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Pediatric mandibular metastasis: A rare finding of neuroblastoma

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ABSTRACT

We present a case of metastatic neuroblastoma to the mandible in an 11-month-old patient presenting with worsening right-sided proptosis and scalp swelling after a fall 2 weeks prior. Initial evaluation with computed tomography of the head demonstrated soft tissue masses centered at the right sphenoid and right mandible. These masses proved to be metastatic lesions from an intra-abdominal neuroblastoma. Review of the literature revealed 20 cases of neuroblastoma metastasis to the mandible over the past 70 years. To our knowledge, our patient is the youngest reported case with asymptomatic mandibular metastasis related to neuroblastoma and the first to be characterized with magnetic resonance imaging.

Case report

Our patient is an 11-month-old baby who presented to an outside hospital emergency department for worsening right-sided proptosis and scalp swelling after a fall 2 weeks prior. Computed tomography (CT) of the head revealed an aggressive soft tissue mass centered at the right sphenoid extending into the orbit, as well as partial imaging of a separate soft tissue mass with internal calcification centered at the right mandibular ramus (Fig. 1). Osseous structures adjacent to these soft tissue masses showed permeative lytic changes with aggressive periostitis (Fig. 2).

Magnetic resonance imaging (MRI) of the head revealed multiple skull base lesions, with involvement of the orbital walls bilaterally and the sphenoid and occipital bones, as well as of the right mandibular ramus. On nonenhanced T1 sequence, the mandibular lesion was slightly hypointense to gray matter (Fig. 3A and B) and demonstrated heterogeneous enhancement after contrast (Fig. 4A and B). Additionally, the mass displayed mildly increased T2 and diffusion-weighted signal (Figs. 5 and 6). Susceptibility images showed multiple hypointense foci with "blooming" within the mandibular soft tissue mass (Fig. 7). No intra-axial tumor was identified. The diagnosis of metastatic neuroblastoma was suggested based on the constellation of findings on both the CT and the MRI

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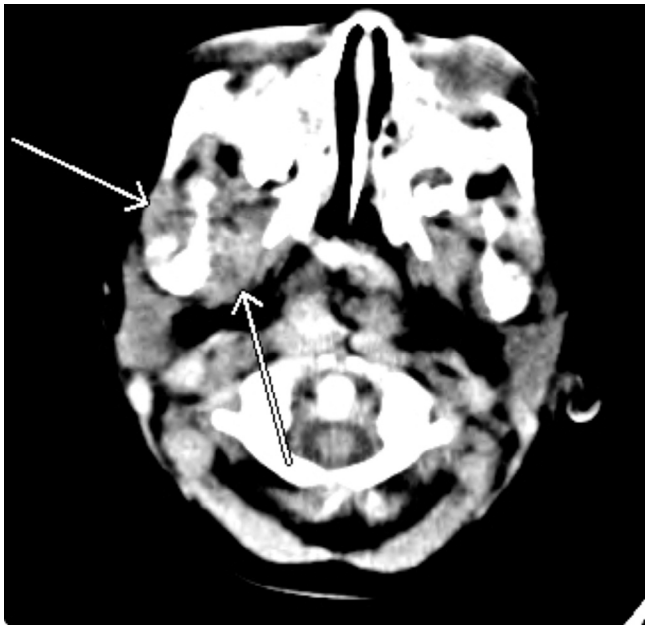

Fig. 1 – Axial computed tomography image shows a soft tissue mass with internal calcification centered on the ramus of the right mandible (arrows).

of the head, and not solely on the findings within the mandibular lesion.

Immediate follow-up CT of the chest and/or abdomen and/or pelvis identified a large abdominal mass arising from the left adrenal gland causing focal mass effect and encasing the abdominal vasculature (Fig. 8). Multiple hepatic hypodense lesions and a left pelvic soft tissue mass were also noted. Urine catecholamines were found to be elevated, consistent with the suspected diagnosis of neuroblastoma. Open biopsy of the abdominal mass and bone marrow aspirate was performed,

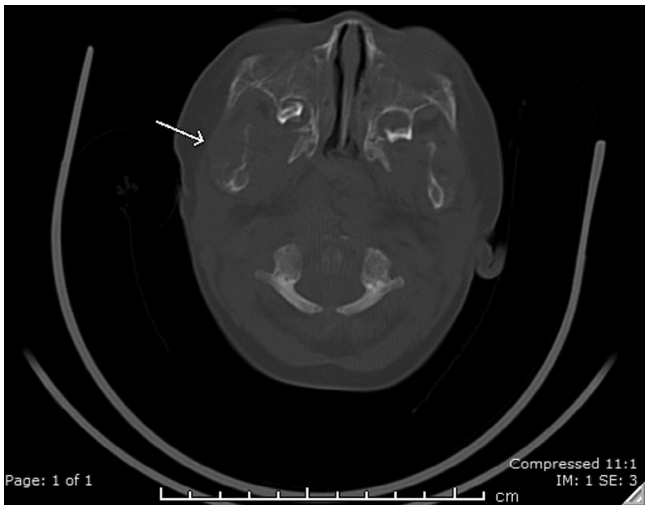

Fig. 2 – Axial computed tomography image with bone reconstruction shows aggressive periosteal reaction centered at the ramus of the right mandible.

revealing high-risk neuroblastoma due to MYCN amplification. Subsequent MIBG (metaiodobenzylguanidine) scan demonstrated radiotracer accumulation within the bilateral lower extremities, as well as in the known lesions of the abdomen and skull base, consistent with metastatic disease (Fig. 9).

The patient began treatment with cyclophosphamide and topotecan. She is currently being followed as an outpatient approximately 6 months later with overall decreased tumor burden.

Discussion

Neuroblastoma is the most common solid tumor in pediatric patients and the third most common pediatric tumor overall [1,2]. Embryologically, neuroblastomas arise from the ectodermal neural crest cells of the sympathetic nervous system, most commonly in the adrenal medulla, which is the most common location of neuroblastoma. In the United States, the incidence of neuroblastoma is approximately 1 of 7000 [3]. Most cases of neuroblastoma occur sporadically, but approximately 1%-2% exhibit familial transmission [1,3,4]. Approximately 80% of patients with neuroblastoma are diagnosed by 4 years of age, with an average age of 22 months [5,6].

Clinically, patients with neuroblastoma present with palpable abdominal abnormalities and/or complaints related to mass effect on adjacent organ systems, such as extremity edema, shortness of breath, and bone pain [6]. When orbital metastatic disease is present, patients can exhibit proptosis, as seen in our case, or orbital ecchymosis. The diagnosis of neuroblastoma is made with elevated urinary catecholamines and histopathological confirmation or bone marrow aspiration [3]. Metastasis is frequently encountered upon initial presentation (60%-70%), and the extent of metastatic lesions is correlated with prognosis and staging [1].

Detailed neuroblastoma staging is beyond the scope of this report, although briefly, staging of neuroblastoma can be performed by 2 means: postsurgically or presurgically. The International Neuroblastoma Staging System (INSS) is more commonly used and relies on surgical pathologic confirmation. This is in comparison to the more recently created presurgical staging and/or risk stratification system, The International Neuroblastoma Risk Group Staging System (INRGSS). The INRGSS relies on image-defined risk factors obtained from cross-sectional imaging and nuclear scintigraphy, as well as bone marrow sampling.

When comparing the 2 staging systems of neuroblastoma, there are notable differences. For instance, the tumor crossing midline is important for INSS but not so for INRGSS. The nodal lexicon also varies between the 2 staging systems: INSS uses the terms ipsilateral and contralateral, whereas INRGSS uses regional and nonregional. Both systems utilize the "S" qualifier for metastatic disease confined to skin, liver, and bone marrow (less than 10% of the total nucleated cell for INRGSS); however, the age cutoff for "S" used in the INSS is 12 months, whereas INRGSS uses an age cutoff of 18 months [1].

Although approximately 50%-60% of cases of neuroblastoma present with metastatic disease, mandibular metastases