# A Case of Intratemporal Rhabdomyosarcoma in a Child Presenting with VII$^{th}$, IX$^{th}$, and X$^{th}$ Cranial Nerve Paralysis

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**Objective: Rhabdomyosarcoma is the most common soft tissue tumor in children, with average age of onset being 5 years, and approximately 70% cases diagnosed below 10 years of age. It accounts for 37% of primary head and neck malignancies in children. Chemotherapy with surgery, and radiation is selected as the primary treatment. We report a rare case of rhabdomyosarcoma in the temporal bone presenting with glossopharyngeal and vagus nerve paralysis as well as facial palsy.**
**Case report: The patient was a 6-year-old boy, and his initial symptom was dizziness followed by facial palsy and hoarseness. Although a severe type of otitis media was suspected in the first clinic, CT and MRI showed a temporal bone tumor with parameningeal extension. Biopsy with cortical mastoidectomy revealed an embryonal-type rhabdomyosarcoma. Pretreatment re-excision was abandoned because of parameningeal involvement. The tumor disappeared after a series of chemotherapy, however, meningeal dissemination occurred, and he eventually died even after an additional administration of anti-cancer agents and intensive modulated radiation therapy.**
**Conclusion: In the case of facial palsy concomitant with other cranial nerve paralysis, care must be taken into neoplastic origin. Early image diagnosis may offer a chance of complete resection in addition to chemoradiotherapy.**

**Key words: embryonal rhabdomyosarcoma, facial palsy, glossopharyngeal nerve, vagus nerve, chemoradiotherapy**

## INTRODUCTION

Rhabdomyosarcoma (RMS) is a malignant tumor that originates from undifferentiated mesenchymal cells. It is estimated that approximately 2.9% of all malignant tumors in children and approximately 90 cases per year occur in Japan. Two-thirds of all RMS cases occurred in children under 6 years of age. Approximately 35% of pediatric RMS cases are reported to occur in the head and neck region, and the most frequently involved site is the orbit (33%), followed by the oral and pharynx (29%), and the face and neck (24%), whereas temporal bones are uncommon (3%) [1]. Parameningeal extension of the head and neck RMS likely involves the cranial nerves, especially the facial, trigeminal, and abducent nerves, but infrequently results in lower cranial nerve disorders [2].

Herein, we report an extremely rare case of RMS in the temporal bone presenting with glossopharyngeal and vagus nerve disorders along with facial nerve paralysis.

## CASE REPORT

The patient was a 6-year-old boy. He had experienced slight dizziness and had a medical examination at the pediatrics department of a nearby general hospital, but no problems were noted. Twenty days later, his parents noticed his left facial paralysis and hoarseness. After 3 days, he complained of left ear pain and visited an ENT clinic. He was treated with antibiotics for the diagnosis of acute otitis media. A few days later, he was diagnosed with left facial nerve palsy and was referred to the Department of Otolaryngology, Tokai University Hospital. The patient had no special medical history.

At the first visit, left facial palsy was assessed as 6/40 points using the Yanagihara scale and House–Brackmann grade 5. Otoscopy of the left ear revealed marked swelling and discharge in the medial part of the ear canal; therefore, the eardrum was not identified (Fig. 1A). A pure tone audiogram revealed a 50 dB conductive hearing loss in the left ear (Fig. 1B). Blood examinations revealed no elevation of the white blood cell count and C-reactive protein level.

Initially, a severe type of acute otitis media (acute mastoiditis) complicated with facial nerve palsy was suspected; however, a temporal bone CT scan revealed soft tissue density with extensive bone destruction from the tympanic cavity to the mastoid. The facial canal bone was completely destroyed, and the posterior skull base was greatly devastated (Fig. 2A). Gadolinium-enhanced MRI revealed a 3 cm-sized irregular mass

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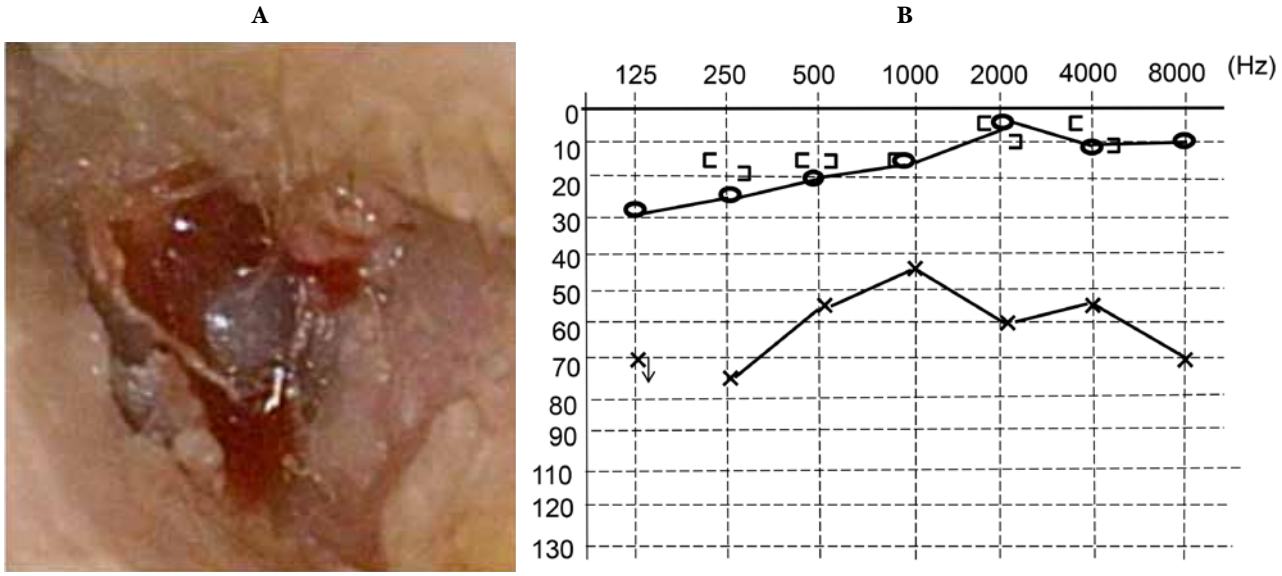


**Fig. 1** Initial ear findings
A. Otoscopy of the left ear revealed swelling and discharge in the medial ear canal.
B. Pure tone audiogram showed 50 dB conductive hearing loss in the left ear.

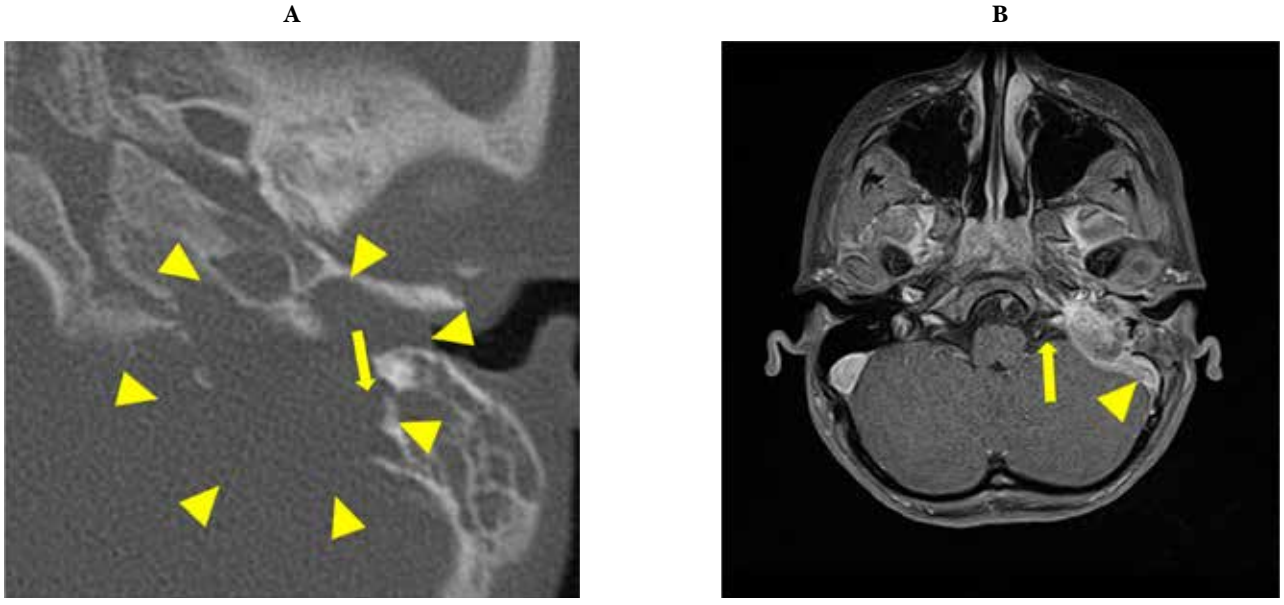


**Fig. 2** Pretreatment images
A. Computed tomography axial views presenting a tumor in the left temporal bone (*) with wide bony destruction of posterior cranial fossa (arrowhead). Arrow indicates uncovered facial nerve.
B. T1 weighted magnetic resonance imaging with Gd enhancement showing tumor (*) invading the sigmoid sinus (arrowhead) and glossopharyngeal nerve and vagus nerve (arrow).

lesion invading the posterior fossa and sigmoid sinus (Fig. 2B). Additional physical examination revealed left soft palate elevation failure and left vocal cord paralysis (Fig. 3), suggesting a combination of glossopharyngeal and vagus nerve paralysis. This was thought to be the cause of hoarseness that had been observed from the onset. Incomplete jugular foramen syndrome was considered to be associated with the tumor extending to the sigmoid sinus through the jugular bulb, which was consistent with the MRI findings. Biopsies were performed under general anesthesia. The tumor was exposed after retroaural incision and was confirmed to have destroyed the mastoid air cells multicentrically (Fig. 4). The histopathological diagnosis was embryonal RMS (Fig. 5).

The parameningeal tumor had advanced into the next stage; therefore, preoperative classification was assessed as stage 3 according to the Japan rhabdomyosarcoma study group (JRSG) classification [3]. Since only biopsy was performed, the postoperative group classification was III, which was evaluated as an intermediate-risk group. These assessments led him to undergo 39 weeks of JRSG chemotherapy (vincristine 0.05 mg/kg, cyclophosphamide 73 mg/kg, actinomycin D 0.045 mg/kg). Local radiation therapy (total 50.4 Gy) was also administered. At 23 weeks of treatment, otoscopy revealed an almost normal left ear canal and a normal eardrum (Fig. 6A). The tumor had

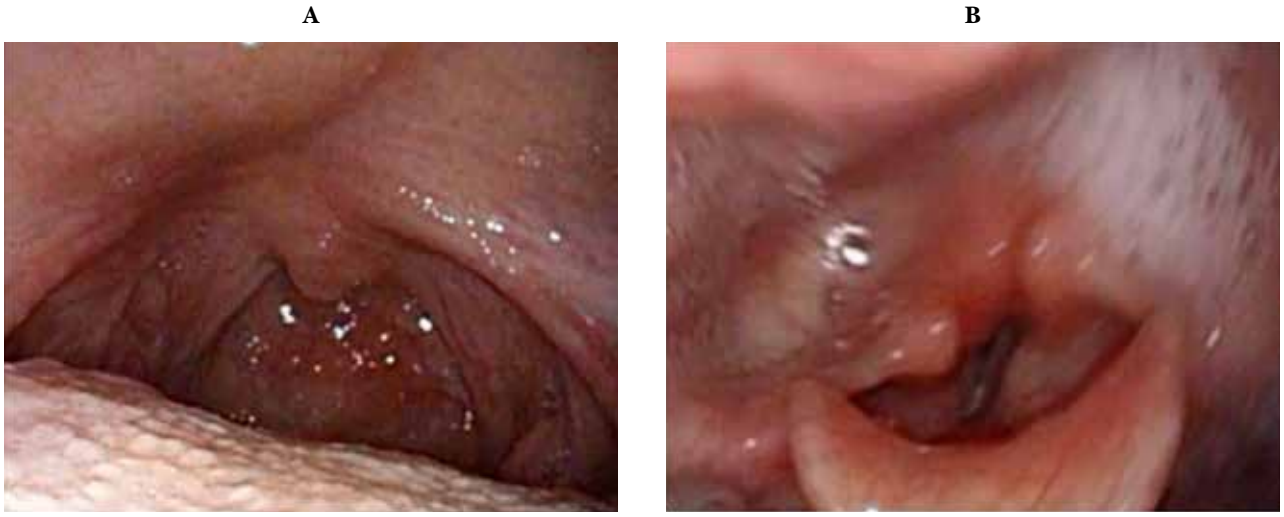


**Fig. 3** Lower cranial nerve paralyses
A. left soft palate elevation failure.
B. left vocal cord paralysis

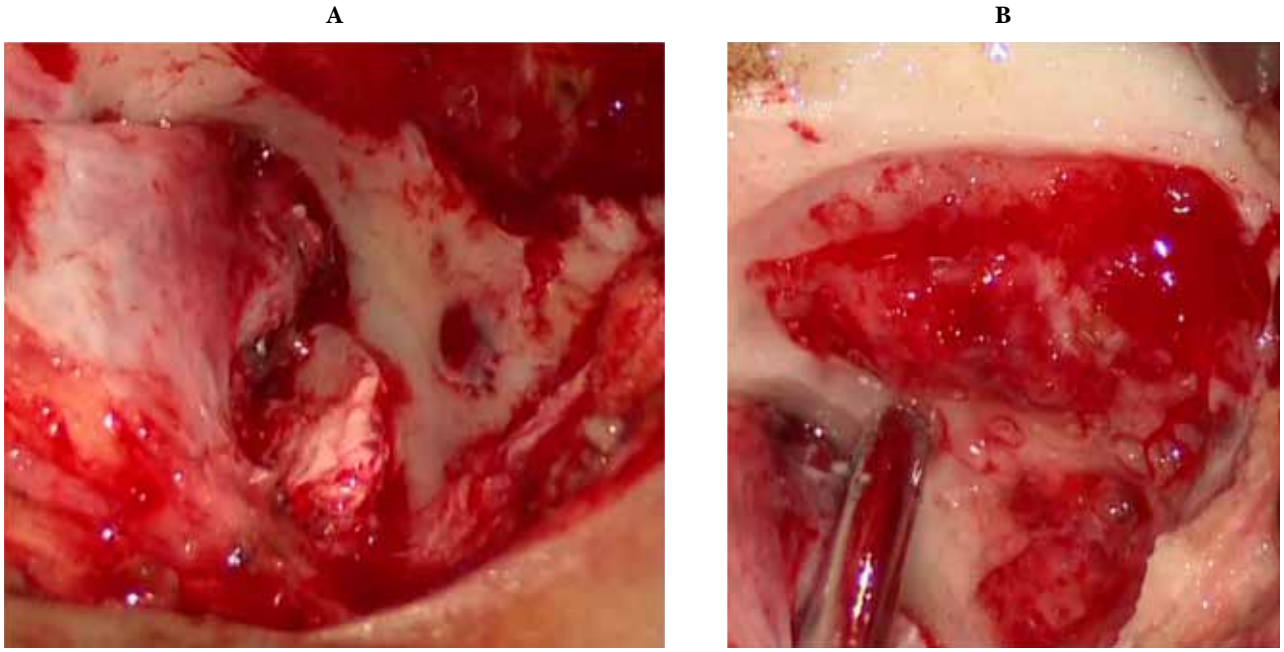


**Fig. 4** Intraoperative findings during tumor biopsy
A. The mastoid cortex bone was destroyed (arrow). EAC: external auditory canal
B. Multicentric pathologic granulation (*) was observed in the ear canal and mastoid cavity after cortical mastoidectomy.

almost disappeared on checking with MRI (Fig. 6B).

However, at 34 weeks, the patient complained of nausea and vomiting. Dizziness was not reported, and gaze/non-gaze nystagmus was not observed. A lumbar tap was performed, and cytological examination of the cerebrospinal fluid revealed a class V malignancy. The patient was then diagnosed with meningeal dissemination of RMS. Although an additional administration of anti-cancer drug (irinotecan 2.2 mg/kg + vincristine 0.07 mg/kg) and whole brain and spinal cord irradiation by intensive modulated radiation therapy were performed, he died 1 year and 5 months after the first visit. The entire clinical course is shown in Fig. 7.

## DISCUSSION

RMS may originate at any anatomical site and occurs predominantly in the head and neck regions, especially in the orbits, skull base, nasal cavity, and nasopharynx, even where there is little or no musculoskeletal tissue. In pediatric cases, approximately 30–40% of tumors occur in the head and neck regions, but the ear and temporal bones are uncommon sites of involvement [1, 4]. Half cases of parameningeal RMS are reported to complicate cranial nerve palsy and cranial nerve disorders II to VII, IX, X, and XII. Above all, facial nerve palsy was most common in 34% of cases, whereas IX and X were relatively rare. The combination of these three nerves, such as this case,

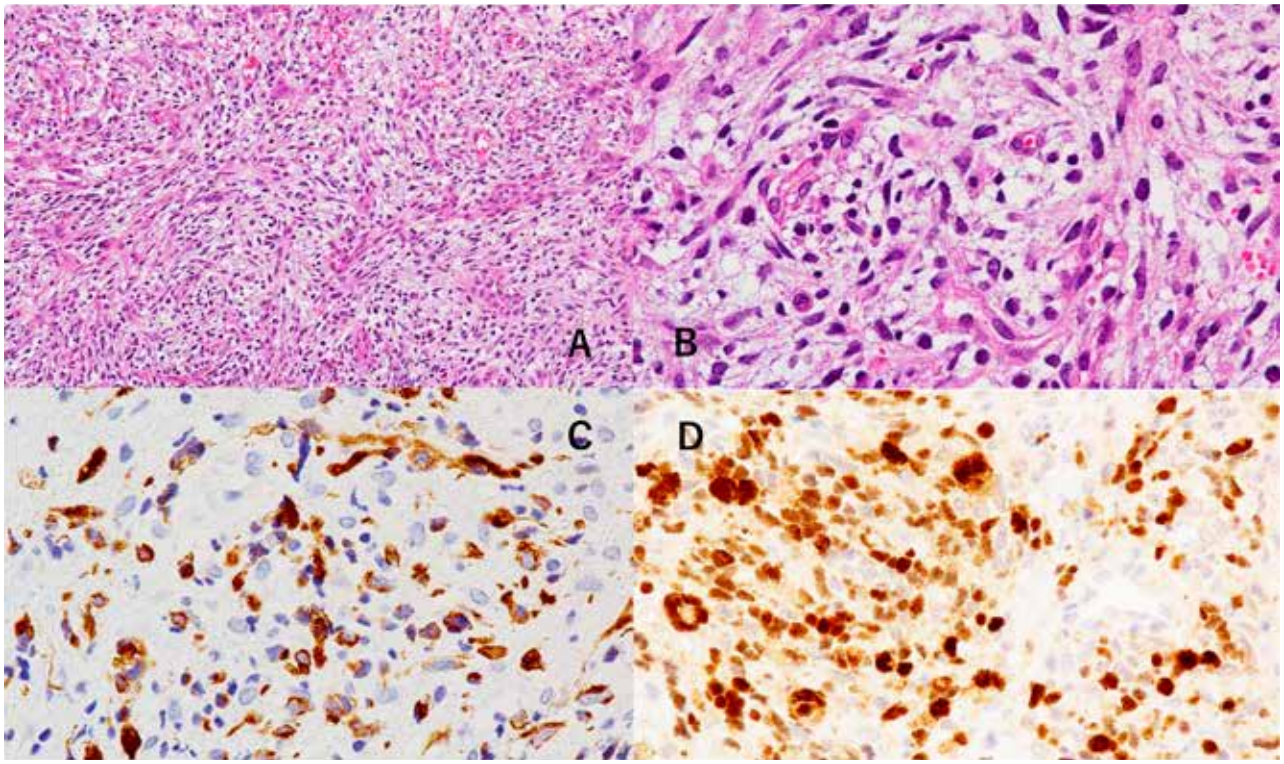


**Fig. 5** Histopathological diagnosis as an embryonal type of rhabdomyosarcoma
A, B. Hematoxylin-eosin stain (A: low power field, B: high power field).
C, D. Immunostains to desmin and myogenin.

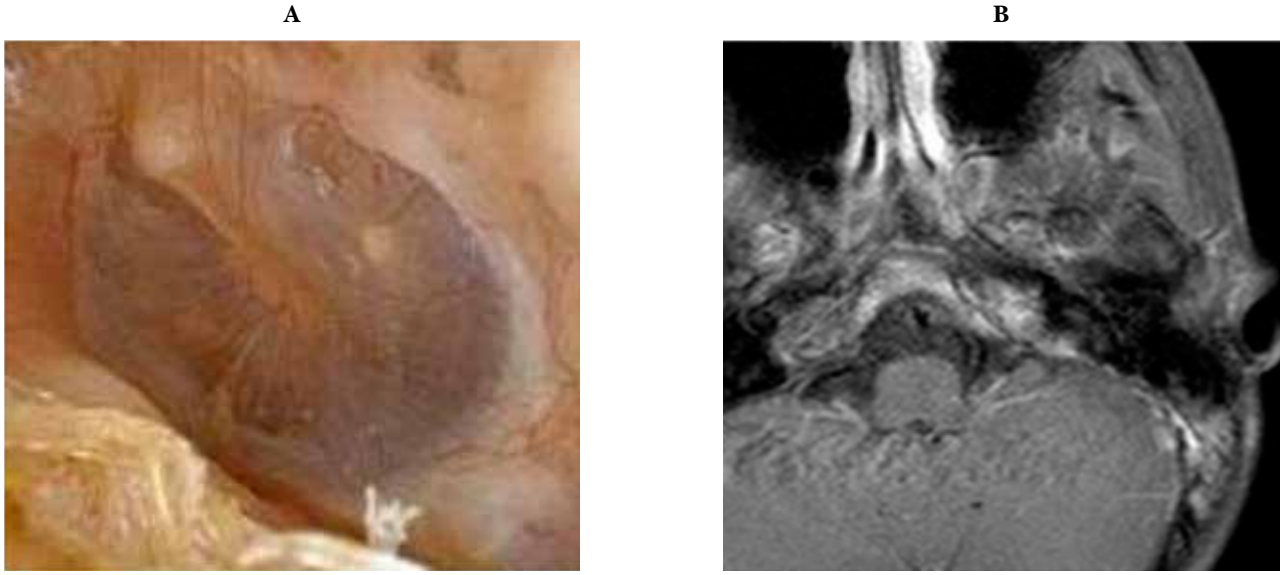


**Fig. 6** Findings at 23 weeks of treatment
A. Otoscopy of left ear clearly showed the normal ear drum.
B. Tumor seemed to disappear in magnetic resonance imaging.

was found in only one case in the literature [2].

Treatment modalities for RMS are selected according to the JRSG preoperative and postoperative stage classifications and the following risk assessment [3]. Multimodality treatment, using a combination of surgery, chemotherapy, and radiotherapy, has been seen to result in better outcomes [3, 4].

Although the role of surgery is limited in the treatment of RMS [5, 6], pretreatment re-excision after biopsy or delayed primary excision following induction chemotherapy, which allows for reduction in radiotherapy dosing, can be considered if complete tumor resection is anticipated [6, 7]. In the case of a malignant tumor localized within the temporal bone with facial palsy alone, a subtotal temporal bone resection may be indicated, even though the patient would lose VII and VIII functions and needs concomitant reconstructive surgery. However, this case initially showed glossopharyngeal and vagus nerve disorders, which are signs of extratemporal parameningeal extension; therefore, surgical intervention seemed to have posed limitations. Further technical advancements are expected [8].

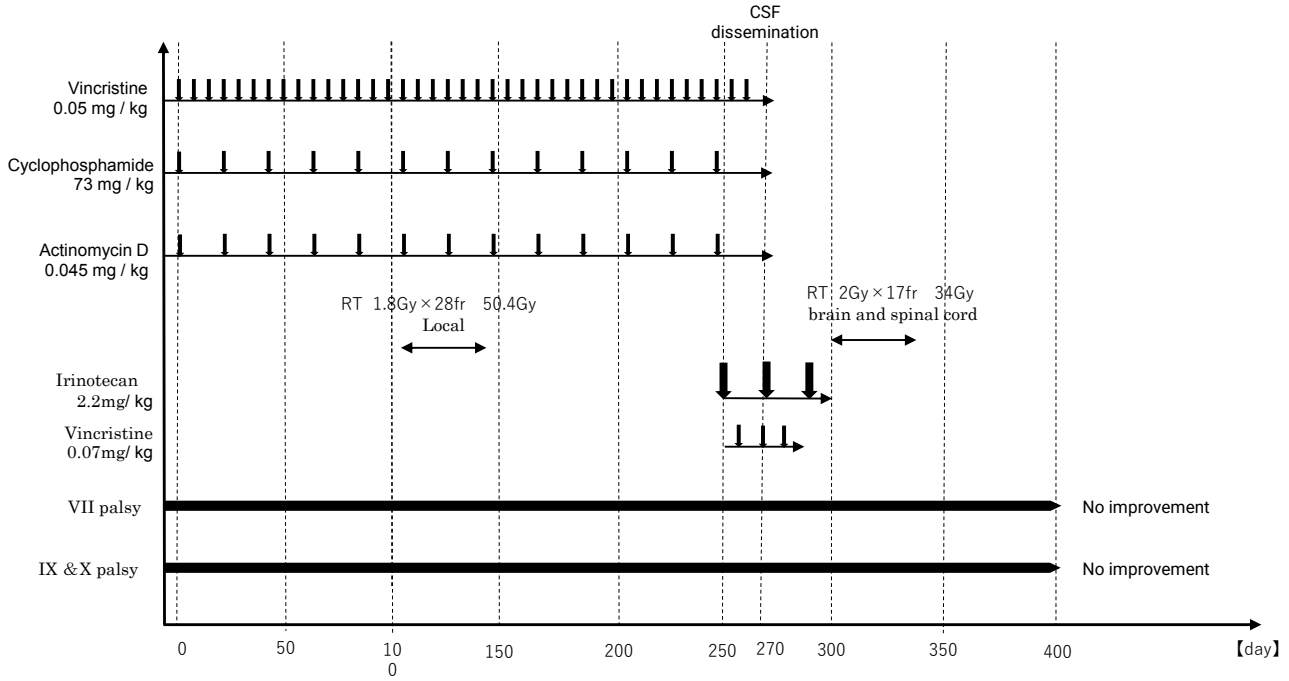


**Fig. 7** Entire course of treatment
After meningeal dissemination in the cerebrospinal fluid, additional chemotherapy and intensive radiotherapy were administered.

During the course of treatment, facial palsy and soft palate/vocal cord paralysis did not improve even after the tumor disappeared. Reportedly, 78% of patients with facial palsy experience paralysis unrecovered after treatment [2]. The reason for no recovery may be tumor infiltration to the nerve trunk itself, which causes total denervation, unlike nerve compression by schwannomas.

According to the features of facial nerve paralysis of neoplastic origin by Jackson *et al.* [9], this case could be initially diagnosed as the tumor origin based on multiple cranial nerve symptoms. In younger children, it is generally difficult to identify subjective symptoms, especially hoarseness and dysphagia; therefore, careful examination of cranial nerve findings is essential. In this case, the involvement of the IX and X nerves had an impact on the decision of treatment in addition to obtaining the diagnosis. If facial palsy concomitant with other cranial nerve paralysis is observed, a tumorous disease is strongly suspected, and earlier imaging should be planned with subsequent biopsy.

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