Case report:

A rare glial tumor entity: Astroblastoma

Meena Patil*, Manjiri Karandikar**, Sarita Garde***

*Assistant Professor. ** Professor, *** Associate Professor
Department of Pathology, B.V. U. Medical College, Pune, Maharashtra, India
Corresponding author: Dr. Meena Patil : Email- meenipatil@yahoo.co.in

Abstract:
Astroblastoma is a rare primary brain tumour comprising < 3% of all gliomas. It usually presents as a well-demarcated, contrast-enhancing mass in the cerebral hemisphere. It occurs in children, adolescents & young adults. Ependymoma & astroblastoma show perivascular pseudorosettes & hyalinisation of blood vessels & hence may pose a diagnostic problem. Herewith we reported a case of a 26 year old female with signs & symptoms of intracranial space occupying lesion. Intraoperative cytology showed perivascular pseudorosettes. A provisional diagnosis of ependymoma was considered. Histopathology showed an astroblastoma.

Key words: Astroblastoma

Introduction:
Astroblastoma is a rare glial neoplasm of unknown origin and uncertain prognosis. It constitutes 0.45 to 2.8% of brain gliomas.

It usually presents as a large, well-demarcated, supratentorial cerebral hemispheric mass. Fronto-parietal region is preferred. Usually younger age group is affected.

Most patients present with intermittent headache, nausea, impaired vision, seizures. It is subtyped as low-grade and high-grade astroblastoma. The lesion is characterised by a papilliform architecture, astroblastic pseudorosettes & perivascular hyaline-isation. Complete surgical removal is the treatment of choice in astroblastomas, although high-grade tumors have a high recurrence rate.

Case Report:
A 26 year old female presented with headache, vomiting & giddiness since 2 days. She had intermittent headache for 2-3 years. Computerised Tomography (CT) scan of brain showed a large, heterogeneous solid to cystic lesion 6.5 x 5.6 x 6 cm. in size in right frontal lobe with mass effect. A diagnosis of glioma was suggested.

Magnetic resonance imaging (MRI) of brain showed, a large 6.2 x 5.5 x 3.6 cm., solid/cystic neoplastic lesion in right fronto-parietal region with involvement of part of body of corpus callosum showing enhancement pattern. It appeared to be in close proximity to right middle cerebral artery & was vascular in nature with some areas of hemorrhage within it. There was no connection of the tumor with any ventricle. Differential diagnosis of Glioma or Glioblastoma multiforme was suggested.

Craniotomy and small corticotomy was done & the tumour was excised. A small intraoperative biopsy was sent for crush/squash cytology for provisional rapid diagnosis. Cytology showed a densely cellular tumour. Tumour cells were arranged around blood vessels showing rosette-like arrangement i.e. pseudorosettes. A provisional diagnosis of ependymoma was suggested. Multiple irregular
grey-white bits of the ICSOL were sent for histopathology.

Histopathology revealed a cellular tumour composed of club shaped cells with short thick cytoplasmic processes with terminally expanded foot plates loosely arranged around blood vessels. Nuclei were hyperchromatic & angulated. Areas between perivascular rosettes were distinctly rarified. IHC for GFAP, S-100 protein & Vimentin was done & it showed positivity for all three confirming glial origin of the tumor. Taking into consideration the clinical history, radiological findings, intraoperative cytology, histopathology & IHC, a final diagnosis of astroblastoma was offered. Absence of necrosis & mitosis suggested that it was a low grade tumour. The patient was sent for chemotherapy but was lost for follow-up.

Discussion:

Astroblastoma is a rare type of glioma comprising <3% of all gliomas. Approximately 40 cases of astroblastoma have been reported in the literature since Bailey and Bucy reported the condition for the first time in 1930. Bailey and Bucy believed that astroblastoma originated from astroblast, an intermediate stage between glioblast and astrocyte. Astroblastomas occur most frequently in young adults, occasionally in children & rarely in infants. Most develop superficially in the cerebral hemisphere but they may occur at other sites as corpus callosum, cerebellum, optic nerves, brain stem & cauda equine. On imaging studies, astroblastomas are typically well-circumscribed, predominantly solid masses. Cystic change may be present, but is usually not extensive. The cell of origin of astroblastoma is not exactly known, but widely accepted to be the astroblast- an intermittent cell between spongioblast & astrocyte. Till recently, the possible origin from tanycyte was considered.

Raised intracranial pressure & seizure episodes are the common clinical presentations. Other signs & symptoms are visual loss, memory disturbances, weakness & altered sensorium. Macroscopically, they are usually well-circumscribed & solid masses with a homogeneous cut surface; although larger examples may show both cyst formation & necrosis. Microscopically, the lesion is characterised by papilliform architecture with radially arranged cellular elements directing unipolar cytoplasmic processes in “cartwheel” fashion towards centrally placed stromal blood vessels. Individual tumor cells are elongated with relatively abundant eosinophilic cytoplasm. Blood vessels often show progressive collagenous thickening & hyalinization. In long standing cases it can lead to partial fibrous obliteration of the neoplastic tissue.

Histopathological appearance of these tumors closely resembles that of an ependymoma. However, astroblastomatous cell processes are shorter, stouter & less tapering than those forming ependymal type pseudorosettes. Ependymomatous pseudorosettes generally lie embedded in a dense fibrillar matrix, whereas the gliovascular structures of the astroblastoma are supported by an intervening population of astrocyte-like tumor cells. Diffuse fibrillary astrocytoma of gemistocytic variety having papillary architecture with perivascular pseudorosettes & papillary meningioma are other important differential diagnoses.

Absence of fibrillar background & shorter, broader & less tapered unipolar cell processes forming rosettes gives important clue that the tumour is not an ependymoma. Besides,
ependymomas usually do not exhibit marked vascular sclerosis. The nuclei of astroblastomas are somewhat larger than those of ependymomas & more uniform than those of diffuse fibrillary astrocytomas. Astroblastomatous cell processes terminate on the target vessels as expanded foot plates while ependymal cell processes become thin or hair-like/fibrillar by the time they terminate on target vessels. Papillary meningiomas, though well-demarcated, cytologically uniform & rich in perivascular, pseudorosettes, are extra axial & dura-based. They show areas of clear-cut meningioma.\\[1\\]

Astroblastomas along with gliomatosis cerebri and polar spongioblastoma are included in neuroepithelial tumors of uncertain origin and are grade IV tumors as per new WHO classification of brain tumors suggested by Stephen B. Tatter; contrary to grade I in WHO 2007 classification of brain tumors.\\[6, 8\\] Bonnin et al. reported two distinct histological types: A low-grade type with better differentiated pattern and favourable postoperative prognosis and a high-grade type showing more anaplastic microscopic features with short postoperative survival. High-grade lesions show focal or multifocal regions of high cellularity, anaplastic nuclear features, elevated mitotic indices, vascular proliferation, and necrosis with pseudopalisading.\\[8\\]

Immunohistochemically, astroblastomas are immunoreactive to GFAP, S-100 protein, & Vimentin. The majority displays a focal cytoplasmic immunoreactivity for EMA. IHC for CAM5.2 & Synaptophysin has not been documented.\\[5\\] These changes are not typical of either ependymomas or astrocytomas.\\[7\\] Natural history of astroblastoma seems to place it in between astrocytoma and glioblastoma. Total resection is the best way of treating an astroblastoma. Regular follow-up is required even in low-grade variants due to unpredictable behaviour. Adjuvant therapy is recommended for high-grade and recurrent cases. Favourable prognosis is almost invariably associated with well-circumscribed tumors which permit total resection of tumor in all grades. In a series of 23 patients reported by Bonnin and Rubinstein, patients with high-grade astroblastomas who did not receive postoperative radiotherapy had a shorter survival time. Caroli et al. reported a high-grade astroblastoma with a 5-year survival without recurrence after total resection, radiation therapy, and Temozolamide usage.\\[8\\]
Cytology 1 and 2

References: