

Swan-Neck Deformity of the Cervical Spine Following Multiple Laminectomies in a Child with Neurofibromatosis

— A case report —

Moon, Myung Sang M.D., Kim, In M.D., Lee, In Ju M.D., Ahn, Yong Pal* M.D.

Dept. of Orthopedic Surgery and Internal Medicine*, Catholic Medical College, Seoul, Korea

= 國文抄錄 =

廣範圍 後弓切除後 發生한 頸椎 變形

가톨릭대학 의학부 정형외과학교실 및 내과학교실*

문명상 · 김 인 · 이인주 · 안동팔*

가톨릭 의과대학 부속 성모병원 정형외과에서 2년 3개월된 소아에 발생한 신경섬유종의 치료 목적으로 시행한 광범위 경추 후궁절제후 경추에 심한 후만변형과 사지에 강직성 마비가 초래된 예를 경험하였기에 증례보고와 함께 간단히 문헌고찰을 실시하였다. 경추의 안정성은 후방인대 및 골성구조가 중요한 역할을 하며 만일 이들 구조물이 수술적으로 광범위하게 손상되면 안정성을 잃게 되어 점차 해당척추에 아탈구 및 진행성 만곡변형이 생기게 되고 이와같은 변형이 생기면 2차적으로 척수 또는 척수신경근이 압박되어 여러가지 신경 증상이 나타난다. 본 예에서도 수술전 사지에 신경 섬유종으로 인한 강직성 부전마비가 있었으며, 수술 후 일시 신경증상의 호전이 있었으나 척추 변형과 함께 다시 악화되었다. 본 예와 같이 신경섬유종으로 추체 전방도달에 의한 추체간골유합술이 곤란한 경우에는 설사 수술 조작이 어려울지라도 후궁절제술과 함께 척추후방유합술을 시행함이 타당하리라고 생각된다.

In spite of many reports outside of Korea about progressive kyphosis and spontaneous subluxation or dislocation following multiple laminectomies, none have been encountered or recorded as yet in Korea, though similar operations are being performed in many hospitals.

The purpose of this case report is to emphasize the importance of the posterior osseoligamentous complex in maintaining the stability of the cervical spine. When this structure is destroyed extensively by surgery, particularly in children, close observation of cervical stability is required in order to

prevent a disabling deformity of the cervical spine.

Case Report

J.C.K., a 2 year 3 month old boy, was admitted to the neurosurgical unit of St. Mary's Hospital, Seoul, with a history of weakness of all limbs, abnormal gait, and a palpable hard mass on the left side in the occipitotemporal area. His mother noticed a peanut sized mass in this area when he was 1 year old and observed a gradual enlargement of this mass and a progressive weakness in all limbs.

Five months prior to this admission, a short leg brace was applied to balance muscles of the lower limbs and to improve the gait. One week prior to this entry, biopsy, from the left neck disclosed a neurofibromatous pattern.

Positive physical findings on admission were; a diffuse hard mass on left side of neck measuring 12×5cm and Café-au-lait spots. Generalized motor weakness, increased deep tendon reflex in the lower extremities, a bilateral positive Babinski and ankle clonus were also observed.

A routine preoperative X-ray of the cervical spine showed a widening of the spinal canal, and an enlargement of the intervertebral foramen in the upper region suggesting the presence of a tumor in the neural canal. A widening of the retropharyngeal space, deviation of the tracheal air shadow suggested another diffuse tumor mass in this region. (Fig. 1)

A cervical myelogram showed a space occupying lesion with a complete block at the

third cervical level. (Fig. 2)

Total laminectomies from the second to fifth cervical vertebrae and excision of the tumor mass were performed. The laminae of the second and third cervical vertebrae were thinned from within. An intradural extramedullary tumor mass extended from the level of the foramen magnum to the third cervical vertebra displacing the cord to one side.

The cord-like tumor mass extended beyond the intervertebral foramen along the nerve root. The main tumor mass and nerve roots were resected; however, neither a remnant of the mass from the anterior aspect of the spine nor the intracranial portion could be excised. Crutchfield tong traction was applied along the long axis of the body following closure of the operation wound. Crutchfield tong traction was continued for 6 weeks; however, after removing it, a mild kyphosis of the neck was noticed. Thus, the patient was transferred to the orthopedic unit for correction of the deformity. Although the widening of the retropharyngeal space was

Fig. 1. Preoperative routine X-ray of the cervical spine showing deviated tracheal air shadow and widening of retropharyngeal and retrotracheal spaces suggesting the location of tumor and enlargement of intervertebral foramen of the upper region suggesting involvement of roots.

Fig. 2. Cervical myelogram showing space occupying lesion with a complete block at third cervical level.

presumed to be caused by another neurofibromatous growth and a stabilization operation was proposed, the parents would not accept another operation. Thus, surgery was not performed.

The postoperative course was characterized by marked improvement of motor activities and neurologic signs.

The patient was observed for 32 months after the operation.

At 16 months postoperatively, X-ray of the neck showed severe kyphosis and subluxation of C₂ on C₃ and C₃ on C₄ but there was no progression of neurologic signs. When he was brought to us 32 months after operation, he was completely bedridden. The neck showed a kyphotic deformity with the face tipping upward. Physical examination disclosed severe spastic quadriplegia in spite of neck support by a collar brace. (Fig. 3)

Discussion

It has been recognized that progressive

kyphosis, spontaneous dislocation or subluxation of the cervical spine can develop following extensive laminectomies in children. Cattell and Clark(1967) feel that skeletal and ligamentous deficiencies, neuromuscular imbalance, and progressive osseous deformity consequent to bone growth after multiple laminectomies may be the causes of deformity and instability of the cervical spine. They also indicate that ligamentous laxity in children may account for the more rapid onset and greater degree of deformity compared with adults. Sim et al (1974) feels that muscle weakness, following prolonged immobilization after operation, also plays a role in developing such deformities.

In the present case, being one of the youngest reported, loss of the osseo-ligamentous complex is presumed to be a primary factor causing the disabling kyphotic deformity of the affected spine and muscle weakness due to resection of nerve roots may play a role in this respect.

The advantages and importance of anterior

Fig. 3. Series of films taken postoperatively showing progressive kyphosis and subluxation of C₂ on C₃ and C₃ on C₄.

rather than posterior fusion is well stressed by previous authors. We presume that posterior fusion is advisable, though there are surgical difficulties, when the anterior aspect of the cervical spine is inaccessible due to presence of the neurofibromatous growth, as in this case.

Conclusion

The posterior osseo-ligamentous complex is

important in the maintenance of the spinal stability. When this structure is widely damaged by surgery, instability may ensue and subsequent kyphosis with subluxation may develop, particularly in children. When an anterior interbody fusion is not indicated due to presence of neurofibromatous mass all along the anterior portion of the cervical spine, decompression laminectomy with simultaneous posterior fusion is presumed to be preferable.

REFERENCES

- Cattell, H.S., and Clark, G.L., Jr.: *Cervical Kyphosis and Instability Following Multiple Laminectomies in Children*. *J. Bone and Joint Surg.*, 49-A: 713-720, 1967.
- Cattell, H.S., and Filtzer, D.L.: *Pseudosubluxation and other normal variations in the cervical spine in children*. *J. Bone and Joint Surg.*, 47-A: 1295-1309, 1965.
- Sim, F.H., et al.: *Swan-Neck Deformity following Extensive Cervical Laminectomy*. *J. Bone and Joint Surg.*, 56-A: 564-580, 1974.
- Tachdjian, M.O., and Matson, D.D.: *Orthopedic Aspects of Intraspinal Tumors in Infants and Children*. *J. Bone and Joint Surg.*, 47-A: 223-248, 1965.