Primary Burkitt’s lymphoma of the frontal bone

Case report

HASSAN H. A. GAWISH, F.R.C.S.

Neurosurgical Unit, University Teaching Hospital, Lusaka, Zambia

The author reports a case of recurrent Burkitt’s lymphoma in the left frontal region with extradural extension. Craniotomy was performed to relieve rapidly progressive intracranial compression. The patient received cytotoxic treatment postoperatively, with remarkable regression of the tumor.

KEY WORDS □ Burkitt’s lymphoma □ hemiplegia

The neurological manifestations of Burkitt’s lymphoma have been described previously. Involvement of the central nervous system has included extradural spinal compression causing acute paraplegia, and intracranial metastases from a primary lymphoma site elsewhere. This report is of a case in which the tumor originated in the left frontal bone, and invaded the extradural space. There were no signs of any other focus in the body.

Case Report

This 8-year-old boy from northern Tanzania was admitted to the University Teaching Hospital, Lusaka, on September 30, 1973. For 5 months he had had a steadily growing mass in the left frontal region.

History. Three months earlier excision of the tumor was attempted at an upcountry hospital in Tanzania. At that time the tumor was about 5 cm in diameter with its center 8 cm anterior to the coronal suture and 7 cm to the left of the sagittal line. At the time of that operation the patient did not show any neurological or other evidence of systemic involvement. At operation a skull defect was found; under this defect an avascular tumor, white in color, was fungating out. The surgeon’s impression was that the patient had a fractured skull with brain fungus. Unfortunately, the tumor was not examined histologically. Two months later he had a recurrence of the tumor, 10 cm in diameter, again without any neurological signs or systemic involvement. He was referred to the University Hospital of Dar es Salaam and then transferred to our hospital a month later.

Examination. On admission the patient was fully conscious without any neurological signs. The tumor was 15 × 17 cm, located in the left frontal region; it crossed the midline to the right, extended posteriorly to the parietal bone, laterally to the zygomatic arch, and anteriorly to 5 cm from the superciliary margin. The tumor was soft, but not tender. The skin overlying it was stretched, with dilated veins. Systemic examination showed a discrete enlargement of the anterior cervical lymph nodes. Chest, heart, and abdomen were normal on gross examination.
Primary frontal Burkitt's lymphoma

Fig. 1. Left: Preoperative lateral skull film showing rarefaction and irregular erosion of the frontal bone. Right: Preoperative anteroposterior skull film showing separation of the sagittal suture and rarefaction of the left frontal bone. The arrows point to the skull defect found at operation.

Laboratory tests were as follows: hemoglobin 13.8 gm/100 ml; white blood cells 9800; neutrophils 27%; monocytes 2%; lymphocytes 55%; eosinophils 16%; sedimentation rate 40 mm/hr, Westergren method. Urea and electrolytes were normal. The high eosinophilic count was explained by the presence of an Ascaris infestation.

Skull films showed separation of sutures and rarefaction of the left frontal bone with an area of bone erosion (Fig. 1). X-ray films of the chest and abdomen, and an intravenous pyelogram were normal. Angiography showed a large left frontal mass displacing the anterior middle cerebral arteries as well as the middle meningeal artery medially and downward (Fig. 2). No blood circulation to the tumor was detected.

By October 3, the tumor had grown rapidly; the anterior margin was then 2 cm from the superciliary margin. The patient started vomiting, and within 12 hours he became drowsy, responding only to pain, with dilated fixed left pupil and spastic right hemiplegia.

Operation. An emergency craniotomy was performed under general anesthetic, using an endotracheal tube. The skin of the forehead was reflected forward through a coronal incision which caused severe bleeding from the skin. The tumor itself was avascular, whitish-blue in color, and very soft. The extracranial portion of the tumor was removed, revealing a bone defect 5 cm in diameter at the center of the left frontal bone. The defect was enlarged by bone nibbling, removing the left frontal bone and sparing a thin rim over the superciliary margin and orbital plate. The superior squamous section of the temporal bone was removed as well as the anterior portion of the left parietal bone and the medial part of the right frontal bone. The tumor was 5 cm thick.

Fig. 2. Preoperative left carotid angiogram, frontal view in the late arterial phase. The anterior cerebral artery (A.C., arrow) is displaced across the midline to the right, and the middle cerebral branches (M.C., arrow) are displaced medially. Note the avascular area underneath the frontal bone.
in places; intracranially it extended forward to the orbital roof, laterally to the lateral end of the sphenoid wing, posteriorly it spread over the anterior part of the parietal lobe and crossed the midline to the right. It was adherent all along its deep surface to the external layer of the dura. A large portion of the tumor was removed; the dura was not opened, and the wound was closed.

Pathological Examination. Laboratory examination of the excised section showed malignant tumor tissue consisting of proliferating lymphoblasts, with large histiocytes scattered through the field giving the characteristic starry-sky appearance of Burkitt’s lymphoma (Fig. 3). Methyl green pyronin stain showed pyroninophilia of the tumor cells.

Postoperative Course. The patient showed steady improvement. He was transferred to Dar es Salaam on the ninth postoperative day fully conscious, with spastic right hemipareses. The wound healed completely. Routine postoperative lumbar puncture showed no tumor cells and normal protein content.

The right hemipareses continued to improve and by mid-December, 1973, he had full motor power of the right leg and improved use of the right arm. He had three courses of Endoxan (cyclophosphamine) by intravenous infusion, the first on the 11th postoperative day with 2-week intervals between each course. Since his operation he has shown no signs of tumor recurrence.

Discussion

The age of the patient, the area he came from, and the rate of tumor growth, are all in keeping with the diagnosis of Burkitt’s tumor. The fact that the patient had a skull swelling for 5 months, without any systemic involvement apart from enlarged cervical lymph nodes, points to the skull vault as the primary focus. Biopsy of the tumor was contemplated but surgery was necessary as an urgent procedure to relieve intracranial compression. The dura was not opened, so as to leave an intact barrier in case the tumor should be found to be malignant. No frozen section was available at that time. Covering the skull defect will be considered after the possibility of recurrence is excluded.

Although there is no definite indication of the origin of the tumor, we can conclude on clinical grounds that it arose from the left frontal bone. This is an unusual site of origin for intracranial Burkitt’s lymphoma. The primary focus is normally in the facial bone or in the abdomen with direct invasion or hematogenous dissemination to the brain.

References

Primary frontal Burkitt’s lymphoma

16. Wright DH: personal communication, 1974