Received: 03.01.2011 / **Accepted:** 07.02.2011
DOI: 10.5137/1019-5149.JTN.4091-11.1

Cerebral Hernia Caused by a Thoracic Surgery for Multiple Schwannomas in a Patient with Neurofibromatosis Type 2

Nörofibromatosis Tip 2 Hastasında Multipl Schwannomlar için Torasik Cerrahi Nedenli Serebral Herniasyon

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ABSTRACT

Schwannoma is a benign nerve sheath tumor composed of Schwann cells. A solitary schwannoma in the thorax is not rare, but a patient with Neurofibromatosis type 2 (NF2) and multiple thoracic schwannomas is extremely rare. We report the case of a 27-year-old woman with NF2 and two large schwannomas in her mediastinum and chest wall. We performed thoracic surgery on the patient, but she developed a cerebral hernia immediately after the operation. We report this case in order to analyze the relationship between the operation and cerebral hernia and find a way to prevent this consequence in the future. We conclude that a cranial MRI must be performed for patients with multiple thoracic schwannomas before the surgery. If there is a tumor inside the cranium, the disease in the brain must be treated first to prevent the occurrence of cerebral hernia or hemorrhage.

KEYWORDS: Schwannoma, Neurofibromatosis type 2, Hemorrhage, Cerebral hernia, Thoracic surgery

ÖZ

Schwannom, schwann hücrelerinden oluşan benign bir sinir kılıfı tümörüdür. Toraksta tek başına bir schwannom nadir değildir ama Nörofibromatosis tip 2 (NF2) ve çok sayıda torasik schwannomu olan bir hasta çok nadirdir. Mediasten ve göğüs duvarında 2 büyük schwannomu olan 27 yaşında bir kadın NF2 hastasını sunuyoruz. Hastada torasik cerrahi yaptık ama ameliyattan hemen sonra serebral herniasyon gelişti. Bu vakayı ameliyat ile serebral herniasyon arasındaki ilişkiyi analiz etmek ve gelecekte bunu önlemenin bir yolunu bulmak için sunuyoruz. Cerrahi öncesinde çoklu torasik schwannomu olan hastalarda bir kraniyal MRG yapılması gerektiği sonucuna vardık. Kraniyum içinde tümör varsa serebral herniasyon veya kanama oluşmasını önlemek için beyindeki hastalık daha önce tedavi edilmelidir.

ANAHTAR SÖZCÜKLER: Schwannom, Nörofibromatosis tip 2, Kanama, Serebral herniasyon, Torasik cerrahi

INTRODUCTION

Schwannoma is a common nervous system disease featuting benign tumor originating from the nerve sheath (7). A solitary schwannoma in the mediastinum or chest wall is not rare, but patients with NF2 and multiple thoracic schwannomas are extremely rare. In NF2 patients, intracranial tumors are potentially dangerous, especially during a thoracic operation to treat a schwannoma. This report details an NF2 patient who developed a cerebral hernia after thoracic surgery. We will discuss the reasons for the hernia and suggest methods to prevent it.

CASE REPORT

Eight years ago, a 27-year-old woman underwent an excision of a schwannoma in her. Two years later, she felt tightness in her chest and shortness of breath after exercising. The symptoms became severe, and she went to the local hospital. An X-ray of her chest showed two shadows of soft tissue in the left superior mediastinum and inferior right lung. Thoracic computed tomography (CT) scans revealed a 7cm, oval-shaped mass in the right anterior chest wall and a 5cm mass in the superior mediastinum (Figure 1A, B). The patient was admitted to our hospital on February 25, 2009, and the diagnosis upon admission was Multiple Schwannomas. Without finding any obvious contraindications, we excised the tumors on the mediastinal and thoracic walls using general anesthesia on March 2, 2009. During the operation, we observed that the left lung was well-developed, and there was no thoracic fluid. A 10 cm×7 cm×5 cm tumor, with a distinct boundary, was found 1 cm superior to the aortic arch, and it was removed after we opened the capsule. Another, similar-sized tumor inside the right thoracic wall was also excised together with some parts of the pleura. This tumor was adjacent to the anterior chest wall, but not arising from or invading the ribs. Rather, it appeared to arise from an intercostal nerve trunk. After the operation, microscopic

examination revealed a structure with interlacing bundles of numerous elongated spindle-shaped cells, with a typical palisading pattern, and the pathology report indicated it was a benign schwannoma (Figure 2). During the operation, the blood pressure was about 120-130/75-80 mmHg, and the patient lost approximately 200 ml of blood. The surgery to excise the schwannoma was successful, but the patient returned to the recovery ward with dysphoria, tachypnea, blood oxygen saturation of 89%, and a heart rate of 167 bpm. We administered drugs to maintain her blood pressure at 110/70 mmHg and managed the symptoms. Her right pupil was dilated to 8 mm, and the left pupil was dilated to 4 mm, without any light reflex. The patient's vital signs were normalized due to treatment, and a subsequent head CT scan showed several tumors and hemorrhaging in the brain (Figure 3). We used a diuretic drug to lower her intracranial pressure while keeping her blood pressure stable. The patient's vital signs were stable, but multiple organs were dysfunctional. Fourteen hours after the operation, she was in a deep coma and had no spontaneous breath and reflexes. Eighteen days later, it was impossible to resuscitate the patient, and her family requested removal of life support.

DISCUSSION

Schwannoma or Neurofibroma

Schwannoma, a benign tumor, is the most common neurogenic tumor in the thorax. It is more precise than its synonymous neurilemoma or neurinoma since this tumor arises from schwann cells (18). According to the World Health Organization, benign schwann cell tumors can be classified as either schwannomas and neurofibromas (17). The former is homogenous tumor, consisting only of schwann cells, while the latter is heterogeneous, consisting of a mixture of proliferating nerve sheath cells (6). In addition, Schwannoma usually has a capsule (11,14). The two different tumors can

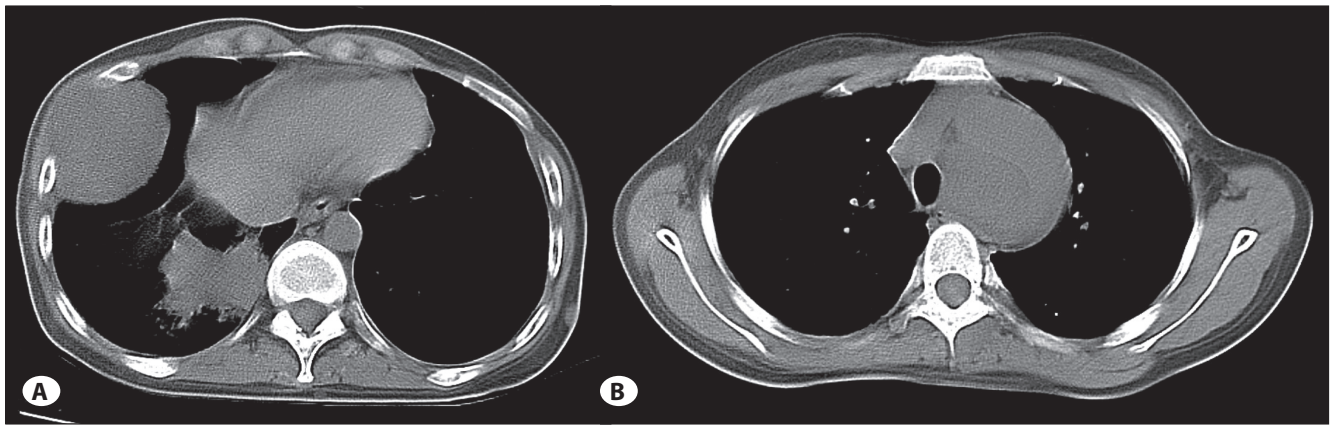

Figure 1: Chest CT scan shows the tumors in the right chest wall **(A)** and in the superior mediastinum **(B)**.

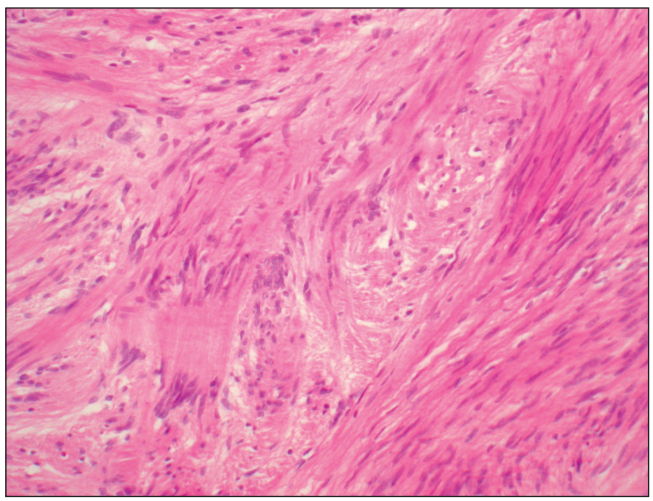

Figure 2: Haematoxylin and eosin stained, original magnification 100. The section of schwannoma reveals densely cellular, spindle-shaped cells in palisading arrangements (Antoni A), myxoid areas of low cellularity (Antoni B) and palisading nuclei surrounding pink areas (Verocay body).

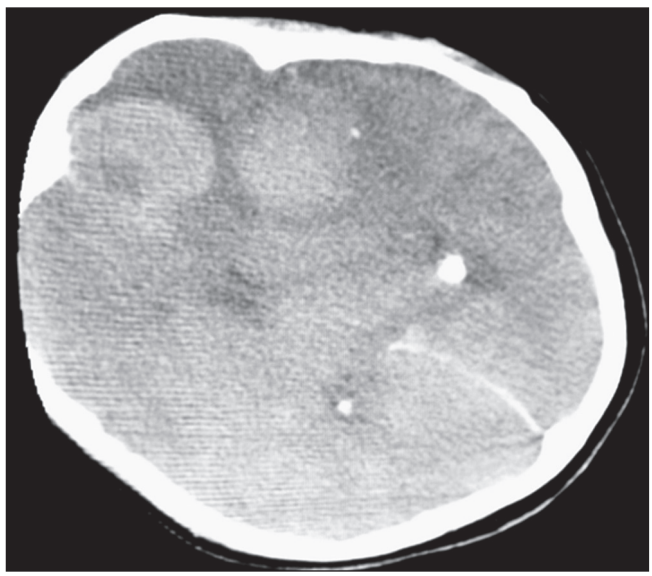

Figure 3: Head CT scan shows several tumors (gliomas or meningiomas) in the brain with hemorrhage.

be discriminated easily by a biopsy of the tumor (17). There are two types of schwannoma: Antoni A and Antoni B (12). The former is composed of spindle-shaped cells that have a compact arrangement in interlacing fascicles or short bundles. Sometimes, Verocay bodies, which are composed of palisading nuclei, are observed. In Antoni B schwannoma, the tumors are loosely composed of reticular fibers, cysts, vascellum, and the Schwann cells in an open network (3). Under the microscope, neurofibroma is usually characterized by stellate and polyhedral cells embedded in a myxoid stroma, and it less commonly has solid sheets of epithelioid cells. Our patient's was well-encapsulated and had typical Antoni A, Antoni B, and Verocay bodies upon biopsy, marking it as a Schwannoma.

Neurofibromatosis 1 or 2

When a patient has multiple schwannomas or neurofiromas, Neurofibromatosis 1 (NF1) and Neurofibromatosis 2 (NF2) should be considered. Sometimes, NF1 patients can feature multiple neurofiromas, and NF2 patients can have multiple schwannomas (9). NF1, an autosomal dominant disease, has a frequency of one in 3,500 live births. There are many typical neroplasia syndromes in NF1, including optic pathway tumors, Lisch nodules, café-au-lait macules, plexiform neurofibroma, freckling in the axillary or inguinal regions, learning disabilities, and a cancer predisposition (2, 10). Neurofibromatosis type 2 is also an autosomal dominant disease and is caused by the mutation of a tumor suppressor gene on chromosome 22q. It affects approximately one in 25,000 individuals and has nearly 100% penetrance for a 60 year old patient (1). The most recent diagnostic criteria for NF2 include: (I) bilateral vestibular schwannomas diagnosed by MRI or (II) a direct relative, such as parent, sibling, or child with NF2, plus (i) unilateral vestibular schwannoma detected before 30 years old or (ii) any two of the following: schwannoma, glioma, meningioma, or juvenile posterior subcapsular lenticular opacity (4). Our patient likely had NF2 because she had two schwannomas in the mediastinum, some gliomas or meningiomas in the brain, and a family history (father) of NF2.

The Causes of Hernia

Schwannoma and neurofiroma are two of the most common mediastinal neurogenic tumors (16). While it is easy to diagnose a tumor in the mediastinum or chest wall, the diagnosing physician should also consider whether the patient also has an intracranial tumor. In NF2, vestibular schwannomas may reach large dimensions before the onset of audiological, vestibular, or facial symptoms (13). In addition, intracranial meningiomas are present in 45–58% of NF2 patients, and they may become large before causing symptoms (1). Failure to diagnose an intracranial tumor without any correlated symptoms can lead to severe consequences following thoracic surgery. First and foremost, during the operation, the locus ceruleus-sympathesis-adrenal medulla system is stimulated, the concentration of noradrenalin, dopamine, and some other catecholamines increase, and the blood pressure increases very quickly. Also, abnormal blood vessels (*i.e.*, displaying overgrowth, vasculopathy, and arterial aneurysms) are frequently seen in the tumors of patients with NF (5,8,15). Therefore, intratumoral hemorrhages can easily occur in the brain during the surgery. Secondly, acoustic neuroma, which is the most common schwannoma in the brain, localizes to the posterior cranial fossa, and hemorrhage at this site is much more likely to cause cerebellar tonsillar hernia. Thirdly, compression of the superior vena cava during the thoracic operation can hamper venous return and increase intracranial pressure. Finally, if the thoracic schwannoma in the posterior mediastinum grows into the vertebral canal, endorachis would be prone to damage and the leakage of cerebrospinal fluid could lead to a decrease in the subarachnoid space pressure. All of these factors likely contribute to the development of a brain hernia via insertion of the tonsil of the cerebellum into the great occipital foramen.

CONCLUSION

Patients who have multiple schwannomas in the thorax are likely to have NF2. Thoracic surgery for these patients is dangerous because of the high possibility for hemorrhage or cerebral hernia. Therefore, cranial MRIs must be performed for these patients to determine if there are brain tumors prior to an operation. If a tumor is present, this tumor must be treated first to prevent complications from thoracic surgery.

DISCLOSURE

The authors report no conflict of interest concerning materials or methods used in the present report or findings specified in this paper. Author contributions to the study and manuscript preparation include the following. All three authors participated in the processes of conception and design; acquisition of data; analysis and interpretation of data; drafting the article; critically revising the article; review of the final version of the manuscript; and approval of the manuscript for submission.

ACKNOWLEDGMENTS

We thank J. R. Skaar for proofreading the manuscript. Sida Qin and Chengcheng Yang contributed equally to this article.

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