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Subependymal giant cell Astrocytoma

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Abstract

Subependymal giant cell astrocytomas (SEGAs) are slowly growing tumours corresponding to WHO grade I. They are intraventricular and usually occur in the setting of tuberous sclerosis complex. They often result in obstructive hydrocephalus. Treatment is usually restricted to surgical resection, recurrences are rare and long term prognosis is excellent. We present a series of three cases.

Introduction

Subependymal giant cell astrocytomas (SEGAs) are benign, slowly growing tumours corresponding to WHO grade I.¹ These present in first or second decade of life, are intraventricular in location and usually occur in the setting of tuberous sclerosis complex (TSC).² However, they are not restricted to this setting. TSC comprises a group of autosomal dominant disorders which are characterized by hamartomatous and benign neoplastic lesions in the CNS and various extraneural tissues.¹ Treatment of SEGAs is usually limited to surgical resection and recurrences are rare.² These are rare tumors. In studies on CNS neoplasms conducted at AKU, SEGAs were extremely rare.^{3,4} Three cases of subependymal giant cell astrocytoma which were resected are reported.

Case Report

Case 1

A 3 years old girl from Quetta presented with a history of seizures and projectile vomiting. C.T. scan showed a bulky, contrast enhancing mass in the wall of the right lateral ventricle near the foramen of Monro. No history of tuberous sclerosis complex was available. The gross specimen was received in fragments measuring 1.5 x 1.5 cms in aggregate. These were submitted entirely. Histology showed a neoplastic lesion composed of large polygonal cells with abundant hyaline pink cytoplasm. The nuclei had fine granular chromatin with prominent nucleoli. Occasional cells showed nuclear pleomorphism and multiple nuclei. Perivascular pseudopalisading pattern was also seen. There was no significant mitotic activity or necrosis and calcification was not seen. The case was diagnosed as subependymal giant cell astrocytoma, WHO grade 1. Clinical work up and correlation to confirm / rule out tuberous sclerosis complex was advised.

Details of surgery and postoperative complications were not available.

Case 2

An 11 year old boy presented with a history of seizures since he was a few weeks old, which persisted despite treat-

ment. He suffered from drop attacks and had delayed milestones. Earlier MR showed bilateral subependymal tubers. There was family history of Tuberous sclerosis complex. The frequency of seizures increased recently and he began to suffer six to eight drop attacks per day. His latest MRI showed a circumscribed intraventricular mass in the right lateral ventricle with extension into the 3rd ventricle near the foramen of Monro with resulting obstructive hydrocephalus.

The specimen consisted of multiple fragments measuring 2.5 x 2.0 cms in aggregate and was submitted in toto. Histopathology showed a cellular neoplasm composed of closely packed large polygonal to spindle cells with eccentric vesicular nuclei having prominent nucleoli and abundant glassy hyaline eosinophilic cytoplasm. Multiple foci of microcalcifications were seen. Mitotic activity was not significant and necrosis was not present. On immunohistochemistry, tumour cells were positive for glial fibrillary acidic protein (GFAP) and S100 protein. Based on histologic and immunohistochemical features, a diagnosis of subependymal giant cell astrocytoma, WHO Grade 1 was made.

The patient was reopened, intra ventricular drainage was performed, and ventriculo peritoneal shunt was placed. The patient is doing well.

Case 3

A 4 year 9 month old male child had permanent perception disorder since birth. He developed gradual weakness, drowsiness, lethargy, and ultimately seizures. He was diagnosed to have tuberous sclerosis. His MRI showed a large, partly calcified contrast enhancing mass arising from third ventricle and extending to right and left lateral ventricles.

The gross specimen consisted of multiple fragments of tissue measuring 8 x 6 x 3 cms in aggregate. Representative sections were submitted in 3 cassettes.

Histopathological sections showed a neoplastic lesion composed of large closely apposed polygonal and rounded cells with glassy, hyaline eosinophilic cytoplasm and eccentrically placed vesicular nuclei with prominent nucleoli. In areas, tumor cells were seen to condense around blood vessels. On immunohistochemistry, tumour cells showed positivity for GFAP and S100 protein. A diagnosis of subependymal giant cell astrocytoma, WHO Grade 1 was made.

The patient was reopened and tumor was excised. No shunt was placed. There were no post-operative complications. Follow up is so far unremarkable.

Discussion

The major CNS manifestations of TSC include distinctive foci of gyral expansion (tubers), hamartomatous subependymal glial nodules and SEGAs, with the latter appearing

evolve from the enlargement of subependymal nodules. Extra-neural manifestations of TSC include cutaneous angiofibromas, subungual fibromas, cardiac rhabdomyomas, intestinal polyps and renal angiomyolipomas.² TSC results from mutations in TSC1 and TSC2 genes located on chromosome 9q and 16p respectively.⁵ The cortical tubers may be detected by CT or MRI.⁶

SEGAs typically arise in the wall of the lateral ventricles and produce obstructive hydrocephalus by increasing intraventricular pressure. On CT / MRI, these are usually seen as large, bulky and variably calcified contrast enhancing masses in the region of the foramen of Monro (resulting in hydrocephalus). They are usually circumscribed and grossly sharply delineated from the neighbouring brain tissue, and are usually anchored to the ventricular wall over a broad front.²

Being sharply demarcated, they either do not or only minimally infiltrate the neighbouring brain parenchyma and do not use the CSF to escape from the ventricles.^{1,7}

On microscopy, due to their discreteness, SEGAs are composed only of tumour cells and a vascular stroma without the incorporated brain parenchyma which is characteristically seen in the fibrillary astrocytomas.² They are uniformly cellular with large round to polygonal cells having pink glassy hyaline cytoplasm and eccentric vesicular nuclei with prominent nucleoli (Figure 1). Clustering of tumor cells and perivascular pseudopalisading are common. Considerable nuclear pleomorphism and multinucleated cells are frequently seen.¹ On immunohistochemistry, there is focal positivity to GFAP, and more strong, widespread positivity for S100 protein, (Figure 2) and some positivity for Neurofilament (NF). This suggests that these tumours are hybrids expressing structural proteins of both glial and neuronal cells.⁸

After surgical resection, post-operative course is uniformly favourable since even large tumours can be excised, and long term prognosis is excellent.² Even lesions with mitoses, microvascular proliferations, and foci of necrosis have a favourable prognosis.^{7,9,10}

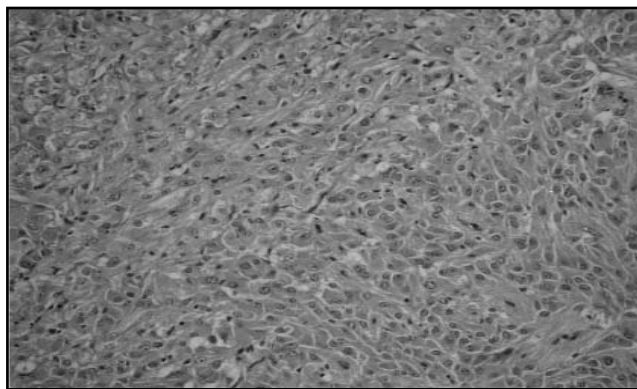


Figure 1. Section showing large polygonal cells with abundant pink cytoplasm and large vesicular nuclei. H & E (X20).

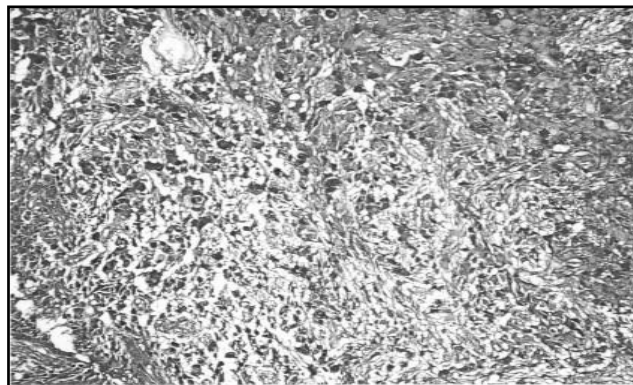


Figure 2. Section showing immunohistochemical staining with S100 protein. (X10)

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