Upon admission to our hospital, she complained of both visual and hearing loss. She also showed a grade II weakness of the right extremities. MRI revealed hydrocephalus and a 32×63×48-mm sized well-enhanced mass around the right thalamus. We also observed severe compression of the third ventricle and diffuse peritumoral edema. An enhanced MRI scan revealed a lobulated, heterogeneous enhanced tumor (Fig. 1). A computed tomography (CT) scan of her brain also showed a well-enhanced mass, as well as multiple lobulated spots with hypodensity within the mass. To decompress her increased intracranial pressure, we underwent operation to remove the mass completely, using a transcortical approach to the posterior horn of the left lateral ventricle. A left parietal craniotomy and a cortisectomy at the superior parietal lobule were performed. The tumor was brown in color, and the margin between the brain parenchyma and supporting tissue was clearly distinguishable. The tumor was hypervascularized, hard, and lobulated, and it was completely removed in piecemeal fashion. A frozen biopsy of the tumor suggested that it was a pilocytic astrocytoma. The tumor was found to be composed of strands or cords of oval and spindle cells embedded in abundant myxoid stroma (Fig. 2A).
Intracranial Myxoid Chondrosarcoma | JH Park, et al.

eral ventricle.

Preoperative imaging methods, including CT and MRI, revealed similar results in previous patients with intracranial extraskeletal myxoid chondrosarcomas. Precontrast CT scans have shown isodensity of tumors in five of the seven previously reported patients; however, it should be noted that the two remaining patients had preoperative tumor bleeding16,17). Most dispersed chromatin, and a moderate amount of eosinophilic cytoplasm that was often finely vacuolated (Fig. 2B). Mitotic figures were rarely observed.

The tumor cells were further examined by immunohistochemistry, and antibodies were used at the dilutions listed. Tumor cells were found to be focally and strongly positive for epithelial membrane antigen (1 : 25, Dako, Glostrup, Denmark) (Fig. 2C), weakly positive for class III β-tubulin (1 : 200, clone TU-20, Genetex, Irvine, CA, USA), diffusely positive for microtubule-associated protein 2 (1 : 200, clone AP18, Neomarkers, Fremont, CA, USA) (Fig. 2D), and positive for vimentin (1 : 250, Zymed, San Francisco, CA, USA) (Fig. 2E). In contrast, the tumor cells were negative for S-100 protein (1 : 1000, Zymed, San Francisco, CA, USA), cytokeratin (1 : 250, Zymed San Francisco, CA, USA), and glial fibrillary acidic protein (GFAP, 1 : 200, Biogenex, San Ramon, CA, USA). Final pathologic analysis of the above results led to a diagnosis of extraskeletal myxoid chondrosarcoma. Postoperative MRI showed no residual tumor. The patient then underwent adjuvant radiotherapy, at a total dose of 6080 cGy, as well as rehabilitation. After six months of treatment, the headache and weakness symptoms had improved to grade IV, but other neurologic deficits, including blindness and deafness, were unchanged.

DISCUSSION

Histologically, three subtypes of cranial and intracranial chondrosarcomas have been described: classic, mesenchymal, and myxoid15,79. Intracranial extraskeletal myxoid chondrosarcomas are extremely rare, with only seven cases previously reported to date2,5,8,13-16). A summary of these previous patients, including imaging results, the surgical extent of the tumor, postoperative radiation treatment, and patient outcomes is shown in Table 1.

Intracranial extraskeletal myxoid chondrosarcomas are thought to originate from the dura, leptomeninges, parenchyma, and choroid plexus2,5,13-16). Our findings in the present case suggest that the tumor originated from the choroid plexus of the lateral ventricle.

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Fig. 1. Imaging findings for the current patient. A: T1-weighted magnetic resonance imaging (MRI) showing homogeneous iso-signal intensity of a 63-mm tumor in the left lateral ventricle along with ventricular dilatation. B: T2-weighted MRI showing a heterogeneous high signal intensity tumor and peritumoral edema. C: T1-weighted enhanced MRI showing strong enhancement of the tumor.

Fig. 2. Histologic features of the tumor in the current patient. A: The tumor consists of strands or cords of oval cells and abundant myxoid stroma (H&E, ×100). B: The tumor cells interconnected to form cords and had relatively uniform oval nuclei and a moderate amount of eosinophilic cytoplasm (H&E, ×400). C, D and E: These tumor cells are positive for epithelial membrane antigen (original magnification ×400) (C), microtubule-associated protein 2 (original magnification ×400) (D), and vimentin (original magnification ×100) (E) by immunohistochemical staining.
We describe a case of intracranial extraskeletal myxoid chondrosarcoma and review the literature on these rare tumors. Despite their malignant nature, these tumors have well-defined margins and are clearly distinct from normal brain tissue. Intracranial extraskeletal myxoid chondrosarcoma has a high rate of leaving calcifications behind and is associated with a low risk of local recurrence. In this report, we present a case of intracranial extraskeletal myxoid chondrosarcoma with complete resection and no recurrence, and we discuss the treatment options for these rare tumors.

**Table 1. The characteristics of previous cases of intracranial myxoid chondrosarcoma**

| Authors          | Reference, year | Age (years)/sex | Location/origin                        | Size                | Surgical extent | Postoperative radiotherapy, dose | CT                  | MRI                  | Postoperative course |
|------------------|-----------------|-----------------|----------------------------------------|---------------------|-----------------|-------------------------------|---------------------|---------------------|----------------------|
| Scott et al.     | (15), 1976      | 39/male         | 4th ventricle, choroid plexus          | Not described       | STL             | Not done                      | Not done            | STL                 | 13 days died d/t ventriculitis |
| Salcman et al.   | (13), 1992      | 28/female       | Left parafalcine & dura of falx       | 70×50×40 mm         | TR              | Not done                      | Isodense enhancement (+) | T1 : hypointense T2 : hyperintense enhancement (+) | 20 months alive and local recurrence |
| Sato et al.      | (14), 1993      | 43/female       | Pineal gland & dura                   | Not described       | PR              | Yes, 6000 cGy                  | Enhancement (+)    | Not done            | 3 years d/tumor progression |
| Chaskis et al.   | (2), 2002       | 69/male         | Right F. cortex                       | Not described       | TR              | Not done                      | Not described       | Enhancement (+)    | 1 months died with septic shock d/t diverticulitis |
| González et al.  | (5), 2002       | 17/female       | Right F-P cortex                      | 23×20 mm            | TR              | No, 6000 cGy RTx was performed after 1st recurrence | Not described       | T1 : hypointense T2 : hyperintense enhancement (+) | 20 months alive, twice had tumor recurrence |
| Im et al.        | (8), 2003       | 43/male         | Left P. cortex                        | 20 mm               | TR              | Yes, 5940 cGy                  | Isodense enhancement (+) | T1 : hypointense T2 : hyperintense enhancement (+) | 3 years alive, no recurrence |
| Sorimachi et al. | (17), 2008      | 37/female       | Pineal region                         | Not described       | 1st : PR 2nd : TR | Not done                      | Mixed dense enhancement (+) | T1 : mixed intensity enhancement (+) | 1st : 13 months recurrence 2nd : 7 months alive, no tumor recurrence |
| Present case     | 2011            | 21/female       | Left lateral ventricle/choroid plexus | 32×63×48 mm         | TR              | 6080 cGy                       | Isodense enhancement (+) | T1 : hypointense T2 : hyperintense enhancement (+) | 20 months alive, twice had tumor recurrence |

**Conclusion**

We describe a case of intracranial extraskeletal myxoid chondrosarcoma and review the literature on these rare tumors. Despite their malignant nature, these tumors have well-defined margins and are clearly distinct from normal brain tissue. Intracranial extraskeletal myxoid chondrosarcoma has a high rate of leaving calcifications behind and is associated with a low risk of local recurrence. In this report, we present a case of intracranial extraskeletal myxoid chondrosarcoma with complete resection and no recurrence, and we discuss the treatment options for these rare tumors.

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**CT:** computed tomography, **MRI:** magnetic resonance image, **STL:** subtotal resection, **FM:** foramen magnum, **TR:** total resection, **CPA:** cerebropontine angle, **PR:** partial resection, **F:** frontal, **F-P:** frontoparietal, **P:** parietal, **d/t:** due to...
recurrence in previous reports. Here, we present a case report of this tumor treated with total tumor resection and adjuvant radiotherapy.

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