Myasthenia gravis associated with fibrous histiocytoma

Sir,

Association of myasthenia gravis with extrathymic malignancies, especially leukemia, reticuloendothelial, and solid tissue malignancy, has been described, abeit rarely.[1]–[3] In a se-

ries of 296 thymectomized myasthenic cases, only 5 patients had extrathymic fibrous histiocytoma and extrathymic malignancies were observed more frequently in thymomatous myasthenia than in nonthymomatous ones.[4]

A 35-year-old male presented with gradually progressive dysphagia for solids for 6-month duration. For the last 1 month, he experienced diplopia especially in extremes of continuous horizontal gaze and occasional jaw weakness. Two weeks prior to hospitalization, he also developed dyspnea. There was no history of a focal or generalized muscular weakness. A 6-cm round, well defined, soft tissue swelling was noted in the left infraclavicular area. There were no other focal neurological deficits. The tensilon test and repetitive nerve stimulation test were suggestive of myasthenia gravis. However, anticholinesterase antibodies were not detected. Antinuclear antibody, RA factor, immunoglobulin levels, and other seromarkers were also negative.

Spiral computerized tomography (CT) scan of the thorax did not reveal any thymic lesion, but an extrathoracic soft tissue mass was observed in the left intraclavicular region [Figure 1]. A subsequently performed CT-guided fine needle aspiration cytology was suggestive of a spindle cell tumor. He underwent enucleation of the mass and the diagnosis of fibrous histiocytoma was confirmed by CD-68 stains, which were positive for histiocyte cells, while the cytokeratin and epithelial membrane antigens were negative [Figures 2 and 3]. Postoperatively his breathlessness and swallowing difficulty improved. He underwent phasmapheresis along with anticholinestenase and immunomodulation therapy and showed gradual symptomatic improvement.

The prominent clinical feature of this patient was pharyngeal weakness followed by diplopia and breathlessness. It was associated with extrathoracic soft tissue swelling in the left infraclavicular region. History, clinical examination and electrophysiological evaluation suggested postsynaptic neuromuscular junction disorder and the tensilon test was positive. The mass, which turned out to be fibrous histiocytoma, was excised. Other possibilities of such presentation are in cases of oculopharyngeal muscular dystrophy, bulbar palsy, and AIDP inflammatory myopathy, but these were ruled out on the basis of clinical course, investigation and response to medication. oma found to have ththymoms an Autoimmune Disease and approximately 10% of patients with myasthenia

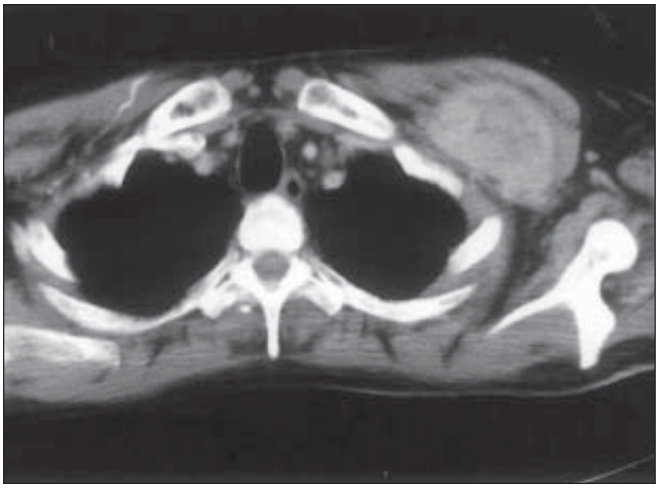

Figure 1: CECT thorax, axial section shows well circumscribed, lobulated chest wall mass with central necrosis in left infraclavicular region

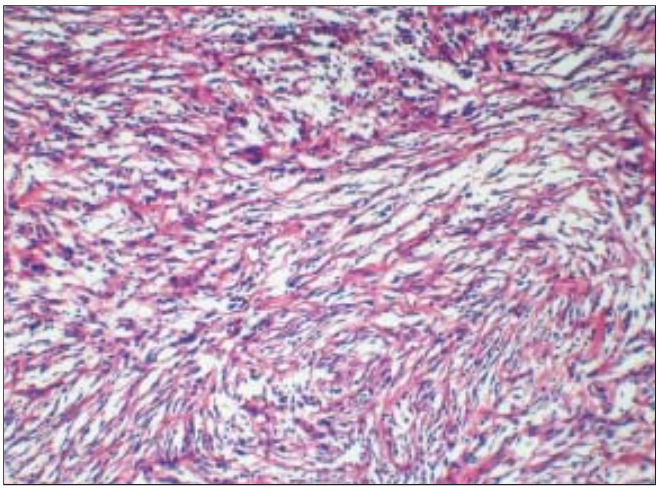

Figure 2: Hematoxin and Eosin stains showing the tumor composed of spindle shape cells arranged in storiform pattern with multinucleated giant cells

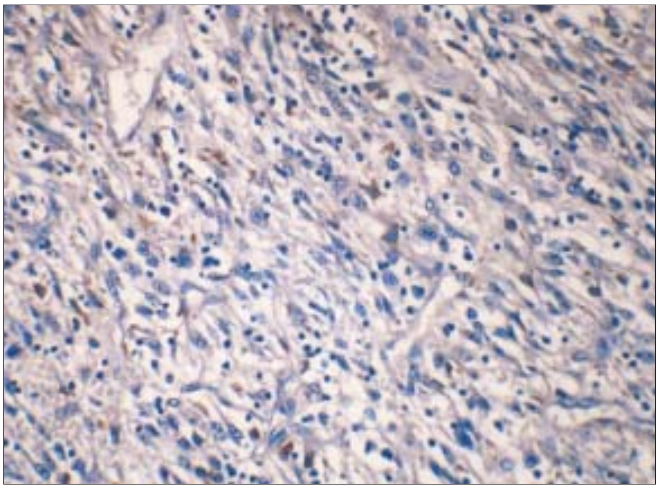

Figure 3: The spindle shaped cells stained positive for CD-68, which is a Histiocyte cell marker for fibrous histiocytoma

In the literature, 59 myasthenic cases with an extrathymic malignancy were found. There were 18 cases of thymomatous myasthenia and 41 nonthymomatous ones. The primary site of extrathymic malignancy was blood-forming organs, namely, 15 cases of leukemia and sarcoma of reticuloendothelial system, breast (eight), lung (six), thyroid (five), digestive tract (four), skin (three), uterus (three), fibrous histiocytoma (three), miscellaneous (eight), and unknown (five).[4]

Thus, myasthenia gravis although not an uncommon neu-

rological disorder, its association with extrathymic malignancy is very rare and the rarest association is with fibrous histiocytoma, *as in our case*. Further autoimmune research studies are needed to establish their association and interaction at the molecular level.

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