INTRODUCTION

Intracranial extraskeletal myxoid chondrosarcoma (EMC) is an extremely rare disease, with only 13 cases reported since its first description in 1972 [1-13]. However, due to lack of precise diagnostic criteria and histologic/genetic evidence of previously reported cases, it is uncertain if all reported cases are true intracranial EMC [14]. We hereby present a new case of intracranial EMC in fourth ventricle. We reviewed previously reported cases of intracranial EMCs to validate their diagnosis and to find out if there are clinical similarities among them.

CASE REPORT

A 36-year-old male presented with dizziness persisting for 2 weeks. MRI of the patient showed well-enhanced mass of fourth ventricle. The tumor showed low signal intensity on T1, high signal intensity on T2-weighted image and showed homogenous enhancement on contrast enhanced T1-weighted image (Fig. 1). Initial impression of diagnosis was hemangioblastoma, but preoperative angiography did not show any staining of the tumor. The tumor was approached with midline suboccipital craniotomy and telovelar approach. The tumor was attached to the choroid plexus of the rhomboid fossa; there was no other attachment to adjacent structures. The tumor was completely removed en bloc (Fig. 2). The whole operation was done under MRI navigation guidance and facial motor evoked potential monitoring.

Microscopic examination of the specimen showed spindle shaped cells with scant eosinophilic cytoplasm on basis of abundant myxoid stroma. Overall pathologic morphology resembled that of a mesenchymal tumor (Fig. 3). Immunohistochemistry (IHC) staining results showed negative in glial markers isocitrate dehydrogenase 1 and glial fibrillary acidic protein (GFAP). Final pathology report was EMC. Immediate postoperative MRI showed no remnant enhancing lesion suggesting complete removal of tumor. The patient recovered well without any significant neurologic deficit, except mild dysphagia and hypesthesia of left thigh. Adjuvant radiotherapy (54 Gy/27 fx) start-
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mas and chordomas from EMCs. Epithelial membrane antigen staining shows positive in most cases of chordoid gliomas, chordomas, and chordoid meningiomas while only 25% of EMCs show positivity. GFAP staining is used to differentiate EMCs from gliomas like pilocytic astrocytoma [17]. Of 13 case reports, 10 studies had at least one IHC result reported. The results are described in Table 2. Neuronal markers such as GFAP, neuron-specific enolase, synaptophysin all showed negative, while vimentin was positive in all tested patients, and most of the patients showed negative in cytokeratin and EMA staining.

Fig. 3. Pathologic findings. A: Low power view (H&E stain, $\times 12.5$) shows a somewhat well demarcated mass. B: In the mass, myxoid stroma (oval circle) is noted (H&E stain, $\times 200$). C: High power view (H&E stain, $\times 400$) shows haphazardly arranged spindle cells with myxoid stroma. D: The GFAP immunohistochemistry shows negative reaction (GFAP stain, $\times 400$). These findings are compatible with extraskeletal myxoid chondrosarcoma. H&E, hematoxylin and eosin; GFAP, glial fibrillary acidic protein.

Fig. 4. Postoperative MRI at 3 months after surgery. T1-weighted enhanced axial (A) and sagittal (B) images show no residual tumor.
| Study            | Sex/age | Location                | Size (cm)          | Extent of resection | Adjuvant treatment                  | Prognosis                                      | Etc                                      |
|------------------|---------|-------------------------|--------------------|---------------------|-------------------------------------|------------------------------------------------|------------------------------------------|
| Scott et al. [1] | M/39    | Fourth ventricle        | N/A                | Subtotal removal (50%) | N/A                                | V-P shunt after 5 days → deceased 13 days postop | Rock-hard consistency, calcification, decalcification needed |
| Smith et al. [2] | M/12    | Left cerebellum         | 3.5×1.0×0.5        | Total removal       | None                                | No recurrence 13 mo                           | Foramen magnum dura, bone not involved   |
| Salcman et al. [3]| F/28    | Left parafalcine        | 7×5×4              | Total removal → reop | 125-I radioactive seeds brachytherapy | Recur 8 mt → reop → no recur 12 mo           | N/A                                      |
| Sato et al. [4]  | F/43    | Pineal region           | N/A                | Subtotal removal    | Radiotherapy (6,000 cGy) + chemotherapy | Deceased 37 mo after diagnosis                | Leptomeningeal seeding                   |
| Sala et al. [5]  | F/55    | Right cerebellum        | N/A                | Total removal       | None                                | Recur 10, 16, 31, 43 mo (reop) → deceased 7 yrs | Right petrous bone origin                |
| González-Lois et al. [7] | F/17 | Right fronto-temporal convexity | Less than 3 | Total removal → reop | Radiotherapy (6,000 cGy)            | Recur 16 mo → reop → recurrence 4 mo         | N/A                                      |
| Chaskis et al. [6] | M/69    | Right frontal           | 1.5                | Total removal       | None                                | Deceased 1 mo                                | Subcortical, parenchymal                 |
| Im et al. [8]    | M/43    | Left parietal           | 1.2×1.0×1.0        | Total removal       | Radiotherapy (5,940 cGy)            | No recur 36 mo                               | Subcortical, parenchymal                 |
| Sorimachi et al. [10] | F/37    | Pineal region           | N/A                | Subtotal removal → reop | None                                | Progression 14 mo → reop → no recur 7 mo    | Tumor bleeding, calcification in 2nd path, S100 (-) → (+) |
| O’Brien et al. [9] | F/26    | Left cerebellon pontine angle | 2.5             | Biopsy → subtotal   | Proton therapy                      | No recur 12 mo                               | 20 wk pregnant                           |
| Park et al. [12] | F/21    | Left thalamus and lateral ventricle | 3.2×6.3×4.8 | Total removal       | Radiotherapy (6,080 cGy)            | No recur 6 mo                                | Parenchymal                              |
| Dulou et al. [11] | F/70    | Left parasagittal       | N/A                | Neoadj RTx → total removal | Chemotherapy (ifosfamide)          | Recur 3 mo → CTx → deceased 10 mo            | Breast cancer patient                    |
| Qin et al. [13]  | F/41    | Left cerebellum         | 3×3×3              | Subtotal removal    | Radiotherapy (4,060 cGy)+ chemotherapy (TMZ) | No recur 20 mo                              | Poor dissection margin, peritumoral bleeding, skull destruction |
| Present case     | M/36    | Fourth ventricle        | 1.6                | Total removal       | Radiotherapy (5,400 cGy/27 fx)      | No recur 3 mo                               | Medulla dorsal surface, no bone involvement |
ed choroid plexus, remaining multipotent mesenchymal cells of skull base or dura mater as possible origins of the tumor. To our knowledge, this case is the second intraventricular EMC reported [12] and first EMC to be histologically confirmed with IHC results that formed inside fourth ventricle.

Optimal treatment of EMCs is still debated. In most of the cases the tumor was separated well from surrounding tissue. In 9 out of 14 cases, including present case, the tumor was totally removed by surgery. However, it is unclear if the extent of removal is associated with lower recurrence or better prognosis. Three cases reported recurrence of tumor after total removal. Adjuvant therapy also varied widely. A recent phase II trial of pazopanib in advanced EMCs showed anti-tumor activity, but it is even more difficult to treat intracranial EMCs because central nervous system permeability needs to be considered [22]. Five cases have undergone adjuvant radiotherapy, of which two with concurrent chemotherapy. Further studies need to be done regarding necessity and efficacy of adjuvant treatment.

In conclusion, we present a rare case of intracranial EMC originated from the choroid plexus inside fourth ventricle. IHC staining results are crucial in differential diagnosis of intracranial EMCs from other intracranial tumors mimicking its pathologic morphology. Optimal treatment of the disease is still debated; more cases and further studies are needed regarding the necessity of radical excision, adjuvant chemotherapy and radiotherapy.

Ethics Statement

The Institutional Review Board of Severance Hospital exempted informed consent due to its retrospective nature and minimal risk for harm to the patient, and this report was conducted according to the guidelines of the Declaration of Helsinki for biomedical research.

Availability of Data and Material

All data generated or analyzed during the study are included in this published article (and its supplementary information files).
Intracranial EMC in Fourth Ventricle

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Conflicts of Interest
The authors have no potential conflicts of interest to disclose.

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