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Primary optic nerve tumors: An experience at tertiary care center

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Abstract
Primary optic nerve neoplasms are less common than secondary involvement of optic nerve by direct invasion from extension of intraocular malignancies. Here we present11cases originated primarily in optic nerve and its contents. We had 1.42% cases of primary optic nerve tumors among all operated cases of CNS tumors. Primary optic nerve tumors include pilocytic astrocytoma (5cases) & a case of pilomyxoid astrocytoma occurring in children. Among the primary optic nerve sheath tumors, meningioma (2cases) was most common along with one case each of schwannoma and AV hemangioma. Meningioma was seen in middle to elderly females while schwannoma in young adult. Primary optic nerve tumor presents with overlapping clinical and radiological manifestation. Hence intraoperative squash smear cytology can be used effectively to give preliminary diagnosis, as it is fairly accurate & established technique for diagnosis. Visual outcome of patients with primary optic nerve tumors depends on the type of the lesion which can be done by confirmative histopathology only.

Keywords: primary optic nerve tumors, squash cytology, pilocytic astrocytoma, meningioma

1. Introduction
The optic nerve is a tract of the central nervous system extending from optic disc to optic chiasm. It is composed of retinal ganglion cells, axon and glial cells. It is completely invested by dura, arachnoid and pia mater with an artery and a vein in the center [1]. These histological structures can be a potential source of various primary neoplasm arising in the optic nerve, although secondary involvement of the optic nerve is more common than primary optic nerve tumors. The secondary involvement of optic nerve may arise by direct invasion from intraocular malignancy like retinoblastoma (29.5%), melanoma, from meningeal carcinomatosis or from distant primary tumors [2]. Primary optic nerve tumors are much rare with optic nerve gliomas account for only 1-5% of intracranial gliomas [3].
Here we present a retrospective case series of eleven primary optic nerve tumors occurring in the span of 10 years.

2. Material and methods
This retrospective observational study was carried out at tertiary care hospital with patient's referral from the period of Jan 07 to Dec 16. During this period, total 752 cases of CNS tumors were operated. Out of this, 11cases (1.42%) were arising primarily from optic nerve and its contents. After taking complete clinical & radiological details, received samples were processed by standard formalin fixation & paraffin embedding method. All the sections were stained with routine H&E stain and reported according to WHO 2007 classification of CNS tumors. Out of eleven cases; four cases were sent for intraoperative cytological diagnosis by squash smear technique.

3. Result
The youngest patient in the study was 3 yrs old boy whereas oldest was 70 yrs. female.

Table 1: Age, sex, affected side wise distribution along with histological diagnosis & WHO grade (2007 classification of CNS tumors) offered in eleven cases of primary optic nerve tumors.

S. No.	Age/sex	Side	H/P diagnosis	WHO grade
1	10yrs/M	right	Pilocytic astrocytoma	Grade I
2	70yrs/F	right	Meningioma	Grade I
3	3yrs/M	left	Pilocytic astrocytoma	Grade I
4	15yrs/M	left	Pilocytic astrocytoma	Grade I
5	4yrs/F	right	Pilocytic astrocytoma	Grade I
6	32yrs/F	left	AV hemangioma	-
7	28yrs/M	right	Schwannoma	Grade I
8	50yrs/F	left	Meningioma	Grade I
9	7yrs/F	right	Pilocytic astrocytoma	Grade I
10	4yrs/F	right	Pilomyxoid astrocytoma	Grade II
11	35yrs/F	left	Meningioma	Grade I

This study showed slight female predominance with M : F ratio of 0.83. Primary optic nerve tumors occurred more commonly on right side (6 cases) than left side (5 cases). The most common primary optic nerve tumor was pilocytic astrocytoma (5 cases) & a case of pilomyxoid astrocytoma. Meningioma (2 cases) was most common among primary optic nerve sheath tumor while one case each of schwannoma & AV hemangioma.
Intra operative squash smear cytology was available in 4 cases with one discordant case on final histopathological diagnosis.

Table 2: showing intraoperative squash diagnosis & its histopathological correlation

No	Intraoperative squash cytological diagnosis	H/P diagnosis	Positive correlation
1	Pilocytic astrocytoma	Pilocytic astrocytoma	+
2	Pilocytic astrocytoma	Pilocytic astrocytoma	+
3	Meningioma	Meningioma	+
4	Tough to smear, inconclusive	AV hemangioma	-

4. Discussion

Primary optic nerve tumors are not very common with optic nerve gliomas account for only 1% -5% of all intracranial tumors [3, 4]. We had 1.42% cases of CNS tumors originating primarily in optic nerve.

The diagnosis of optic nerve tumors can be done by either computed tomography (CT) scan and or magnetic resonance (MR) imaging. With imaging primary optic nerve gliomas present as fusiform intraorbital, intraconal, intradural homogenously solid well defined mass with proptosis of corresponding globe. (Fig. 1) whereas primary optic nerve sheath tumors, most common among is meningioma on imaging typically have a `tram -track' configuration whereby contrast enhancing tumor is seen along the side of optic nerve [5]. (Fig. 2) We had similar imaging findings for primary optic nerve tumors.

Fig 1: MRI showing intraorbital, intraconal fusiform solid mass in optic nerve with proptosis of right globe in glioma

Fig 2: CT scan image showing 'tram-track 'appearance of optic nerve in meningioma

Due to symptomatic and imaging mimics among the narrow range of optic nerve tumors, a confirmatory diagnosis can be done by biopsy & thereby histopathology. Among the primary optic nerve glial tumors, we had 5 cases of pilocytic astrocytoma (WHO grade I) and a case of pilomyxoid astrocytoma (WHO grade II), both are in the categories of low grade gliomas. All 5 cases presented with slowly progressive unilateral proptosis, occurring in the age ranging from 3 yrs. to 15 yrs. Our findings were mirroring with [6, 7].

Pilocytic astrocytoma (PA) typically showed elongated spindle shaped piloid astrocytes arranged in biphasic pattern. Degenerative changes in the form of Rosenthal fibers and eosinophilic granular bodies were seen almost every case matching with the histomorphological features of PA elsewhere in CNS [8]. According to Dutton *et al* pilocytic astrocytoma comprise of 1.5% to 3.5% of all CNS tumors and 66% of all optic nerve tumors [4]. We had 1.42% cases of pilocytic astrocytoma among all CNS tumors; contributing to 45.45% of all optic nerve tumors matching the findings of Dutton *et al*. PA is considered as a low grade gliomas which mostly affect children and adolescent, but is known to reoccur after decades of removal of primary optic nerve tumor [9]. So it can cause diagnostic dilemma with common optic nerve tumors occurring later in the life.

A single case of pilomyxoid astrocytoma (PMA) showed monophasic angiocentric arrangement of monomorphic, bipolar tumor cells against myxoid background. Rosenthal fibers, granular bodies & biphasic pattern were typically absent in PMA differentiating it from PA [10, 11]. (Fig.3)

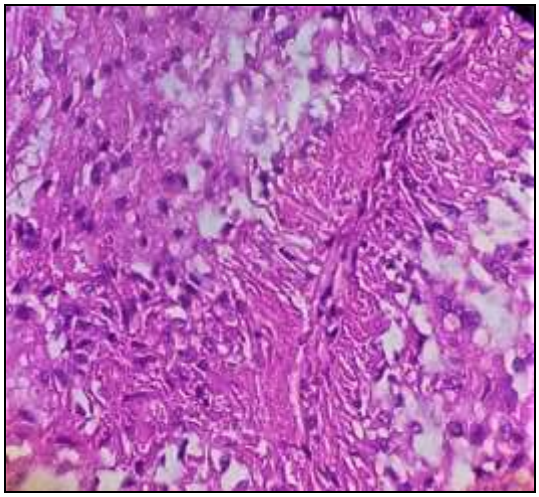

Fig 3: Microphotograph showing angiocentric arrangement of monophasic bipolar cells against myxoid background in Pilomyxoid astrocytoma (100X).

Among the range of primary optic nerve sheath tumors, we had meningioma (WHO grade I) the most common (3cases) with one case each of schwannoma (WHO grade I) & AV hemangioma. Primary optic nerve meningioma presented as unilateral proptosis in middle aged females (35 yrs. to 70yrs) with peak at 5th decade. Tissue diagnosis mostly showed syntitial & transitional meningioma [12]. According to Dutton *et al* optic nerve sheath meningioma is the second most common primary optic nerve tumor and represents for one third of all optic nerve tumors [13]. We had 3 cases of

meningioma among 11cases of primary optic nerve tumors contributing to 27.27% of all mirroring findings of Dutton *et al* about optic nerve sheath meningioma. Though rare, schwannoma are known to occur as optic nerve sheath tumor [14]. We had one case of schwannoma occurring in 28 yrs. male presented as eccentric growth in optic nerve on imaging. On microscopy it showed hyper cellular (Antoni A) & hypo cellular (Antoni B) areas with verrocay bodies. (Fig.4)

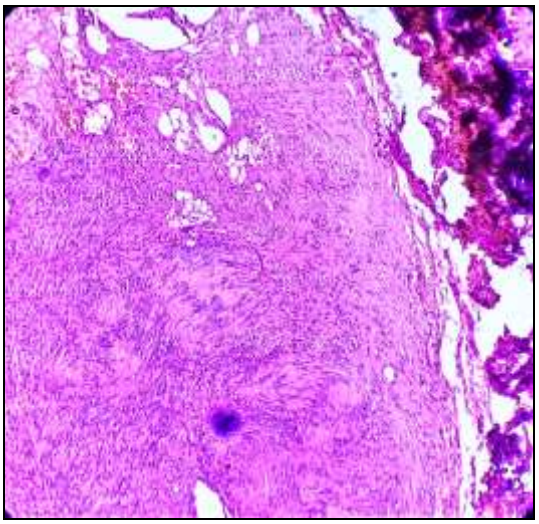

Fig 4: Microphotograph showing verrocay bodies in schwannoma (40X).

Out of four cases, 3 cases of intraoperative squash smear cytology were correlated well with final histopathological diagnosis. Tissue of pilocytic astrocytoma smeared easily, showed a tumor comprising of rod shaped nuclei with bipolar long piloid cytoplasmic process. Rosenthal fibers, granular bodies and thick walled vessels are seen in absence of mitotic activity. (Fig5) A case of meningioma was soft to smear, showed polygonal to spindle cells with oval nuclei showing intra nuclear pseudo inclusion correlated with histopathological diagnosis. (Fig. 6) A single case of AV hemangioma was tough to spread and was inconclusive on squash cytology [15].

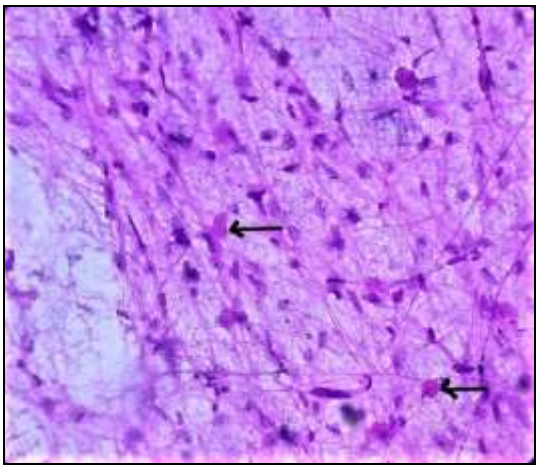

Fig 5: Microphotograph bipolar cells with granular bodies in Pilocytic Astrocytoma (100X)

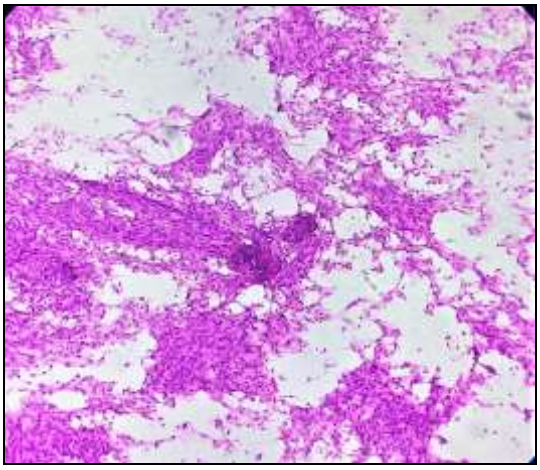

Fig 6: Microphotograph of squash smear of meningioma (100X)

Biopsies from the optic nerve may present with diagnostic challenge owing to small size of tissue sample. Another reason could be optic nerve gliomas are infiltrating through the pia causing thickening of the arachnoid cells creating diagnostic difficulties between gliomas &meningioma [16]. In such situations tumor cell arrangement, Rosenthal fibers, granular bodies, psammoma bodies & verrocay bodies are of diagnostic importance.

Though primary optic nerve tumors may have varied biological nature, final visual outcome depends on the nature of the lesion [17]. Both gliomas and meningioma have poor prognosis for vision. With recurrence, involvement of the chiasm and especially of the hypothalamus or third ventricle, prognosis for life falls significantly for gliomas. However unlike gliomas, prognosis in meningioma for life is excellent [18].

6. Conclusion

Primary optic nerve tumors are less common than secondary involvement of optic nerve by direct invasion from intraocular malignancy. Most of the primary optic nerve tumors present with overlapping clinical and radiological manifestation. Hence intraoperative squash smear cytology can be used effectively to give preliminary diagnosis, as it is fairly accurate & established technique for diagnosis. Visual outcome of patients with primary optic nerve tumors depends on the type of the lesion which can be done by confirmative histopathology only.

7. Acknowledgment: none

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