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# Ependymoblastoma: a clinical review

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## Abstract

Ependymoblastoma is a malignant rarely reported neuroectodermal tumor. The authors describe a further case of cerebral ependymoblastoma and examine the clinical-prognostic aspects of this tumor in the light of the published data.

**Keywords:** Ependymoblastoma, primitive neuroectodermal tumor.

## 1 Introduction

Ependymoblastoma is a rare, highly malignant, neuroectodermal tumor, histologically distinctive and well-differentiated from anaplastic ependymoma [4, 6, 12]. Ependymoblastomas have a remarkably uniform appearance, consisting of an arrangement of densely-packed sheets of small cells intersected by numerous thin-walled blood vessels and forming numerous rosettes. The tumor cells are poorly differentiated, oval or spindle-shaped, with ill-defined cytoplasm and wispy polar processes. The nuclei are round to oval and contain coarse chromatin nodes. Mitotic figures are numerous. The most characteristic feature is the presence of ependymal rosettes and tubules lined with columnar cells: the rosettes are composed of cells forming multiple layers; mitotic figures are frequently demonstrable in a juxtaluminal position [4, 6, 7, 10, 12–14].

Ependymoblastoma, first described by BAILEY and CUSHING in 1926 [2], was identified as a distinct tumor by RUBINSTEIN in 1970 [14]. Since then, 40 more cases have been reported, 27 complete with clinical details [1–14].

We report a further case of ependymoblastoma, bringing the total of detailed reports to 28, and analyze the clinical-prognostic aspects in this group of patients.

## 2 Case report

This 5-year-old girl was hospitalized with a 2-month history of gait difficulty accompanied by headache and vomiting during the second month. A CT scan showed a median, hyperdense lesion invading the IV ventricle with moderate, homogenous enhancement; triventricular hydrocephalus was also present (Figure 1). The patient presented ataxia and bilateral papilledema at neuro-

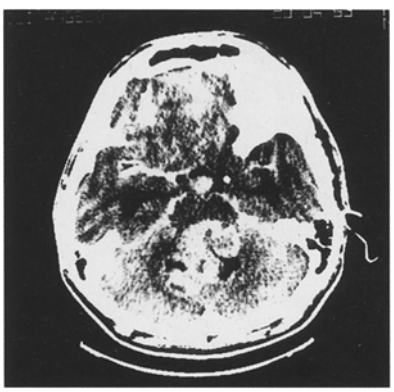


**Figure 1.** CT scan after contrast enhancement showed a cerebellar hyperdense lesion.

logical examination. She was submitted to surgery and an apparently total removal of a grayish, fleshy and circumscribed tumor invading the IV ventricle was achieved. Histologically, the tumor was hypercellular and composed of cells with hyperchromatic nuclei and small central-lumen multilayered rosettes and canals (true rosettes). In the more compact areas, undifferentiated hyperchromatic cells were present and mitoses were numerous. Histological diagnosis was ependymoblastoma (Figures 2, 3 a, b).

The child's parents did not give permission for further chemo- or radiotherapy and she died 3 months after surgery from a recurrence.

## 3 Discussion

In 82% of cases, ependymoblastoma manifests during the first 5 years of life (range: birth to 36 years; median 3 years); in rare cases it was congenital (3 cases; 11%) or appeared in adulthood (3 cases; 11%). Males and females was affected similarly (M : F ratio; 1.1 : 1). Clinical history was generally brief (range 1 week to 7 months; median 3 months). Symptoms and signs are reported in figure 4.

All ependymoblastomas described were intracranial: in 20 (71%) the tumor was situated within the supratentorial compartment, in 7 (25%) in the subtentorial one, and in 1 (4%) in both with infiltration of the tentorium (9). Seventy-nine percent of ependymoblastomas involved the ventricular cavities, particularly when they were situated in the subtentorial compartment (100% vs 60%). This finding sustains the hypothesis that the tumor derives from primitive ventricular cells [10]. From a neuroradiological point of view, ependymoblastoma does not present any characteristic features differentiating it from other primitive neuroepithelial tumors [1, 5]. At CT and MRI, its appearance is that of a dishomogenous (85%) or hyperdense (25%) lesion with dishomogenous (75%) or homogenous (25%) enhancement. Differential diagnosis should include ependymoma, medulloblastoma, astrocytoma, and choroid plexus papilloma or carcinoma.

In our case, the site of the tumor (posterior cranial fossa) and its neuroradiological appearance (hyperdense tumor with moderate enhancement) ini-

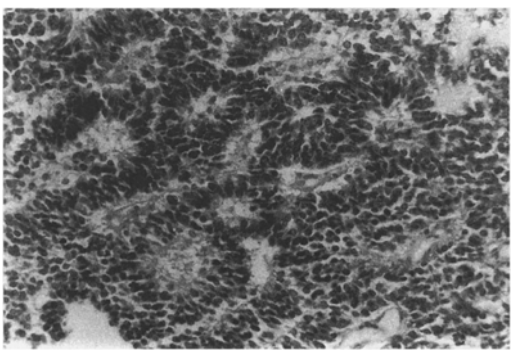
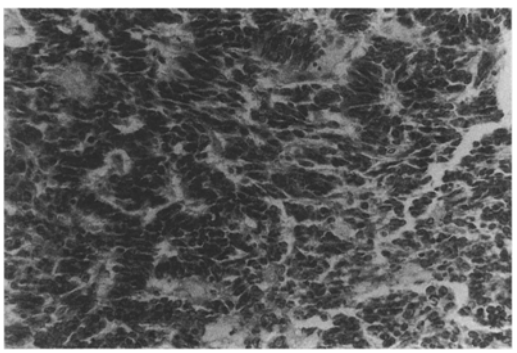
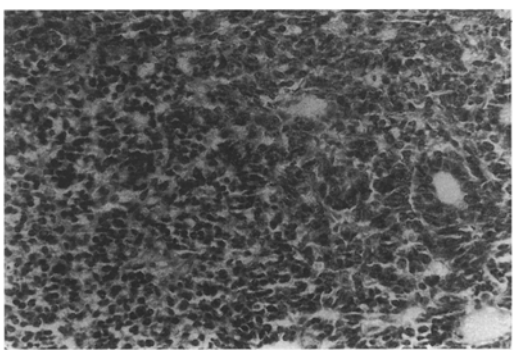


**Figure 2.** Photomicrograph showed uniform tumor cells with rosettes and mitotic figures. Rosettes possessed multiple layers of nuclei and mitosis. H & E, × 200.



a



b

**Figure 3 a, b.** The neoplasm composed of uniform neuroepithelial cells (3a) forming perivascular pseudorosettes, ependymoblastic rosettes (3b). H & E, × 165.

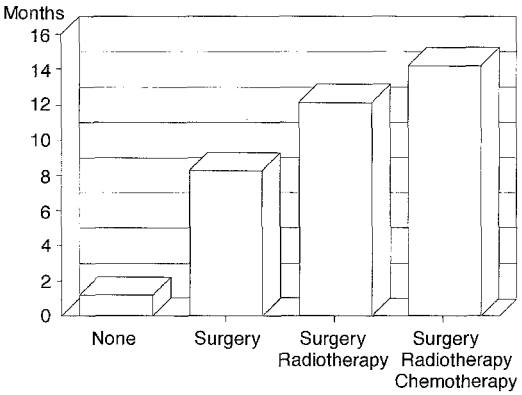



**Figure 4.** Treatment and median survival in 28 cases of ependymoblastoma.

tially led us to believe that the lesion was a medulloblastoma.

Macroscopically, this tumor presents as a well-defined (85% of cases) solid (90% of cases) lesion, varying in size from 1 to 11 cm. In 14 cases (50%), there was leptomeningeal tumor infiltration, massive in 5 of them (36%).

The tumor in our case was well-defined, solid and 5 cm in diameter, and did not present any leptomeningeal involvement.

Surgical exeresis was performed in 22 cases (79%): gross total removal was achieved in 10 (56%) and partial removal in 12 (44%). Postoperative radiotherapy was performed in 17 cases (61%); total dosage was 20–25 Gy to the brain, 40–50 Gy to the posterior cranial fossa, and 20–25 Gy to the spinal cord. Chemotherapy was also administered to 4 patients (14%), using vincristine and ACNU. Of the 28 cases examined, 5 (18%) received no treatment.

Average survival in the 28 cases considered in this study was 8.1 months (range 1 week to 15 months). Only one patient reported by MORK [10] survived for nine years but ependymoblastoma was only diagnosed after a third operation, eight years after clinical onset, following which the patient survived one more year.

The cause of death was always tumor regrowth, reported in all the cases studied. One patient (4%) presented tumor regrowth associated with two small lung metastases. In three patients (11%) there was a CSF dissemination of the tumor and other spinal lesions were observed.

Prognosis was not influenced by the type of treatment performed (Table I): it was worth noting, however, that survival was always longer than average in patients treated by surgery, radiotherapy, and chemotherapy. In a recent study, RICCARDI [11] reported five cases of tumor remission in children with primitive neuroectodermal brain tumor (one of which was an ependymoblastoma) all of whom received a combination of surgery, chemotherapy (carboplatin), and radiotherapy. We think that further therapeutic studies are essential to improve the prognosis for patients with ependymoblastoma.

**Table I.** Symptoms and signs of ependymoblastoma in 28 cases

| | | |
|---|---|---|
| Headache | 28 | 100% |
| Vomiting | 28 | 100% |
| Papilledema | 28 | 100% |
| Cranial vault modifications | 15 | 54% |
| Hemiparesis | 21 | 75% |
| Seizures | 10 | 36% |
| Aphasia | 8 | 29% |
| Ataxia | 7 | 25% |
| Cerebellar deficit | 7 | 25% |

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