Extramedullary hematopoiesis masquerading as a cranial (clivus) tumor

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ABSTRACT

Extramedullary hematopoiesis (EMH) is a well-known phenomenon occurring during fetal development. In the postfetal condition, EMH is commonly associated with hematologic conditions including chronic myeloproliferative or lymphoproliferative disorders, leukemias, and chronic and inherited anemias. We report an unusual location for EMH that masqueraded as a cranial tumor.

KEYWORDS Clivus tumor; extramedullary hematopoiesis; polycythemia vera

Extramedullary hematopoiesis (EMH) is defined as hematopoiesis occurring in locations other than the bone marrow medullary space. It is a well-known phenomenon found during the fetal embryonic/developmental period, occurring principally in the liver and spleen. When found during the postfetal condition, EMH is usually associated with a hematologic condition, including chronic myeloproliferative and lymphoproliferative disorders, leukemias, and chronic and inherited anemias, and may be found in other nonhepatic/splenic sites. We report an unusual case of EMH that occurred in the sella turcica/clivus area of the central nervous system that was originally favored to be an extradural cranial tumor.

CASE STUDY

A 57-year-old woman with known hypertension, neurofibromatosis, and polycythemia vera presented with progressive bilateral vision loss. Magnetic resonance imaging revealed a transpatial mass thought to originate from the sella turcica (*Figure 1a*). The mass measured approximately 5 cm with regional osseous remodeling, and given its presumed origin from the clivus the differential diagnosis favored a clival chondroma, with pituitary macroadenoma, giant cell tumor, plasmacytoma, and lymphoma being additional considerations. A biopsy showed a collection of hematopoietic elements containing erythroid, myeloid, and megakaryocytic

cells (*Figure 1b*). Blasts were not increased with a CD34 immunostain, ruling out a granulocytic sarcoma. The tissue was considered to represent a collection of extramedullary hematopoietic tissue. Given the patient's history of polycythemia vera, a subsequent bone marrow biopsy was performed, which documented a marrow consistent with polycythemia vera that had evolved to the fibrotic stage (*Figures 1c, 1d*). The patient was treated with a course of low-dose radiotherapy with resolution of the mass and her visual symptoms.

DISCUSSION

The major challenge in this case was to establish a diagnosis of the mass in the sphenoid sinus/clivus area. The finding of nonmalignant hematopoietic tissue representing EMH was somewhat of a surprise, because a tumor was strongly suspected and cranial EMH is an unusual location. With this finding of EMH, the patient's history of polycythemia vera was recalled, and a subsequent bone marrow examination was consistent with the fibrotic stage of polycythemia vera.

EMH represents the production of blood elements outside of the bone marrow, which in the postfetal stage usually results from an inadequate production of blood cells. The most frequent causes of EMH include myelofibrosis, metastatic tumor replacing the bone marrow, leukemia, sickle cell disease, and thalassemia.¹ The spleen is the main site for

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Figure 1. (a) Heterogeneous T2 hyperintense mass appears centered within the clivus (white arrowhead), fills and expands the sphenoid sinus (white arrow), and lifts the sella turcica (black arrowhead). T2 hypointense margin along the periphery of the marrow (black arrow) represents thinned but intact bone, suggestive of a long-standing process. (b) Clival mass that represents nonmalignant hematopoietic tissue. Hematoxylin and eosin $\times 200$. (c) Bone marrow biopsy showing findings consistent with polycythemia vera progressing to fibrotic phase. Hematoxylin and eosin $\times 400$. (d) Reticulin stain of bone marrow biopsy showing +4/4 reticulin fibrosis (fibrotic stage of polycythemia vera). Hematoxylin and eosin $\times 200$.

EMH, with the liver and lymph nodes being frequent secondary sites, but unusual locations have been reported in the thyroid, pancreas, endometrium, kidney, adrenal glands, pleura, prostate, and breast.² EMH in the central nervous system is rare, and only eight cases were documented between 1988

and 2010 in a retrospective study.³ Most were autopsy cases. Symptomatic EMH in the central nervous system is a rarity.³ EMH in the central nervous system most often occurs in the cranial meninges,⁴ followed by the spinal meninges and paraspinal tissue.⁵ Rarely, brain tumors will have foci of EMH.^{6,7} Our case is unusual in that the EMH was thought to represent a primary cranial tumor. The precise pathogenesis of EMH in the central nervous system is not known, but our case was associated with a hematopoietic disorder known to be associated with EMH. When EMH is found in older children and adults, an underlying hematopoietic condition should be sought if not already recognized.

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