

Pineal nongerminomatous malignant germ cell tumor

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Case series

2015

We retrospectively analyzed 17 patients treated for pineal NGMGCTs between 1986 and 2007 at the University of Niigata. **RESULTS:** Twelve patients underwent total or subtotal resection of their tumor via the occipital transtentorial approach. Five patients underwent partial resection, and four of them later underwent total resection by salvage surgery. After surgery, eight patients were treated with combined radiochemotherapy including whole-brain irradiation, two received radiation monotherapy, one had chemotherapy with local irradiation, and six were treated with chemotherapy alone. The median follow-up period for surviving patients was 179 months. The 10-year overall survival and progression-free survival rates for the radiochemotherapy group were both 75.0 % (two patients had a recurrence and died); the rates for other adjuvant therapies were 77.8 % (two died) and 22.2 % (seven had a recurrence), respectively. Radiochemotherapy was significantly associated with an increased rate of progression-free survival compared with the other adjuvant therapies ($p = 0.0396$). **CONCLUSIONS:** For pineal NGMGCTs, initial treatment strategies including gross total resection of the tumor before or after whole-brain irradiation and chemotherapy provided good therapeutic outcomes. Obtaining complete remission of the primary tumor, irrespective of the timing of surgical resection (i.e., before or after adjuvant therapies), or complete response by neoadjuvant radiochemotherapy during an initial treatment appears to be essential for improving therapeutic outcomes of intracranial NGMGCTs ¹⁾.

2014

Qi et al. retrospectively reviewed the records of 143 patients with nongerminomatous pineal region tumors surgically treated via an occipital transtentorial approach between 2000 and 2011. The tumor was small (<2 cm) in 14.7 % of patients, medium (2-4 cm) in 52.4 %, and large (>4 cm) in 32.9 %. **RESULTS:** Gross total tumor removal was achieved in 91.6 % of patients, subtotal in 7.0 %, and partial in 1.4 %. Histological diagnosis was nongerminomatous germ cell tumor in 41.3 %, pineal parenchymal tumor in 14.7 %, glial tumors in 28.7 %, and miscellaneous in 15.4 %. The overall complication and mortality rate was 18.2 % and 0.7 %, respectively. Permanent morbidity occurred in 5.6 % of patients, including hemianopsia in 3.5 % and Parinaud syndrome in 2.1 %. Hydrocephalus was resolved in 82.1 % without surgery for the CSF diversion. Sixty-eight patients with malignant tumors underwent radiotherapy; 35 also received adjuvant chemotherapy. One hundred thirty patients were successfully followed up with a mean duration of 43 months. Finally, 86.9 % of the patients achieved a favorable functional outcome ($mRS \leq 2$), 3.1 % had an mRS score of 3, 1.5 % had an mRS score of 4, and 8.5 % had died ($mRS = 6$). **CONCLUSIONS:** Radical surgery was recommended as the optimal treatment for nongerminomatous pineal region tumors. Favorable results could be achieved by experienced neurosurgeons. Hydrocephalus could be cured by radical tumor removal in the majority of cases. The occipital transtentorial approach was indicated for most pineal region tumors, but surgeon's preference and experience should also be considered. New understanding of the arachnoid membranes of this region may be helpful for tumor resection ²⁾.

1994

3 children with malignant, marker-positive pineal nongerminomatous germ cell tumors treated with a 'sandwich' protocol. Here, we report on the long-term survival of these children. Preoperative chemotherapy consisted of two courses of bleomycin, etoposide, and cisplatin. En bloc resection of the tumors via the supracerebellar, infratentorial route was performed immediately after decline of tumor marker levels. Postoperatively, two courses of vinblastine, ifosfamide, and cisplatin were applied, followed by craniospinal irradiation. The patients showed no major neurological deficits and no evidence (neuroradiologically or with regard to tumor marker levels) of recurrence of disease after 66, 71, and 78 months, respectively. We propose this regimen for children with tumors of the pineal region in whom the tumor markers are positive. It should be started without histological classification of the tumor to avoid possible spillage of malignant tumor cells to the cerebrospinal fluid ³⁾.

1)

Jinguji S, Yoshimura J, Nishiyama K, Yoneoka Y, Sano M, Fukuda M, Fujii Y. Long-term outcomes in patients with pineal nongerminomatous malignant germ cell tumors treated by radical resection during initial treatment combined with adjuvant therapy. *Acta Neurochir (Wien)*. 2015 Oct 20. [Epub ahead of print] PubMed PMID: 26482943.

2)

Qi S, Fan J, Zhang XA, Zhang H, Qiu B, Fang L. Radical resection of nongerminomatous pineal region tumors via the occipital transtentorial approach based on arachnoidal consideration: experience on a series of 143 patients. *Acta Neurochir (Wien)*. 2014 Dec;156(12):2253-62. doi: 10.1007/s00701-014-2224-4. Epub 2014 Sep 23. Erratum in: *Acta Neurochir (Wien)*. 2015 Feb;157(2):349. PubMed PMID: 25246142.

3)

Knappe UJ, Bentele K, Horstmann M, Herrmann HD. Treatment and long-term outcome of pineal nongerminomatous germ cell tumors. *Pediatr Neurosurg*. 1998 May;28(5):241-5. PubMed PMID: 9732256.

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