

A CASE OF GARCIN'S SYNDROME ASSOCIATED WITH CARCINOMA OF THE MIDDLE EAR

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ABSTRACT

The unilateral involvement of all or nearly all of the cranial nerves, associated with various conditions, was first described by Garcin in 1926. Different conditions such as tumors or inflammations are known to be causes of this syndrome. The primary carcinoma of the middle ear is one of causes of the syndrome of Garcin. In Japan, we have had no report of this syndrome caused by the tumors of the middle ear. A 54-year-old right-handed male with this syndrome who began to complain of an occlusive feeling in the right ear in August, 1966 and ultimately developed nearly total unilateral involvement of the cranial nerves on the right side was written. There was no evidence of increased intracranial pressure, and the X-ray film of the basal view showed invasion of the tumor. Typical findings of Garcin's syndrome were emphasized in this report.

The unilateral involvement of all or nearly all of the cranial nerves, associated with various conditions, was first described by Garcin in 1926¹⁾. He presented three criteria for the diagnosis of this rare syndrome.

At the present time, the "syndrom paralytique unilateral global des nerfs craniens" is characterized by 1. unilateral involvement of all cranial nerves, 2. absence of any signs of involvement of motor or sensory systems in extrimities, 3. absence of any signs of increased intracranial pressure, 4. abnormal X-ray findings of the base of the skull²⁾.

Many authors have described the following conditions as the cause of this syndrome: nasopharyngeal tumor, sarcoma of the base of the skull or meninges, middle ear tumor, meningioma, neurinoma, chordoma, myoma, cholesteatoma, craniopharyngioma, brain-stem glioma, lymphangioma and luetic or tuberculous meningitis^{3) -10)}.

On the other hand, it is believed that the primary carcinoma of the middle ear is relatively rare^{11) 12) 13)}.

It may cause various neurological syndrome, such as Gradenigo's syndrome, Vernet's syndrome and Schmidt's Syndrome^{14) 15)}, however, Garcin's syndrome

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secondary to the carcinoma of the middle ear has been very rare. In Japan, 17 cases of Garcin's syndrome have been reported, none of which were due to the primary tumor of the middle ear¹⁶⁾.

We experienced a case of Garcin's syndrome in which the primary carcinoma of the middle ear was considered to be the cause of this syndrome.

OBSERVATIONS

Clinical—The patient, a 54-year-old right handed man, was admitted to the Oto-rhino-laryngology service of the Tohsei Hospital on February 27, 1967, with the chief complaint of the right fronto-temporal head ache.

The Past and Family History: Noncontributory.

The history of his complaints began in August, 1966 on having occlusive feeling of the right ear. Then he noticed gradual onset of severe paroxysmal sharp throbbing pain over distribution of the first branch of the right trigeminal nerve. The pain lasted for minutes or hours and between attacks the patient complained of dull head ache on the right side. He also developed episodes of vertigo around the time of admission.

Neurological examination at this time revealed the following abnormalities: deviation of the tongue to the right, increased deep tendon reflex on the right side with positive Chaddock's sign. Blood pressure was 170/120 and the electrocardiogram showed prolongation of P-Q interval with suspected left ventricular hypertrophy. The chest film and the upper gastrointestinal series were unremarkable. A lumbar puncture showed opening pressure of 170 mmH₂O, protein 100 mg/dl, sugar 75 mg/dl, cell count 5/mm³ consisted of lymphocytes. Serum protein, icterus index, T.T.T., Z.T.T., alkaline phosphatase and urinalysis were within normal limits. C.B.C. was as follows: erythrocytes 356×10^4 , leucocytes 4,900 with eosinophilia and neutrophilia, platelets 144,000 and hemo-

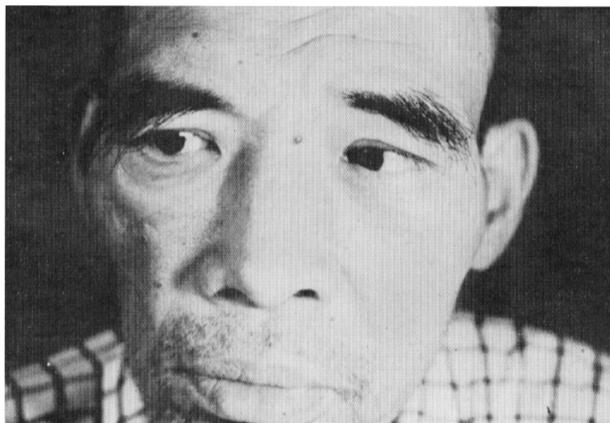


FIG. 1

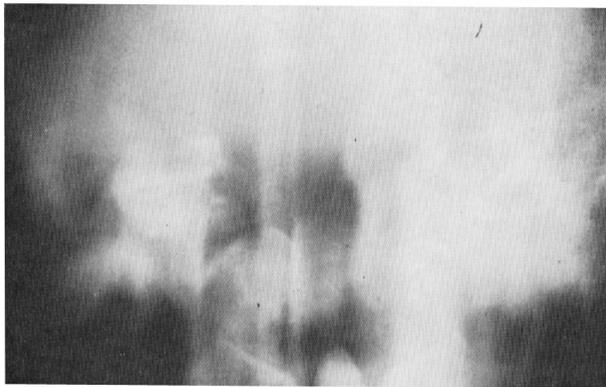
globin 71%. Kidney function tests were normal. The electroencephalogram was also normal.

On May 9 his trigeminal neuralgia was relieved temporarily by injection of alcohol to the right Gasserian ganglion. On June 23 Co⁶⁰ was begun. On July 21 neurological consult was obtained and revealed the following abnormalities: some constricted visual field in the right eye, pallor of the right optic disc with arteriosclerotic changes of the fundi, diplopia on right lateral gaze and up or downward gaze, sensory deficit over the right face with the depressed right corneal reflex and atrophy of the right masseter muscle, the right abducens palsy, the right facial nerve palsy of peripheral type, hearing loss of the right ear, paralysis of the bilateral soft palate with deviation to the left on phonation, the right vocal cord palsy, atrophy of the bilateral trapezius and the right sternocleidmastoid muscle without fasciculations and the tongue deviating to the right with some atrophy on the right side (Fig. 1).

Neurological examination was otherwise negative except for generalized weakness of muscles without myasthenic phenomenon. Physical examination was also unremarkable with the exception of marked emaciation and many firm lymphadenopathies around the neck without tenderness.



FIG. 2



R

FIG. 3

L

The skull X-ray showed abnormal density around the right petrous pyramid in the basal view (Fig. 2), bony defect of the right middle and posterior fossa in tomograms (Fig. 3).

Neurological examination of this patient was essentially unchanged until his death October 10, 1967.

Pathological—Metastasis was found in the lymphnodes of the neck, the liver, the bone marrow and the spleen. There was sclerosis in the coronary arteries and atheromatous changes with ulcers in the aorta. There was congestion of the lower lobes of the lungs. No primary malignancy was seen in the digestive, cardiovascular, respiratory and genitourinary systems.

Neuropathological examination revealed a large tumor mass extending anteriorly and posteriorly from the right petrous pyramid, occupying the right middle and posterior cranial fossa and reaching the foramen magnum (Fig. 4).

No neoplastic invasion to the right orbit was found but the sella turcica

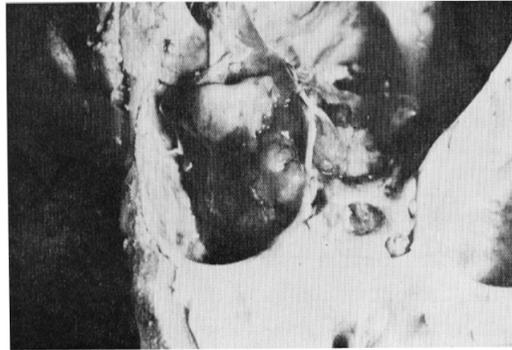


FIG. 4

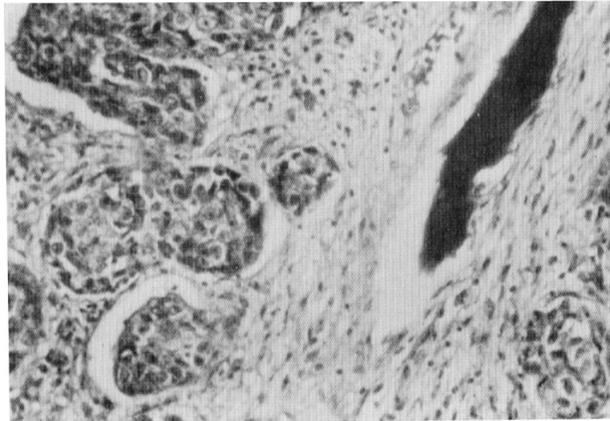


FIG. 5

was partially involved. Adhesion was demonstrated between antero-inferior aspect of the right temporal lobe and the tumor. A small area of brownish discoloration was seen over the ventral surface of the pons in its caudal part. Marked arteriosclerotic changes were found in the vertebulobasilar system.

Microscopically the tumor consisted of groups of malignant epithelial cells without cornification—They tended to form cell nests with proliferation of connective tissue (Fig. 5). The small area of brownish discoloration mentioned above was a small metastatic focus of neoplasma. The other metastatic focus was demonstrated in the subependymal area of the third ventricle on the right side near the posterior commissure. Though the cranial nerves on the right side were severely damaged, parts of the second, third and fifth cranial nerves carefully examined and showed some demyelination and degeneration of the

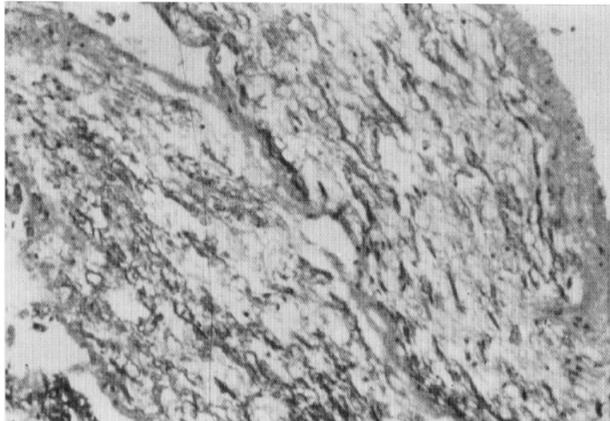


FIG. 6. The 3rd cranial nerve. B.P.A.S.

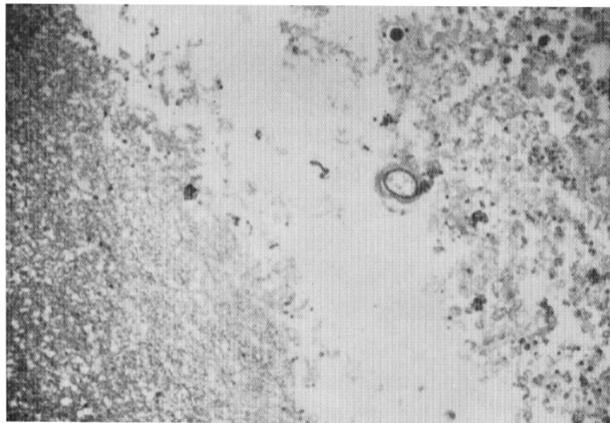


FIG. 7. The 5th cranial nerve. B.P.A.S.

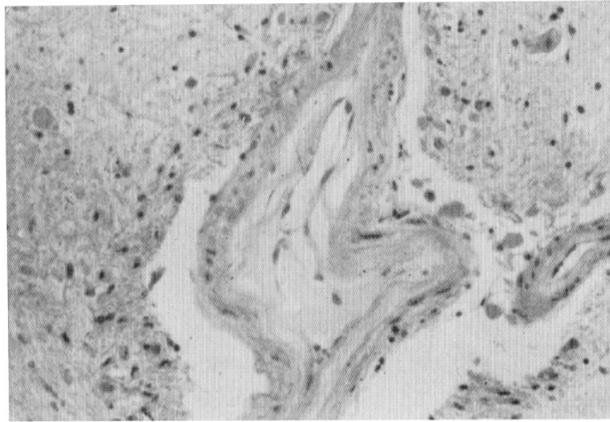


FIG. 8

axons (Fig. 6 and 7). A small focus of softening with old hemorrhage was present in the posterior limb of the left internal capsule. Hyalinization of the wall of a small vessel was seen in the area of the softening and associated with proliferation of glial cells and eosinophilic changes of some neurons here (Fig. 8).

COMMENT

Since Garcin presented a new syndrome of unilateral global involvement of the cranial nerves, many cases of this rare syndrome have been reported. Most of them do not strictly fulfill the clear-cut picture drawn by Garcin. As pointed out by Spiegel⁹⁾ and Hamaguchi⁵⁾, it is possible that the tumor encroaches upon the brain or the other side of the base of the skull and gives rise to cerebral signs or bilateral cranial nerve signs, if the patient lives long enough. There seems to be some discrepancy between opinions about the definition of Garcin's syndrome. Some of the authors agree on making diagnosis of Garcin's syndrome without evidence of pure total unilateral involvement of the cranial nerves, while others emphasize four criteria elaborated by Garcin.

Of 17 cases of Garcin's syndrome reported in Japan, only three cases showed total unilateral involvement of the cranial nerves. The case presented here demonstrated involvement of the all cranial nerves on the right side except for the olfactory nerve. Though the oculomotor and trochlear nerves were not completely affected clinically, the pathological findings mentioned above can prove some involvement of these nerves. The patient received an injection of alcohol to the Gasserian ganglion as a possible disturbance of the right trigeminal nerve. However, his severe right frontal head ache associated with horizontal diplopia long before the injection and prompt relief of pain after

blocking of the ganglion can be considered as other evidences of encroachment upon the right trigeminal nerve by neoplasma.

Another diagnostic problem as Garcin's syndrome in this case is that the patient had transient hemiparesis before starting his trigeminal neuralgia. This can be undoubtedly explained by the fact of a small area of old encephalomalacia in the left internal capsule compatible with his clinical history. Therefore, the right hemiparesis of this patient is an incidental complication and has no connection with his cranial nerve palsy. It is possible to state that the case in this report well fulfills, though not complete, four criteria described by Garcin.

It has been well known that the primary malignant tumor of the temporal bone including carcinoma of the middle ear is rare¹²⁾. Furstenberg estimated the incidence at 1 in 20,000 of aural conditions¹⁷⁾. Mattick and Mattick, working in the cancer clinic, found only 10 cases of intrinsic aural carcinoma in 3,000 tumors of all kinds¹⁸⁾. Tucker reviewed 89 cases of the middle ear carcinoma and stated their rarity¹⁹⁾. According to the statistics in the United States, there is one carcinoma of the middle ear per 4,000~5,000 cancer patients.

Autopsy finding of this case revealed a primary carcinoma in the right temporal bone. It is difficult to determine whether this malignant neoplasma started from the middle ear or mastoid cells. Clinically it is apparent that the patient initially began to have occlusive feeling in the right ear. Therefore, it is most likely that the epidermoid carcinoma started from the middle ear.

SUMMARY

A 54-year-old right-handed male complained of occlusive feeling in the right ear in August 1966. Ultimately the patient developed nearly total unilateral involvement of the cranial nerves on the right side. There was no evidence of increased intracranial pressure and the X-ray films of the skull revealed invasion of a tumor to the base of the skull. These findings well correspond to the criteria for diagnosing the syndrome of Garcin. The syndrome of Garcin has been reported very rarely by primary carcinoma of the middle ear. None of the reported cases in Japan were due to the primary tumors of the middle ear or mastoid cells. This is the first case of Garcin's syndrome secondary to the malignant tumor originated from the middle ear in this country.

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