Bilateral Oculomotor Nerve Schwannomas Presenting With Ptosis And Oculomotor Nerve Palasy- A Rare Presentation Of Neurofibromatosis-2

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INTRODUCTION
Neurofibromatosis-2 previously also known as central neurofibromatosis is characterized by bilateral acoustic schwannomas. Other cranial nerve schwannomas such as 5th, 7th, 9th, 10th are also known to occur but they generally occur in combination with 8th nerve schwannomas (1-3). Oculomotor nerve schwannomas are a rare clinical occurrence. Bilateral oculomotor nerve schwannomas are extremely rare occurrence with no case reported in literature. We report a rare case of bilateral oculomotor nerve schwannomas presenting with ptosis and third nerve palsy and discuss management options in such patients.

CASE REPORT
A 40 year old male presented in our out patient department with gradually progressive ptosis of right eye of one month duration. There was no history of trauma, headache or history of diabetes mellitus or hypertension. On careful examination patient was found to have right sided oculomotor nerve palsy. Examination of the rest of the cranial nerves revealed decreased sensations on left half of the face, weakness of muscles of mastication on left side. Examination of rest of the cranial nerves was normal. Patient also had spasticity in all four limbs with exaggerated deep tendon reflexes. Planters were extensor bilaterally.

A provisional diagnosis of right sided intracranial mass lesion was made and a contrast MRI brain was ordered, which revealed bilateral 3rd nerve tumours with a massive left sided 5th nerve tumour. MRI also revealed empty sella and enlarged choroid plexus in fourth ventricle. A diagnosis of multiple cranial nerve schwannomas with NF-2 was made. Two days following admission patient complained of radicular pain in dorsal region with increased weakness in lower limbs. MRI spine revealed an intra dural extramedullary mass in dorsal region causing cord compression at D-4 level.

Since patients’ paraplegia was progressive he underwent D4-5 laminectomy with microsurgical removal of IDEM mass. Histopathology was suggestive of neurofibroma. This was followed by microsurgical removal of left 5th nerve mass in two stages Intracranial mass was removed by extended middle cranial fossa approach and extra cranial portion was removed via osteoplastic maxillotomy approach. Post operatively the patient had mild deterioration of 5th nerve function. His spasticity and weakness improved remarkably Since bilateral oculomotor nerve tumors were small and their removal is generally indicated for large tumours causing mass effect, patient was explained the prognosis and it was decided to follow these tumours clinically and radiologically.
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Figure 1
Table 1: Showing diagnostic criteria of NF-2

<table>
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<tr>
<th>Criteria</th>
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<td>1. Bilateral eighth nerve mass seen with appropriate imaging techniques (e.g., CT or MR imaging)</td>
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<td>2. NF-2 in a first-degree relative, and either a unilateral eighth nerve mass or two of the following neurofibroma, meningioma, glioma, schwannoma, or juvenile posterior subcapsular lenticular opacity</td>
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Figure 2
Fig 1. Contrast Enhanced Axial image of the patient showing small bilateral 3rd nerve tumors.

Figure 3
Fig 2. Coronal contrast enhanced T1 weighted images showing bilateral 3rd and massive 5th nerve tumors.

Figure 4
Fig 3. Contrast enhanced saggital images showing tumor along the course of third nerve.
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**Figure 5**
Fig 4. Sagittal T2 weighted image showing empty sella and enlarged choroid plexus in fourth ventricle.

**Figure 6**
Fig 5. Contrast enhanced axial images showing 5th nerve tumor in middle and posterior fossa.

**Figure 7**
Fig 6. Showing right sided ptosis.
**DISCUSSION**

Neurofibromatosis 2 (NF2) is a rare autosomal dominant disorder with bilateral vestibular schwannomas as a defining feature. Patients with NF-2 also develop other cranial, spinal, peripheral schwannomas, cranial and spinal menigiomas, and cataracts. Other associated intracranial lesions are non neoplastic calcifications with involvement of the choroid plexus being most common. Cerebellar and cerebral cortical calcifications may also be seen(4). The diagnostic criteria for NF-2 has been reviewed in table 1. Though schwannomas other than 8th nerve schwannomas are known to occur but isolated third nerve schwannomas are a rare presentation of NF-2. Though our patient didn’t have 8th nerve schwannomas but he had bilateral 3rd nerve ,left fifth nerve schwannoma, a neurofibroma at D-4 level in spine and a brother who was operated for vestibular schwannoma and had multiple intracranial menigiomas. Hence our patient fulfilled criteria 2 given in table for NF-2. Apart from these our patient had empty sella and enlarged choroid plexus of fourth ventricle.

In Rubinstein’s review of an autopsy series of patients with NF-2 (5), there were seven patients with bilateral acoustic schwannomas, six with tumors of other cranial nerves and multiple menigiomas, and five with spinal ependymomas. One patient in Lichtenstein’s review (6) had bilateral acoustic schwannomas; multiple neuromas, ependymomas, and menigiomas in the spine; and an astrocytoma in the cerebellum. Kramer (7) described a patient with bilateral tumors of cranial nerves V, VII, VIII, and XI; several intracranial menigiomas; and numerous spinal neuromas. However in none of these series described patients with bilateral oculomotor nerve schwannomas with or without vestibular schwannomas.

About 38 cases of isolated third nerve schwannomas have been reported in literature. It is noteworthy that preoperative ocularmotor dysfunction was manifested in 29 cases out of 38. The tumor was located in the orbit in four cases (solitary orbital type), in the subarachnoid space in 17 cases (cisternal type), in the cavernous sinus in 12 cases (cavernous type), extending from the cavernous sinus to the cistern in five cases (cisternocavernous type). Surgical intervention was conducted in 34 cases and most of them underwent total or subtotal resection of the tumor(8). However, ocularmotor function was not improved or recovered postoperatively in the majority of those cases. Hence surgery is generally indicated only for large tumors with intractable symptoms and, if surgery is conducted in the case of small or less symptomatic tumors involving the cavernous sinus, the surgical strategy should be changed based on the operative findings to prevent further neurological damage. It would be possible to observe the natural course of the tumor in the cavernous sinus using MR imaging until the tumor becomes large or stereotactic radiosurgery may be an alternative to radical resection(8).

Conceivably, two clues leading to the diagnosis are an oculomotor paresis and tumor location along the course of the oculomotor nerve, especially in its cisternal portion(8).

In the present case, both the clues were present. The patient had ptosis on right side and a well defined enhancing mass along the course of oculomotor nerves in their cisternal portion on both the sides. Also in our case patient had a large 5th nerve schwannoma on left side as well as spinal neurofibroma but there was no evidence of 8th nerve tumours. Thus our case points out the fact that third nerve schwannomas with gradual onset third nerve palsy though rare, can be a presenting feature of neurofibromatosis -2 in...
CONCLUSIONS

Neurofibromatosis-2 is an autosomal dominant disorder characterized mainly by bilateral acoustic schwannomas. A high frequency of cranial nerve tumors other than acoustic neuromas was observed in few of the series of NF-2 but they have been mainly confined to cranial nerves 5th 7th 9th 10th and occasionally 12th. However most of these cases have been described in patients with acoustic schwannomas. Oculomotor nerve schwannomas both in isolation and in patients of NF-2 are rare. Only a handful of cases have been described in literature. However as illustrated in our case bilateral oculomotor nerve schwannomas presenting with gradually progressive ptosis and third nerve palsy can be a rare presentation of NF-2.

Treatment of these rare tumors is governed by their size. Small asymptomatic tumors are better followed up clinically and radiologically because surgery for these tumors is generally followed by deterioration of 3rd nerve function and is generally reserved for tumors causing mass effect. It would be possible to observe the natural course of the tumor clinically and radiologically until the tumor becomes large in size. Stereotactic radiosurgery may be considered an alternative to radical resection.

References
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