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AJNR Am J Neuroradiol 1985, 6 (1) 15-18

<http://www.ajnr.org/content/6/1/15>

This information is current as of May 9, 2025.

Radiologic Characteristics of Primitive Neuroectodermal Tumors in Children

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Radiographic findings are reviewed in 31 children with primitive neural ectodermal tumors seen at the Hospital for Sick Children from 1962 through 1983. Seventeen children were initially evaluated with computed tomography (CT). Ten of these had both CT studies and angiography. The tumors were large, irregular, typically iso- to hyperdense, and showed dense, heterogeneous contrast enhancement. Cysts were present in 65% and calcifications in 71% of cases. The angiographic findings were nonspecific, ranging from avascular to markedly vascular. Although these tumors were usually found in the cerebral hemisphere, particularly the frontal lobes, two cases are included of tumors arising elsewhere: one in the pineal and one in the suprasellar region. Epidemiologic data are reviewed.

Historically, central nervous system tumors are categorized by location and/or histologic type. Primitive neuroectodermal tumors were first described by Hart and Earle [1] in 1973. They described a group of supratentorial tumors that closely resembled microscopically the germinal or matrix cells of the primitive neural tube. The tumors were at least 90%–95% undifferentiated. Formerly, these tumors have been identified as cerebral neuroblastomas, undifferentiated gliomas, cerebral medulloblastomas, and small cell tumors. Since originally described, over 100 cases have been reported [2–9]. Becker and Hinton [3] recently categorized and described the pathology of primitive neuroectodermal tumors and reported on 15 primitive neuroectodermal tumors from the Hospital for Sick Children. Their criteria were those used in this report and are the same as those of Hart and Earle [1]. This cerebral tumor is classified by histologic type and is irrespective of the site of origin. Although medulloblastoma is now considered to be a primitive neuroectodermal tumor of the posterior fossa [3], our report is limited to those tumors outside the posterior fossa. We recently reviewed tumors at the Hospital for Sick Children and accumulated 31 cases that fall into this category. These cases were seen from 1962 through 1983. Radiologic examinations were available for analysis in 22. Five patients were studied with angiography and 19 with computed tomography (CT); of these 17 were studied at the time of initial presentation. Our radiologic description of the primitive neuroectodermal tumor is based on this latter group.

Materials and Methods

The 31 patients comprised 19 boys and 12 girls. This agrees with the original article of Hart and Earle [1] of a slight male predominance. The age range was 3½ months to 17 years. More than 50% of the children were seen by age 3, and 70% by age 6 (fig. 1). The symptoms at presentation included headache and irritability (14 cases), nausea and vomiting (18), seizures (eight), ataxia (four), and hemiparesis (six). Papilledema was demonstrated in 15 cases. Several types of therapy were used. These were a combination of subtotal to total surgical excision, radiation of the cranial vault with an average dose of 5000 cGy, radiation of the cranial vault and spinal cord with an average dose of 5000 cGy to the cranium and 3000 cGy to the spinal area, and various chemotherapeutic regimens. The modes of therapy

Received January 25, 1984; accepted after revision May 29, 1984.

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AJNR 6:15–18, January/February 1985
 0195–6108/85/0601–0015 \$00.00
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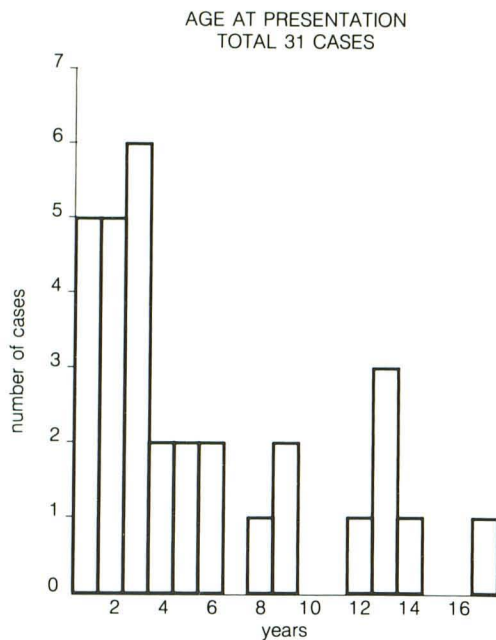


Fig. 1.—Ages at initial presentation in 31 children with neuroectodermal tumors.

TABLE 1: Models of Therapy in Children with Neuroectodermal Tumors

Type of Therapy	No. of Cases	Outcome
No surgery; radiation and chemotherapy	2	Both died, at 2½ and 4¾ years
Biopsy	2	Both died, at 2 and 14 months
Surgery, radiation to cranium only	9	Three died, at ½, 5, and 30 months; five alive, at 3½, 5, 6, 6¾, and 17 years; one lost to follow-up
Surgery, radiation to head and spine	11	Six died, at 5, 8, 11, and 16 months and 4 and 5 years; five alive, at 6, 9, and 12 months and 9½ and 11 years
Surgery, incomplete radiation treatment	3	All alive, at less than 6 months
Surgery with radiation and chemotherapy	4	Two died, at 15 and 22 months; two alive, at 16 and 34 months

and results are shown in table 1. Follow-up was possible in all but one case. Of the 30 cases, 15 died, and the range of survival from diagnosis of the tumor to death was several weeks to 5¼ years. Autopsies were performed in nine patients. All showed evidence of residual tumor, with five of the nine showing evidence of CSF spread of tumor. The other 15 patients were alive at the time this report was prepared. It should be noted that three of the 15 patients were diagnosed within the previous 6 months. However, five had survived longer than 5 years (fig. 2).

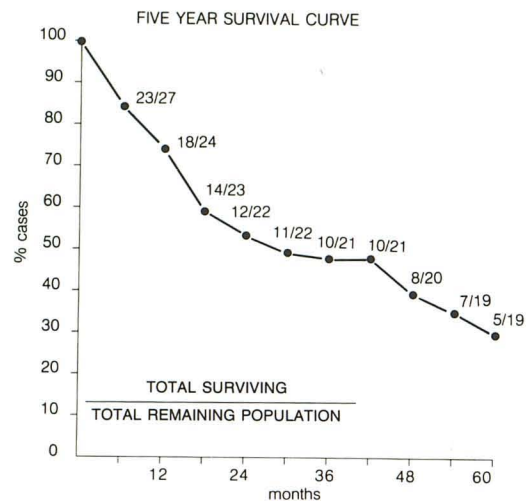


Fig. 2.—5 year survival rate in 31 children with neuroectodermal tumors.

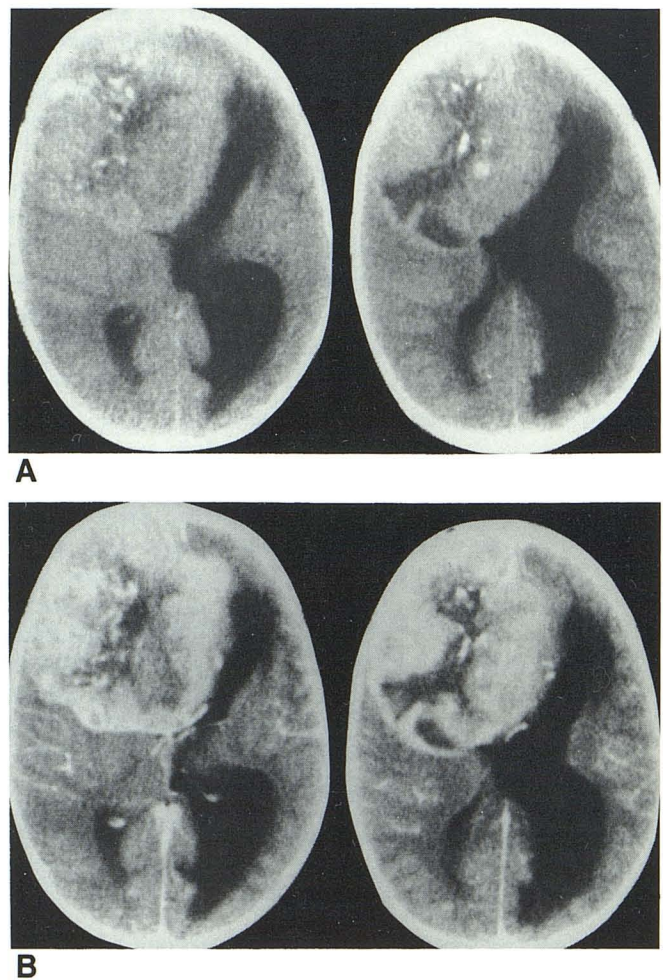


Fig. 3.—Typical CT appearance of primitive neuroectodermal tumor. **A**, Unenhanced CT scans. Large right frontal mass primarily hyperdense with hypodense regions found to be necrotic at operation. Contour of tumor is irregular. Multiple calcifications. No edema. **B**, Enhanced CT scans. Dense heterogeneous contrast enhancement in nonnecrotic parts of tumor.

Radiographic Findings

The location of the tumor was as follows: frontal lobe (10 cases), pineal (six), temporal lobe (four), hemispheric (three), parietooccipital (three), frontoparietal (two), thalamic (one), suprasellar (one), and parietal lobe (one). This corresponds with the findings of Ganti et al. [9], in which most tumors were superficial in the lobes, rather than in the thalamus and basal ganglia.

The tumors were large. The mean greatest diameter was 6.5 cm on CT, with nine of the 17 being 7.5 cm or larger. The solid part of the tumor was variable in appearance. It was typically iso- to hyperdense, occasionally with irregular regions of hypodensity, probably necrosis. Cysts (65%) were common, as were calcifications (71%), which are often extensive. A thin halo of edema was frequently visible.

Dense, but heterogeneous contrast enhancement was nearly always present. Unlike Ganti et al. [9], who found nine of 13 cases with intracranial subarachnoid metastases on CT, none of our cases demonstrated this finding. The larger tumors were often irregular in shape. The CT size and ap-

pearance of these tumors can be summarized as aggressive, and often quite different from other supratentorial tumors (fig. 3).

Although the primitive neuroectodermal tumor is microscopically the same as medulloblastoma, most do not appear to be the same radiologically. Both are typically iso- to hyperdense on unenhanced CT, but medulloblastomas have cystic (16%) and calcific (25%) components in only a few cases and tend to have a more homogeneous enhancement [10].

Unlike the CT findings, angiography was quite nonspecific. Six tumors were avascular, three had a mild tumor blush, and six were moderately vascular (fig. 4).

These features were typical of many of the tumors, which most often were of the cerebral hemispheres. However, we encountered several primitive neuroectodermal tumors mimicking other tumors because of an unusual location. One tumor arose in the pineal region (fig. 5). The amount of calcification in this tumor precluded any demonstrable contrast enhancement. Incidentally this was the only tumor to have spinal subarachnoid metastasis when first seen. This tumor could not be differentiated radiographically from other pineal tumors. By older criteria this tumor would be diagnosed pathologically as a pineoblastoma, but Becker and Hinton [3] include this in the primitive neuroectodermal tumor group. Another unusual location was the suprasellar region (fig. 6). This tumor had such characteristic calcifications, hyperdensity on plain CT and minimal associated edema, that the probability of a primitive neuroectodermal tumor was raised before operation.

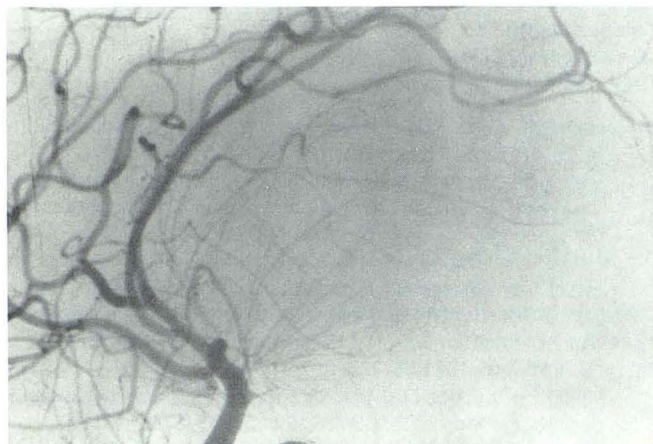
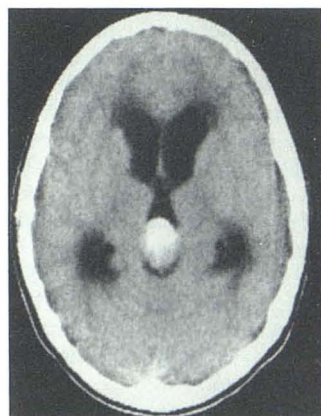


Fig. 4.—Angiographic features of primitive neuroectodermal tumor. Lateral projection of internal carotid injection. Moderate vascularity of large mass. This was the most vascular of the 15 tumors examined by angiography.

Discussion

We reviewed the tumors at the Hospital for Sick Children and evaluated 31 tumors as classified by Becker and Hinton [3] as primitive neuroectodermal tumors. Clinically most children had symptoms and signs of increased intracranial pressure. These children were treated with combinations of surgery, radiation therapy, and chemotherapy. The best survival was seen in those receiving surgery and radiation to the cranium or both the cranium and the spinal axis. The most common location of the tumor was the frontal region. Our

Fig. 5.—Primitive neuroectodermal tumor of pineal region. Unenhanced CT scan. Markedly calcified mass of pineal region causing moderate hydrocephalus.



5



6 A



6 B

Fig. 6.—Primitive neuroectodermal tumor of suprasellar region. A, Unenhanced CT scan. Hyperdense solid part of mass with small hypodense cyst. Multiple calcifications. No edema. B, Enhanced CT scan. Dense, homogeneous contrast enhancement of solid tumor and some enhancement of cyst wall.

TABLE 2: CT Features of Neuroectodermal Tumors in Children

Finding	No. of Cases (%) (n = 17)
Calcifications	12 (71)
Cyst(s)	11 (65)
Iso- to hyperdense on plain CT	12 (71)
Heterogeneous contrast enhancement	13 (76)
Edema, 0-1+	14 (82)
Hydrocephalus, 0-1+	11 (65)

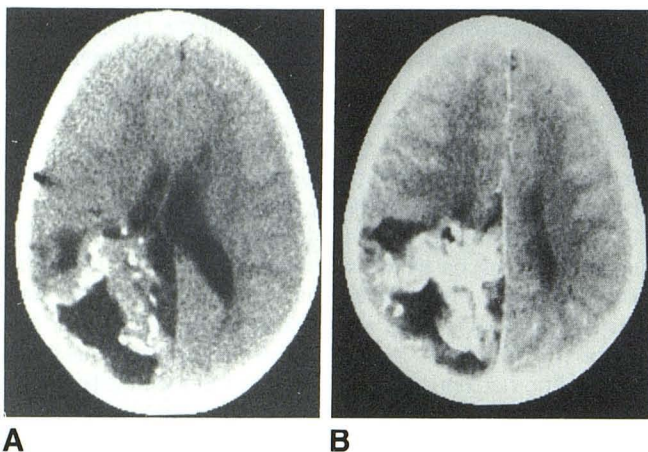


Fig. 7.—6 years after biopsy and radiation therapy. **A**, Unenhanced CT scan. Hyperdense solid part of tumor, irregularity of mass shape, cystic region of hypodensity, and multiple calcifications. No associated edema and minimal hydrocephalus. **B**, Enhanced CT scan. Dense, heterogeneous contrast enhancement with associated cyst. Tumor was unchanged on CT after 4½ years.

report indicates five children surviving more than 5 years with this tumor. Previously only Duffner et al. [4] had reported a patient with a primitive neuroectodermal tumor surviving as long as 39 months. Clinically these children suffer from the side effects of surgery and radiation. The reasons for this are believed to be their frequent young age, the immature brain being especially affected by radiation, and the large amount of cerebrum often involved. Posttreatment symptoms range from seizures and hemiparesis to delayed speech, learning disabilities, mental retardation, and diabetes insipidus.

The CT features of this tumor are summarized in table 2. Several recent articles have described cerebral neuroblastoma [11, 12]. These appear similar radiographically to our cases and suggest some overlap. Previously reported neuroblastomas were isodense to hypodense on precontrast CT [11]. We have found that most primitive neuroectodermal tumors are iso- to hyperdense on CT.

Ten of the 12 cases that were diagnosed more than 6 months before this study and are alive have had follow-up CT examinations. Three of these showed no CT evidence of residual tumor. These three were alive 7 months, six years, and 11 years, respectively, after diagnosis. Three showed

radiographic evidence of residual tumor that appeared stable. Although radiation necrosis could not be excluded, the most closely followed case (fig. 7) showed residual tumor similar to the original tumor diagnosed 6 years before. The other four showed signs of progression of tumor. Our longest survivor was lost to follow-up in 1979, but clinically had no sequelae other than seizures. This represents a survival of 17 years. No CT was performed on this patient.

In summary, primitive neuroectodermal tumors often have a quite characteristic CT appearance. They are large masses most commonly of the frontal lobes that on the unenhanced part of the CT scan show calcifications in 71% and cystic components in 65% of cases. They all enhance with contrast material and usually have only minimal associated edema. The tumor can also occur outside the cerebral hemispheres, as occurred in our series in the pineal and suprasellar regions. Previously reported tumors have a poor prognosis; however, in two of our cases, there was no evidence of tumor on CT at a follow-up of 6 and 11 years after diagnosis.

ACKNOWLEDGMENT

We thank Laurence E. Becker, Head, Division of Neuropathology, University of Toronto, for help in manuscript preparation.

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