Melanotic neuroectodermal tumor of infancy in the skull: CT and MRI features

A 1-month-old girl presented with swelling in the right temporal region. The mass was 40 × 40 mm. CT showed a slightly high-density mass that had destroyed the right temporal and sphenoidal bones (Fig. 1). T1-weighted MRI images showed that the mass had mildly high signal intensity in the periphery (Fig. 2), whereas T2-weighted MRI images showed low signal intensity in the corresponding areas of the mass (Fig. 3). The tumor was markedly enhanced with contrast medium, apart from the central area.

The patient underwent craniotomy. Macroscopically, the inside of the mass was blackish-brown. Photomicrographs of the tumor showed a biphasic pattern with large melanin-containing epithelioid cells arranged in alveoli or tubules around an island of small cells in a dense fibrous matrix. Immunohistochemistry was positive for HMB-45 (melanoma-associated antigen). The diagnosis of melanotic neuroectodermal tumor of infancy (MNTI) was made. Three months after surgery, follow-up CT revealed recurrence of the tumor. The recurrent tumor was removed completely. The patient has been followed-up for 17 months without recurrence so far.

Figure 1 CT shows that the mass has destroyed the sphenoidal and temporal bones. Note that bony spicules are evident (arrow).

Figure 1 Le scanner montre une destruction des structures osseuses temporales et sphénoïdales. À noter la présence de spicules osseuses (flèche).

Figure 2 T1-weighted image shows that the signal intensity of the tumor in the periphery is slightly high (arrow).

Figure 2 L’IRM pondérée en T1 montre un signal discrètement élevé en périphérie de la tumeur (flèche).

Figure 3 T2-weighted image shows that the signal intensity of the tumor is low in the periphery, but that of the central part is very high.

Figure 3 L’IRM pondérée en T2 montre un signal hypo-intense de la tumeur en périphérie et très élevé dans sa partie centrale.
MNTI is a rare neoplasm that, when it occurs, often forms during the first year of life [3]. The tumor usually develops in the head and neck regions, particularly in the maxilla, followed by the skull, mandible and brain [3]. Despite their rapid local growth potential, MNTIs are generally classified as benign neoplasms.

Hyperostosis and osteogenesis with prominent spicules of bone on CT underscore a characteristic finding suggesting the diagnosis of MNTI [1]. In the current case, T1-weighted images revealed mildly high signal intensity, probably because melanin causes the T1-shortening effect. Differential diagnoses include Ewing’s sarcoma, osteosarcoma, eosinophilic granuloma, lymphoma, leukemia and metastasis from neuroblastoma [2]. Clinical information (such as patient’s age and multiplicity of lesions) as well as the radiological features can be helpful in the diagnosis. Knowledge of this rare entity, including its CT and MRI features, is important for making a proper diagnosis and providing appropriate treatment strategies for patients.

References


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Spontaneous chronic subdural hematoma of the posterior fossa

The posterior fossa is an unusual location for chronic subdural hematoma in adults without a history of trauma. The most common associated factor is coagulation abnormality. To our knowledge, fewer than 20 cases similar to ours have been reported in the literature [6]. We describe a post-surgical case diagnosed by MRI.

A 38-year-old woman was admitted to our department because of progressive, pronounced dizziness and headaches, without any previous history of head injury. Her relevant past medical history included systemic lupus and antiphospholipid syndrome. On admission, the patient was awake, alert, and complained of severe vertigo and was unable to walk unaided. The neurological examination revealed cerebellar signs and gait ataxia associated with bilateral papillary edema. Blood tests showed a spontaneous low prothrombin rate (45%).

The MRI study demonstrated a right subdural space-occupying collection at the convexity of the cerebellar hemisphere, with compression of the fourth ventricle. This collection was hyperintense and heterogeneous on both T1- and T2-weighted MRI sequences (Figs. 1-3).

After the coagulation profile was normalized, the patient underwent surgical evacuation of the hematoma through a suboccipital right craniectomy. Chocolate-colored fluid gushed out under pressure. No source of bleeding could be identified. The subdural space was rinsed...