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INTRATHORACIC MENINGOCELE*

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AN INTRATHORACIC meningocele is a saccular protrusion of the dural sac through an enlarged intravertebral foramen; it projects paravertebrally into the thoracic cavity, displacing the posterior pleura. Seventy per cent of the intrathoracic meningoceles reported in the literature occurred in patients with neurofibromatosis.[1,3,8–11] The posterior mediastinal mass observed in the survey chest roentgenograms is often erroneously interpreted as neurofibroma.

The purpose of this paper is to emphasize this entity and to present a case report.

REPORT OF A CASE

An asymptomatic 44 year old woman was admitted to the Jefferson Medical College Hospital because of an abnormal survey chest roentgenogram. The systemic review disclosed no abnormalities. The patient was a nonsmoker. There was no family history of neurofibromatosis, skin tumors or café au lait spots. A slight bilateral hearing loss was of undetermined etiology.

The results of laboratory studies were within normal limits. Skull roentgenograms revealed no abnormalities. An examination of the chest disclosed a large mass in the posterior mediastinum with multiple scalloped erosions of the upper thoracic vertebrae (Fig. 1, *A*, *B* and *C*). The mass was initially thought to be a neurofibroma.

A thoracic myelogram showed the opaque medium filling the large mass noted on the roentgenograms. Several small meningoceles not identified on the survey roentgenograms also filled with the opaque material (Fig. 2, *A* and *B*). The rest of the subarachnoid space was normal. Fluoroscopy and cine studies revealed no pulsations in the intrathoracic meningocele.

No treatment was administered because of the absence of symptoms, and the patient was discharged, to be followed by serial chest roentgenography. No change had been noted after 6½ months, at the time of this report.

DISCUSSION

The first case of intrathoracic meningocele was reported by Pohl[12] in 1933. The etiology has not been established. The meningocele is probably the result of a neural arch derangement with herniation of meningeal structures through the defect. Constant pulsations may erode the bone progressively.[2,7,13]

Intrathoracic meningoceles have been found in patients from 2 days to 68 years of age.[3] Most of them are discovered in the fourth and fifth decades. There is no sex predominance.[1,8,14]

The majority of intrathoracic meningoceles present on the right side, and less than 10 per cent are bilateral.[8,14]

Intrathoracic meningoceles which pulsate may enlarge over a period of 2 to 11 years,[1,2] while those which do not pulsate may not change in size or shape. One would expect meningoceles to reflect the pulsations of the subarachnoid space;[4] however, Byron *et al.*[2] reported a meningocele which did not pulsate and did not enlarge in a 2 year period. Unfortunately, myelography was not performed, and the mediastinal mass was believed to be an intrathoracic neurofibroma. Uncontrollable bleeding during thoracotomy led to the patient's death. Autopsy revealed a meningocele adherent to the surrounding pleura, vertebral bodies and ribs, which explained the lack of pulsation. We believe that the lack of pulsation is a reliable sign of stability.

The meningocele in our patient did not pulsate or respond to the Valsalva and Müller maneuvers in spite of direct communication with the subarachnoid space, and we concluded that it was stable because of fibrous tissue formation peripherally. No change in size of the lesion was noted during 6 months of observation.

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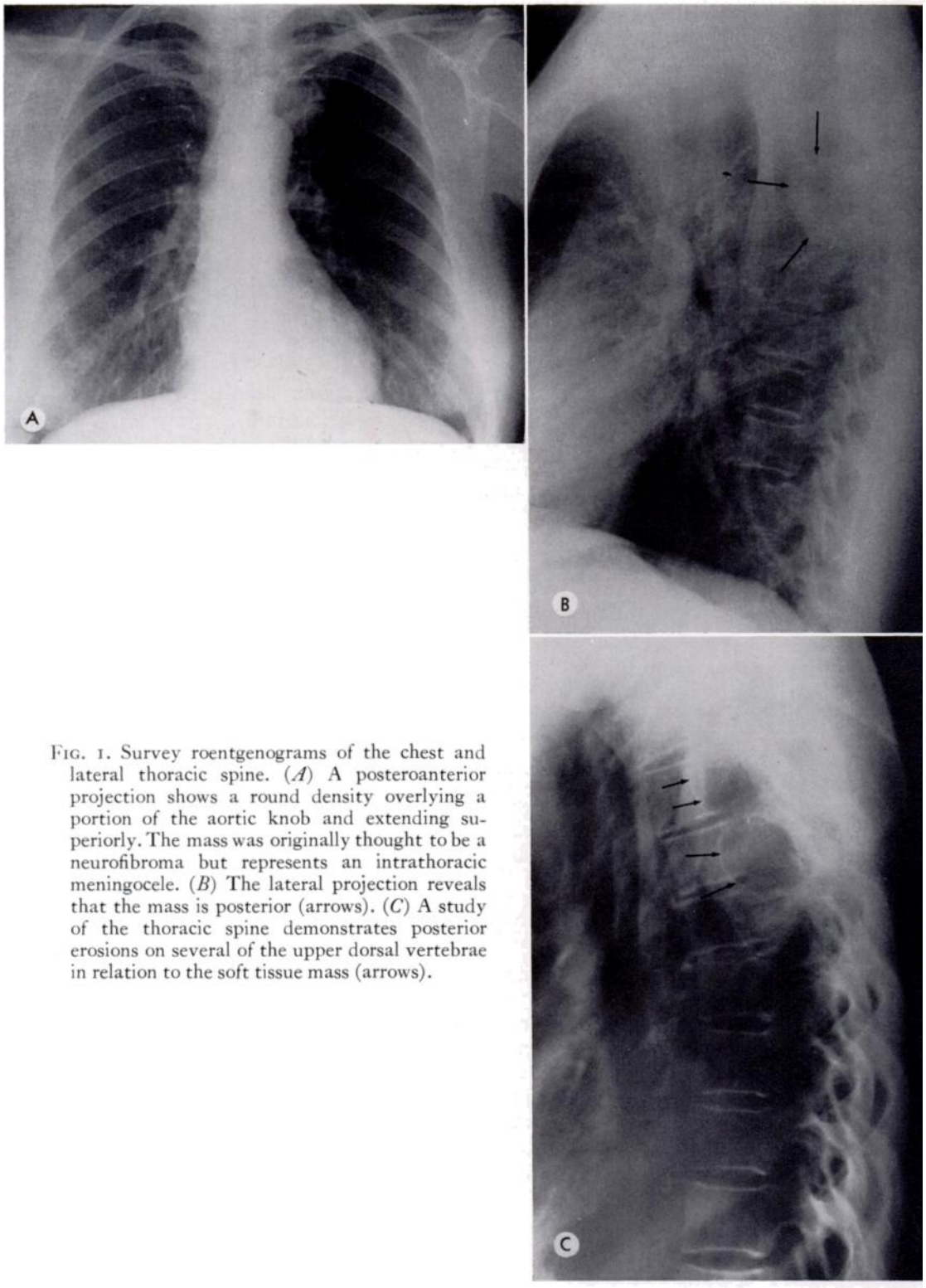

FIG. 1. Survey roentgenograms of the chest and lateral thoracic spine. (*A*) A posteroanterior projection shows a round density overlying a portion of the aortic knob and extending superiorly. The mass was originally thought to be a neurofibroma but represents an intrathoracic meningocele. (*B*) The lateral projection reveals that the mass is posterior (arrows). (*C*) A study of the thoracic spine demonstrates posterior erosions on several of the upper dorsal vertebrae in relation to the soft tissue mass (arrows).

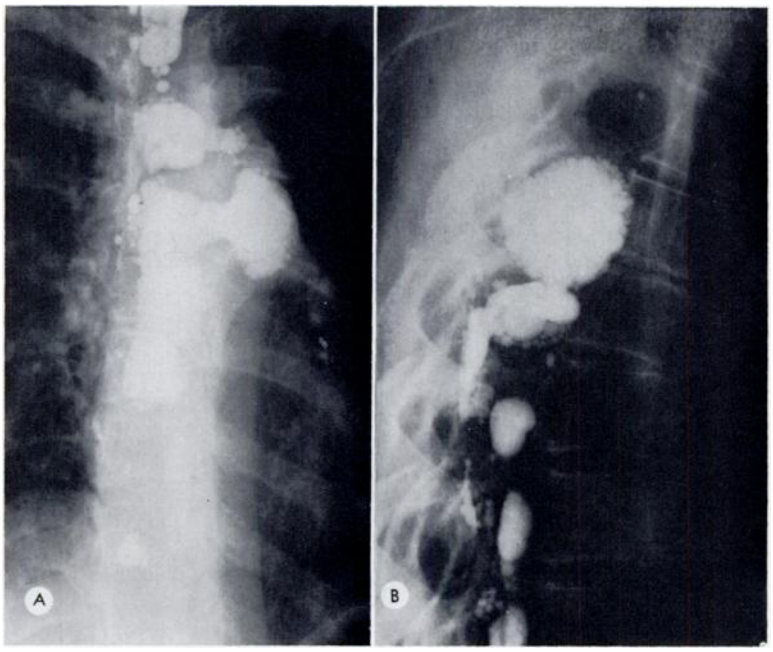

Fig. 2. (*A*) Anteroposterior and (*B*) lateral roentgenograms taken during myelography reveal the direct communication of the meningocele to the subarachnoid space. Several small meningoceles are located above and below the largest one. The meningoceles are not completely filled.

Patients with lateral meningocele are usually free from neurologic symptoms. Occasionally, pressure on adjacent structures causes pain. In contrast, neurofibroma commonly produces neurologic signs and symptoms, the result of the intraspinal component which affects the spinal cord and nerve roots.[8,10,11] Pain may also result from pressure on the intercostal nerves.

A mediastinal mass in a patient with neurofibromatosis is more often a meningocele than a neurofibroma.

The roentgen features of meningoceles are: intravertebral foramina enlargement, pedicle erosion, increase of interpediculate measurements, dorsal erosion of vertebral centra and well defined paravertebral soft tissue mass. Since neurofibroma and meningocele cysts have similar roentgenographic characteristics,[3,5,6] myelography is essential for roentgenologic diagnosis.

TREATMENT

Surgery is to be avoided unless there are progressive manifestations or growth of a meningocele. Postoperative spinal fluid fistula is a common complication; the larger the meningocele, the greater the incidence of the complication. Lack of pulsations of a meningocele suggests an adequate fibrous capsule or adhesion which should prevent progressive growth and symptoms.

Some workers advocate early surgical intervention, in the absence of symptoms, in view of the fact that these meningoceles may grow progressively, with increasing risk.[14] We believe, however, that in asymptomatic patients, surgery is contraindicated if a meningocele is non-pulsating. Progressive enlargement of the mass, symptoms and bone erosion are indications for surgery.

SUMMARY

1. A case of intrathoracic meningocele is reported with a brief review of the literature.

2. Fluoroscopic and cine roentgenologic studies failed to demonstrate pulsations or change in size of the lateral meningocele. These findings are believed to be evidence

of stability resulting from peripheral fibrosis.

3. No change was noted in the patient reported herein 6½ months after the diagnosis was made.

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