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Original Article

Correlation of MRI Findings in Optic Pathway Glioma with the Presence of Neurofibromatosis

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ABSTRACT

Background: *Optic nerve glioma (also known as optic pathway glioma) is the most common primary neoplasm of the optic nerve. The tumor can arise anywhere along the optic pathway from behind the globe to the occipital cortex. Optic pathway gliomas (OPGs) are associated with high rate of visual morbidity and mortality. Studies have shown no specific clinical histologic or neuro imaging features to differentiate aggressive from non-aggressive OPGs.* ***Purpose:*** *The biological behaviour of optic pathway glioma is unpredictable and it is not clear if specific anatomical patterns may be of use in prognosis of the OPGs. The prognosis is reportedly better in OPGs associated with neurofibromatosis (NF). The purpose of the study was to compare the MRI findings between patients with NF, with those without NF and to determine prognostic imaging signs if any.* ***Material and Methods*** *- MRI studies of 41 patients with OPG (21 with NF and 20 without NF) were reviewed at presentation and at follow up. Statistical bivariate analysis was used to compare the size and extension of tumor between patients with and those without NF.* ***Results:*** *Orbital component of the optic nerve was most commonly involved in patients with NF (65%) with optic chiasma being the most common site of involvement (90%) in the non NF group. Extension beyond the optic pathway at diagnosis was more frequent in the non-NF group. In patients with NF, the morphological appearance of the optic nerve was preserved with the tumor being of limited size as compared to the patients in the non-NF group. Statistically significant correlation between MRI features and growth pattern of glioma was absent.* ***Conclusion:*** *As per our study we found that more than half the NF patient's tumor size remained stable in contrast to less than 5% of the non-NF group. Although no statistical correlation was found between MRI imaging and the biological behaviour of the tumor, we can infer that NF OPG is a separate entity from non-NF OPG with different prognostic features requiring a customised approach as per the type.*

Key words: *Glioma, Optic Pathway, Neurofibromatosis.*

INTRODUCTION

Optic pathway glioma (OPG) is a slow growing tumor typically affecting young children out of which 30% have NF[1] with better prognosis. OPG in children are almost always low grade asteocytoma (Barkovich JA). About 20 to 30% of patients experience visual impairment and even death.[2] Most of the neuro imaging studies of glioma are based on CT findings. In this study we have described the MRI studies of OPG, to find the distinguishing features between patients with and without NF. We also intended to find the prognostic signs on MRI , that will be of value in decision making for treatment. The treatment consist of surgery, chemotherapy, and radiations with no strict criteria for starting treatment.

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MATERIALS AND METHODS

Only patients for whom neuroimaging was available at presentation, before initiation of any kind of treatment or surgical intervention, were included in the study. Hospital records of HBT medical college, Mumbai were searched from May 2014 to June 2016, 41 eligible patients were identified of which 21 were NF and 20 without.

The images obtained before initiation of any kind of treatment were evaluated for interval changes in the size and extension of tumor. All patients underwent at least one T1- weighted and T2 weighted MR sequence. A detailed study of optic pathways, either axial n coronal was always included. Contrast agent (gadopentetate dimeglumine 0.1 to0.2 mmol/kg)was used.

Four states are defined. 1)stable no change in size or extent of involvement of visual pathways. 2)minimally enlarged, unequivocal enlargement in one plane only or an increase of less than 25% in two planes 3)enlarged an increase in size of over 25%in two planes or further extension and 4)reduced, shrinkage of more than 25%in two planes. The serial imaging studies were reviewed to determine the outcome of treatment (medical or surgical) and the imaging findings at the presentation were correlated with the long term findings. The histologic reports were also reviewed.

RESULTS

There was no significant difference between the patients with and without NF in regard to age or sex. Symptoms were present at diagnosis in all the children without NF (100%)but in only 32%of those with NF(p<.ooo1). Within the NF group the age of the symptomatic children was significantly younger (4.5+/- 2 years)than that of the asymptomatic children (6.0+/-3years) (p<.05). Involvement of the optic pathways by site at diagnosis is summarized in table, the most common site of involvement in the NF group was the orbital nerve, noted in 66%of the patients, as compared with only 32% in the non NF group(p<.005) Among the non NF children, the chiasma was involved in 90%,as compared with 62% in NF group (p<.005). Extension beyond the optic tract at diagnosis was uncommon in NF group. No correlation was found between the presence of orbital involvement and visual symptoms. In the NF group ,no correlation was found between the location of the tumor and the type of the symptoms. The presence of cystic components in the tumor was significantly more common in the non NF patients. In most of the NF patients (18/20,90%) the tumor did not affect the original shape of the optic pathway and appeared on images as a thickening of the optic nerve and chiasma. Both the maximal diameter and the volume of the tumor were significantly greater in the non NF group. Hydrocephalus was found in none of the children with NF but in 47% of those without NF. There was no correlation between the size of the mass and presence of hydrocephalus.

DISCUSSION

Most studies to date have failed to distinguish between OPG in patients with NF with those without NF and the few studies that have methodically compared these two groups relied mostly on CT findings. Therefore it remains unclear whether there are specific morphologic or anatomic patterns that can help predict the prognosis of OPG and whether NF–OPG is a distinct entity from non- NF-OPG.

The present study yielded several morphologic features that distinguish NF-OPG from non NF-OPG. In patients with NF the most common site of involvement was the optic nerve. the tumor was smaller than in the non-NF patients. The original shape of optic pathway was preserved, and cystic components were uncommon. In the non-NF group, the chiasma and hypothalamus were the most common site of involvement, the tumor was mass like, and cystic components were frequently seen, as was extension beyond the optic pathways. The prognosis was also significantly different: half the NF patients remained stable compared with only 5% of non NF patients. However no correlation was found between imaging features and prognosis.

Studies in the literature seem to indicate that OPG patients with and those without NF differ significantly by tumor location, with involvement of optic nerve being much more common in NF group. Hoffman et al found that virtually all their patients with orbital tumor had NF and Listernick et al reported that isolated involvement of the optic nerve occurred only in the patients with NF. The chiasma involved more often in non NF OPG. Involvement of the chiasma alone is also more likely in the absence of NF. Indeed, in the various series of OPG with NF involvement of the chiasma alone was rare. We found that patients with NF apparently have less diffuse involvement of the optic pathways although the difference from the non NF group was not significant . However isolated optic nerve or chiasmatic involvement was rare in both the NF (n-2)and non NF(n-1)groups. Therefore on the basis of MR findings we suggest that localised gliomas may be less common than previously thought. Several studies refer only to involvement of optic tracts with some reporting a higher prevalence in the patients with NF and others describing a higher occurrence in those without NF, we did not find any significant difference between the NF and non NF groups. Assessment of involvement of the optic tracts by tumor is difficult, because the increased signals intensity on T2 weighted MR images can be caused by edema as well as by direct tumor extension. A noteworthy findings in the present study was the different shape and size of the tumor in the two groups. In the patients with NF the involved chiasma was thickened although its contour was preserved, whereas in non NF group, the tumor was mass like.

The symptomatology of the two groups also differed significantly. All the non NF patients were referred for examination because of symptoms, whereas only 32% of the NF patients were symptomatic at presentation(p<.0001). The smaller dimensions and the relative sparing of the hypothalamus in the NF patients may account for this findings, the lag between the onset of the symptoms and imaging study was much longer in the non NF group, (up to 4 years), whereas NF patients examined immediately (p<.0005) the lower rate of occurrence of symptoms combined with the shorter interval between symptoms, onset and diagnosis in the NF group may reflect a bias in referral patterns, with a lower threshold of suspicion in NF patients.

Table 1: Involvement of optic nerve by site

Group	Orbital nerve	Chiasma	Hypothalamus	Tracts	Beyond optic pathways
NF (n= 21)	13(65)	12(62)	5(25)	7(36)	0
Non-NF (n=20)	7(32)	18(90)	14(70)	9(47)	13(69)
P value	<0.005	<0.005	<0.001	N.S.	<0.001

Table 2: Size and extension of OPGs

Group	Before treatment			Reduction	After treatment	
	Stable	Enlarged	Minimal Enlargement		Postoperative Enlargement	Post chemotherapy Enlargement
NF (n= 16)	8	2	2	2	0	0
Non-NF (n=15)	1	5	0	0	7	4
P value	<0.001	<0.001	<0.001		<0.001	<0.001

Table 3: Symptoms of presentation of children with OPG

Symptomatic	NF 6	Non-NF 20	P value <0.0001
Visual symptoms	3	14	<0.0001
Non-Visual symptoms	4	11	<0.0001
-precocious puberty	1	2	
-Failure to thrive	2	1	
-Devolopmental delay	1	-	
-Diabetes insipidus	-	1	
-Hemiparesis	-	2	
-Increased ICT	-	4	
-Torticollis	-	1	

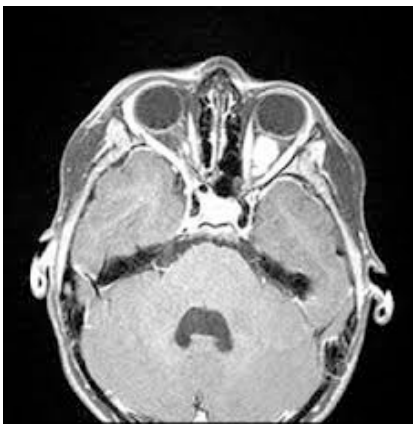

Fig 1: This axial, post-contrast image demonstrates enhancement of the fusiform, kinked shaped optic nerve tumor

A significant correlation was found between hypothalamic involvement and the occurrence of endocrinologic symptoms (such as failure to thrive and precocious puberty)and signs of increased intracranial pressure, interestingly, orbital involvement did not correspond to visual symptoms, possibly because infants and even young children tend not to complain of visual disturbances.

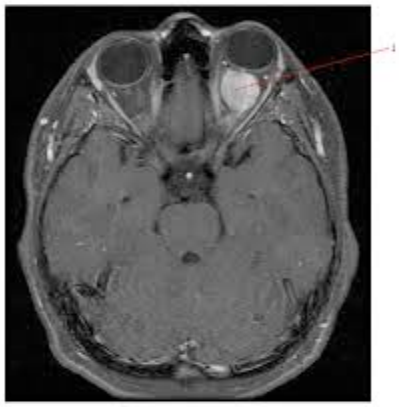

Fig 2: The axial MR below shows a unilateral optic nerve glioma

One of the most striking features differentiating NF- OPG from non NF-OPG is the long term behaviour of the tumor. In our study,15% of the NF group showed minimal tumor enlargement, and 51%remained stable, in the non NF group, there was a clear propensity for growth in 95%of patients (p<.001).Although earlier studies (Dutton, McCullough and Ellsworth),as well as in 1994 review by Sutton reported that the presence of NF has no influence on prognosis of OPG ,now a days it is accepted that NF acts as a protective factor. OPGs have been found to enlarge in 9%, 12 %,13%,and 22% of patients with NF(Listernick, Janss, and Charrow respectively), and in 54%if isolated intraorbital lesions are excluded (Packer), as compared with 63%,68%and 100% of patients without NF it is of interest that spontaneous regression of the tumor was observed in 3 of our patients with NF. This has been described previously in NF grade histologic findings in 3 of our patients, there was leptomeningial spred .

The location of tumor has also been reported to be related to long term out come in our study, as single site involvement was rare, no such correlation to be found.

CONCLUSION

We found no correlation between the imaging parameters analysed and the biological behaviour of OPG associated with NF. This observation may indicate that neuroimaging is not contributory to the prediction of outcome in this group of patients. OPG in patients without NF differs significantly from NF-OPG in both imaging features and prognosis. Non NF OPG and NF OPG are apparently distinct entities, each warranting a specific diagnostic, clinical, and therapeutic approach.

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