

Neuroblastoma: case involving metastases to the mandible

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Abstract

Neuroblastoma is the fourth most common solid tumor of childhood. The most common symptoms of the malignancy are abdominal masses and abdominal pain. Metastases to the head and neck region are relatively common and thus of interest to dentists. The treatment of choice is surgical excision of the tumor followed by radiation therapy to the surgical site. The diagnosis, treatment, and clinical course of a 2½-year-old black male with rare metastatic lesions of the mandible is presented.

Homer Wright first described neuroblastomas as malignant tumors of childhood that are derived from neural crest tissue and generally are located at any anatomical site along the craniospinal axis.^{1,2} Neuroblastomas are the fourth most common solid tumors in children³ and are the most common neoplasms to be identified at birth. Of children who develop neuroblastomas, 25% are affected by age 1 year and 75% by age 5 years.^{2,4-6} The purpose of this paper is to report a patient with a neuroblastoma involving rare metastases to the mandible.

Literature Review

The most common presenting symptoms of neuroblastomas are abdominal masses and abdominal pain frequently accompanied by anorexia, weight loss, low grade fever, and generalized aches and pains.^{2,7} The adrenal gland is the most common site of origin, followed by the retroperitoneal tissues. The tumors are usually unilateral with no predilection for either side, although bilateral cases have been reported. The tumors are highly cellular, homogeneous masses of small, regular cells as large or slightly larger than lymphocytes with ill-defined cytoplasm and dark-staining nuclei. The distinctive rosettes described by

Wright are a spheroid group of nuclei enclosing a delicate cobweb tangle of fibrillary material. Rosettes are present in 15-50% of the specimens.^{1,8} Elevated levels of catecholamines in the urine are considered diagnostic.⁴

Neuroblastomas of adrenal origin grow toward the body midline. Secondary retroperitoneal masses may engulf the lumbar vertebrae and pelvic brim. A right-side tumor is likely to invade the liver, a left-side tumor is likely to invade the pancreas. Kidneys also may be affected. Neuroblastomas frequently metastasize. The liver, skeleton, and lymphatic glands are involved most frequently. In the skeleton, the orbits, temporal bone, and cranium are the most common sites of metastases.^{1,9-13} The potential doubling time of the neuroblastoma is 3-5 days.¹⁴

The prognosis for patients with neuroblastomas is related to the extent of involvement when the diagnosis is made. Children with localized tumors often are cured after surgical resection. Chemotherapy offers some control of advanced or disseminated tumors, but there has not been a significant improvement in the survival rate of these patients.^{2,15,16}

The preferred treatment for children with neuroblastomas consists of surgical removal of the tumor followed by radiation therapy to the operative site. This usually can be accomplished with localized tumors and will result in an 85-90% cure rate. If complete removal is not possible, considerable benefit may result from the removal of the greater part of the tumor followed by radiation therapy in a manner similar to that for Wilm's tumor. Palliative radiation therapy doses of 400-600 rads is effective in controlling bone pain due to metastases. Chemotherapy for advanced neuroblastoma is unsatisfactory. Cyclophosphamide, vincristine, doxorubicin, and dacarbazine have been used in various combinations.^{2,15}

Case Report

AK was a 2½-year-old black male when he presented at the University of Kansas Medical Center with a history of fever, lethargy, anorexia, and weight loss. He also had progressive lower extremity weakness over the preceding 3-4 weeks. The mother had noted an enlarging mass in the left temporal area for the same duration. He was in Lagos, Nigeria when the symptoms presented and was evaluated by physicians there. AK's mother decided to return to Kansas City and the patient was admitted to the Medical Center.

Medical Evaluation

Physical examination revealed a large 2½-year-old black male. The patient was very lethargic during the evaluation. The pupils were equal in size and reacted to light; the ears appeared normal; and the throat was normal and without erythema. Evaluation of the temporal mass revealed a 6×6 cm mass that was firm and tender to palpation. The lungs were clear. Cardiovascular examination revealed a normal sinus rhythm with a Grade II/VI systolic ejection murmur. The abdomen was distended and tender. The lower extremities demonstrated marked edema. His neurological evaluation revealed decreased muscle control in the lower extremities and an inability to sit. Laboratory results were within normal limits except for an increased erythrocyte sedimentation rate and a decreased hemoglobin and platelet count. These findings were consistent with other reports in the literature of advanced, diffuse neuroblastomas.

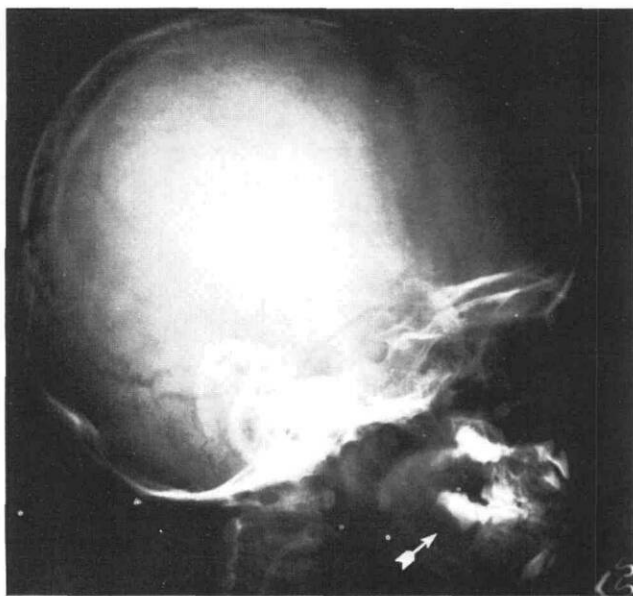


FIG 1. Lateral cephalometric radiograph demonstrating mandibular bone destruction from the neuroblastoma.

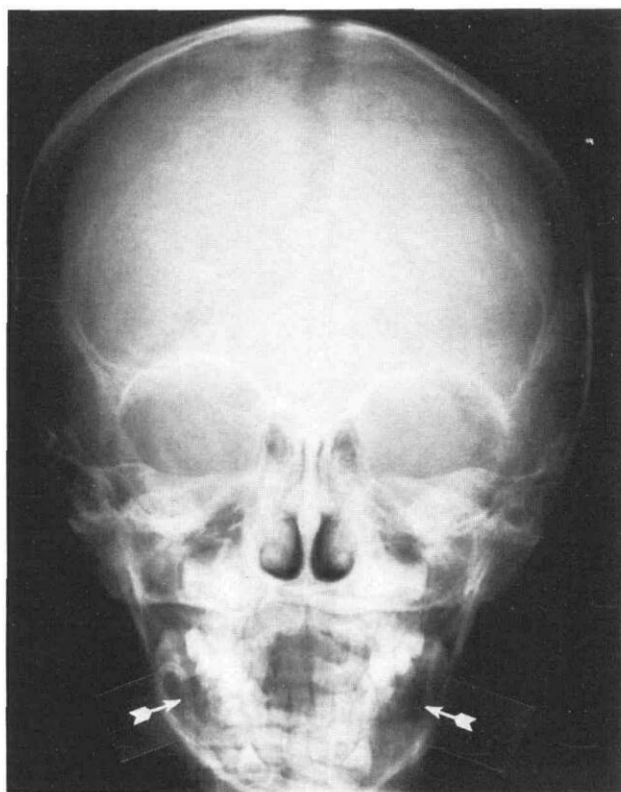


FIG 2. A-P cephalometric radiograph revealing bilateral mandibular radiolucencies in molar area.

Radiographic Evaluation

Ct scans of the abdomen revealed a right suprarenal mass. Bone scans revealed multiple areas of increased uptake of radiopaque Technicium 99m including the temporal area, coronal region, mandible, thorax, and pelvis, contributing to the tumor diagnosis. Radiographs also revealed involvement of vertebral bodies at approximately T4-T12.

Dental Evaluation

The patient's chief oral complaint was an ulcerated lesion in the lower right quadrant. Visual examination revealed a normally erupting primary dentition. There was a 3×3 cm ulcerated lesion of the gingiva between the mandibular right second primary molar and the mandibular right first permanent molar. The first permanent molar was displaced distally and vertically and obviously was erupted prematurely secondary to the tumor. The location of the lesions in the mandible was consistent with previous reports. Metastases to the mandible in cases of neuroblastomas are rare. Only 12 cases of metastatic mandibular neuroblastomas and 3 cases of primary mandibular neuroblastomas have been reported.^{17,18}

Periapical and panoramic radiographs revealed

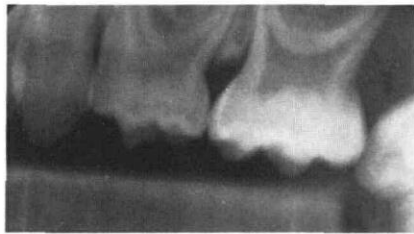


FIG 3. (left) Dental radiograph demonstrating bone destruction distal to mandibular right second primary molar and the premature eruption of the mandibular right first permanent molar.

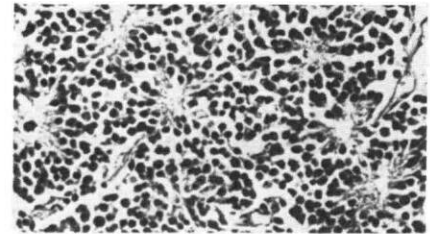


FIG 4. (right) Microscopic view of rosettes of Homer Wright from: Russell DS, Rubinstein LJ: Pathology of Tumours of the Nervous System. London; Edward Arnold LTD, 1959 p 252.

multiple radiolucent lesions of the mandible. The lesions were bilateral.

Treatment and Clinical Course

Bone marrow, skeletal, and soft tissue biopsies revealed malignant infiltrate consistent with the diagnosis of stage IV neuroblastoma. Radiation therapy and chemotherapy were established as the course of treatment due to the extensive proliferation of malignant cells. A 7-day course of cyclophosphamide and a 1-day course of doxorubicin were administered. Three thousand rads of radiation therapy were delivered to the head and spine over a 2-week period. The left temporal swelling improved markedly; however, the paralysis of the lower extremities persisted. Anemia developed secondary to hemorrhage in the oral tumor. Multiple blood transfusions of packed red cells and platelets were administered. The mandibular lesions continued to increase in size. The oral ulceration remained unresolved. Palliative radiation therapy treatment to the mandible (1500 rads over 10 days) was instituted. The disease continued to spread and the patient died 5 months after diagnosis of the neuroblastoma. The cause of death was listed as cardiac failure secondary to bleeding and acidosis as a result of a neuroblastoma.

Conclusion

The report of a 2½-year-old black male with neuroblastoma metastases to the mandible was presented. This report emphasizes the need for early diagnosis of malignant lesions because of their rapid rate of duplication. The late diagnosis in this patient prevented the possibility of surgical excision of the tumor and led to death in a short time. This discussion also provides the dentist with another consideration in the differential diagnosis of oral ulcerative lesions and radiolucent mandibular lesions in children.

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