Giant Malignant Granular Cell Tumor (GCT) of the Posterior Mediastinum

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A 64-year-old woman was admitted to our hospital complaining of progressive dyspnea, fatigue, and cough. Physical examination showed dullness on percussion and decreased breath sounds in the lower zones of the right hemithorax. The chest radiograph revealed a huge right basithoracic mass. On computed tomography, a well-circumscribed and heterogeneous mass that was 15 × 10 cm in size, with necrosis and calcification was visualized in the right hemithorax (Fig. 1). A magnetic resonance imaging study demonstrated a huge heterogeneous mass with peripheral enhancement, internal necrosis, and areas of cystic degeneration located close to the vertebral column, but showed no evidence of intraspinal extension (Fig. 2). The patient underwent surgery by right lateral thoracotomy muscle sparing, through the fifth intercostal space. The giant mass produced compression to the lower lobe of the lung, and seemed to be attached to the sympathetic chain (Fig. 3*A*), but there was no evidence of involvement of mediastinal structures. The tumor was completely excised en bloc with the sympathetic nerve (Fig. 3*B*), and seemed to be a solid, encapsulated, white-to-yellow mass 15 × 10 × 8 cm in size. Histopathologic examination revealed sheets of rounded or polygonal cells separated by thin bands of hyalined collagen. Tumor cells had eosinophilic granular cytoplasm with indistinct cytoplasmic borders. Nuclear pleomorphism was present and some nuclei had prominent nucleoli with high nuclear-to-cytoplasmic ratio. There were mitotic figures and coagulative necrosis (Fig. 3*C*). Immunohistochemical studies showed strong diffuse reactivity of the cells for CD68, neuron-specific enolase, vimentine, and S100 (Fig. 3*D*). On the basis of these findings, the diagnosis was malignant granular cell tumor (GCT). Postoperative course was uncomplicated, and the patient was discharged on the fourth postoperative day. GCTs are usually benign, uncommon, and clinically silent neoplasms of Schwann cell origin, which may occur at any age, irrespective of race and sex, but are more frequent between the fourth and sixth decades in women. The malignant variant is very rare, estimated to account for no

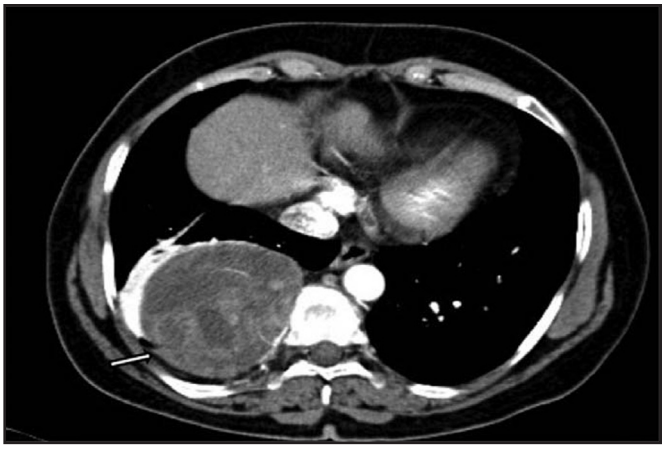

FIGURE 1. Computed tomography scan showing a well-circumscribed and heterogeneous mass with areas of cystic changes in the posterior mediastinum (white arrow).

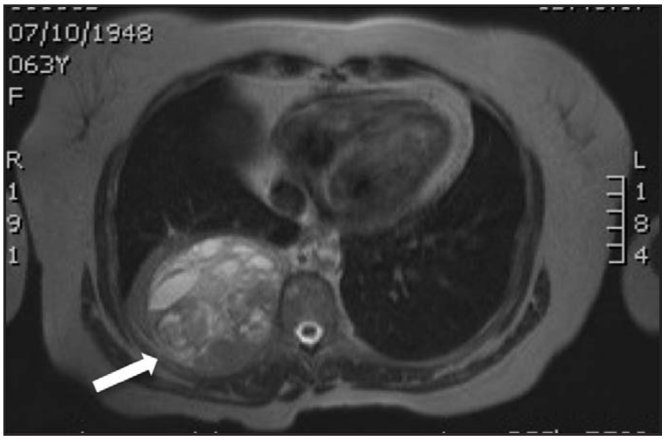

FIGURE 2. Magnetic resonance imaging study demonstrating a huge heterogeneous mass with peripheral enhancement, internal necrosis, and areas of cystic degeneration located close to the vertebral column (white arrow).

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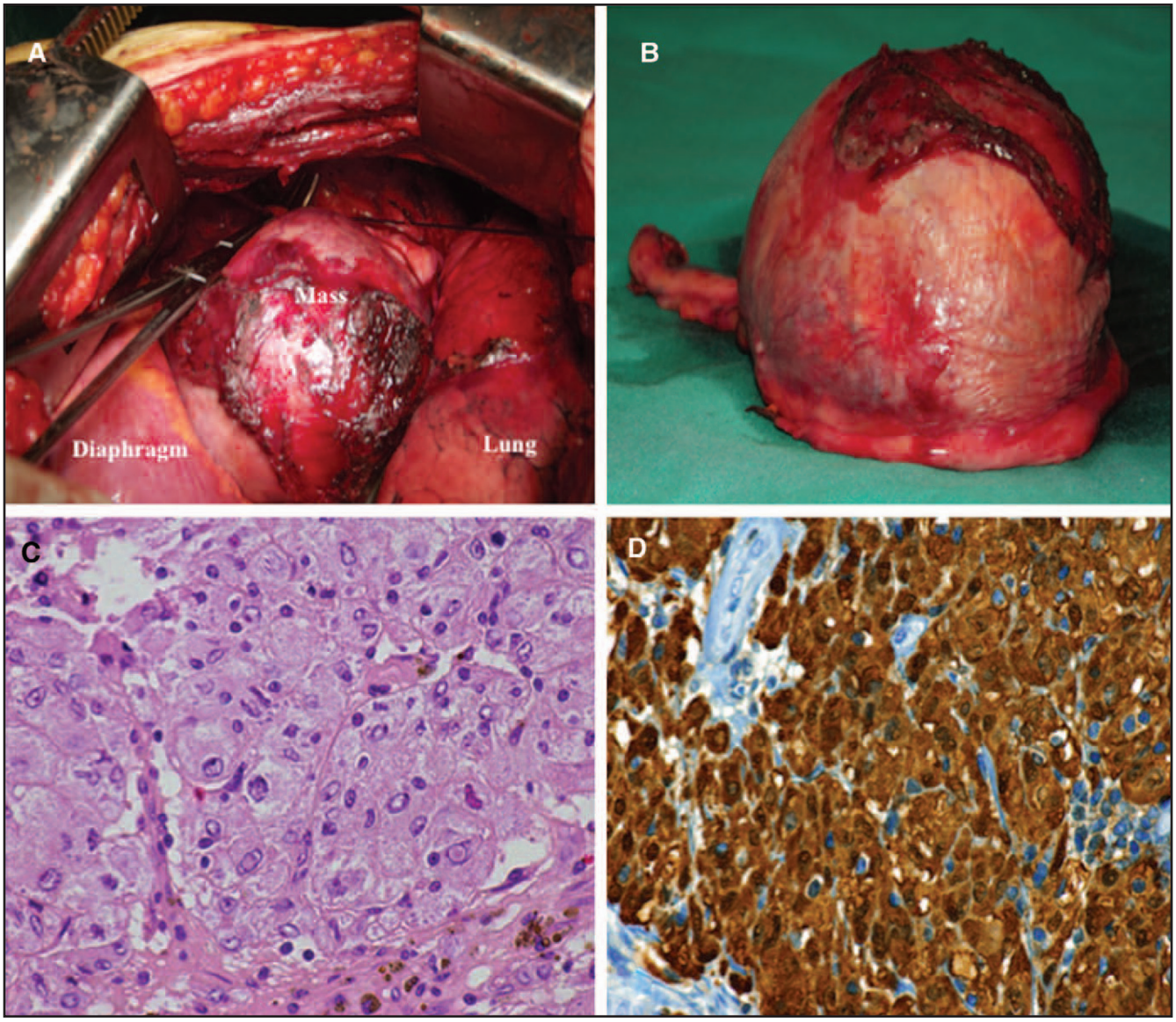

FIGURE 3. *A,* Intraoperative picture of tumor after right thoracotomy. The mass seems to be attached to the sympathetic chain. *B,* Resected mass en bloc with the sympathetic nerve. *C,* Histologic picture showing a proliferation of cells, with irregular nuclear borders and well-defined single nucleoli. The cells have eosinophilic granular cytoplasm with indistinct cytoplasmic borders (hematoxylin and eosin stain, original magnification 200×); (*D*) Immunohistochemical stain shows strong positivity for S100 (original magnification 200×).

more than 1% to 2% of all GCTs, and only 0.2% of all soft-tissue sarcomas. There have been no reports suggesting any benefit of chemotherapy or radiation therapy for this rare tumor. Complete surgical excision of the mass is the standard procedure of treatment[1,2].

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