Case Report

Non Infantile Desmoplastic Astrocytoma: a case Report

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Received: 11 September 2015 Initial Review: 14 October 2015 Accepted: 24 October 2015 Published Online: 23 November 2015

ABSTRACT

Desmoplastic infantile astrocytomas (DIA) are huge cerebral masses with cystic and solid components in infants with a benign course. Less than 50 cases of have been reported in the literature and most of the cases are children below 24 months. We are presenting a rare case of DIA which presented at the age of 4 years with vomiting and headache in neurosurgical OPD. He was diagnosed with a huge left temporal parietal tumor with midline shift which on histopathological examination was confirmed as DIA. Patient was operated and had good recovery and was followed up for two years.

Keywords: *Astrocytoma, Benign Tumors, Desmoplastic, Infantile, Pediatric Tumors*

Desmoplastic infantile astrocytomas (DIA) are huge cerebral masses with cystic and solid components which are usually found in infants. Van den Berg et al first described Desmoplastic infantile ganglioglioma (DIG) in 1987 which was quite similar to the DIA described by Taritatu in 1984 [1,2]. Less than 50 cases of DIA have been reported in the literature. In India these were first described in 2005 by Bala Subramanian et al and since then various reports have come up [3]. The tumors follow a benign course even though the MRI imaging suggests aggressive tumor. Most of the cases reported in literature are children below 24 months. Here, we are presenting a rare case of DIA, which presented at the age of 4 years. Only 8 such cases with late presentation are described in literature.

CASE REPORT

A 4-year-old male child presented with vomiting, headaches and excessive irritability of 2 months duration for which the child was being conservatively managed at a local hospital. There was no history of trauma, fever, convulsions or unconsciousness. There was no history of contact with tuberculosis. The child was born full term with normal vaginal delivery. The child had a large head since birth, and the parents were advised to show to a physician, but as the child had no symptoms; hence, parents did not consult anybody.

On admission, child was conscious, irritable with signs of raised intracranial pressure (heart rate – 74/ minute, blood pressure – 124/86 mm-Hg, and respiratory rate – 26/minute). General examination revealed a large head with head circumference of 52 cm (95th percentile), child was dehydrated and irritable. His fundus examination revealed bilateral papilloedema. His central nervous system examination revealed increased tone with exaggerated deep tendon reflexes on right upper and lower limbs. Babinski sign was positive on right side. No cranial nerve palsy was noted. He was admitted in ICU and treatment was started with intravenous antibiotics, mannitol, fluids and supportive measures.

Routine hematological and biochemical test were within normal range. MRI brain showed a large intra axial mass. T1 weighted images showed that the mass had variegated appearance with midline shift and left lateral ventricular extension. There were areas of cystic

degeneration and the right ventricle was enlarged due to pressure at foramen monro (figure 1 A & C). On contrast enhanced pictures, the solid parts of the tumor were strongly enhancing while cystic and degenerated parts were hypo intense (figure 1 B & D). The tumor extended to left temporo parietal and occipital region with hydrocephalus and midline shift.

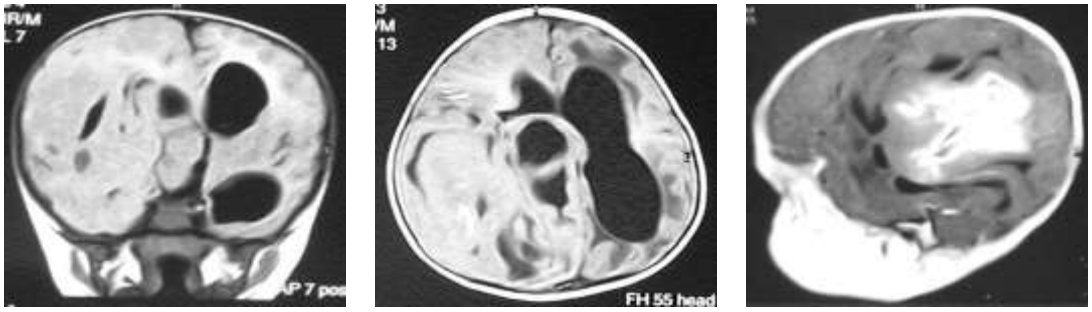

Figure 1 - A, B figures show T1 weighed images showing temporo parietal occipital variegated mass having solid and cystic components with midline shift. C figures suggest intense enhancement of contrast further differentiating degenerated parts and solid parts of tumor with midline shift.

A left temporo-parieto-occipital craniotomy was performed. The solid tumor measured about 6cmx5cmx8cm and had dural attachment. It was firm and relatively avascular with rubbery consistency at places. The cystic areas inside the tumor were gelatinous with yellowish fluid accumulations. The whole mass was excised completely and complete hemostasis achieved. A Right ventriculo-peritoneal shunt was done a few days after surgery because of developing hydrocephalus.

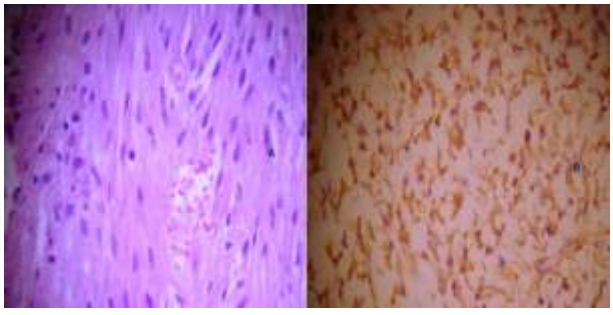

Figure 2 – A Photomicrograph showing spindle shaped pleomorphicatrocities in desmoplastic stroma along with prominent blood vessels (HEx200). B Photomicrograph GFAP positive atrocities (GFAPx100)

Histopathology showed that the tumor had marked desmoplastic component with neoplastic astrocytes. Mild cellular pleomorphism and little mitosis were seen. Primitive neuroepithelial elements were seen, as isolated nodular aggregates of undifferentiated small cell population (Fig. 2). Immunohistochemistry showed strong reaction with glial fibrillary acidic protein suggesting intense desmoplastic material. The picture was consistent with desmoplastic infantile astrocytoma.

DISCUSSION

Taratulo is credited with first reporting DIA in 1984 [1]. Van den Berg in 1987 had classified these into 2 types DIA and DIG depending upon the presence of ganglionic cells [2]. Later, both jointly classified as desmoplastic infantile ganglioglioma (DIG) and kept in WHO grade I tumor as these are superficial, benign tumors of infancy of neuroepithelial cell origin [4]. There are around 60 cases of DIA reported in literature [5]. Mostly the tumors present in infants under the age of two years, and only few (6-10) cases of these tumors are reported in children above 2 years of age [5].

DIA constitute 0.5-1% of cases of total intracranial tumors [5]. In most of the cases, these are mixed in nature having both solid and cystic components and only two cases are reported with purely solid components [6]. Our case had 90% solid with only 10% cystic areas. The common presentation is of huge masses with cystic and solid areas. These commonly occupy frontal and parietal cerebrum and only a few involve occipital lobe as seen in our case [7,8]. Atypical tumors with diencephalon, hypothalamus, and suprasellar location have also been described. Many tumors with bilateral location have been described [7]. The patients present with features of raised intracranial pressure and at times cranial nerve defects [7]. All these patients have similar computerized tomography scan and magnetic resonance imaging characteristics.

Histological, the tumor has been well described in literature. These are classified as grade I by WHO classification, which means they are low grade with

excellent prognosis and less chances of recurrence (4). These tumors are characterized by their intense desmoplastic cells and have been further divided into DIA, DIG and pleomorphic Xanthoastrocytoma [9]. The differentiating features among the three are DIA have no ganglion cells, DIG with ganglion cells on immunohistochemistry and intracellular lipid in pleomorphic variety [10,11]. Ganglion cells if present are positive for neurofilament and synatophysin in (DIG). Further these tumors have low mitotic rate and neoplasia.

There are reports of large series of these patients treated with excision and responding well to treatment with no recurrence [12-15]. Partial resections of some deep-seated tumors have been given adjuvant chemotherapy with limited success [14]. The tumors commonly occur in frontal or parietal lobes, with a solid component closely attached to the dura, which intensely enhances with contrast, surrounded by a parenchyma cystic component and mass effect. These tumors are amenable to surgical excision because of their surface location and distinct firmness with clear demarcation from the surrounding normal brain. No adjuvant therapy is recommended after total excision. Long-term prognosis after total surgical excision has been excellent.

CONCLUSION

Desmoplastic astrocytoma is a rare benign tumor of infancy, but rarely, it may present in elder children also. Hence, it should be kept in differential of brain tumors in children. The tumor has excellent prognosis, if detected in time and excised completely.

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How to cite this article: Malhotra A, Malhotra M, Punia RPS. Non-infantile desmoplastic astrocytoma - A case report. Indian J Case Reports. 2015; 1(4): 71-73.

Conflict of interest: None stated, Funding: Nil