A rare case of cerebellar anaplastic pleomorphic xanthoastrocytoma

ABSTRACT

Pleomorphic xanthoastrocytoma (PXA) is a low-grade glioma comprising 1% of all astrocytomas with an extremely rare anaplastic counterpart usually found in young adults. These tumors are most often cerebral in origin and their presentation in the elderly signifies poor prognosis. As these tumors are an important differential of glioblastoma, diagnosing them accurately is essential for management. We present a 68-year-male with positive cerebellar signs and clinico-radiological impression of cerebellar metastatic deposits, subsequently diagnosed as cerebellar PXA with anaplastic features. The case in discussion is unique in its age, site, and grade of presentation, with key histological features rebuking the clinical and radiological diagnosis of metastasis. The rarity and ambiguous management protocol of these tumors make their documentation an important addition to the existing literature with emphasis on possibility of late presentation and at sites other than the cerebrum.

KEY WORDS: Anaplastic, cerebellum, pleomorphic xanthoastrocytoma

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INTRODUCTION

Pleomorphic xanthoastrocytoma (PXA) is a rare glioma of the central nervous system, with the anaplastic variant being extremely uncommon. It was first described as a unique entity in 1979 constituting <1% of all astrocytic neoplasms.[1,2] It mostly affects children and young adults, with a median patient age of 22 years at diagnosis.[2] We present a case of a 68-year-male with a cerebellar lesion diagnosed as PXA with anaplastic features.

CASE REPORT

A 68-year-old male patient was admitted to our institute with complaints of difficulty in walking. He had a history of seizures with no history of vomiting, visual disturbance, or relevant familial history. The patient was hypertensive for 10 years and on neurological examination had right cerebellar signs like gait ataxia.

Preoperative magnetic resonance imaging scan of the brain revealed a 5 cm × 5 cm × 3.4 cm irregular, mildly enhancing space-occupying lesion in the right half of the cerebellum with tiny cystic areas. The lesion was isointense to hypointense on T1W1 and hyperintense on T2W1 and fluid-attenuated inversion recovery. Computed tomographic (CT) scans of the brain revealed hypodensities in the right cerebellar hemisphere with hyperdensities in the periphery. Diagnosis of metastatic tumor of unknown primary was made on imaging and tumor decompression surgery was performed.

Gross examination showed multiple gray brown-to-white soft-tissue pieces altogether measuring 4.5 cm × 3.5 cm × 1 cm. The tissue was processed in its entirety.

Microscopic examination showed fragments of tumor comprising highly pleomorphic cells and multinucleated giant cells with cytoplasmic vacuolations in a fibrillary background with adjacent unremarkable cerebellar parenchyma [Figure 1a and b]. Spindle cells were seen intermingled with multinucleated giant astrocytes and large multinucleated xanthomatous cells. Eosinophilic hyaline droplets were also observed among the tumor cells. Reticulin fibers were present. Mitotic count was up to 6 per 10 high-power fields [Figure 2]. There was no evidence of microvascular proliferation or necrosis. On immunohistochemistry, tumor cells were diffusely

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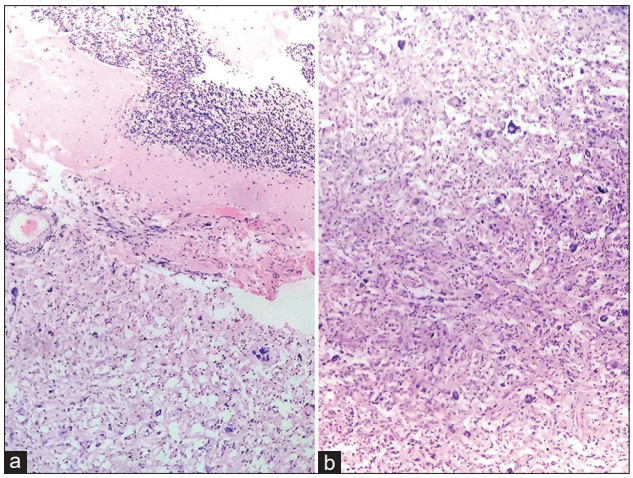

Figure 1: (a) Sections show fragments of cerebellar tissue along with tumor tissue showing highly pleomorphic cells and multinucleated giant cells with cytoplasmic vacuolations in a fibrillary background. Spindle cells are intermingled with multinucleated giant astrocytes (H and E, ×40). (b) Sections show fragments of cerebellar tissue along with tumor tissue showing highly pleomorphic cells and multinucleated giant cells with cytoplasmic vacuolations in a fibrillary background. Spindle cells are intermingled with multinucleated giant astrocytes (H and E, ×100)

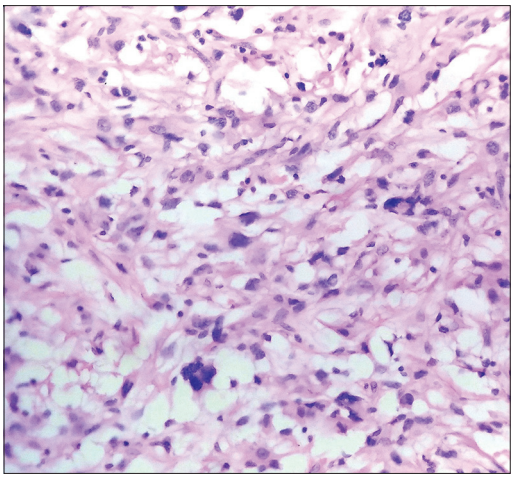

Figure 2: Sections show large, multinucleated xanthomatous cells having intracellular lipids pushing cytoplasmic organelles and glial filaments to the periphery are noted. Eosinophilic hyaline droplets are also observed among the tumor cells (H and E, ×400)

positive for glial fibrillary acidic protein (GFAP), S100P, and vimentin but negative for CD38, CD68, and pancytokeratin. Foamy macrophages surrounding some tumor cells were positive for CD68. The Ki67 labeling index was 30% indicating a high proliferative index. Hence, a diagnosis of anaplastic PXA was rendered.

The patient received external beam radiotherapy postoperatively and is under close follow-up since then.

DISCUSSION

PXA is an astrocytic tumor most commonly found in the cerebral hemispheres affecting males and females equally.[2]

Seizures, epilepsy, and focal neurological deficits are common forms of presentation.[3] It may progress to higher grades. Cerebellum is one of the rare sites of origin of this tumor, and if occurs at all, the patient presents with headache, dizziness, or cerebellar signs as was in our case.[4]

By definition, PXA is comprised of large pleomorphic and frequently multinucleated cells, spindle and lipidized cells, a dense pericellular reticulin network, and numerous eosinophilic granular bodies. PXA tumor cells are neoplastic astrocytes, but there is often neuronal differentiation.[3]

Anaplastic PXAs have similar microscopic features as PXA except for brisk mitotic activity and marked areas of necrosis. Thus, PXAs with >5 mitoses per 10 high-power fields are considered anaplastic PXAs and are bracketed under the WHO grade III category.[5] They have a poorer prognosis, with only 20 reported so far.[4] The tumors are strongly immunoreactive for GFAP, and the absence of microvascular proliferation and pseudopalisading necrosis helps to distinguish it from glioblastoma.

Our case had mitotic counts of six per ten high-power fields and a Ki67 labeling index of 30% indicating aggressive behavior of the tumor.

CT scan appearance of the tumor is solid-cystic to variable with lesions showing hypodense to hyperdense areas as was also observed in the case under discussion.[6]

Although the management is not standardized, it has been observed that a gross total resection has a better outcome rather than decompression surgery. Thus, the surgeon needs to have an accurate diagnosis and categorization of the WHO grade to enable a complete resection as subtotal resection coupled with high mitotic count and necrosis augers a bad prognosis for the patient.[2]

Neurofibromatosis 1 is associated with pilocytic astrocytomas and sometimes rarely with cerebral pleomorphic astrocytomas.[4] These tumors are also associated with mutations like BRAF V600E and sometimes with loss of SMARCB1.[5]

The differential diagnoses of PXAs are ganglioglioma, epithelioid glioblastoma, desmoplastic infantile ganglioglioma, and malignant fibrous histiocytoma.[6]

CONCLUSION

The rarity and ambiguous management protocol of these tumors make their documentation an important addition to the existing literature with emphasis on possibility of late presentation and at sites other than the cerebrum. In addition, the uncommon age and site of presentation itself

confer it unique for reporting so as to improve the clinical and pathological knowledge of these tumors,

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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