Bilateral Apical Intrathoracic Masses Associated with von Recklinghausen's Disease*

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This 50-year-old black woman with von Recklinghausen's disease was first noted to have a right apical intrathoracic mass on a chest roentgenogram in 1964. By 1966, bilateral apical masses were noted. Thyroid scan was normal in 1972. In April, 1976, the patient was admitted to the hospital with a three-month history of dull, right upper chest pain that radiated around her back and down her right arm. Her skin was marked with multiple café-au-lait spots and cutaneous neurofibromas. Marked kyphoscoliosis of the upper thoracic and lower cervical spine was present. Cervical spine films showed bilateral flattening and elongation of the lower neural foramina. Posteroanterior and lateral chest roentgenograms (Fig 1 and 2) were obtained.

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Diagnosis: Meningoceles

Figures 1 and 2 demonstrate smooth, round, bilateral apical densities with the larger right-sided mass producing tracheal deviation to the left. Postero-anterior chest roentgenogram after opake myelography (Fig 3) demonstrates layering of the contrast material in both of the apical intrathoracic masses. Figure 4, a myelographic spot film, shows the separate origins of the two meningoceles from the subarachnoid space of the cervicothoracic spinal canal. Figure 5, an oblique spot film, demonstrates multiple thoracic meningoceles at various levels above T5.

Since the masses were small and produced no pulmonary symptoms and the patient's pain was controlled with mild analgesia, she was discharged for follow-up with interval chest roentgenograms.

The differential diagnosis of round, smooth thoracic masses in von Recklinghausen's neurofibromatosis includes schwannomas of the vagus nerve, phrenic nerve, or brachial plexus, ganglioneuromas of the sympathetic chain, and lateral meningoceles. The definitive diagnosis of meningocele requires myelography. Skeletal abnormalities that predispose to meningocele occur in one-half to two-thirds of patients with von Recklinghausen's disease.1 However, meningocele is uncommon and particularly so in the thoracic region. Only 76 cases of intrathoracic meningocele had been reported up to 1975, 70 percent of which were associated with von Recklinghausen's disease.2 Meningoceles may be present at almost any age, but is most common in the 30-to-60 year range in both sexes. In their 1969 review, Miles et al8 recorded a preponderance of right-sided meningoceles. They could document only eight cases with multiple lesions, five of which were bilateral. von Recklinghausen's disease was present in 64 percent of their patients.

The syndrome of neurofibromatosis, kyphoscoliosis, and intrathoracic meningocele was described by Nanson.4 Kent and Blades5 could obtain no correlation between posterior mediastinal neurogenic tumors and von Recklinghausen's disease. This finding led subsequent authors to suggest that a posterior mediastinal mass in a patient with von Recklinghausen's disease, particularly in the presence of characteristic musculoskeletal abnormalities of the spine and thorax, is likely to be a thoracic meningocele.6 Sixty percent of the cases of Miles et al8 were asymptomatic and discovered by chance.

Suggested etiologies for the meningocele include trauma, dural dysplasia, cystic degeneration of neurofibroma, elongation of nerve root sleeves, and regional dysplasia.

These lesions are generally small and asymptomatic so that surgery becomes necessary only as progressive enlargement and restriction of pulmonary function develop.8 A water-tight closure of the dural defect following resection of the lesion is necessary to prevent a substantial morbidity and even mortality which may result from leakage of cerebrospinal fluid into the chest.7 Empyema, meningitis, and hemorraghe are other common complications.

REFERENCES