Cerebellar liponeurocytoma: a novel report from Nigeria in a 6-year-old girl, and review of literature

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Abstract

Liponeurocytoma is a newly defined clinical entity predominantly seen in the cerebellum as a slow-growing tumor. In this report, we present the case of a 6-year-old Nigerian girl with a liponeurocytoma, and review of literature.

Case Report

Patient is a 6-year-old girl who presented with headache and vomiting, both of four weeks duration. Headache was insidious, dull, predominantly left sided, worse in the morning associated with vomiting which relieves it. She also had associated gait disturbance. Neurological examination showed a fully conscious female child with intact high cerebral function. She has truncal ataxia, otherwise the cerebellar function was preserved. Other systems were unremarkable.

Cranial computed tomography and magnetic resonance imaging revealed left cerebellar tumor with obstructive hydrocephalus (Figures 1 and 2). She had an initial right frontal Venticulo-peritoneal shunt for cerebrospinal fluid diversion, and late left retrosigmoid suboccipital craniotomy with microscope assisted gross total resection of left hemispheric tumor one week later under general anesthesia. The excision was confirmed by radioimaging after surgery (Figure 3).

Intraoperative finding was soft greyish to yellowish mass with partly cystic consistency and a fairly well defined brain-tumor boundary. There was a leash of tumor vessels that bled briskly. She had fever and mild anemia post-surgery and these were managed. She was discharged from the hospital two weeks later with a Karnofsky score of 90% and is still being followed up in the out-patient, while awaiting possible radiotherapy if her condition does not remain stable. Histology showed an extensively lipidized tumor composed of sheets of mature adipose tissue with some neurocytic cells in the background minority with hyalinized blood vessesls. No mitosis was seen. Immunohistochemistry was positive for S-100 and desmin consistent with lipocystic medullocytoma.

Discussion and Conclusions

Liponeurocytoma is a relatively newly defined clinical entity predominantly found in the cerebellum as a slow-growing tumor located predominantly in the cerebellum,[1] old synonyms include lipomatous medulloblastoma, lipidized medulloblastoma, medullocytoma, neurolipocytoma, lipomatous glioneurocytoma, and lipidized mature neuroectodermal tumor of the cerebellum. In 2000, the World Health Organization (WHO) pathology and genetics of tumors of the nervous system,[1] described cerebellar liponeurocytoma as a distinct entity from medulloblastoma in terms of prognostic, epidemiological and clinical aspects. This rare tumor is WHO grade I–II, generally with an accordingly indolent behavior. Since 1978, when liponeurocytoma was first described, there have been more than 40 reported cases.[1]

Our report is a novel case in literature not only in Nigeria but also in the whole of Africa, to the best of our knowledge. There have been 43 other reports of liponeurocytoma in literature.[1] The patients' ages ranged from 4 to 69 years, with a median of 49 years. Our index case is 6 years old female. Ten (24%) of the lesions were found in patients aged 30 years and younger, and 32 (76%) of the lesions were found in patients older than 30 years. There were 22 female (52%) and 20 male (48%) patients. Thirty-six lesions (86%) were found within the cerebellum, and 6 (14%) were found in a supratentorial location. The average follow-up for patients undergoing surgery for liponeurocytoma was 48 (range 0-192) months. There have been a number of recurrences with a mean time from diagnosis to first local recurrence presentation of 10.6 (range 10-12) years.[1] Surgical resection is recommended as the initial treatment modality for patients with liponeurocytoma to establish a diagnosis. The rare occurrence and diagnosis of this type of tumor as well as its variable appearance on imaging can make radiological diagnosis challenging.[2]

There is no consensus regarding the treatment of liponeurocytoma, specifically whether chemo- or radiotherapy is a necessary part of

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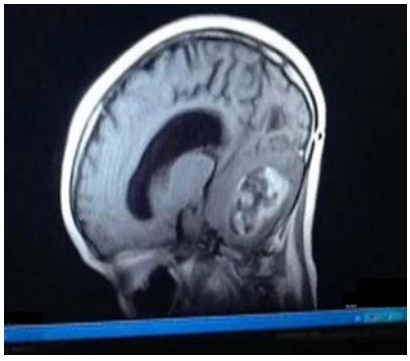

Figure 1. Tumor with dilated lateral ventricle.

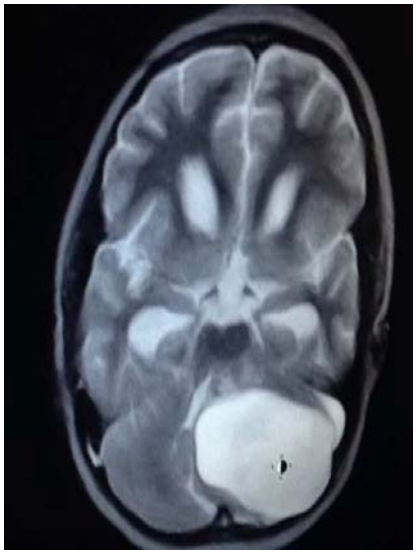

Figure 2. Contrast with enlargement of both ventricles and contrast enhancing tumor.

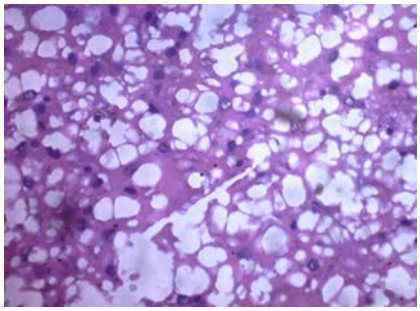

Figure 3. Histology ×400 showing lipocystic neurocytoma of the cerebellum.

the postoperative treatment regimen. Intraoperative finding was soft greyish to yellowish mass with partly cystic consistency and a fairly well defined brain-tumor boundary. There were leash of tumor vessels that bled briskly. She had smooth recovery from anesthesia and was monitored in the intensive care unit over the night. The post-surgery scan is shown in Figure 4. She had fever and mild anemia post-surgery and these were managed. She was discharged from the hospital two weeks later with a Karnofsky score of 90% and is still being followed up in the outpatient.

We recommend longer follow-up periods. Most follow-ups in the literature are shorter than 5 years, a period during which the tumor may still be in a silent phase, starting to regrow. However, rather than exposing the patients to the risks and side-effects of radiotherapy, without any evidence to support its usefulness in preventing recurrence, we recommend reoperation on a recurrent tumor, which has only a slightly more aggressive histology than the primary presentation, with the option of using adjuvant radiotherapy at this time.[3] Further studies regarding the natural history of this lesion are warranted.[4]

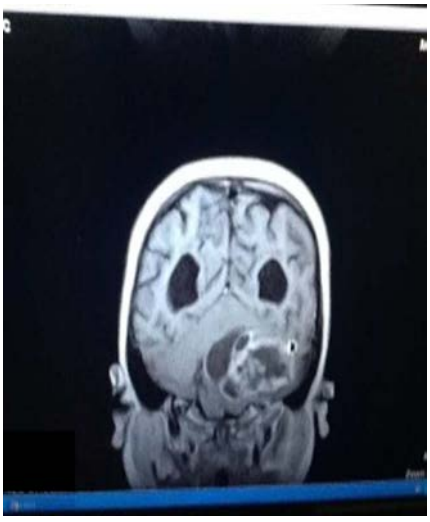

Figure 4. Post surgery assessment.

In conclusion this lesion appears universal though rare and is essentially the same as those seen in other parts of the world.

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