Melanotic neuroectodermal tumor involving the cranium in infancy

Case report

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A case of the cranial form of a melanotic neuroectodermal tumor is reported in an infant, and similar cases reviewed in seven other patients. The predilection of this tumor for the anterior fontanel region, its rapid invasion of adjacent bone, and its predominance to date in Negro males are documented. The prognosis appears to be excellent after simple excision and electrocautery of the dural extension. The probable origin and pathological features are discussed.

KEY WORDS • melanotic neuroectodermal tumor • infants • anterior fontanel

Melanotic neuroectodermal tumors are considered rare benign pigmented lesions, occurring in infancy and most frequently involving the maxilla.4,6,5,14,19,20,23,5,28,29,30 This tumor may also originate in the mandible,1,5,15,30,32 epididymis,10,11,34 thigh,31 mediastinum,23 and cranium,1,2,7,9,17,26,27 Rapid growth, invasion of contiguous bone, and adherence to underlying dural sinuses are characteristic features of the cranial form of this unusual tumor. We are reporting the successful management of a case of melanotic neuroectodermal tumor which arose in the anterior fontanel and have reviewed seven other reported cases where the cranium was involved.

Case Report

A small lump was first noted at the anterior fontanel in a 3-month-old Negro boy who was under medical treatment for bronchitis. Examination on May 1, 1969, disclosed a firm, nontender, fixed mass 1 cm in diameter, over which the scalp was freely movable. Skull films showed only a soft tissue shadow corresponding to the lesion. The tumor increased rapidly in size, and by June 30 measured 4 cm in diameter and had completely obliterated the anterior fontanel (Fig. I left). An abortive attempt was made to excise the lesion locally, under the mistaken impression it was an epidermoid cyst. When a pigmented tumor invading the surrounding bone was encountered, the lesion was merely biopsied. Postoperatively, a pneumoencephalogram was performed, but the air injected only filled the subarachnoid space.

Operation. On July 15, 1969, the mass was excised by a circumferential craniectomy through normal bone margins. The tumor was adherent to the dura over the sagittal sinus at the anterior fontanel. The dural layers were split in an attempt to remove the tumor; however, frozen section showed tumor cells in the layer of excised dura. The remaining dura was cauterized prior to an in-
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FIG. 1. Left: Skull film June 30, 1969, showing marked bone proliferation in the region of the fontanel and tumor extension posterior to the coronal suture. The mass measured 4 cm in diameter and obliterated the fontanel. Subarachnoid air is present from an attempted pneumoencephalogram. Right: Skull film 22 months after cranieotomy. The surgical defect measures 6 x 5 cm. The bone margins show no evidence of recurrence.

tracranial exploration which proved negative.

Postoperative Course. Recovery was uneventful, and the infant was discharged within 2 weeks. When last seen at 2 yrs 3 mos of age after a 22-month follow-up, the child showed no clinical or roentgenographic evidence of recurrence and was developing normally. The postoperative cranial defect measured 6 x 5 cm (Fig. 1 right).

Pathological Examination. The excised specimen measured 5.8 cm in diameter with a thickness of 2.6 cm. The central portion containing gross tumor had a firm gritty consistency and measured 4.2 cm in diameter. Its cut surface appeared grayish-yellow interspersed with areas of diffuse dark pigmentation. Two cell types arranged in a fibrocollagenous stroma were characteristic of microscopic features. The first resembled a neuroblastic cell with its hyperchromatic nucleus and indistinct agranular cytoplasm; these cells were arranged in clumps or strands frequently outlined by large cuboidal epithelial-like cells. The second type, the epithelioid cell, had a nucleus with a fine vesicular chromatin pattern and cytoplasm containing pigment granules. These epithelioid cells lined nests of cells of the first type, forming the alveolar-like pattern characteristic of this lesion (Fig. 2).

Discussion

Less than 100 cases of melanotic neuroectodermal tumor have been reported in infants and these in various locations.6,8,14,19,20,22,25,29,30 The histogenesis of this tumor is uncertain, as reflected by the nomenclature used.

The term used by Krompecher10 in 1918 to introduce this tumor was "congenital melanocarcinoma of the alveolar process." Mummery and Pitts24 in 1926 related it to an odontogenic origin by the term "melanotic epithelial odontoma." Halpert and Patzer12 in 1947 chose to designate this lesion a "retinal anlage tumor" because of its resemblance to the primitive optic vesicle. The term "melanotic progonoma" was introduced by Stowens20 in 1957 because of its postulated origin from atavism of sensory neuroectoderm.

The most recent contributions to the histogenetic theories suggest an origin from the neural crest because of high urinary vanilmandelic acid levels and electron microscopic studies.6,22,25,26

Melanotic neuroectodermal tumor of in-
Melanotic neuroectodermal tumor of skull

Fig. 2. Left. Low-power photomicrograph of tumor showing dense fibrous stroma surrounding nests and strands of two distinct cell types. The first type is a small cell with a dark-staining hyperchromatic nucleus, clusters of which are frequently surrounded by the second type of cell, the large cuboidal epithelial-like cell. A characteristic alveolar-like pattern appears in the central part of the figure. H & E, ×100. Right. High-power view of the alveolar-like pattern. The nuclei of the large cuboidal epithelial-like cells have a fine vesicular chromatin network. The distinct cytoplasm contains areas of abundant pigment granules, giving the tumor its gray-black appearance. These cells are lining the clumps of smaller cells which have dark-staining hyperchromatic nuclei and indistinct cytoplasm. There are no pigment granules present in these small cells, which resemble neuroblastic cells, and their nuclei are generally round or ovoid; some appear very dark-staining and spindle-shaped. The spindle-shaped cells are thought to represent differentiation of the more immature cells into polar neurons. The fibrocollagenous stroma is also shown. H & E, ×400.

fancy is considered benign, despite its exceptionally rapid growth and invasion of surrounding bone. The marked bone proliferation demonstrated in our case has also been observed in another case involving the anterior fontanel, and in one at the maxilla. Metastatic lesions have not been reported except in a stillborn infant with a maxillary tumor resembling a melanotic progonoma. This case appears an exception to the behavior of melanotic neuroectodermal tumor of infancy, if it is indeed correctly classified. Simple excision is the treatment of choice, and in such cases where complete removal is impossible, x-ray therapy has been shown effective. Recurrence results from either incomplete removal or multicentric origin.

The melanotic neuroectodermal tumors of infancy involving the cranium were located in the anterior fontanel in seven cases, and in the right mastoid area overlying the lateral sinus in one (Table 1). All patients were less than 1 year of age; six were males and, of five reports recording the race, four were Negro. These statistics, limited however by their small number, would suggest that males, particularly Negro males, are affected more frequently than females. No deaths were attributed to the tumor itself. The average size of the tumors involving the cranium was 4.6 cm. All were adher-
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**TABLE 1**

Summary of cases of melanotic neuroectodermal tumor in infancy

<table>
<thead>
<tr>
<th>Author, Year</th>
<th>Nomenclature</th>
<th>Patient's Age, Sex, Race</th>
<th>Tumor Location</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clarke &amp; Parsons</td>
<td>retinal anlage</td>
<td>9 mos, M Caucasian</td>
<td>anterior fontanel</td>
<td>14 mos</td>
</tr>
<tr>
<td>Kuhn, et al. (1954)</td>
<td>heterotopic pigmented retinoblastoma</td>
<td>3 mos, F</td>
<td>anterior fontanel</td>
<td>—</td>
</tr>
<tr>
<td>Davis (1962)</td>
<td>retinal anlage</td>
<td>3½ mos, M Negro</td>
<td>anterior fontanel</td>
<td>died postop</td>
</tr>
<tr>
<td>Reyes, et al. (1964)</td>
<td>retinal anlage</td>
<td>4 mos, F Negro</td>
<td>anterior fontanel</td>
<td>—</td>
</tr>
<tr>
<td>Ashley (1964)</td>
<td>melanotic &quot;adamantinoma&quot;</td>
<td>newborn, M</td>
<td>right mastoid</td>
<td>16 mos</td>
</tr>
<tr>
<td>Neustein (1967)</td>
<td>melanotic progonoma or retinal anlage</td>
<td>6 mos, M Negro</td>
<td>anterior fontanel</td>
<td>—</td>
</tr>
<tr>
<td>Allen, et al. (1969)</td>
<td>retinal anlage</td>
<td>4 mos, M</td>
<td>anterior fontanel</td>
<td>7 yrs</td>
</tr>
<tr>
<td>Gilmor &amp; Mealey (1972)</td>
<td>melanotic neuroectodermal tumor of infancy</td>
<td>5 mos, M Negro</td>
<td>anterior fontanel</td>
<td>22 mos</td>
</tr>
</tbody>
</table>

ent to the underlying dural sinus at the time of surgery. Because of their proximity and adherence to the dural sinus, these neoplasms have been interpreted as a possible hamartomatous growth of the arachnoid villi. The more recent evidence implicating the neural crest, however, makes this hypothesis unlikely. The longest follow-up report has been 7 years without recurrence or sequelae, indicating a good prognosis after simple excision. The dural involvement in our case was satisfactorily treated by electrocautery which seems safer than ligation and excision of the sinus when the tumor extends posterior to the coronal suture.

References

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