Diagnosis of Intrathoracic Meningocele*

HEINO LAITINEN, M.D., F.C.C.P. and MARTTI TURUNEN, M.D., F.C.C.P.

Helsinki, Finland

Intrathoracic meningocele is looked upon by many as a rare tumor of the thoracic cavity. Its diagnosis is usually not made preoperatively. Among the twenty-odd cases reported in the literature there are no more than 4 diagnosed with certainty without thoracotomy (Schüller and Uiberall 1938, Head 1949, Cross and co-workers 1949, Cmyral 1952, and Scars, Clayton and Siebel 1958); to these is to be added the case now reported. Intrathoracic meningocele is very often accompanied by von Recklinghausen’s cutaneous neurofibromatosis and characteristic skeletal changes. If, in addition, radiographic examination reveals an intrathoracic tumor, there is every reason to suspect the presence of a meningocele. On the other hand, when there is reason for such suspicion, it is not too difficult to find out, by means of myelography or puncture, whether the cyst communicates with the subarachnoid space or not. The diagnostic difficulties are mainly due to the fact that intrathoracic meningocele is seldom encountered even in large hospitals and doctors consequently have little experience with it.

In the past two years, two cases of intrathoracic meningocele have been treated at First Surgical University Clinic, Helsinki. In one of these cases, published previously by one of us (M.T. 1952), the diagnosis was not established until operation. In the other case, the diagnosis was confirmed preoperatively by air myelography. Since this case exhibited all the typical features of intrathoracic meningocele, the following report of it will be found justified.

CASE REPORT

A man aged 56 years, admitted to the clinic on Feb. 2, 1953. As long as he could remember he had had reddish nodules and brownish patches on his skin. After the age of 20, he became aware of increasing spinal deformity. For the past 10 years he had suffered from breathlessness and cough, particularly on effort and in cold weather. The dyspnea had aggravated lately, and the patient had been hardly capable of work. There had also been back pain between the shoulder blades on exertion. Eight years before admission x-ray examination of the chest had revealed an intrathoracic tumor, interpreted as of benign character.

Physical Examination.—The patient was small and slender, in a fair general condition. The skin of the trunk, head, arms, and thighs showed reddish nodules ranging in size from a pin-head to a cherry, and cafe-au-lait patches. The neck was short and the cervical spine greatly deformed. The thoracic spine was also deformed, with kyphoscoliosis about the middle (Fig. 1). The lungs were markedly emphysematous, but otherwise thoracic and abdominal examination revealed nothing pathologic. Neurologic examination also yielded a negative result. The blood sedimentation rate was 14 mm. in an hour. All the other routine laboratory examinations gave a normal result.

X-ray Examination of the Chest.—The seventh and eighth thoracic vertebrae were wedged, giving rise to an acute kyphosis with the apex at the seventh vertebra and to slight scoliosis convex to the left at the same level. There was well-marked scalloping on the posterior and left lateral aspects of the sixth to ninth vertebral bodies, but the intervening disc spaces were unaffected (Fig. 3 b). The left pedicles of the

*From The First Surgical University Clinic, Helsinki, Director: Professor P.E.A. Nylander, M.D., F.C.C.P.
seventh and eighth vertebrae were completely destroyed (Fig. 3a), the spinous processes of these vertebrae turning backwards (Fig. 3b) and to the right (Fig. 3a). The left intervertebral foramina of these vertebrae formed one large opening (Fig. 3b). The seventh intercostal space was increased in width at its dorsal ends, and the related ribs were eroded. The seventh rib showed an upward subluxation at its vertebral end (Fig. 2b). A rounded, well-circumscribed, homogenous tumor, measuring 10 by 8 cm, was seen in the posterior mediastinum left of the spine, at the level of the sixth to the ninth thoracic vertebrae (Fig. 2a). The lungs were emphysematous, but otherwise the chest showed nothing abnormal.

Since the presence of von Recklinghausen's disease and of skeletal changes of the chest suggested that the tumor in the posterior mediastinum was a meningocele, air myelography was carried out. Sixty cubic centimeters of cerebrospinal fluid were withdrawn by lumbar puncture and replaced by air. With the patient lying on his right side, and the upper part of the trunk lifted up, the air was seen to rise into the sac-like tumor of the thoracic cavity and to form a semilunar air bubble along its
upper contour (Fig. 4, a and b). The location of the air bubble varied according to the patient's posture (Fig. 4 c). When he lay on his left side, the air was seen to return to the subarachnoid space. Later, the presence of air could be established in the enlarged ventricles of the brain.

Since the progressive troubles of the patient, particularly the back pain, were evidently due to the meningocele, excision of the tumor was carried out (Prof. P. E. A. Nylander). The approach was made through the bed of the resected fifth rib, and the thin-walled cyst the size of a tennis ball was detached from its surroundings. It communicated with the spinal canal through a pedicle the lumen of which was passable by a finger. The meningocele was excised, and the resultant dural opening was closed by dural and pleural flaps. A drain was placed into the pleural cavity, and the wound closed in the usual way. The patient's convalescence was uneventful.

Pathologic Report.—The tumor was a thin-walled sac with a smooth inner surface. Histologically, the wall consisted mainly of proliferative arachnoidea. Some thinned dura could also be seen, partly attached to the arachnoidea, partly separate.

Discussion

In this case of intrathoracic meningocele all the pathologic features were present which are generally taken to characterise this rare cystic tumor of the posterior mediastinum (cf. Pohl 1933, Mendelsohn and Kay 1949, Byron and co-workers 1949, Ciaglia 1952, Kessel 1952, Turunen 1962, Cmyral 1952). Among these, von Recklinghausen's neurofibromatosis of the skin was most striking. It seems to be almost regularly present in conjunction with intrathoracic meningocele, that in more than a half of the reported cases von Recklinghausen's disease has also been diagnosed. Reddish, often pedunculated nodules and cafe-au-lait patches on the skin and a radiographically detected rounded, well-circumscribed tumor constitute such a remarkable association that a suspicion of intrathoracic meningocele is fully justified.

Characteristic skeletal changes lend support to such a suspicion. In the case now reported and in most of those published previously, spinal deformity extended up to the neck (Fig. 1b). The most characteristic of these changes was thoracic kyphoscoliosis at the level of the tumor. The
FIGURE 4A

FIGURE 4B

FIGURE 4C
semilunar erosions in the posterior and left lateral aspects of the vertebrae related to the meningocele were particularly interesting. Their shape (Fig. 3 b) resembled the scalloping caused by an aneurysm in the anterior vertebral aspects, and the assumption readily suggested itself that changes of pressure in the subarachnoid space involved the meningocele (e.g., Sengpiel and co-workers 1948) and eroded the bone of the vertebral surfaces without affecting the elastic intervertebral disc. Dolley (1949) has pointed out the similarity in the etiology of vertebral erosions caused by an aneurysm and those caused by a meningocele.

In the case here reported, the left side pedicles of the vertebrae adjoining the meningocele had become almost completely destroyed and the pedicles on the right side showed some erosion. On the left side, therefore, the intervertebral foramina formed one large opening. The spinous processes and the related neural arch turned backwards and towards the right. The spinal canal was greatly distended at this point, and a sagittal tomogram taken of this part of the thoracic spine (Fig. 3 b) had a great resemblance to the post-mortem preparation described by Ameulle and co-workers (1948) in which the spinal canal and dural sheath were extensively dilated in the vicinity of the meningocele.

Von Recklinghausen's neurofibromatosis, certain skeletal changes, and intrathoracic meningocele constitute such a well-established association that it seems most unlikely that they have no causal correlation with one another. There are writers, such as Sengpiel and co-workers (1948) and Ciaglia (1952), who believe that an intrathoracic meningocele is the result of congenital malformation, though most authorities seem to ascribe it to neurofibromatosis. Brook and Lehman (1924) showed that periosteal neurofibromas produce osseous changes of the type which Pohl (1933) thought to be responsible for the rise of a meningocele. Cross and co-workers (1949), on the other hand, think that a meningocele arises in a diseased dura and that the osseous changes only provide favorable circumstances for its appearance.

The case here reported seems to allow a few assumptions concerning the causal correlation between neurofibromatosis, skeletal changes, and intrathoracic meningocele. First of all, the patient reported having had the cutaneous lesions of neurofibromatosis since early childhood, but not having noted the progressive spinal deformity until his twenties. The intrathoracic tumor was not recognized until the patient was 48 years of age. This suggests that the primary disease was neurofibromatosis. The extensive erosions of the posterior vertebral surfaces and of the pedicles, which had produced a dilatation of the spinal canal (Fig. 3), suggested a disease of the dura which made it too weak to resist the pressure in the subarachnoid space. The protrusion of the sac-like dilatation of the intra-spinal meninges through the enlarged intervertebral foramen and widened intercostal space into the thoracic cavity seems to have been a later process. The case, thus, lends support to the view concerning the causative mechanism of intrathoracic meningocele advanced by Cross and co-workers.
The presence of von Recklinghausen's disease and of certain changes of the spine and ribs suggests but does not prove that an intrathoracic tumor discovered radiographically is a meningocele, for other cystic tumors and neurofibromas of the posterior mediastinum may be responsible for similar skeletal changes. The diagnosis of intrathoracic meningocele has, therefore, to be checked by finding out whether the tumor is a thin-walled cyst connected with the subarachnoid space. Schüller and Uiberall (1938) injected lipiodol into the spinal canal and noted that it found its way into the meningocele. Byron and co-workers (1949) and Cmyral (1952) injected some contrast medium into the meningocele, from which it flowed into the subarachnoid space. Head (1949) carried out simultaneous puncture of the tumor and of the lumbar spinal canal and found that the removal of part of the cerebrospinal fluid decreased the pressure in the tumor. Cross and co-workers (1949) used air myelography and found that air moved from the subarachnoid space into the tumor.

In the case reported in this paper the diagnosis of intrathoracic meningocele was also confirmed by air myelography. This proved to be an easy diagnostic method. The air passed easily into the meningocele, where its presence was easily recognizable in the radiograms. It was also possible to watch the passage of air from the meningocele into the subarachnoid space and back by altering the patient's posture. Air myelography seems to be a highly suitable method of confirming the diagnosis of intrathoracic meningocele. It is only when the lumen of the pedicle is obstructed that air myelography is of no use; the diagnosis has then to be confirmed by thoracotomy.

SUMMARY

The correlation between neurofibromatosis, certain skeletal changes, and intrathoracic meningocele is discussed and attention is called to the close association between these conditions. The significance of air myelography is pointed out and an interesting case reported.

RESUMEN

La correlación entre neurofibromatosis, ciertos cambios esqueléticos y el meningocele intratorácico se discute y se llama la atención sobre la estrecha relación existente entre estas condiciones patológicas. Se señala la significación de la mielografía con aire y se relata un caso.

RESUME

Les auteurs discutent la relation qui unit la neurofibromatose, certaines altérations du squelette, et le méningocèle intrathoracique. Ils attirent l'attention sur les rapports étroits qui associent ces différents éléments. Ils mettent en évidence la valeur de la myéographie gazeuse, et rapportent un cas intéressant.

Bibliography will appear in reprint.