Case Reports

Intrathoracic Meningocele

Two Additional Cases of This Rare Entity

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Reviews of the published reports of intrathoracic meningoceles by Hillenius and by del Buono and Osácar listed 46 cases. It is possible that one or two of the patients have been recorded more than once but the rarity of this condition prompted us to report 2 cases seen at our hospitals.

Case Reports

Case 1, BVAH 46711. A 37-year-old white married female was admitted to the Boston Veterans Administration Hospital on Oct. 20, 1959 because of a tumor of the left arm. She had a history of multiple lumps beneath the skin for 24 years. Twenty years and again 19 years prior to admission biopsies had shown neurofibromata. Eleven years before admission a neurofibroma was removed from her mid-dorsal back; the operative report was obtained and indicated that a finger of the tumor had extended into the head of one rib but the pleura had not been entered and dissection did not extend into the vertebral canal. The patient had no neurologic symptoms.

Examination. There were several hundred soft, pliable, nontender subcutaneous tumors scattered over her body. The lesion for which she entered was purplish, sessile, soft and 6x6 cm. in size, on the exterior surface of the upper part of the left arm. There was a well-healed surgical scar paravertebrally at D7, on the left. Roentgenograms of the chest and spine showed a left posterior mediastinal mass, measuring 4x5 cm. (Figs. 1 and 2). Adjacent to the mass the vertebral ends of the 5th and 6th ribs were spread apart and there was local erosion of the lower margin of the 5th rib and the upper margin of the 6th rib. Scoliosis of the thoracic spine was present with the convexity to the left and the apex of the scoliosis at the D6 level. The pedicles of the vertebral bodies were thinned and the interpediculate spaces were increased between D3 and D7. Lateral laminographs showed the pedicles intact on the right (Fig. 3). On the left (Fig. 4) the pedicle of D5 was very thin, showing erosion of its inferior surface and the pedicle of D6 was narrowed because of erosion of its superior surface. There was posterior scalloping of the bodies of D4, D5 and D6 which was most marked on the left side of the vertebral bodies. The dorsal spine was kyphotic and the discal spaces between D5 and D7 were narrowed anteriorly and minimal spurring and sclerosis were present at the anterior margins of the vertebral bodies adjacent to the narrowed discal spaces.

On myelography, the Pantopaque collected in a sac which extended from the mid-portion of D8 through D6 (Fig. 5). Even with the patient's head lowered nearly 80 degrees only a few drops of Pantopaque went along

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Case 1. (Left) Laminagram through the right pedicles of the thoracic vertebral bodies. The pedicles are normal.

(Right) Tomograph through the left pedicles. The pedicles of T5 and T6 are thin and there is posterior excavation of T4, T5 and T6.

the right side of the canal and into the cervical region. When the patient was placed on her left side most of the contrast material went into a meningocele (Fig. 6) which extended through D5–D6 intervertebral foraminal opening.

Course. The cutaneous lesion of the arm was excised and a diagnosis of acanthosis was made. As the patient had no symptoms from the meningocele, it was felt that surgical intervention was not warranted at that time. We have attempted follow-up of her case, without success.

Case #, U.H. #755-557. R.A., a 33-year-old white female was admitted to University Hospitals on Oct. 11, 1960 because of pain and spasticity of her legs.

Approximately 6 years previously, she had noted pain between her shoulders, and a film of the chest showed a mass in the left upper lung. Myelography was done and the proper diagnosis of intrathoracic meningocele was made. A thoracotomy for removal of the lesion was complicated by drainage of cerebrospinal fluid which required a second thoracotomy. Four years later she had pain in the buttocks and left sciatic pain, and 1 year before admission to University Hospitals there developed progressive numbness and weakness of first the left and later both legs. At a private hospital, beginning in June, 1959, she underwent a series of three dorsal laminectomies with removal of “granulation” tissue and a “cystic lesion of the dura.” Her symptoms continued to progress.

Past history is remarkable in that a tumor of the neck was removed in childhood. The patient’s mother had neurofibromatosis.

Examination. There were multiple obvious neurofibromata. There was a severe spastic paraparesis with a sensory level at D8.

Myelography demonstrated a total block at D5 and two large pools of Pantopaque, one of which communicated with the pleural cavity. There were also a dorsal kyphoscoliosis, widening of the dorsal interpedicular spaces and posterior vertebral scalloping at D5–D6.

Course. At laminectomy a cystic lesion was found, extending beyond the previous site of laminectomy. This was excised and the dura mater was opened. The patient, initially worsened by the procedure, gradually recovered to walk with braces.

Discussion

The records of patients with intrathoracic meningoceles are not all complete but 33 of 46 patients from the Hillenius and del Buono and Osácar papers had neurofibromatosis, as did another patient reported by Gremmell et al. and as did our patients. Therefore, at least 36 of 49 patients have had neurofibromatosis. The common radiographic features have been a posterior mediastinal mass, enlarged intervertebral foramina, localized enlargement of the spinal canal mani-
cranial nerves, most often II and VIII, and gliomas or meningiomas involving the brain or brain stem. Although neurofibromas are one of the most common intraspinal tumors, the majority do not extend outside the canal and those that are "dumbbell" in type rarely cause changes in more than one vertebral body. In the series of 46 intraspinal neurofibromas reported by Broager\(^1\) extended through intervertebral foramina. In only 2 of these could the extraspinal extension be seen on a film of the chest. One of these 2 patients had posterolateral excavation of the spine and only a single vertebral body was eroded. Two of Broager's patients had cutaneous neurofibromata and another patient had multiple intracranial tumors as well as multiple cervical hourglass neurinomas. Kent \textit{et al.}\(^9\) found only 4 patients with neurofibromatosis in 105 patients with mediastinal neurogenic tumors. Scoliosis, common with intrathoracic meningoceles, is less frequently associated with intraspinal and mediastinal neurofibromata. Carey \textit{et al.}\(^4\) reviewed the roentgenographic changes in 94 patients with mediastinal tumors at the Mayo Clinic. Some scoliosis occurred in 7 of these patients. No asymptomatic posterior mediastinal neurofibroma with the radiographic findings commonly associated with intrathoracic meningoceles has ever been reported; therefore, there is little reason for confusion between the two entities.

\textbf{FIG. 5. Case 1.} Pantopaque collected in a sac. The patient's head is lowered 60 degrees.

Fested by an increased interpediculate measurement and excavations of the posterior vertebral body and kyphoscoliosis. The meningocele occurs most often at the apex of the scoliosis on the convex side. Occasionally, as in our Case 1, the ribs are spread and have erosion of their adjacent margins at the site of the meningocele.

The diagnosis of a posterior mediastinal neurofibroma has often been made in patients with intrathoracic meningoceles and cutaneous neurofibroma; and surgical treatment with this presumptive diagnosis has occasionally resulted in meningitis and the patient's death. However, as pointed out by previous authors,\(^5\) posterior mediastinal tumors are uncommon in patients with neurofibromatosis. Hagelstam\(^7\) found 2 patients with mediastinal neurofibromas in 94 cases of neurofibromatosis. In a monograph by Crowe \textit{et al.}\(^5\) on the study of the clinical as well as the genetic aspects in a series of 223 persons with neurofibromatosis, no mention was made of tumors of the mediastinum or spinal canal. The common tumors of the central nervous system seen in their patients were neurofibromas of the

\textbf{FIG. 6. Case 1.} Pantopaque in the intrathoracic meningocele. With the patient on her left side the Pantopaque left the sac and went readily into the meningocele.
Summary

Two additional cases of intrathoracic meningocele are reported, bringing the total known cases to 49, 36 of the patients having neurofibromatosis. A comparison is made between this entity and intraspinal neurofibroma.

References