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AJNR

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AJNR Am J Neuroradiol 1993, 14 (3) 537-539

<http://www.ajnr.org/content/14/3/537>

This information is current as
of April 18, 2024.

Congenital Clivus Chordoma

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Summary: The authors report a case of a congenital clivus chordoma that caused cranial nerve palsy and hydrocephalus within a few days after birth. The tumor was well demonstrated by sonography, CT, and MR; the preoperative diagnosis was histologically confirmed after subtotal resection.

Index terms: Clivus; Chordoma; Brain neoplasms, in infants and children; Pediatric neuroradiology

Chordomas are rare malignant tumors that arise from notochordal remnants and have a predilection for the upper and lower ends of the neural axis. In children, chordomas are very unusual: the youngest patient with an intracranial chordoma described in the literature, to our knowledge, was 4 months old (1). We report a case of congenital clivus chordoma that caused cranial nerve palsy and hydrocephalus.

Case Report

Within the first days after birth, a female newborn presented with torticollis, muscular hypotonia, and acrocyanosis. Examination revealed permanent convergent strabismus, drooping left angle of the mouth, and dysphagia, indicating involvement of cranial nerves VI, VII, XI, and XII. The anterior fontanelle was tense. Sonographic examination showed a tumor below the third ventricle in front of the pons (Fig. 1). Computed tomography (CT) demonstrated a hyperdense midline tumor at the skull base extending into the mesencephalic region. The brain stem was displaced and the cerebral aqueduct compressed with resulting obstructive hydrocephalus (Fig. 2A). Bony destruction of the dorsal part of the clivus in the region of the sphenooccipital synchondrosis was visualized on high-resolution CT (Fig. 2B). T1-weighted axial and sagittal magnetic resonance (MR) images at .5-T demonstrated a tumor of low signal intensity. After administration of Gd-DTPA, a homogeneously enhancing tumor was seen apparently rising from the clivus. The sagittal images revealed

that the cranial parts of the tumor reached the dorsum sella and compressed the ventral portion of the pons. The lesion extended down to the fourth cervical vertebra (Fig. 3). Our preoperative diagnosis was chordoma or chondrosarcoma.

When the patient was 4 weeks old, the tumor was subtotally resected by a posterolateral approach. The ventrocaudal part of the tumor was not identified at surgery, therefore, total extirpation was impossible. Histologically, there were groups of vacuolated so-called physaliphorous cells. Only minimal amounts of mucoid extracellular substance could be found. Focally, highly pleomorphic and polychromatic nuclei were seen. Several tumor cells showed a positive immunoreactivity with the proliferation marker Ki-67 and thus supported the impression of an enhanced proliferative activity.

Discussion

Chordomas are believed to develop from notochordal remnants and to have a predilection for



Fig. 1. Congenital clivus chordoma. Sonographic examination showed a tumor (arrows) below the third ventricle in front of the pons.

Received January 10, 1992; accepted after revision August 11.

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Fig. 2. A, CT revealed a hyperdense midline tumor at the skull base displacing the brain stem. The compression of the cerebral aqueduct causes an obstructive hydrocephalus as the enlarged inferior horns of the lateral ventricle shows.

B, High-resolution CT shows bony destruction of the dorsal part of the clivus in the region of the sphenoooccipital synchondrosis.

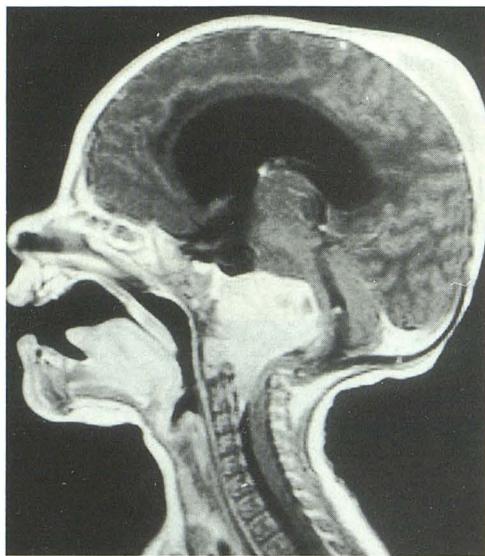
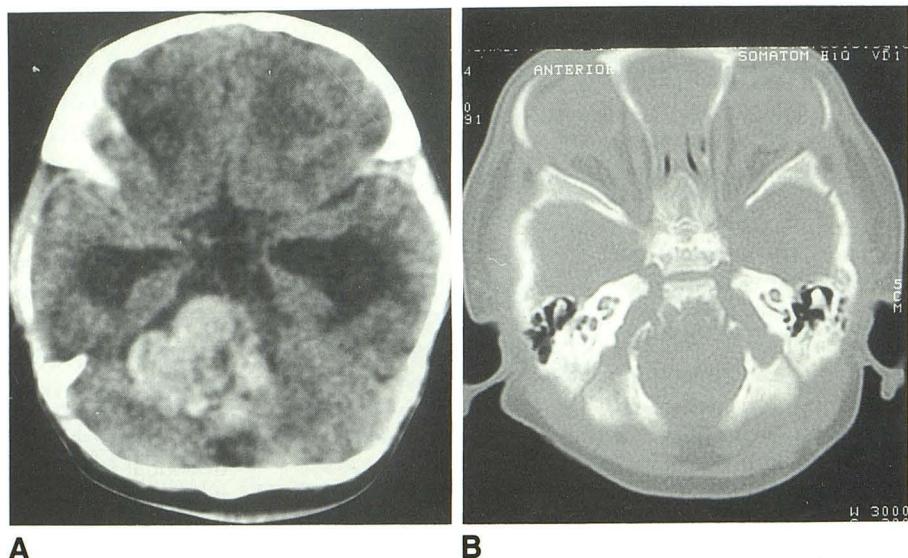


Fig. 3. Contrast-enhanced sagittal MR image, 500/20/2: Homogeneous tumor of high signal intensity apparently arising from the clivus. The rostral parts reached the dorsum sella and the caudal part extended down to the fourth cervical vertebra. Hematoma of the scalp (cephalhematoma) is seen as an incidental finding.

the upper and lower ends of the neural axis (2–4). Although slow-growing, chordomas are locally invasive, destroying bone and infiltrating the soft tissues with a 10% to 20% incidence of metastases (5). In children, chordomas are very unusual; most of them are found at the skull base, arising from the clivus in the region of the sphenoooccipital synchondrosis (6). This is a report of a

clivus chordoma presenting within a few days after birth.

A recently published review (1) listed a total of 58 cases of chordomas in patients 16 years old and younger. Thirty-seven of these (64%) were located at the skull base, the youngest reported patient being 4 months old. Matsumoto et al (1) pointed out that there seems to be a distinct clinical and pathologic picture in the infant and young child. Typical signs of cranial chordomas in children younger than 5 years are long tract signs, lower cranial nerve involvement, increased intracranial pressure, and extension of the tumor through the foramen magnum into the cervical spine. These are the same clinical findings as described in our case. Furthermore, chordomas in very young patients seem to metastasize more frequently (1). In fact, similar to the 4-month old child described by Matsumoto et al, the histopathologic picture of our case suggests an increased proliferative activity. Despite this and the large size of the tumor there were no metastases found in our case.

With CT the chordoma appears as a soft-tissue mass with areas of calcification, moderate contrast enhancement, and bone destruction. MR imaging is of great value in defining both extension and site of origin of these tumors. In our case, the sagittal projection is especially helpful in case of planned surgery (7). On T2-weighted images and after administration of Gd-DTPA, this kind of tumor typically reveals a high signal intensity. Other tumors to consider with a similar appearance are chondroma and chondrosarcoma.

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