Sphenoid Sinus Carcinoma with Intramedullary Spinal Cord Metastasis and Syringomyelia - Report of A Case -

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= Abstract =

Purpose: Primary sphenoid carcinoma is rare. It accounts for 0.3% of all primary paranasal sinus malignancies. Because of the rarity of sphenoid carcinoma, large series of patients with outcome and survival statistics are currently unavailable. So we followed up the 1 case of sphenoid sinus carcinoma treated in our hospital and reported the course of the disease.

Case report : In a review of case reports and small series of patients, 2-year survival was 7%. Our case is alive at 29 months after diagnosis of sphenoid sinus carcinoma.

Intramedullary spinal cord metastasis (ISCM) is an unusual complication of cancer. In our case rapidly progressive paraparesis and urinary retention developed at 25 months after diagnosis of sphenoid sinus carcinoma. MRI of the thoracic spines showed the intramedullary spinal cord tumor mass at T3 and T4 level with accompanying syringomyelia.

Here we report a case of ISCM associated with syringomyelia which has developed after primary sphenoid sinus carcinoma with a review of literature about the clinical behavior and treatment of this lesion.

Key Words : Sphenoid sinus carcinoma, Intramedullary spinal cord metastasis, Syringomyelia

INTRODUCTION

Primary sphenoid carcinoma is rare and has an insidious onset. It accounts for 0.3% of all primary paranasal sinus malignancies. The most frequent histological type is squamous cell carcinoma. Undifferentiated carcinoma is rare. Because of the rarity of sphenoid carcinoma, large series of patients with outcome and survival statistics are currently unavailable. Current therapy utilizes both irradiation and chemotherapy. In a review of case reports and small series of patients, 2-year survival rate was 7%[1].

Intramedullary spinal cord metastasis (ISCM) is an unusual complication of cancer. Chason and co-workers prospectively examined the entire neuraxis at necropsy in 1066 patients with disseminated cancer[2]. ISCM were delineated in 10 of 200 patients with central nervous system metastasis; overall, intramedullary metastases were recognized in less than 1% of their carefully studied patients with cancer. In the review by Grem et al. more than 80% of patients with ISCM

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died within 3 months[3]. Radiation therapy combined with corticosteroid administration remain the therapeutic modalities of choice.

Syringomyelia is defined as cerebrospinal fluid (CSF) dissection through the ependymal lining to form a paracentral cavity. The syringomyelia occurring with an ISCM diagnosed during life time has been reported rarely.

We report here a case of an ISCM associated with syringomyelia which has developed after primary sphenoid sinus carcinoma with a review of literature about the clinical behavior and treatment of this lesion.

A CASE REPORT

A previously healthy 64-year-old man complained of suddenly developed diplopia and headache. He intermittently visited the local clinic for 3 months, but the symptoms were not improved. So he visited the department of neurosurgery of the Chosun University Hospital. The visual field was slightly constricted. The confluent lymph nodes which sized from 1 cm (4 lymph nodes) to 2 cm (1 lymph node) were palpable at left mid-jugular area. Neurologically he was in good orientation and able to walk by himself. The left eyeball deviated medially due to the palsy of left abducent nerve.

The skull films showed ill defined bony destruction in the sella and clivus area with diffuse haziness of the sphenoid sinus(Fig. 1). MR images of the brain revealed a large well defined iso and low signal intense mass in the sphenoid sinus on T1-weighted image(T1WI)(Fig. 2A) and heteroge-

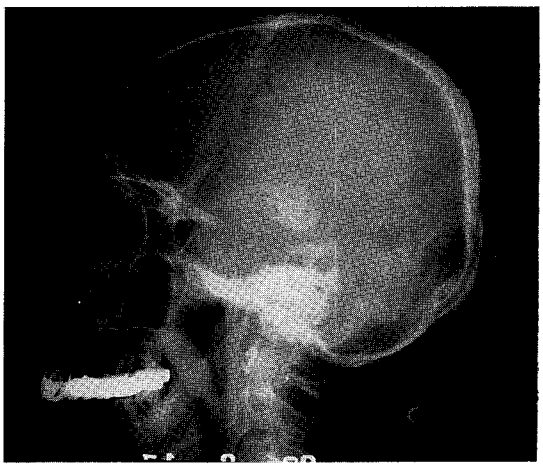

Fig. 1. Lateral Skull Film. It shows ill defined bony destruction in the sella and clivus area with diffuse haziness of the sphenoid sinus.

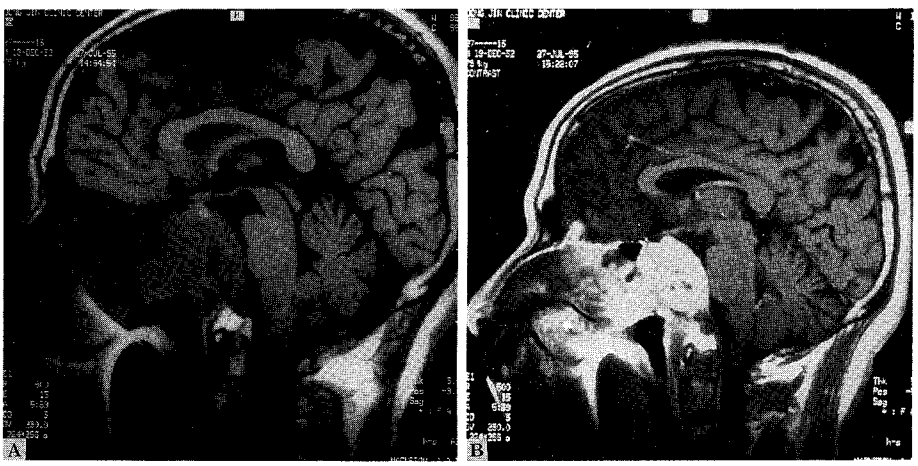

Fig. 2. MR Images of Brain before Radiation Therapy. Sagittal T1-weighted MR image (2A) shows iso and low signal intense large mass in the sphenoid sinus extending into the nasal cavity, nasopharynx, and clivu. Compression of the pons and prepontine cistern are seen. Gd-enhanced sagittal T1-weighted MR image (2B) shows heterogenous enhancement of mass.

nous enhancement of the mass on Gd-enhanced T1-weighted image (Fig. 2B). The mass extended into the nasal cavity, nasopharynx and clivus.

The diagnostic trans-sphenoidal biopsy was performed on August 19, 1993 and revealed undifferentiated carcinoma. The mass was not removed surgically, so the neurosurgeon referred him for palliative radiation therapy. At 4 weeks after biopsy, radiation therapy was started using ^{60}Co gamma-ray. He was treated with right and left lateral parallel-opposed fields (11×19 cm) which encompassed the sphenoid sinus mass, nasopharynx and upper neck including palpated midjugular lymph nodes. The lower neck nodes were treated through an anterior appositional field (19×6 cm) with a midline block. A field reduction of upper neck fields (6×10 cm) was done and left mid-jugular lymph nodes were included in the anterior lower neck field (19×14 cm) after 39.6 Gy. A dose of 50.4 Gy, in 1.8-Gy fractions, was delivered. Even though the main primary tumor was seen in the sphenoid sinus, the radiation therapy was done involving upper and lower neck nodes because of the extension into the nasopharynx and left mid-jugular lymph node area. The primary tumor and involved neck nodes received 64.8 Gy and 77.3 Gy, respectively. The optic nerves and optic chiasm were spared as much as possible not to exceed their tolerance doses.

During the course of radiation therapy, the headache decreased after 16.2 Gy and diplopia was slightly improved and size of lymph nodes decreased after 32.4 Gy. He tolerated the radiation therapy well except mucositis. At 1 month after completion of radiation therapy, the previously noted lymph nodes disappeared. Because a small whitish mass was seen in the nasopharynx after radiation therapy, a course of chemotherapy with 5-FU and cisplatin was done at 1 month after radiation therapy. At 4 months after radiation therapy, the limitation in left lateral gaze of left eye disappeared and the mass at the nasopharynx was not seen. He complained of dry mouth and diplopia after radiation therapy. At 6 months after radiation therapy, he suffered from toothache of left upper second premolar and first and second molar teeth. He performed extractions at a local dental clinic. The extraction sites were healed without problem.

At 10 months after diagnosis of sphenoid sinus carcinoma, he complained of the intermittent left temporal headache. MRI of the brain was taken at that time. The previously seen spenoid sinus mass was regressed. At 19 months after diagnosis, MRI of the brain and nasopharynx was taken again. It showed markedly decreased mass in the sphenoid sinus (Fig. 3). At 22 months after diagnosis, he felt sudden pain on his knee joint and low back pain radiating to left leg. The plain radiographs of lumbar spines and left knee showed the finding of degenerative spondylitis with multiple disc pathology and osteoporosis with multiple compression fractures in lumbar spines. Two months later the bone scintigram was taken due to aggravating pain and it revealed only the hot uptake at the sphenoid sinus area. He visited the neurosurgeon and MRI study was recommended but wasn't taken due to patient's delay.

One month later, at 25 months after diagnosis

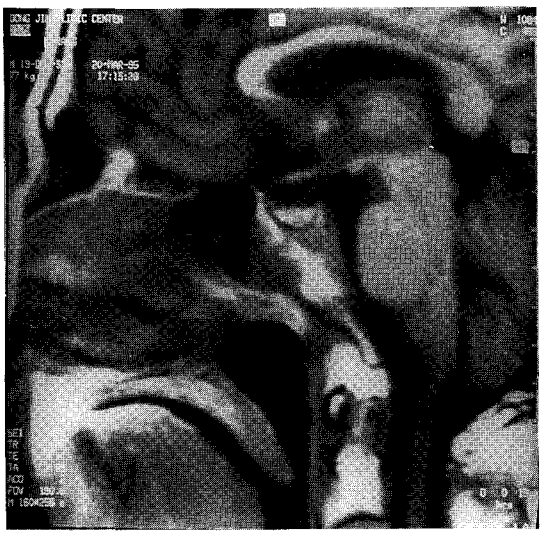

Fig. 3. MR Image of Brain at 19 Months after Diagnosis. It shows markedly decreased mass in the sphenoid sinus.

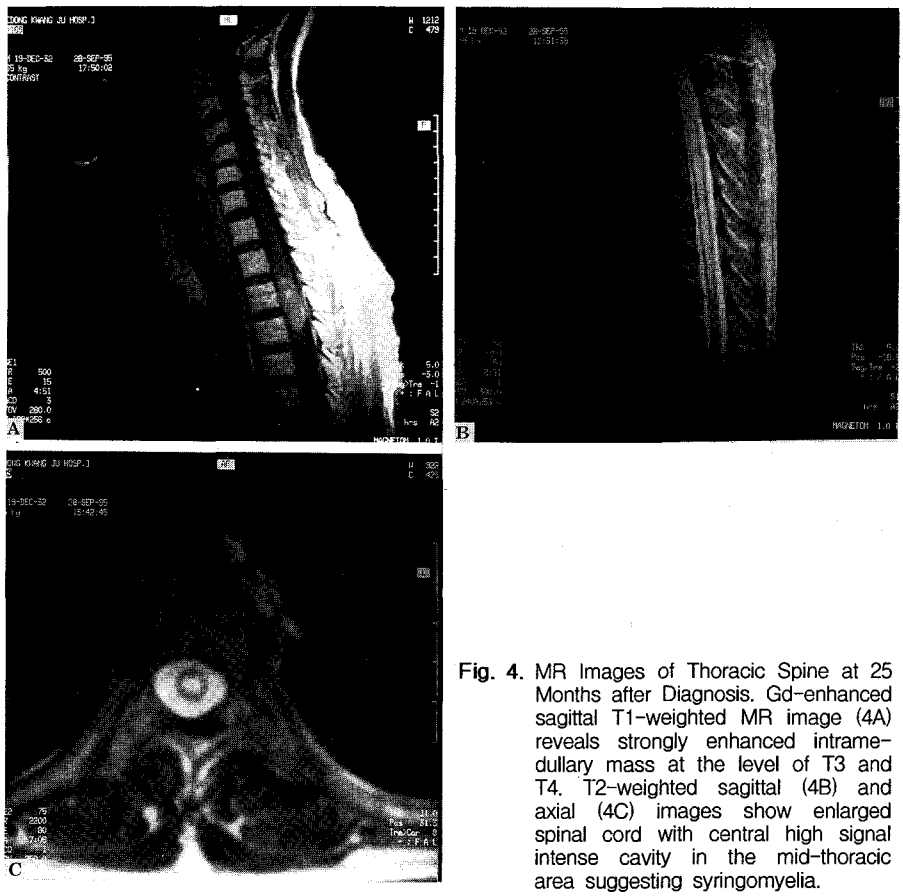

Fig. 4. MR Images of Thoracic Spine at 25 Months after Diagnosis. Gd-enhanced sagittal T1-weighted MR image (4A) reveals strongly enhanced intramedullary mass at the level of T3 and T4. T2-weighted sagittal (4B) and axial (4C) images show enlarged spinal cord with central high signal intense cavity in the mid-thoracic area suggesting syringomyelia.

of sphenoid sinus carcinoma, he felt severe pain radiating to both legs for a week, and then rapidly progressive upper back pain, paraparesis, and painful urinary retention for 3 days. Nuerologic examination revealed paraparesis, hyperesthesia below T3 level, bilaterally increased knee jerk. MRI of the thoracic spines showed intramedullary spinal cord mass at T3 and T4 level with accompanying syringomyelia (Fig. 4A, 4B & 4C). Because the metastatic tumor was intramedullary, the surgical approach was not considered. He was treated with steroid and palliative radiation therapy. The total dose was 40 Gy/ 20 fractions in 27 days with the posterior-anterior field (6×12 cm) from the C7 to the T7 level. The voiding difficulty disappeared after 10 Gy, and paraparesis was improved progressively. He could walk with a cane but upper back pain was persistant after radiation therapy, and he is alive at 29 months after diagnosis of sphenoid sinus carcinoma.

DISCUSSION

Malignancies of the paranasal sinuses comprise between 0.2 and 2 % of all human cancers. Maxillary and ethmoid carcinomas are the most common, frontal and sphenoid carcinoma are held to be quite rare.

By Harbison et al. three cases of sphenoid sinus carcinoma observed personally and 39 identified in the English literatures were reviewed. A review of series from 6 large centers showed that only 4 of 1271 cases of paranasal sinus malignancies originated in the sphenoid sinus.

Thus, sphenoid sinus carcinoma appears to constitute only 0.3% of all paranasal sinus malignancies[1].

Because the early symptoms may be vague and nonlocalizing, sphenoid sinus carcinomas may be far advanced before they are diagnosed. Its symptoms and signs are nonspecific until the sinus wall is penetrated. Once breached, specific neuro-ophthalmological symptoms and signs ensue, resulting from involvement of anatomically contiguous structures. Motor deficits may include ocular muscular palsy associated with tumor involvement of oculomotor, trochlear, and abducent nerves by extension into the cavernous sinus and superior orbital fissure. Sensory deficits associated with cranial nerve invasion within the cavernous sinus usually involve the ophthalmic and maxillary divisions of the trigeminal nerve, with the development of anesthesia or paresthesia. Depending on the area of tumor extensions from the sphenoid sinus, initial cranial nerve involvement may vary from an isolated abducent nerve paralysis or visual loss to an all-encompassing sphenocavernous syndrome that may or may not be accompanied with proptosis.

Although abnormalities were identified by plain radiography, polytomography and computerized tomography, diagnosis of sphenoid carcinoma requires direct biopsy. Treatment, principally with radiotherapy supplemented by chemotherapy, has been disappointing with most patients dead by three years[1].

The reason for the low incidence of malignancy in the sphenoid sinus is not known. It may be related to the paucity of glandular elements in its mucosa or to its location, where, outside the mainstream of the respiratory tract, it is protected from inhaled carcinogens.

There were twice as many men as women in collected series of cases[1]. The age range was wide-from 3 to 78 years. The sixth decade was the most common time of presentation. The most frequent histological type was squamous cell carcinoma (15/42 cases), followed by transitional cell carcinoma (7/42 cases). Only 2 cases were undifferentiated carcinomas among 42 cases[1].

More recently Spiro et al. retrospectively analyzed 105 patients with squamous cell carcinoma of the nasal cavity and paranasal sinuses over a 16-year period. The primary tumor was located in the maxillary sinus in 65 patients (62%), the nasal cavity in 27 (26%), the ethmoid sinus in 11 (10%), and the sphenoid sinus in 2 (2%)[4].

Carcinoma of the sphenoid sinus produces deep constant headache, radiological signs of sphenoid sinus destruction and extraocular paralysis, usually without cervical node metastasis. Radiotherapy offers palliation and some limited prospect of cure, especially if there is no bony destruction of the base of the skull[5].

Because of the rarity of sphenoid carcinoma, large series of patients with outcome and survival statistics are currently unavailable. In a review of case reports and small series of patients, 2-year survival was 7%[1, 6].

In our case, the response to primary radiation therapy began early and was good. Even though lymph nodes (1-2cm) were palpated and the palsy of left abducent nerve was seen, he has lived without local recurrence for 25 months. And then intramedullary spinal cord metastasis developed suddenly. The response to palliative radiation therapy on the metastatic tumor area was also good, so the voiding difficulty and paraparesis were improved after radiation therapy.

Intramedullary spinal cord metastasis is an unusual cause of myelopathy, especially when compared with the common problem of extramedullary metastasis[7]. In Edelson's series intramedullary metastases accounted for only 6 of 175 (3.4%) symptomatic metastases to the spinal cord[8]. The most common primary site for intramedullary metastases is the lung, representing 40 to 50% of the total, followed by the breast, colon, skin (melanoma), lymphatic system (lymphoma), and the kidney[9].

Neither the patients' symptoms nor the neurologic signs distinguished intramedullary from the more common extradural metastases. Jellinger et al. found that the full neurologic deficit developed

in less than 1 week in 9 patients, in less than 1 month in 20 patients, and in less than 6 months in 18 patients[9]. This rapid clinical progression correlated with the tumor necrosis, which is a pathological sign of rapid growth in a neoplasm. Radiographically, vertebral involvement is less common in intramedullary metastases.

How the tumor reaches the spinal cord is not clear. Hematogenous spread via the arterial and the vertebral venous system has been proposed. However, direct extension from nerve roots and the subarachnoid space also appears to be an important mechanism[8].

In this case the primary site is sphenoid sinus, so it represents the possibility of ISCM from the sphenoid sinus carcinoma. Also we couldn't find any other sites of distant metastases except spinal cord in this case.

The clinical data and imaging studies of 12 patients with intramedullary metastases were reviewed retrospectively to see if these lesions had a typical radiographic appearance and to determine the sensitivity of the various radiologic examinations[10]. The lesions were identified antemortem by either myelography, CT, MRI, and/or intraoperative spinal sonography (IOSS). Metrizamide myelography and CT demonstrated a definite intramedullary mass in 9 of 11 patients. MR detected not only enlargement and abnormal signal in the cord but also clinically unsuspected brain lesions. These various imaging procedures showed that cord metastases were often more extensive than anticipated clinically. Spread of tumor into the spinal and intracranial subarachnoid space was common. Imaging of the entire spinal canal and brain, preferably with MRI, is therefore recommended to aid in diagnosis, prognosis, and treatment.

Grem et al. reviewed 55 cases of ISCM including 50 cases from English literature[3]. Radiation therapy combined with corticosteroid administration offered the only effective palliation. The recognition of intramedullary spinal cord metastasis was an omnious finding. Intramedullary spinal cord metastasis generally occurred in the setting of widespread systemic and intracranial disease, but occasionally was the only site of relapse. More than 80% of patients died within 3 months.

Heightened awareness of this entity may lead to early diagnosis at a stage when neurologic deficits are reversible and, it is hoped, more effective palliation can be achieved[11].

Foster et al. reported two cases of syrinx associated with an intramedullary spinal cord metastasis diagnosed during life for the first time in 1987, because intramedullary metastasis usually occurs in the context of rapidly fatal disseminated disease[12]. Pathologically, hydromyelia is simply ependymal lined distension of the central canal, whereas syringomyelia is defined as CSF dissection through the ependymal lining to form a paracentral cavity[13]. Because even the histologic distinction between hydro- and syringomyelia is sometimes difficult and because their appearances on imaging studies are usually indistinguishable, these entities are often grouped under the term hydrosyringomyelia. This term describes any pathological cavity that occupies the substance of the spinal cord, whether or not it is continuous with the central canal. Nearly 80% of syrinxes are noncommunicating i.e., they are separated from the fourth ventricle by a syrinx-free segment of spinal cord. Noncommunicating syrinxes occur with both Chiari I and Chiari II malformations, as well as with acquired disorders such as spinal cord trauma, intramedullary tumors, and extramedullary compressive lesions. Neoplastic syrinxes occur mostly in the cervical spinal cord[14]. The classic MRI finding in syringomyelia is an enlarged cord with a central or slightly eccentric fluid-filled cavity that parallels CSF in signal intensity. A sharp interface between the normal cord and syrinx is typical. Increased signal intensity around the syrinx on T2WI probably represents cord gliosis, edema, or myelomalacia.

In our case, rapidly progressive upper back pain, paraparesis, and urinary retention developed at 25 months after diagnosis of sphenoid sinus carcinoma. MRI of the thoracic spine showed the intramedullary spinal cord mass at T3 and T4

evel with accompanying syringomyelia.

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= 국문초록 =

접형동암 환자에서 척수공동증을 동반한 척수내 전이
- 증례 보고 -

조선대학교 의과대학 치료방사선과학교실, 진단방사선과학교실*

오 윤 경 · 김 영 숙*

목 적 : 접형동에 발생하는 일차성 암은 매우 드물어 전체 부비동 악성종양의 0.3%를 차지한다. 접형동암이 드물게 발생되므로 많은 환자들을 분석하여 얻은 치료결과나 생존기간의 통계가 지금까지도 보고되지 않았다. 이에 본원에서 치료를 시행했던 접형동암 환자 1예의 치료경과를 보고하고자 한다.

증례 보고 : 일부 증례보고들과 소수의 환자들을 대상으로 했던 연구들을 재검토한 한 보고에 따르면 2년 생존율이 7%였다. 본 증례는 접형동암으로 진단된 후 지금까지 29개월동안 생존하고 있다.

또한 척수내 전이는 암환자에서 드문 합병증인데 본 증례에서는 접형동암으로 진단된 후 25개월 만에 갑자기 하지 마비증세와 배뇨곤란을 호소하여 흉추부위를 MRI촬영한 결과 척수내 전이성종양을 3번 4번 흉추부위에서 관찰할 수 있었고 척수공동증을 그 이하부위에 동반하고 있었다.

저자들은 접형동암환자에서 척수공동증을 동반한 척수내 전이소견을 보인 1예를 경험하였기에 문헌고찰과 함께 보고하는 바이다.