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Case report

Cerebrospinal fluid leakage and Chiari I malformation with Gorham's disease of the skull base: A case report

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ABSTRACT

Background: Gorham's syndrome is a rare bone disorder characterized by massive osteolysis of unknown etiology. There are no reports of comorbidity involving cerebrospinal fluid (CSF) leakage and Chiari I malformation with Gorham's syndrome. Here, we report an unusual case of an acute presyrinx state complicated by bacterial meningitis due to CSF leakage and Chiari I malformation associated with Gorham's disease of the skull base.

Case presentation: A 25-year-old woman with Chiari I malformation associated with Gorham's syndrome presented with aggressive paresthesia following bacterial meningitis. Axial magnetic resonance imaging (MRI) and computed tomography (CT) cisternography revealed CSF leakage in the right petrous apex. A presyrinx state was diagnosed based on the clinical symptoms and MRI findings. With resolution of the bacterial meningitis, the spinal edema and tonsillar ectopia also improved. Surgical repair of the CSF leakage was performed by an endoscopic endonasal transsphenoidal approach to prevent recurrence of meningitis. The postoperative course was uneventful.

Conclusion: Skull base osteolysis in Gorham's syndrome may induce Chiari I malformation and CSF leakage. We should pay attention to acute progression of clinical symptoms because Gorham's syndrome may predispose to development of Chiari I malformation and may be complicated by CSF leakage.

1. Introduction

Gorham's syndrome is a rare bone disorder characterized by massive osteolysis and lymphangiomatosis of unknown etiology. Approximately 200 cases have been reported to date. Only 3% of cases with Gorham's syndrome have had skull base involvement [1]. Chiari I malformations associated with Gorham's syndrome are extremely rare and have been reported in only 7 cases to date [1–3]. In addition, cerebrospinal fluid (CSF) leakage and recurrent meningitis has been described in only 5 cases [4–7]. Most importantly, there are

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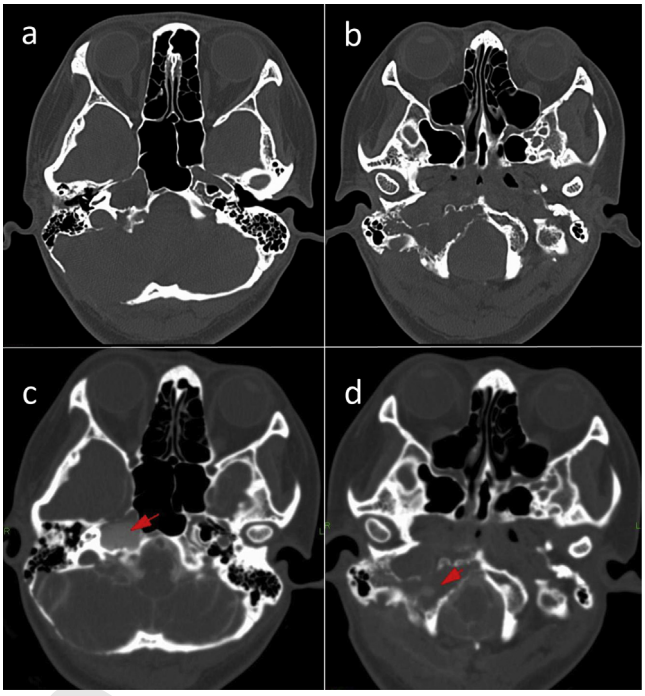
no reports of combined cases of CSF leakage and Chiari I malformation with Gorham's syndrome. Here, we report an unusual case of an acute presyrinx state complicated by bacterial meningitis due to CSF leakage and Chiari malformation associated with Gorham's syndrome involving the skull base.

2. Case presentation

A 25-year-old woman had been treated for meningitis 5 years earlier in another hospital. Computed tomography (CT) revealed a lytic lesion of the right petrous apex, craniocervical junction, and right occipital bone. Biopsy of the osteolytic lesion in the right occipital bone was performed. The histopathological diagnosis was Gorham's syndrome (Fig. 1a, b). A fistula of the dura mater in the right petrous bone was expected due to a past history of meningitis. However, there was no pneumocephalus and no obvious CSF leakage. Therefore, the patient was treated with intravenous antibiotics for 1 month and discharged without any neurologic complications.

The patient was followed up subsequently for 5 years and showed no symptoms. She was then transferred to our hospital due to relocation. The patient was followed up as an outpatient, until she developed a headache with high fever for 3 days. The headache became progressively worse, and she developed impaired consciousness with a Glasgow Coma Scale score of 14 (E3 M5 V6). Physical examination revealed a stiff neck and Kernig's sign was positive. No CSF rhinorrhea was evident. There was low-tone hearing loss in the right ear. Laboratory tests revealed a leukocyte count of 17,700 cells/μl with 98% polymorphonuclear cells, and C-reactive protein (CRP) of 0.09 mg/dl. Lumbar puncture revealed a normal opening pressure of 10 cm H_2O (normal: 5–15 cm H_2O). CSF cytology demonstrated increased white blood cells up to 7160 white cells/μl (98% were polymorphonuclear cells), elevated levels of protein (658 mg/dl; reference range: 15–45), and low glucose concentration (<1 mg/dl; reference range: 40–70 mg/dl). These findings suggested bacterial meningitis. CT revealed a lytic lesion of the right petrous apex and craniocervical junction (Fig. 2a, b). CT cisternography showed leakage of contrast medium in the right petrous apex and occipital condyle (Fig. 2c, d). Axial magnetic resonance imaging (MRI) revealed CSF leakage in the right petrous apex (Fig. 3a). Development of mild tonsillar ectopia was evident on sagittal fluid-attenuated inversion recovery MRI, with the tip of the cerebellar tonsils protruding 7 mm below the foramen magnum with a C2-5 mild high-intensity area (Fig. 3b). The patient has hospitalized with bacterial meningitis. Ceftriaxone 12 g/day and vancomycin 1.25 g/day were started.



Q2 **Fig. 2 – CT and CT cisternography on admission. Noncontrast axial CT. Thinner skull base and partially lytic lesion in the right petrous apex and occipital bone (a, b). CT cisternography (3 h after intrathecal injection of iotrolan) revealing asymmetric CSF leakage (red arrow) at the right petrous apex (c) and occipital condyle (d). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)**

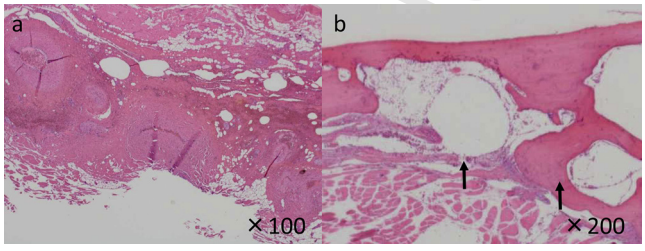


Fig. 1 – Biopsy of osteolytic bone showing fibrosis, vascular and lymphatic channels. Hematoxylin and eosin, ×100 (a). Biopsy of osteolytic bone showing proliferation of thin walled vascular channels (arrow) and osteolytic bone. Hematoxylin and eosin, ×200 (b).

On hospital day 3, the patient developed paresthesia. Sagittal MRI revealed upper spinal edema with syringomyelia, with the tip of the cerebellar tonsils protruding 20 mm below the foramen magnum on midline sagittal images (Fig. 4a, b). MRI of the cervical spine revealed swelling of the cervical cord at the C2-T4 level, with an intramedullary hyperintensity on T2-weighted images (T2WI) and hypointensity on T1 weighted images (T1WI). The diagnosis was a presyrinx state due to exacerbation of tonsillar ectopia associated with intracranial hypertension due to bacterial meningitis or bacterial myelitis. A CSF culture yielded *Streptococcus agalactiae*. We started penicillin G 12 g/day and concentrated glycerin 400 ml/day. We decided to follow up by continuing the antibiotic and concentrated glycerin. We scheduled foramen magnum decompression in case the symptoms did not improve. The meningitis and other symptoms were treated conservatively.

On hospital day 15, MRI revealed improvement of the swelling of the cervical cord and tonsillar ectopia (Fig. 4c, d).

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