

# Intraosseous noninvoluting congenital hemangioma of the mandible in a neonate

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Hemangiomas are benign tumors of the capillary endothelium involved in varied clinical and pathologic entities. Congenital hemangioma is quite rare compared to infantile hemangioma. Hemangiomas rarely occur in bone. Intraosseous hemangiomas are most commonly found in the vertebral column and skull. The mandible is the most frequently affected skull bone. Intraosseous hemangiomas commonly present in the second decade of life. Here, we report a neonate with noninvoluting congenital hemangioma of the mandible. To our knowledge, she is the youngest patient reported in the literature.

**Key words:** intraosseous hemangioma, mandibular hemangioma, congenital hemangioma, neonate.

Hemangiomas are benign tumors of the capillary endothelium involved in varied clinical and pathologic entities. Although infantile hemangioma occurs frequently, the congenital type is quite rare. Hemangiomas most commonly occur in the skin and subcutaneous tissues. Tissues other than skin and visceral organs including liver and intestine are less often involved. Hemangiomas of bone are uncommon. Intraosseous hemangiomas generally occur in vertebra and skull bones, mostly in the maxilla and mandible<sup>1,2</sup>. Here, we report a newborn baby with congenital hemangioma of the mandible.

## Case Report

A 14-day-old female infant was admitted to our hospital with the complaints of a swelling on the right side of the chin. Physical examination revealed a huge mass localized on the right mandibular region. The mass was immobile, fixed to the mandible, relatively firm, and bluish in color (Fig. 1). The hematological and biochemical studies were normal. On direct radiograph, an expansive multilocular, lucent mandibular mass, soap bubble-like lesion with well-defined margins was demonstrated. Computed tomography



Fig. 1. Huge mass localized on the right mandibular region.

(CT) scan demonstrated the expansive mass with hypo-attenuating internal texture in the right aspect of the mandible. The anterior and posterior cortical margins of the lesion were indistinct or destructed (Fig. 2). On magnetic resonance image (MRI), the mass was isointense on T1-weighted spine-echo and hyperintense on T2-weighted turbo spine-echo images compared



Fig. 2. CT scan demonstrated the expansile mass lesion with hypoattenuating internal texture in the right aspect of the mandible. The anterior and posterior cortical margins of the lesion were indistinct or destroyed.

with muscle and enhanced dramatically after contrast administration (Fig. 3). The lesion extended through the perimandibular soft tissue and oral cavity. A biopsy from the mass was performed. The histopathological examination revealed a tumor constituted by capillary-sized vessels that were in lobular form separated by fibrous septa. There was no cytological

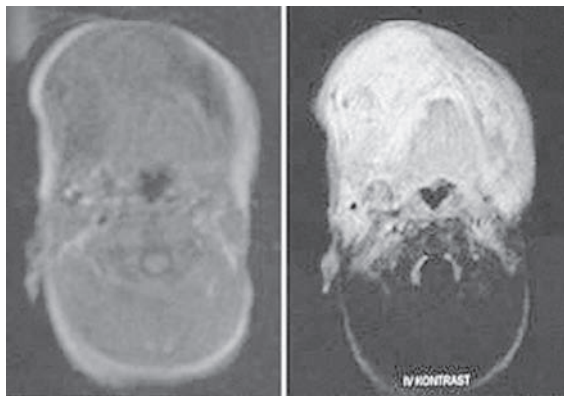


Fig. 3. On MRI, the mass lesion was iso-hypointense on T1-weighted images compared with muscle and enhanced dramatically after contrast administration. The lesion extended to the perimandibular soft tissue and oral cavity.

atypia and/or mitotic figures (Fig. 4). It was diagnosed as capillary hemangioma of the mandible. Presence of the fibrous septa indicated congenital type. Systemic steroid treatment (prednisolone 2 mg/kg/day, peroral [p.o.]) was initiated. Since no response was observed, steroid therapy was cancelled at the end of two weeks. At the end of the one-year follow-up, the tumoral mass remained stable.

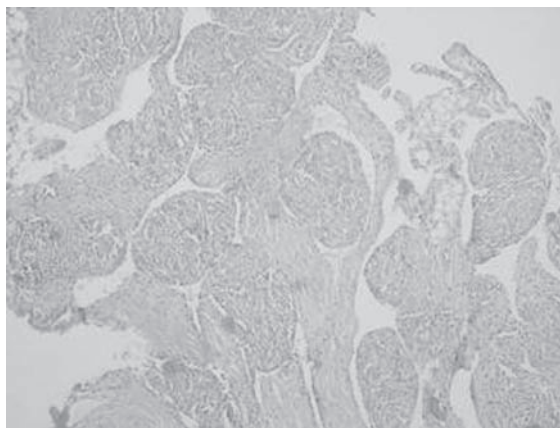


Fig. 4. Lesion comprising capillaries lined by endothelial cells, which are separated into lobules by fibrous stroma (hematoxylin & eosin, x100).

## Discussion

Benign vascular tumors of infancy represent a number of distinct entities with diverse clinical and histopathologic features. They are classified as infantile hemangiomas, congenital hemangiomas, tufted angiomas and kaposiform hemangioendotheliomas (KHE)<sup>3-5</sup>. Infantile hemangioma is the most common type that usually develops in infants within their first few months. It enlarges rapidly until 9 to 12 months of age and then begins to involute. Approximately 50% of cases involute by the age of 5 years and 90% by the age of 9 years. Congenital hemangioma is quite rare compared to infantile hemangioma. Congenital hemangioma has two forms: rapidly involuting (RICH) and noninvoluting (NICH) type. Both have similarities in appearance, location and size. The fully developed lesion is present at birth. The noninvoluting form does not regress, while RICH involutes within six months<sup>5,6</sup>.

Histopathologically, hemangiomas are a benign proliferation of endothelial cells forming capillaries. In infantile hemangiomas, tumor tissue is separated into lobules with normal

tissue elements, while the separation between the lobules is constituted by fibrous tissue in congenital hemangiomas. Infantile hemangiomas highly express glucose transporter isoform 1 (GLUT1), Fcγ receptor II, merosin, and Lewis Y antigen. Other vascular neoplasms (e.g., congenital hemangiomas, KHE, tufted angioma) do not stain positively for any of these antigens<sup>3</sup>.

In our case, the tumor was present at birth, and neither enlarged nor involuted during the one-year follow-up. Although an immunohistochemical study could not be performed, we accepted our case as NICH based on its clinical and pathological features.

Intraosseous hemangioma is a rare tumor that accounts for 0.2% - 0.7% of all bone tumors<sup>7</sup>. It is most commonly found in the vertebral column and skull. The jaw is the most frequently affected skull bone<sup>7,8</sup>. Two-thirds of hemangiomas of the jaw are found in the mandible. Osseous hemangiomas are twice as common in females as males<sup>2</sup>. These lesions usually manifest with progressive, painless facial swelling. Intraosseous hemangiomas commonly present in the second decade of life<sup>2</sup>, and have rarely been reported in childhood<sup>9</sup>. Our case was presented at birth. To our knowledge, she is the youngest reported patient in the literature.

There are no specific diagnostic appearances of intraosseous hemangiomas on plain radiography. If the hemangioma is small, no radiographic evidence may be present. If the tumor is big, non-specific osteolytic appearance such as "honey comb," "soap bubble", "sun burst," or "sun ray" pattern may be seen<sup>8</sup>, as observed in our case. CT may reveal destruction of the bone and extension of the tumor to adjacent soft tissues<sup>10</sup>. An associated soft tissue component may be demonstrated by MRI. Intermediate or high signal on T1-weighted images and high signal on T2-weighted images<sup>11-13</sup> were observed. In our case, MRI findings were similar to those reported in the literature. Since the tumor destructed and invaded peripheral soft tissue and mandible, it resembled a malignant tumor.

Since hemangiomas are slow-growing benign tumors, treatment was generally not indicated. Systemic or intralesional corticosteroids are the mainstay of the treatment for large hemangiomas causing life-threatening complications and disfigurement. Angiogenesis inhibitors such as interferon can be used in selected cases<sup>4</sup>. Surgical excision is the preferred treatment for deeply located hemangiomas such as intramuscular and intraosseous tumors. Prognosis after complete excision is excellent, and recurrence is usually rare<sup>14</sup>. Our case did not respond to corticosteroid treatment. Surgical excision and reconstruction was planned after one year of age.

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