Malignant Granular-cell Myoblastoma of the Posterior Mediastinum

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Malignant granular-cell myoblastoma is an uncommon tumor with only 13 previously reported cases. We report a 59-year-old woman with a malignant granular-cell myoblastoma in the posterior mediastinum. Histologically, the tumor shows a definite transition from benign granular-cell histology to malignant spindle-cell sarcoma. Only the spindle-cell component is noted in the metastatic bone lesion. A neural origin is proposed for this tumor.

Granular-cell myoblastoma is a common tumor that arises in many and varied sites. These tumors are almost invariably benign although local recurrences may occur particularly after incomplete excision. Multifocal origin has also been observed. The tongue is the most frequent site of origin of the benign lesion. Because of the rarity of malignant granular-cell myoblastoma, and its unusual location, the report of a 59-year-old Caucasian woman with a malignant granular-cell myoblastoma arising in the posterior mediastinum is presented.

Case Report

A 59-year-old Caucasian woman was admitted to the Thomas Jefferson University Hospital on May 12, 1968 for investigation of an abnormal chest x-ray findings which showed a large neoplastic mass involving the right hilum and mediastinum, with several small parenchymal nodules in each upper lobe. Physical examination was unremarkable, except for wheezing and dullness to percussion over the right posterior lung field.

A bone survey showed radiolucent areas of osteolytic destruction in the left ischial ramus, the skull, the left anterior rib cage, and the L 5 vertebral body. An upper gastrointestinal series demonstrated extrinsic compression of the esophagus, but no intrinsic mucosal irregularity.

Histologic sections from a rib biopsy showed a metastatic spindle-cell sarcoma. Thoracotomy was performed on May 22, 1968. Right posterior lateral mediastinal tumor was observed. It was adherent to the descending aorta, esophageal adventitia, posterior trachea, right mainstem bronchus, and anterior vertebral bodies. The bulk of the tumor mass was excised along with a 1.5 cm segment of adherent esophagus. Postoperatively, the patient's dyspnea and wheezing were relieved and she was discharged on June 3, 1968.

The patient was re-admitted on May 12, 1969 because of recurrence of the posterior mediastinal mass and symptoms of esophageal compression. The patient expired at home in August, 1969. Postmortem examination was not performed.

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Figure 1. Malignant granular-cell myoblastoma. Tumor in this area is indistinguishable from benign lesion (H and E × 100).

Figure 2. Malignant granular-cell myoblastoma. Zone of increased cellularity and nuclear atypism (H and E × 100).

Figure 3. Malignant granular-cell myoblastoma. Sarcomatous area of posterior mediastinal mass (H and E × 100).

Surgical Specimen

The tumor consisted of a 13 x 6 x 5 cm yellowish-white, partially necrotic mass, which had a lobular appearance and glistened on its cut surface. Microscopically, most of the lesion was composed of large pink cells with granular cytoplasm and eccentrically placed nuclei (Fig 1). The parenchyma was haphazardly intersected by fibrous trabeculae and the tumor demonstrated a fine sinusoidal vascular network. The cytoplasmic granules gave a positive PAS reaction. Merging imperceptibly with this pattern were zones where the nuclei became larger, more irregular, and hyperchromatic.
(Fig 2). The number of nuclei per microscopic field also increased.

Adjacent to these areas were zones of malignant spindle-cell tumor (Fig 3). The cytoplasm lost its granular character and the nuclei became elongated and spindled; growing in a fascicular sarcomatous pattern. Numerous mitoses were seen in these areas. These areas were similar to the rib biopsy material.

**DISCUSSION**

Malignant granular-cell myoblastoma is an extremely rare neoplasm. There have been 13 cases of malignant tumor previously reported and four cases of probable malignancy. Sites of the malignant lesion include urinary bladder, eyelid, subcutaneous tissue, breast, colon and vulva. No malignancy has been reported in the tongue. The age range of patients varies from 21 to 82. Like benign granular-cell myoblastoma, there is a definite predominance in women (11 women to 3 men) in incidence. Metastases are seen via lymphatics and blood stream to involve lungs, liver, bone, brain, spleen and lymph nodes. Eleven of the reported 13 malignant cases died secondary to or had metastasis at this writing.

The present case fulfills the diagnostic criteria of Ross et al for malignant granular-cell myoblastoma. Much of the tissue is histologically indistinguishable from its benign counterpart, and transition from these areas to frank spindle-cell sarcoma is seen. No hint of granular-cell histology can be seen in the metastatic bone lesion.

Since the original description of granular-cell myoblastoma, there have been many theories of origin for this lesion. Most investigators, using electron microscopic and histochemical techniques, presently favor the Schwann cell as the origin of these tumors.

The posterior mediastinum is a common site for both benign and malignant neural tumors. The origin of the present case, from this location, suggests that it too may be of neural derivation.

**REFERENCES**


**ANNOUNCEMENTS**

**Eighth Annual Arizona Chest Symposium**

The Eighth Annual Arizona Chest Symposium will be held at the Ramada Inn, Tucson, March 17-19, in conjunction with a program at the University of Arizona College of Medicine. The Arizona Chapter of the ACCP is one of the sponsors of the symposium. For further information, please write W. Curtis Wilcox, M.D., 5301 East Grant Road, Box 6067, Tucson 85712.

**Symposium on Diseases of the Chest**

A three-day postgraduate course on diseases of the chest will be presented by The Fleischner Society in Montreal, Quebec, May 29-31. The course will be interdisciplinary and will consist of lectures and seminars on topics of current interest to radiologists, internists and surgeons concerned with diseases of the chest. While emphasis will be given to roentgenology, the program will include a number of sessions dealing with the physiologic, pathologic and clinical aspects of chest diseases, particularly as they relate to roentgenology. Further information may be obtained from Dr. Robert G. Fraser, c/o Canadian Association of Radiologists, 1555 Summerhill Avenue, Montreal 109, Quebec, Canada.