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Malignant transformation of supratentorial clear cell ependymoma.

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Recurrence of clear cell ependymoma is not a rare condition, but malignant transformation of clear cell ependymoma has not yet been well presented. The authors report a 44-year-old man who presented with progressive right hemiparesis. A brain tumor in the left frontal premotor area was removed and an initial pathological diagnosis of oligodendroglioma was made. The tumor recurred 4 months later, and reoperation of the tumor and adjuvant local radiotherapy were performed. The patient subsequently underwent surgical removal of recurrent tumors on another four occasions (6 times in total) during a period of 11 years and finally died of the original disease. Histopathological studies of all surgical and autopsy specimens were carried out. The first and second surgical specimens did not contain any ependymal rosettes or pseudorosettes, and thus a diagnosis of oligodendroglioma was made. However, the third surgical specimen showed pseudorosettes. At this time, the tumor had an ultrastructural appearance compatible with ependymoma. Thereafter, the recurrent tumors showed anaplastic features such as nuclear pleomorphisms and necrosis with pseudopallisading. The autopsy specimens resembled a feature of glioblastoma but the tumor was sharply demarcated from the surrounding parenchyma.

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