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GIANT INTRATHORACIC MENINGOCELES ASSOCIATED WITH CUTANEOUS NEUROFIBROMATOSIS TYPE I

Case report

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ABSTRACT - *Background:* Intrathoracic meningocele is a rare pathology, almost always associated with neurofibromatosis type I and with a few cases related in the literature. In the majority of cases cysts are small or asymptomatic, and the surgery is indicated when big or symptomatic cysts are present. We report a case of giant intrathoracic cysts surgically extirpated through out thoracotomy. *Case:* A 59-year-old male with familiar Von Recklinghausen´s disease which developed thoracic radicular pain after a fall. On examination he presented some difficulty in walking fast and dyspneia on small efforts. The chest plain x-ray showed the presence of 3 huge left side intrathoracic cysts (10 to 15cm). The patient was submitted to a surgical treatment and complete extirpation of the cysts was performed through a left side thoracotomy. During the surgery a fourth smaller cyst was detected and also extirpated. Evolution was uneventful and the patient remains well in these last 12 years. This finding of intrathoracic cysts related to neurofibromatosis type I is rare and is probably unique in the literature the presence of 4 huge cysts in one side of the thorax.

KEY WORDS: intrathoracic meningocele, neurofibromatosis type I.

Meningoceles intratorácicas gigantes associadas a neurofibromatose tipo I: relato de caso

RESUMO - Meningocele intratorácica é patologia rara e, quando presente, está quase sempre associada a neurofibromatose tipo I. Há poucos casos descritos na literatura e em sua maioria destes são de cistos pequenos ou assintomáticos, sendo a cirurgia indicada quando são grandes ou sintomáticos. Relatamos o caso de um homem de 59 anos com a doença de Von Recklinghausen familiar, com cistos gigantes extirpados cirurgicamente através de toracotomia.

PALAVRAS-CHAVE: meningocele intratorácica, neurofibromatose tipo I.

The intrathoracic meningocele associated to neurofibromatosis type I(NF I) was first described by Phol in 1933[1], who reported the case of a 47 year-old female initially diagnosed as neurogenic tumor. Up to 1992, 134 cases of intrathoracics meningoceles were described[2], associated to neurofibromatosis type I in 68,8% of them[3]. It is a cystic formation of the posterior mediastin, originated by a sacular protrusion of the meninge in the thoracic cavity through the intervertebral foramen pathologically dilated, or by a bone defect in a thoracic vertebra. Macroscopically, it is an unilobulated cystic formation, with vascularized wall, containing clear and crystalline fluid, with scarce cells, characteristics of the cerebrospinal fluid (CSF). Microscopically, the wall is similar to the duramater. The patients are usually, asymptomatic, although, diffuse or over the spine thoracic pain, radicular pain, headaches, persistent coughs during speech, can be present[4]. A possible complication is the spontaneous rupture[3,5,6]. Usually, the patients present with scoliosis, kyphosis, kyphoscoliosis, hypotrophy of the paravertebral muscles. There is a slight predominance in females in the 5th decade(40.3 years as medium age). However, there are cases described from 2 months to 73 years. The diagnosis and complete study of this pathology imposes: the chest plain X-ray, the myelography, computed tomography (CT) and

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magnetic resonance image (MRI) of the thorax and spine. Neurofibroma,neuroblastoma and ganglioneuroma should be included in the differential diagnosis of a mass in the posterior mediastinum. The surgery with a total extirpation through a thoracotomy or laminectomy is indicated in the presence of cysts with intrathoracic organs compression.

This case cames with all the inherent characteristics of this pathology, differing from the literature by the number and presence of giant cysts(10 to 15 cm), certainly the largests described. Another 16 cases of thoracic meningocele published from 1992 to 2002, 12 of them related to NF I were rewied.

CASE

A 59 years-old male come with complaint of radicular pain over the middle of right hemithorax, after an accidental fall to the ground. The pain became persistent, with some relief after the use of common analgesics. He also had the sensation of arrested legs when walking, fatigue and dyspneia, even during small efforts. The patient had a thoracic kyphoscoliosis with concavity to the right, "café au lait" spots and presence of diffuse cutaneous molluscus mainly over the thorax. On neurological examination, muscular power was preserved on the four limbs, keen reflexes in the legs, no Babinski sign. The gait was slightly stiff, with some difficulty to walk fast or to run. The deep and superficial sensations, and sphincter control were normal. The pain was in D7 and D8 thoracic dermatomes on the right side. He had previous diagnosis of NF I like his sister. Plain X-ray of thoracic spine, CT and MRI of thorax and spine were done, showing the three cystic tumors, whose sizes varied from 10 to 15cm, placed one upon each other, filling out part of the mediastin and almost all the left hemithorax (figs 1-3). There was a great enlargement of the intervertebral foramen on the left side related to the cysts. Taking in account the previous diagnosis of NF I, the presence of intrathoracic meningoceles was postulated. A myelotomography was performed showing a free communication of the subarachnoid space with the cavity of the cysts.

The opening was made by a thoracic surgeon who performed a left side thoracotomy. After deflating the left lung, the cysts were exposed. The medial cyst was punctured, and aspiration showed a limpid and colorless fluid, with reduction of its volume, and of the others as well. A fourth smaller cyst was present between squeezed the medium and the superior one. Opening the middle cyst, a great amount of cerebrospinal fluid (CSF) came out, allowing the vision of an enlarged intervertebral foramen, and the spinal cord inside the spinal canal. The cyst was extirpated after closure of its base with cotton stitchs. The same was made with the two others. The smaller one, that had about 5 cm diameter, was also dissected, closed in its base and resected, but, its wall was very thin, CSF was coming out through the small holes of the sutures.

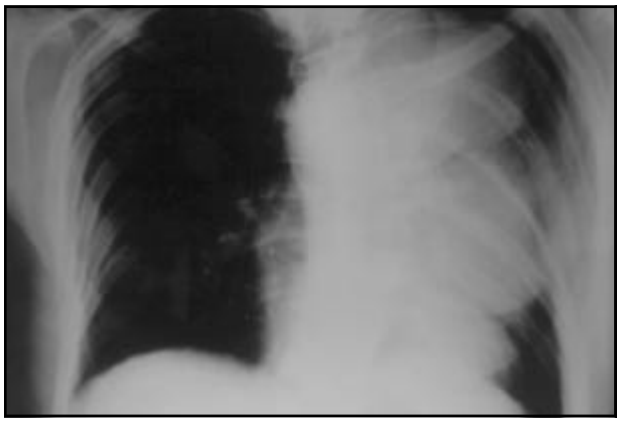

Fig 1. A plain X-ray of the thorax showed big tumors lesions on the left hemithorax.

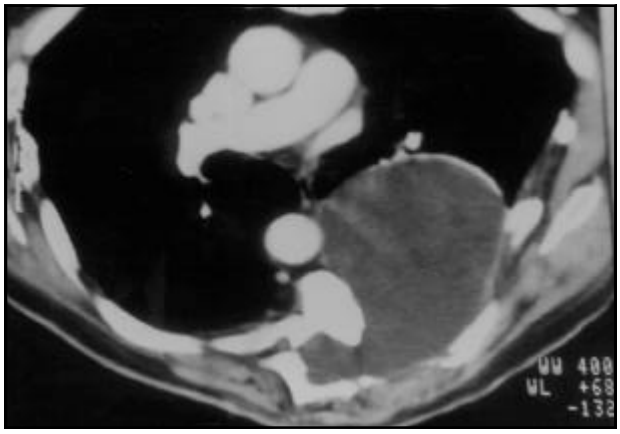

Fig 2. A transversal thorax CT scan showed a big cystic mass on the left hemithorax with continuous at the spinal canal.

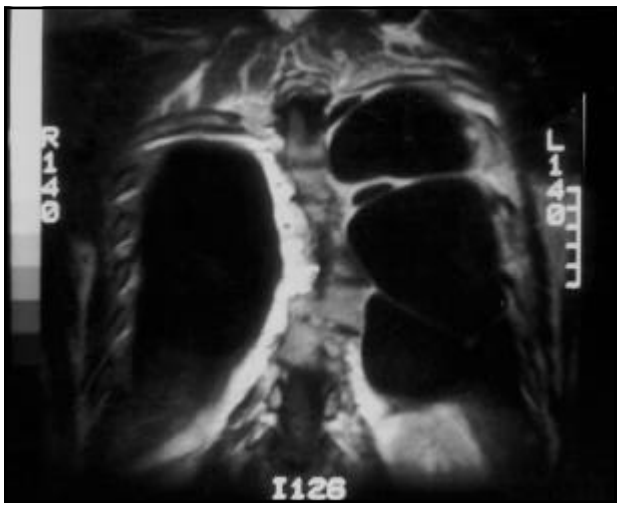

Fig 3. A coronal thorax MRI showed a three big cystic tumors on the left hemithorax.

So, it was necessary to use hemotastic and biological glue over the holes. Using the Valsalva maneuver there was no leak of CSF.

The patient had a satisfactory evolution and immediate improvement of the radicular pain and freedom to walk,

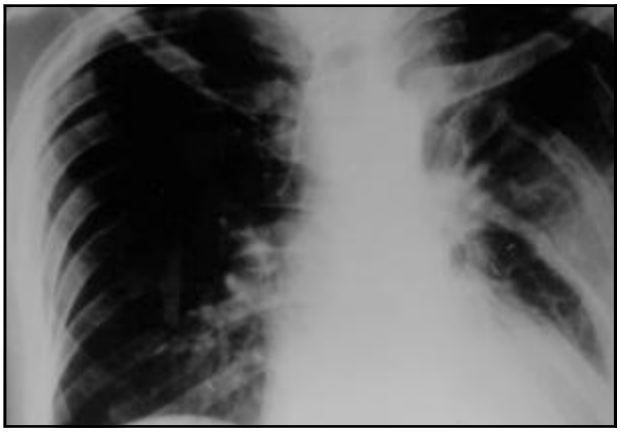

Fig 4. A post-operative plain X-ray with a follow-up in 10 years after surgery without tumor.

and no more dyspneia. A thoracic drainage was maintained for 4 days. The post operative thorax CT, showed the absence of cysts and normal aspect of both lungs. He went home in the 12th post operative day. A follow up was made in these last 10 years after the procedure, showing that the patient remained well, without any complaint and normal exams(Figs 4,5).

DISCUSSION

The presence of the thoracic meningocele in association to diffuse NF occurs in 68,8% of the cases, according to Rubin and Stratmeyer[7]. Intrathoracic meningocele association with other tumors like feocromocitoma, gliomas of the brain stem and of the optical nerve, is also reported[2]. The intrathoracic meningocele could be considered as a frustrated form of the von Recklinghausen's disease[8,9], and only 22.4% of the intrathoracic meningocele is an isolated pathology. Martelli et al[2], associate the presence of these cysts to traumas and microtraumas at dorsal level that could produce a displacement of the meninge in the anchorage point of the intercostal nerve. Sengpiel[10] believes that it can be an anomaly of the duramater in the point of involvement of the nervous root in the intervertebral foramen. The more accepted etiopathogenesis however, is the dural displasia in patients with neurofibromatosis and enlargement of the intervertebral foramen[2]. In these patients the pleural traction through the negative intrathoracic force during inspiration and the pulsation of the CSF pressure would provoke evagination of the subarachnoid space through the intervertebral foramen[2,11]. The age of most common occurence is between 30 and 60 years, although cases with only two months of age[3] and another with 73 years[12] were described. Sakai et al.[13] described in 1996, a case of intrathoracic meningocele associated to dermoid cyst in a lactant, with only 18 days old[14]. This pathology has a slight

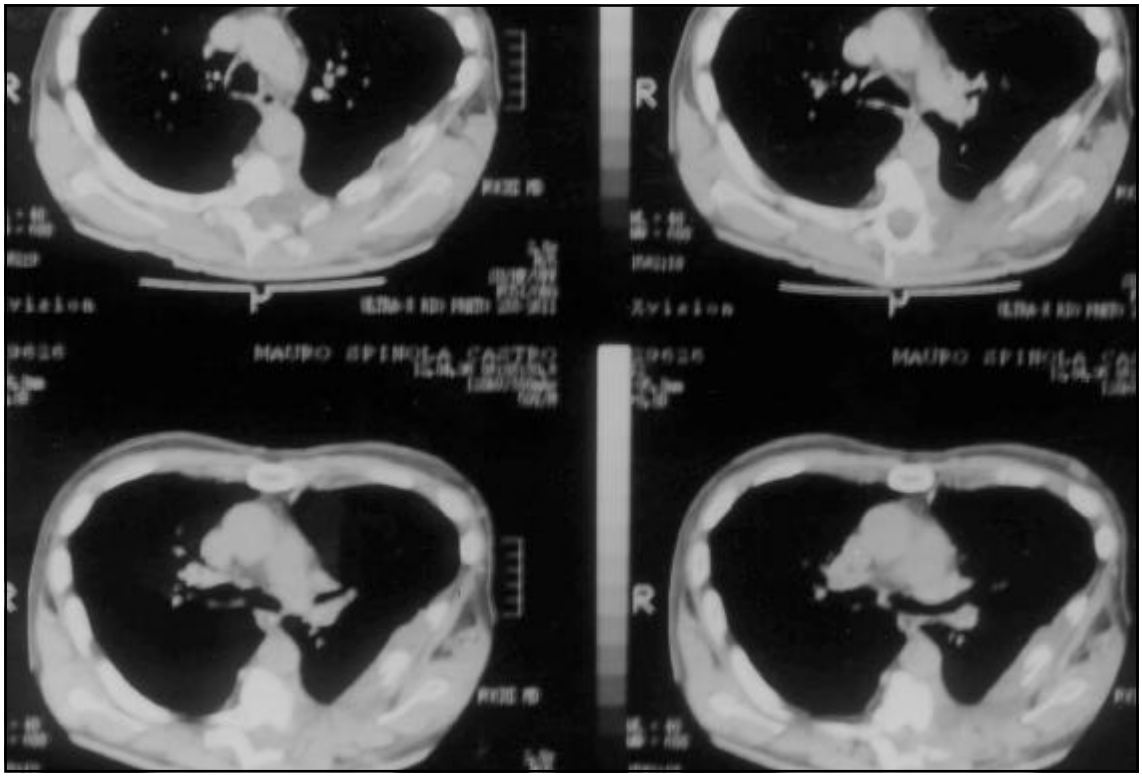

Fig 5. A post-operative transversal thorax CT scan with a follow-up in 10 years without tumors lesions.

Table1. Neurofibromatosis typel with intrathoracic meningoceles 1992-2002.

Author	Year	Sex	Age	NF	Symptom	Treatment
Shibuya	1992	Male	40 years	no	No	Surgery
	1992	Male	46 years	yes	No	Expectant
	1992	Male	50 years	yes	No	Expectant
	1992	Fem	47 years	yes	No	Surgery
Yotsumoto	1992	Fem	18 years	yes	Thoracic pain	Surgery
Madonado	1992	Male	52 years	no	Gastric discomfort	Surgery
Auge	1993	Fem	51 years	yes	No	Surgery
Sakamoto	1994	Fem	47 years	yes	No	Surgery
Sonoda	1994	Fem	26 years	yes	No	Surgery
Doi	1995	Male	41 years	no	Thoracic pain	Surgery
Rainov	1995	Fem	33 years	yes	Thoracic pain+neurol deficit	Surgery
	1995	Male	53 years	yes	Thoracic pain	Surgery
Sakai	1995	Male	18 days	?	Dermoid cyst	Surgery
Zamponi	1996	Male	51 years	yes	No	Surgery
Gut	1996	Male	60 years	yes	No	Surgery
Oide	1999	Male	50 years	yes	Dyspneia	surgery

predominance in females. The area with greater number of cases is the thoracic region, but these cysts has also been reported in the cervical[15] and lumbar regions[16].

This rare entity, whose real incidence is difficult to calculate because of the shortage of symptoms, is incidentally discovered during the performance of a plain X-ray of the thorax. The radicular or diffuse pain, can be present due to distortion or compression of the nerve roots or their ganglia. The pain can also appears due to the bone deformity and distension of the articular capsule, rotation of vertebra with subluxation. In our case, the patient developed thoracic pain on the right after a small fall on the ground, in the opposite side to the meningoceles, which disappeared immediately after the cysts extirpation. He also presented deficiency on walking fast or running, probably due to some cord impairment by pressure in the place of the cysts. Is important to say that after surgery, the patient could walk freely. In 134 cases collected in the literature until 1992 by Martelli et al.[2], 85.8% showed a single lesion, more frequently located in the right hemithorax , nine cases of bilateral cysts and eight with multiple lesions. According to Wilhelm[9], the diagnosis is easier when the cysts are present in the right hemithorax, once in the left they can be obscured by the heart and aorta images. Bone structural alterations such as kyphoscoliosis, escalloping of the vertebra and enlargement of the foramen, can be present in 70% of the patients with intrathoracic meningoceles[2,17,18]. These alterations can also be present in 9 to 38% of the cases with NF without meningoceles[2].

The myelography can show dural ectasia and image of the contrast inside the cyst by the communication with the subarachnoid space. The CT with or without intrathecal contrast can show the vertebral anomalies and the meningocele containing CSF. MRI shows the meningocele, with the same signal as the CSF and delimits a better relationship between the cyst and the cord. After 1992 we found another 16 cases described in the literature, (Table1)[11-14,17,19-25], with intrathoracic meningoceles, but only 12 of them had relationship with NF. Among these 12 cases with NF, 6 were females and 6 males, ages varying from 18 to 60 years, most of them complaining of thoracic pain. Only 2 patients were not operated.

Macroscopically, the meningocele is a cystic formation of variable diameters, and exceptionally multiloculated. In the literature, the cysts were usually smaller than those found in our patient. They were

large cysts (10 to 15cm diameter) and one smaller with only 5cm, responsible for the patient's dispneia. The cyst wall was thick, vascularized and its cavity had continuation to the subarachnoid space, through the enlarged intervertebral foramen. In our case there was just the cystic wall in continuation with the spinal membranes.

Patients without symptoms must be followed and operated only in case of a growing lesion[11]. The most common approach is through a laminectomy, with an intradural repair of the cyst, bringing the advantage of avoiding the thorax drainage, but this approach is inadequate for the larger lesions. For the small or medium sizes meningoceles, the postero lateral extradural approach is recommended[26]. The transthoracic access is indicated for big lesions, because it offers a larger field, with a small chance to damage the cord.

In the literature review, Martelli et al.[2] found the meningopleural fistula as the most common surgical complication, observed in 7.5% of the cases, and among the 134 cases reviewed, 10 died, the last one in 1969. Recurrence were reported in two cases, both affected by a large meningocele[27,28], remaining displasic duramater after surgery and the authors advise the use of a mielotomography just after operation in order to avoid it. In our case, CT was performed immediately after surgery, during the follow up and the last one 7 years later, all them showing normal pictures.

In conclusion, intrathoracic meningoceles are benign and rare lesions, with intimate relationship to diffuse neurofibromatosis, present in 68.8%. The disease is frequently asymptomatic and incidentally diagnosed after a plain chest x-ray. For the complete diagnosis is necessary the performance of CT and MRI. In case of small cysts or asymptomatic patients, the management can be clinical acomplishment and the surgery is indicated for the big cysts or symptomatic cases. The best surgical procedure must be the complete extirpation of the cyst. The cerebrospinal fluid fistula is a common post operative complication occurring in 7.5% of the cases.

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