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Diffuse intrinsic pontine gliomas in adults: A retrospective study

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Abstract

Background: Brainstem gliomas (BSG) constitutes very small proportion in adults brain tumors with pons as most common location. There is significant paucity in literature for adult diffuse intrinsic pontine gliomas (DIPG). **Objective:** In this study, we attempt to review the outcomes of DIPG in single institute. **Methods:** We performed a retrospective chart review of adult DIPG from last 8 years (2010-2018) in a tertiary institute. DIPG was defined as expansile lesions involving more than 50% of the greatest diameter in the pons. **Results:** We found a total 46 patients with the diagnosis of adult BSG. Based on the definition, 23 patients with adult DIPG qualified to be included in the study. The median age was 32 years (IQR: 22-41), with a sex ratio of 16/7 (M/F). Cranial palsies were found in 17 (73%) patients. The median duration of symptoms was 6 months. On magnetic resonance imaging (MRI), contrast enhancement was found in seven (30%) patients. Biopsy was done in five patients. Median follow up was 11 months (IQR: 7-15). Median overall survival (OS) was 15 months (95%, CI 8.3-21.6). Fourteen patients had succumbed to death at the latest follow-up, and seven patients were alive. Median OS for the patients with age less than 40 years and more than 40 years was 7 and 22 months, respectively (p = 0.016). Rest of the variables did not effect OS significantly. **Conclusion:** Adult DIPG's significantly differs from pediatric counterparts in clinical characteristics, as well as OS. Age was the only factor which was significantly associated with survival in our study. Long-term studies with molecular profiling may help in further characterizing these lesions.

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Full Text

Brainstem gliomas (BSG) are rare and constitute less than 2% of all brain tumors in adults.[1],[2],[3]. They

have a bimodal age distribution with peak at 4–13 years in children and fourth decade in adults.[2],[4] BSGs cause clinical symptoms due to direct involvement of the cranial nerve nuclei, compression of the cerebellar peduncles, and distortion of the fourth ventricle. Diffuse iIntrinsic pontine gliomas (DIPG) is an entity that commonly occurs in children constituting 10–20% of primary pediatric tumors. It is a fatal type of pediatric brain tumor with 90% deaths within 2 years of diagnosis.[5] DIPG is often diagnosed based on clinical and radiological criteria with biopsy reserved for cases that are managed in centers equipped for molecular profiling and research.[6] Radiologically, they involve more than 50% of the greatest dimension/axial diameter of the pons.

Adult BSGs are very rare and behave differently than children in both clinical characteristics and prognosis.[2],[7] There is scarcity of literature on adult BSGs, though some studies have tried to address the issue and majority are retrospective studies.[1],[2],[4],[7],[8],[9],[10],[11],[12] The previously published retrospective studies include all the BSG from focal to diffuse, enhancing and nonenhancing, low grade to high grade, and from biopsies to nonbiopsied cases.[11] From the previous studies age, ethnicity, pathological grade, duration of symptoms, enhancement pattern in MRI, performance status, presence of K27M mutations, radiotherapy, and chemotherapy have shown to have association with survival.[10],[12] As the previous studies are heterogenous, we have analyzed the clinical characteristics and prognosis of adult patients treated in our institute who are more than 18 yrs of age with specific diagnosis of DIPG.

Materials and Methods

We did a retrospective chart review of our database for all the intrinsic BSG with age more than 18 years, from the year 2010–2018. Radiologically, we defined adult DIPG as expansile lesions involving more than 50% of the greatest diameter in the pons. All focal lesions like pilocytic astrocytomas were excluded. Lesions arising from adjacent areas like midbrain, medulla, and cerebellar peduncles extending into pons were excluded. Lesions with doubt in diagnosis underwent extensive workup to rule-out lymphoma, metastasis, inflammatory disease, and demyelination. Radiation planning: patients received RT dose of 54 Gy in 30 fractions prescribed and optimized by intensity modulated radiotherapy or volumetric modulated arc therapy to cover 95% of PTV by 95% of the dose. Critical structures optic apparatus, bilateral cochlea, temporal lobes, and normal brain are contoured for dosimetry.

The demographic details, clinical presentation, signs, and symptoms were collected from the case files of the patients. Radiological data were reviewed individually by two individual radiologists, with discussion and agreement of radiological diagnosis of DIPG. Overall survival (OS) was defined as the period from date of diagnosis, defined in most cases by date of first MRI or CT scan showing a brainstem mass, until death. Data whichever was not normal in distribution, median is analyzed for quantitative variables. Other clinical signs and symptoms were analyzed as descriptive. Kaplan-Meir survival (Log-Rank test) was used for assessing median survival overall and with perspective to age, sex, contrast enhancement, predominant location, KPS score, and presence of hydrocephalus.

Results

There were 46 patients with the diagnosis of adult BSG during the study period. Based on the definition, 23 patients with adult DIPG qualified to be included in the study. The median age at diagnosis was 32 years (IQR: 22-41), with majority being males (M: F: 16:7). Cranial nerve palsies were found in 17 (73%) patients. The median duration of symptom was 6 months. Long tract signs were found in 10 (43%) patients. Ataxia and cerebellar signs were found in 10 (43%) and raised ICP signs in 6 (26%) patients [Table 1]. The median KPS score was 70 (IQR: 60–70). Out of 23 patients, 34.7% were of KPS less than 70 (n = 8/23). On radiological assessment with magnetic resonance imaging (MRI), tumor was hypointense in 19/23 patients, rest being isointense, and all were T2 hyperintense [Figure 1]. The predominant location was pons in all the cases with 5 patients having tumor exclusively in pons, 10 had extension to medulla, 4 towards midbrain, and one having leptomeningeal dissemination. Ten patients had extension towards middle cerebellar peduncle [Figure 2].

Patchy, faint contrast enhancement was found in seven (30%) patients. Fourth ventricular distortion was found in 13 (56%) patients and hydrocephalus in 8 (34%) patients. Basilar artery encasement was found in 5 (21.7%) patients [Table 2].{Table 1}{Table 2}{Figure 1}{Figure 2}

Biopsy was done in five patients. Anaplastic astrocytoma was found in 3/5 patients, and one patient had diffuse astrocytoma. H3K27M mutation was not analyzed in these patients. One patient's biopsy was inconclusive. All patients underwent radiotherapy dose of 54 Gy in 30 fractions. Three patients underwent ventriculoperitoneal shunt for hydrocephalus (n = 3/23, 13%).

Two patients were lost to follow up. Median follow up was 11 months (Range: 4.1 months: 48.7 months, IQR: 7–15) [Figure 3]. Fourteen patients had succumbed to death at the latest follow-up, and seven patients were alive. Median OS was 15 months (95% CI 8.3–21.6) [Table 3]. Median OS for the patients with age less than 40 years was 22 months and for patients with age more than 40 years was 7 months (p = 0.016) [Figure 3]. Median OS for patients with KPS less than 70 was 11 months (SE: 3.3) and 48 months for patients with KPS more than 70 (p = 0.47). OS was 15 months and 11 months in patients with contrast enhancement and nonenhancing tumors, respectively (p = 0.82). OS with respect to sex (Males = 17 months and females = 15 months) and presence of hydrocephalus were not significant (p = 0.446 and P = 0.57, respectively). OS time based on the location was as follows [Figure 4]: Pons exclusively – 8 months (SE 4.59), Pontomedullary – 17 months (SE 3.58), pontomedullary with diffuse extension – 7 months (SE-1.66). The difference in survival times among different locations was not significant (p = 0.24). Three patients had received chemotherapy and radiotherapy simultaneously.{Table 3}{Figure 3}{Figure 4}

Discussion

BSGs in adults are very rare, and heterogeneous group of brain tumors vary with regard to underlying pathology, radiographic appearance, clinical course, and prognosis.[11] Although considered to be aggressive, the prognosis of this condition in adults has been reported to be better than the childhood counterparts. There are no standard guidelines for treatment and need for biopsy dictating that the treatment protocol has not been established.[8] The previously published studies are all retrospective in nature, and they have often included all types of adult brainstem lesion including focal and diffuse as well as enhancing and nonenhancing lesions.[2],[4],[8] Though including all BSGs, majority of tumors in the studies were predominantly pontine, and some even have shown that the location is of prognostic significance.[2],[8] The radiological appearance in these diffuse BSGs is similar to the DIPG occurring in children but has been reported to have a totally different prognosis.[8] So, it is important to identify patients with DIPG in adults and study these patients separately.

In our study, the median age of adult DIPGs was 32 years, with IQR of 22–41. Age range was from 18 to 60 years. The median age in other major series was around 30–40 years.[2],[13],[14],[15],[16] Few series have even reported BSGin age more than 75 years.[2],[13] Reithmeir et al. in their study of 101 histopathologically proven BSG reported that patients with age more than 40 had relative risk to die by 1.7 times than their younger counterparts.[4] Guillmao et al. in their multicenter series reported that patients with age >40 years had lesser survival chances.[14] In our present cohort, we have six patients with age more than 40 years. The median survival time for patients with age more than 40 years is 7 months compared to 22 months for patients age less than 40 years (p = 0.016) [Figure 3]. The finding is similar with published literature. Theeler et al. in their retrospective series reported 14.2 months median survival time for patients with age more than 60 years.[8] Majority of the published series showed that the prognosis being worse for patients with age >40 years,[4],[7] although most of the cases were not proven histologically, variable prognosis, may be due to incidence of more malignant lesions in older age groups. Babu et al. in their series of histologically proven malignant brainstem gliomas, with patients older than >60 years had median OS of 13.5 months.[1] These survival times not only depend on the age but also on the socio-economic status of the health care and various other factors.[1],[10],[17]

Majority of the patients presented in our series with multiple cranial nerve palsies. The median duration of the symptoms are 6 months in our series. In children, the symptoms have more rapid progression than adults

worsening the chances of survival. The most common cranial nerve palsy in our series is lower cranial nerves (8/23) with facial nerve being next common nerve involved in our series (5/23). Multiple cranial nerves were involved in 12/23 patients. Long tract signs and ataxia, with cranial nerve deficits, lower the KPS significantly. Low KPS (<70) has been associated with worse prognostic factor.[4] Eight patients in our series had KPS less than 70, with median OS of 11 months (p = 0.47). Although not statistically significant, results agree with above-mentioned studies. Eight patients had hydrocephalus in which three patients required cerebrospinal fluid diversion.

Contrast enhancement was present in seven patients in our series. Out of five patients who underwent biopsy of the lesion two with anaplastic grade, and one with diffuse astrocytoma, did not have contrast enhancement. Contrast enhancement of the tumor is considered as more malignant grade.[18] In our series, the median OS time difference (Contrast enhancement: 15 months, noncontrast – 11 months, P = 0.82) did not reach statistical significance. Szychot et al. in their study correlated the outcome of paediatric DIPG with tumor texture.[19] Tumors with more homogeneous texture are associated with worse prognosis. Ahmed et al. correlated the MRI findings with histologically proven brainstem gliomas, in which contrast enhancement can be found in even in low-grade brainstem gliomas.[20]

Location of the tumor predicts prognosis and influences the survival. As our series is exclusively DIPG, the most common location is pons with extension to surrounding structures. The most common extension of the lesion in our series is pontomedullary region. Supratentorial extension is seen in 4/23 patients and leptomeningeal metastasis in one patient. Babu et al. in their study reported worse outcome in patients with midbrain location.[1] Kesari et al. in their study reported significant impact of tumor site with tumor progression.[2] In their experience, multifocal gliomas followed by diffuse pontine lesions had higher tumor progression with decreased OS time (Multifocal gliomas – 75% progression, 33 months. Diffuse pontine – 74% progression, 59 months). Theeler et al. reported worse outcome correlated with location as well.[8] Median OS with pontine lesions is 25 months. In our series, we have median survival time of 15 months. Median OS for patients exclusively in pons was 7 months in our series and pontomedullary was 17 months (p = 0.361) [Figure 4], although a direct comparison is not possible because of ethnicity variations, socioeconomic health care patterns, variable location, and extension of the lesions. Overall majority of series correlate with pontine location to poor prognosis and less OS.[1],[4],[8]

Biopsy in pediatric DIPG is still debated. In a recent survey involving SIOPE-brain, tumor group opined to do a biopsy within a clinical trial,[21] whereas Williams et al. opined to biopsy in all suspected cases of DIPG in children given that availability of tissue leads to better understanding of the biology.[22] In adults with DIPG, biopsy or surgery is indicated with some atypical features like focal, exophytic nature, contrast enhancement, and suspected treatable diseases like infections, lymphoma etc. In this study, we had excluded all the cases which had atypical features and included only cases which fit the diagnosis of DIPG similar to how diagnosis is made in pediatric population. Though there is no single feature that can exclude other pathologies, we have previously described diagnosis and treatment of brainstem tuberculomas.[23] Lymphomas occurring solely as diffuse lesions in the brainstem is very rare and though imaging is not specific a dramatic response to steroids is always a case in lymphoma.[24] All cases were meticulously investigated to rule out other possible diagnosis in our series. Brainstem lesions with exophytic components are usually resectable providing the advantage of histological diagnosis, clinical improvement as well as decreased tumor volume load. In our series, basilar artery is encased in five cases and cerebellar peduncle is involved in 10 cases [Table 2]. We advise resection seldom in DIPG after negotiating the risks and benefits. However, in our series, we did biopsy only in five cases. Some studies have also reported using DTI that can provide information on displacement of the tracts that may determine respectability.[25]

In our series, five patients underwent biopsy, though they did not have any atypical features. The decision for biopsy was taken by treating surgeons. There are various series, reporting the variable outcomes based on the histology in adult brainstem gliomas.[26] In patients with exophytic component causing significant mass effect, resection can give biopsy with histological diagnosis as well as some symptomatic relief. Modes of acquiring pathological diagnosis can be done by open craniotomy and stereotactic surgery. In Salmaggi et al., in their series of 34 patients of brain stem gliomas, five patients underwent biopsy of the lesion.[13] Dellaretti et al. correlated histological diagnosis with MRI findings.[18] Out of 96 patients with brainstem lesions, most common tumor is low-grade glioma. In their series, focal lesion is a significant factor for diagnosing nontumor pathologies. Theeler et al. reported 146 patients of BSGwith biopsy of 100 cases;[8] most common diagnosis

was anaplastic astrocytomas followed by glioblastomas. The median OS was 32.1 months in their entire cohort. The median survival of the patients in the cohort of radiologically diagnosed was 40 months. In our study, we included exclusively DIPG cases and biopsy done in five cases. Among the five cases, three had a histological diagnosis of AA and one case diffuse astrocytoma.

Previous studies have shown that histological diagnosis correlated with the outcome of OS. Biopsy of the tissue also gives the opportunity for molecular profiling. Overall, the morbidity of the biopsy in BSGwas found to be 9%.[27] In our series, the OS was 15 months. There were no complications due to biopsy procedure in our series. OS did not differ between radiologically diagnosed and histologically diagnosed patients. All patients were advised radiotherapy. Chemotherapy is proved to be useful in adult DIPG as well as with patients of RT failure in low grade DIPG.[12],[28] Reyes-Botero et al. in their study reported the outcomes that 15 adult patients of radiologically or histologically diagnosed recurrent low-grade brainstem gliomas, who underwent chemotherapy after RT failure, with an estimated median PFS after temozolomide was 9.5 months and an OS of 14.4 months.[12] Three patients underwent chemotherapy with temozolomide along with radiotherapy. In our series, no patients underwent chemotherapy after RT; hence, a direct comparison could not be done. Radiation related adverse effects were found in one patient, who expired during the second course of the RT. In two patients, because of the leptomeningeal spread new onset paraparesis was present following the completion of radiotherapy. Tinkle et al. in their recent study reported improved neurological symptoms and OS with low dose cumulative RT.[29] In our study, we could not do progression free survival analysis as we didn't decompress the lesion. The present study also is not devoid of limitation due to retrospective nature. The major drawback of the study is small number of patients and lack of molecular data.

Conclusion

Adult DIPGs significantly differ from pediatric counterparts in clinical characteristics, as well as survival. Age was the only factor that affected the survival in our study. Robust data regarding ideal modality of treatment as well as prognosis is still lacking. Although biopsy and availability of tissue samples provide with the opportunity of molecular profiling and working towards newer treatment protocols, adult DIPG with typical features can be advised radiotherapy without histological diagnosis. Long-term studies with large sample sizes, concentration on imaging characteristics, and molecular profiling will help to improve the outcomes.

Ethical approval

All procedures performed in this study were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed Consent

For this type of study formal consent is not required.

Financial support and sponsorship

Nil.

Conflict of Interest

All authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest or nonfinancial interest in the subject matter or materials discussed in this manuscript.

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Figure 1: It shows the magnetic resonance imaging (MRI) in a 26-year-old male with diffuse intrinsic pontine glioma (DIPG). (a) T2-weighted axial image with heterogeneously hyperintense expansile lesion involving the ventral pons with partial encasement of basilar artery. (b) It shows the lesion to be heterogeneously hyper on FLAIR. (c) T1-weighted image with lesion being hypointense and (d) showing no enhancement in the lesion after administering Gadolinium contrast

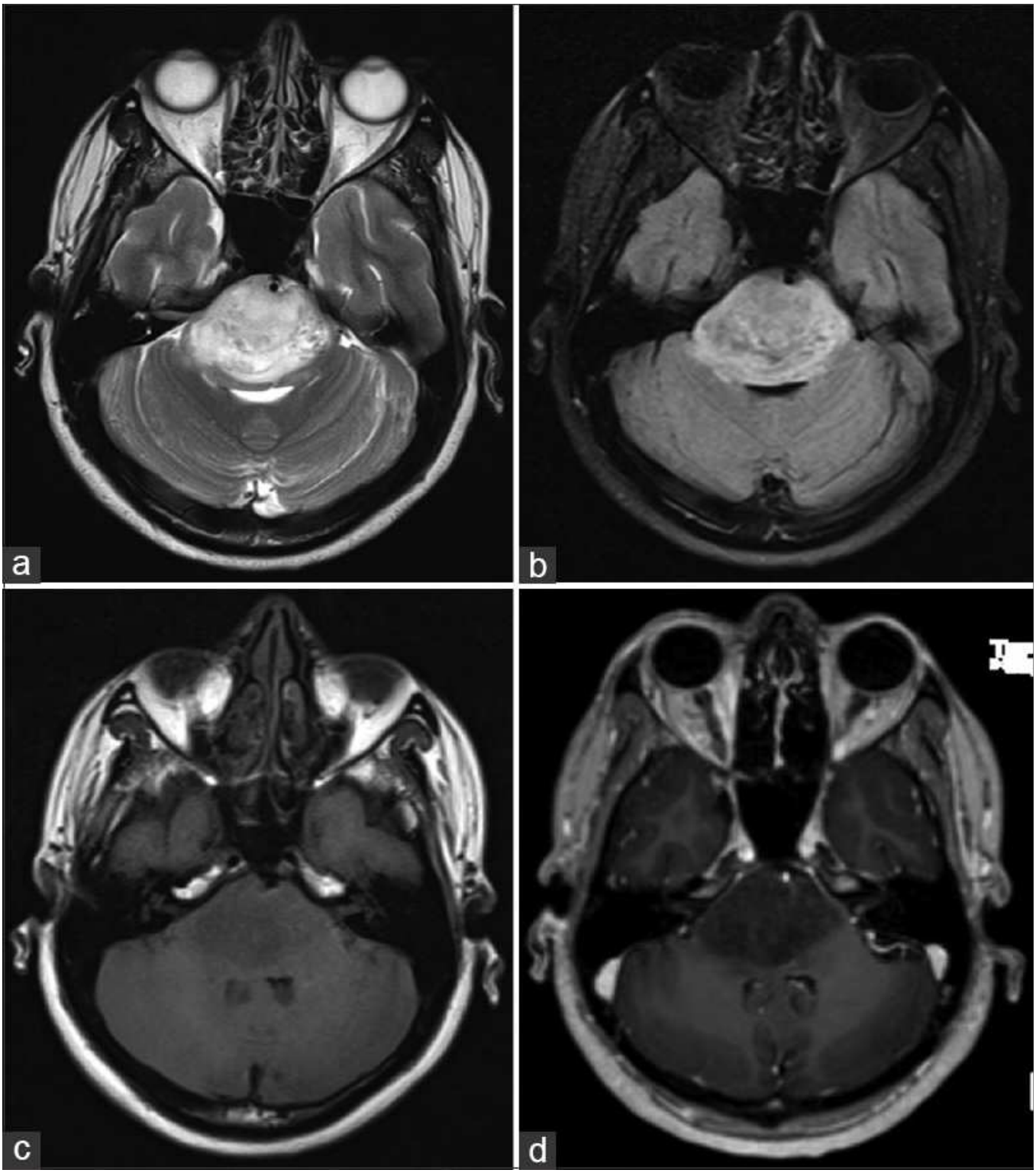

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Figure 2: It shows the magnetic resonance imaging (MRI) in a 24-year-old male with diffuse intrinsic pontine glioma (DIPG). (a) T2-weighted sagittal image showing large expansile hyperintense ventral pontine mass. (b) Ti-weighted post contrast image with encasement of basilar artery. (c) Axial FLAIR image and (d) Axial T2-weighted image showing large hyperintense lesion at the level of internal auditory meatus

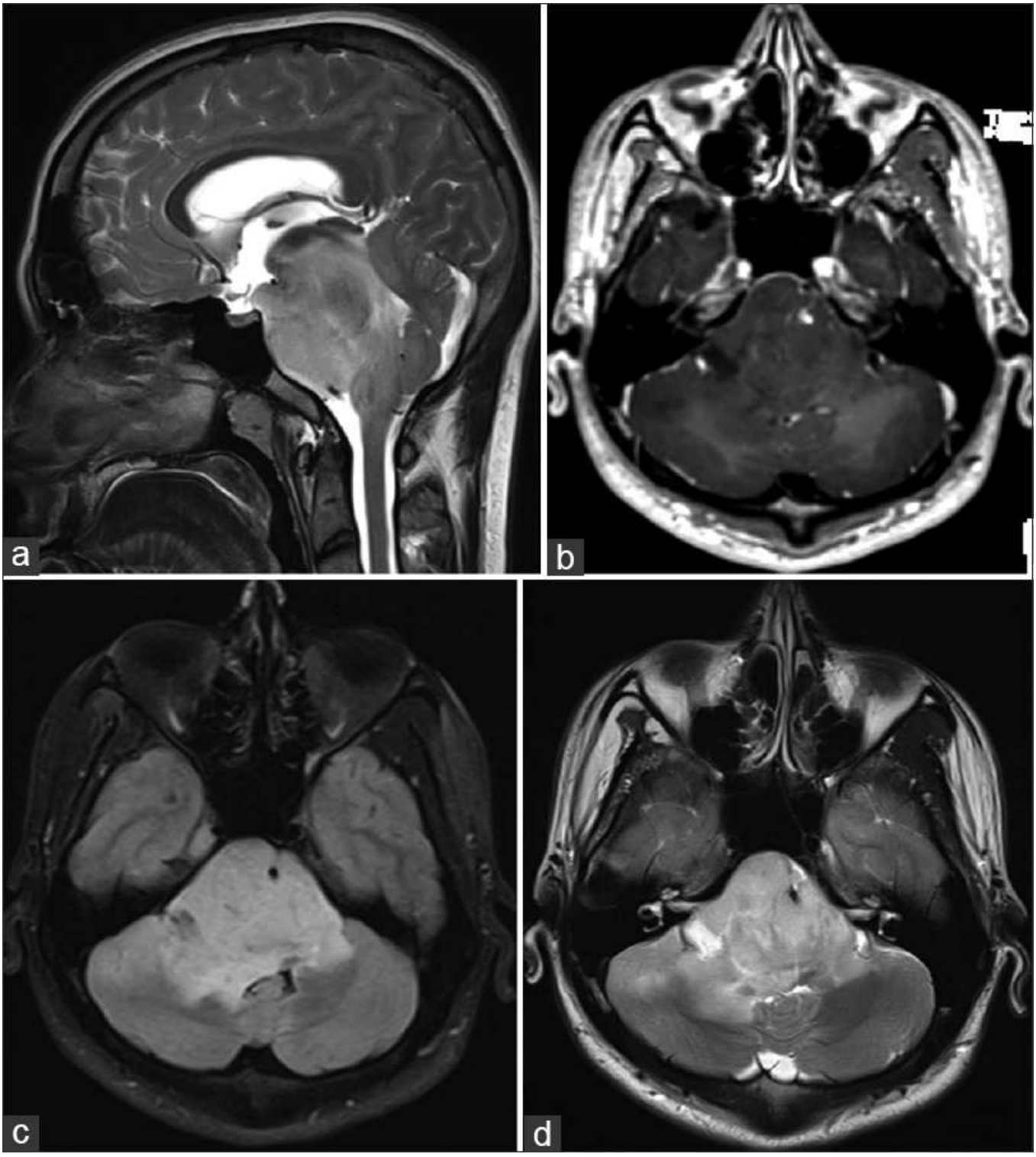

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Figure 3: Kaplan–Meir survival curve showing median overall survival and median overall survival stratified based on age

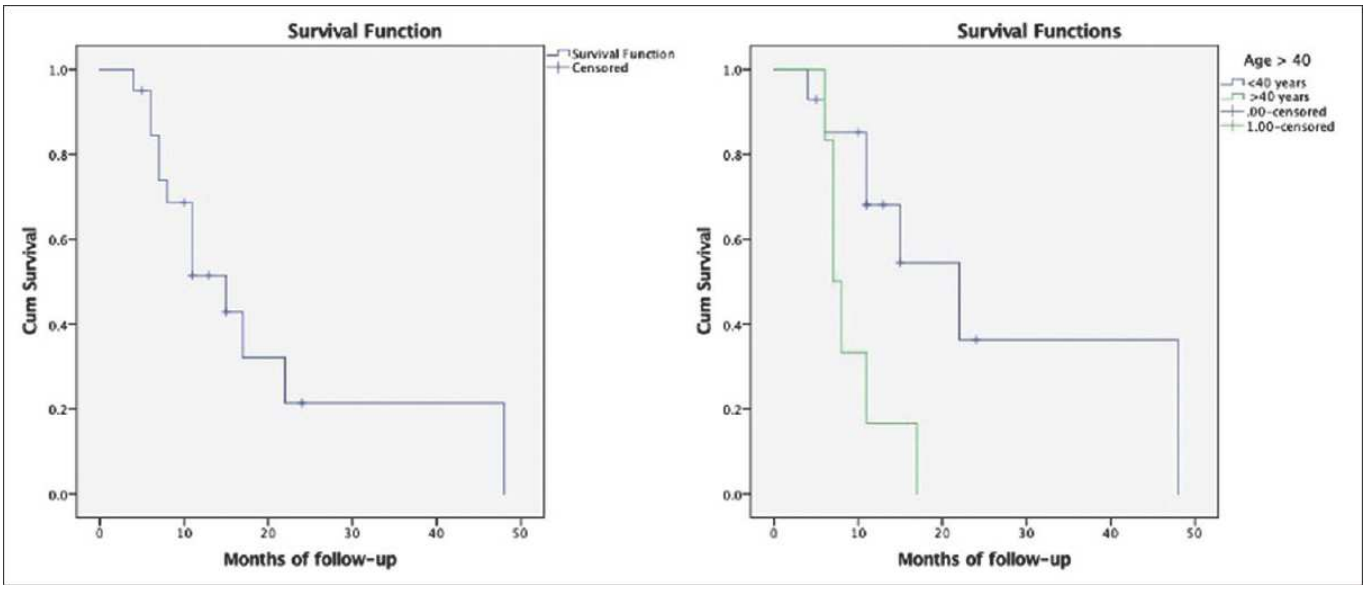

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Figure 4: Pictorial representation of survival time with respect to predominant location of diffuse intrinsic pontine glioma (DIPG)

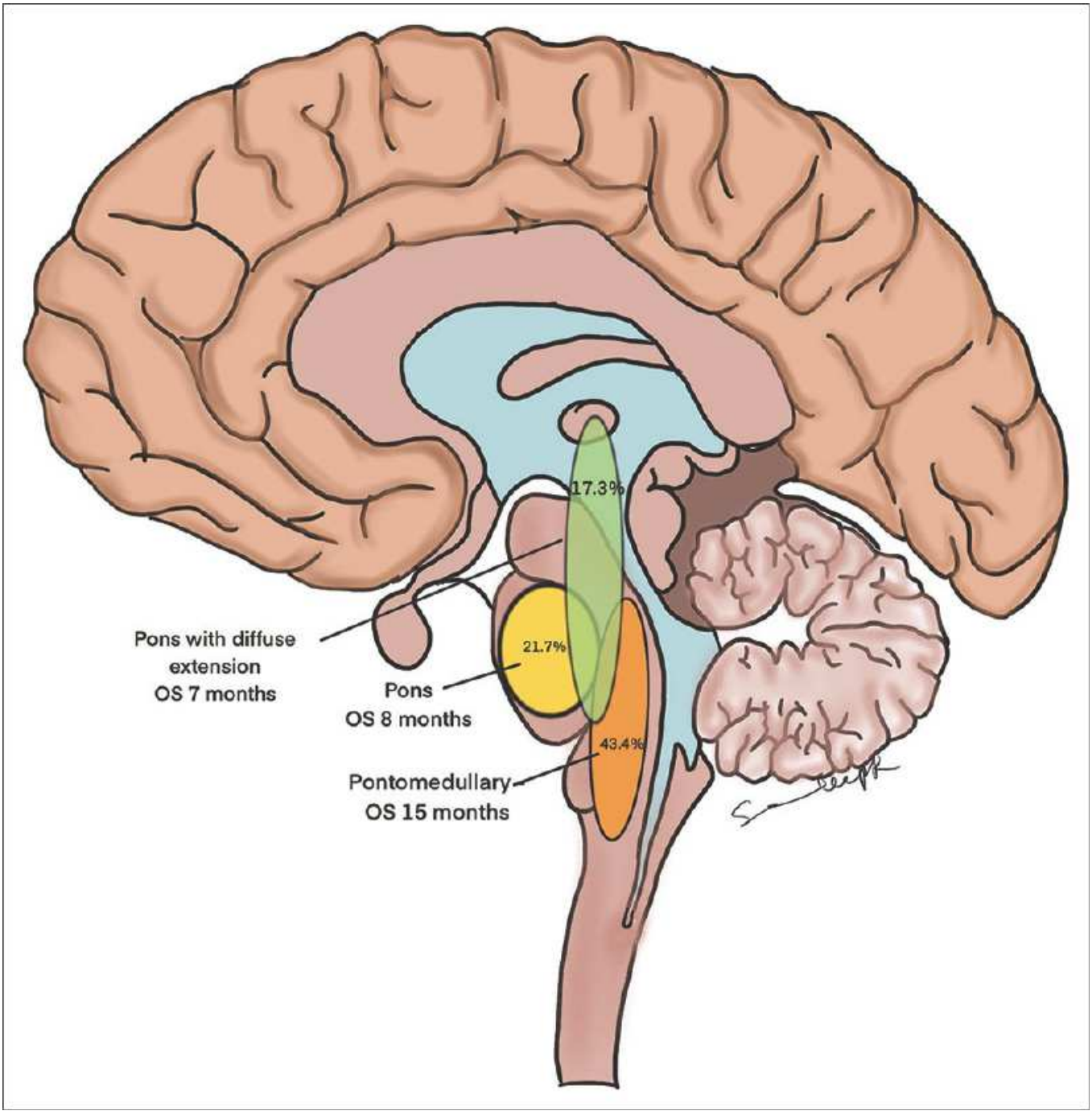

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Table 1: Demographic and clinical details of the adult patients with diffuse intrinsic pontine glioma, described in the study

Patient characteristics	Distribution
Age	Median age: 32 years IQR: 22-41 Mean age: 32.39 (SE: 2.68)
Sex ratio	16: Males, 7: females
Cranial palsies	17/23 patients
Duration of symptoms	Median: 6 months Mean: 13.13 (SE: 5.15)
Long tract signs	10/23 (43%)
Extraocular movement disorders	05/23 (21.7%)
Cerebellar signs	10/23 (43%)
Raised ICP signs	06/23 (26%)
KPS	Median: 70 (IQR: 60-70) KPS <70: *n*=8/23 (34.7%)

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Table 2: Radiological details of the adult patients with diffuse intrinsic pontine glioma, described in the study

Imaging Characteristics	Distribution
T1	Hypo: *n*=19/23 (82%)
	Iso: *n*=4/23 (17.3%)
T2 Hyper	*n*=23 (100%)
Contrast enhancement	*n*=7/23 (30%)
Fourth ventricle distortion	*n*=13/23 (56.5%)
Cerebellar peduncle involvement	*n*=10/23 (43.4%)
Hydrocephalus	*n*=08/23 (34.7%)
Basilar artery involvement	*n*=05/23 (21.7%)
Predominant location	Pontomedullary – 10/23 (43.4%)
	Pons – 5/23 (21.7%)
	Pons with extension into thalamus, cerebellar peduncle – 4/23 (17.3%)
	Extension into upper cervical spinal cord – 4/23 (17.3%)
	Leptomeningeal metastasis – 1/23

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Table 3: Pathology and follow-up details of the adult patients with diffuse intrinsic pontine glioma, described in the study

Pathology and Follow-up details	Distribution		
Follow-up	Median: 11 (IQR: 7-15)		
	Mean: 13.10 (SE: 2.18)		
RT	*n*=23/23		
Histopathological diagnosis	*n*=5/23 (21.7%)		
	Details: Anaplastic astrocytoma (*n*=3)		
	Diffuse astrocytoma (*n*=1)		
	Inconclusive biopsy (*n*=1)		
Median survival time (Kaplan Meir)	Mean: 19.640, SE: 4.503 (95% CI – 10.814-28.466)		
	Median: 15.00, SE :3.404 (95% CI – 8.329-21.671)		
	Age <40 years: 17 cases	22 months	*P*=0.016
	Age >40 years: 6 cases	7 months	
	Contrast enhancement	15 months	*P*=0.82
	Non-contrast enhancement	11 months	
	Sex		
	Male	17 months	*P*=0.446
	Female	15 months	
	Location		
	Pons	8 months	*P*=0.361
	Ponto medullary	15 months	
	Pontomedullary with diffuse extension	7 months	
	KPS		
	>70	48 months	*P*=0.478
	<70	11 months	
Alive	7/23 (30.4%)		
	Lost to follow up: 2		