of the angiogenesis patterns[11], immunoexpression of programmed death ligand 1[12], molecular profiling[13] and even detection of proteomic signatures[14].

***Conflicts of Interest*:** None.

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