Inflammatory myofibroblastic tumor (IMT) is a rare benign neoplasm. It is a challenging disease because the symptoms and radiologic findings are diverse and nonspecific. Although pulmonary IMT is the most common form, pleural origin is an extremely rare clinical entity. Nuchal fibroma (NF) is another rare benign neoplasm. We report herein a case of pleural IMT with concomitant NF in a 15-year-old girl. To the best of our knowledge, this is the first report suggesting an association between IMT and NF, and our case had the largest reported intrathoracic IMT. Moreover, we found a possible association between IMT and increased CA-125 levels.