

## Cranial Vault Lymphoma

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An 80-year old woman presented with a 6-month history of enlarging masses involving the left neck and the back of her head. Physical examination showed an 18 cm x 18 cm x 9 cm mass in the occipital region and a 9 cm x 14 cm violaceous mass in the left neck. Computed tomographic scans of the head and body showed a large mass arising from the occipital scalp extending into the parietal area. There was a 1 cm area of bony erosion through the left occipital bone, with the underlying brain tissue showing contrast enhancement (Figure 1). The left sided neck mass extended from the left parotid gland down to the level of the vocal cords. There were no other significant findings. Biopsy of the occipital mass was consistent with diffuse large B-cell non-Hodgkin's lymphoma. Based upon her clinical presentation and radiographic images, it was felt that her lymphoma had arisen from the scalp area with subsequent spread into the cranial vault, left occipital lobe of the brain and cervical lymph nodes.

The treatment plan was to give 3 cycles of cyclophosphamide, doxorubicin, vincristine, and prednisone combination chemotherapy (CHOP) followed by involved field radiation. She had a dramatic response to the first cycle of CHOP, with near complete resolution of her bulky left neck lymphadenopathy and occipital mass. She was left, unfortunately, with a sizable area of defect in her posterior scalp as well as a 1 cm hole in the calvarium, exposing the dura mater (Figure 2). She subsequently underwent a second cycle of CHOP chemotherapy resulting in further reduction in the size of the lymphoma mass. A trapezius myocutaneous flap was inserted to repair the cranial defect. However, she began to have neurologic and functional decline four weeks later and was found to have progressive disease in the cerebellum. She and her family opted for comfort care and she died shortly thereafter.

Involvement of the cranial vault is an unusual manifestation of aggressive non-Hodgkin's lymphoma. In patients without HIV infection, it is usually a localized disease. Ten patients with non-HIV related primary cranial vault lymphoma have been reported in the literature.<sup>1-9</sup> Complete remission was achieved in 8 of 10 patients with median remission duration of 7+ months (range, 1-72\*). Three of these patients were still disease-free after 2 years. Of the 10 patients, 5 received whole brain radiation alone, 4 received systemic chemotherapy plus whole brain radiation, and one received whole brain radiation and intrathecal methotrexate. Complete remissions were achieved in 3/5, 4/4, 1/1 and patients respectively. While combined modality treatment including anthracycline-based systemic chemotherapy and involved field radiation therapy is considered the standard for localized aggressive non-Hodgkin's lymphoma, these reports suggest that durable remission of primary cranial vault lymphoma can be achieved by radiation therapy alone. In patients with primary cranial vault lymphoma who are unlikely to tolerate systemic chemotherapy, single modality treatment with radiation is a reasonable initial therapy.

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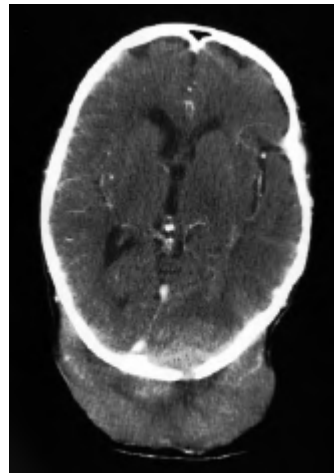


Figure 1.

Figure 2.

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