Paraneoplastic Papilloedema in a Child with Neuroblastoma

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Abstract
Non-metastatic neurological disease complicating neuroblastoma is well recognized. Gross papilloedema in the absence of intracranial disease as initial manifestation of neuroblastoma is reported in adults. We report for the first time a case of bilateral papilloedema in a child with neuroblastoma in the absence of intracranial disease and hypertension.

Key words: Neuroblastoma, Papilloedema

Introduction
Neuroblastoma in children usually presents with symptoms and signs which reflect the location of primary and metastatic disease. It can also present with non-metastatic disease such as opsoclonus or cerebellar ataxia.\(^1\) Initial presentation with gross papilloedema in the absence of intracranial disease is reported in adults.\(^2\) We report a child with neuroblastoma who presented with bilateral gross papilloedema in the absence of intracranial metastasis and hypertension. This complication has not been previously described in children, in the literature.

Case Report
A 6-year-old boy presented with 4 months history of fever associated with headache, nausea, vomiting, loss of weight and appetite. He had blurring of vision. He had no history of contact with tuberculosis. On examination, he was irritable, lethargic and cachectic. His weight was 14 kg and height was 110 cm. His vitals were stable with blood pressure of 100/70 mm of Hg. He had pallor. His fundus examination revealed constriction of visual field, decreased color perception and marked bilateral papilloedema. He had muscle wasting, increased tone, and brisk reflexes in lower limbs.

Laboratory investigations showed hemoglobin of 6.5 gm/dl, total leukocyte count of 8,300 cu. mm with differential of N 61%, L29%, M8%, E2% and platelet count of 5,82,000 cu.mm, and ESR was 137 mm in one hour. His serum LDH was 1100 U/L, uric acid was 2.9 mg/dl and CPK was 40 U/L. His rheumatoid factor and ANA were negative and total serum complement was 80 %. Tuberculosis workup including Mantoux test and gastric juice for AFB was negative. HIV serology was negative. Bone marrow biopsy showed fibrotic marrow which was extensively infiltrated by tumor composed of nests of medium-sized cells round to oval nuclei with stippled chromatin and moderate amounts of granular eosinophilic cytoplasm set in a fibrillary background. There were no rosettes. Normal haemopoietic elements were not present.

His 24-hour urinary VMA was 56 mg in 650 ml [normal 1.0-8.0 mg/24h]. Ultra sonogram of abdomen revealed a mass in the left adrenal gland. His CT brain revealed normal eyes, orbits and optic nerves. There was no evidence of ventricular dilatation, mid-line shift or a space-occupying lesion in the brain.

He was diagnosed to have neuroblastoma in view of bone marrow showing malignant round cell tumor with elevated VMA in urine and mass in left adrenal gland. The child was started on IV Mannitol following which his headache was better. The parents of the child were counseled about his disease and were advised chemotherapy. But they opted to take treatment in their...
native place, so he was discharged at request.

**Discussion**

Neuroblastoma accounts for 8 to 10% of all childhood cancers.\(^3\) It is one of the small round cell tumors of childhood. They arise from any site along the sympathetic nervous system chain. The presenting signs and symptoms of neuroblastoma usually reflect the location of primary, regional and metastatic disease. Unusually, children with neuroblastoma can present with Horner’s syndrome, limping and irritability associated with bone and bone marrow metastasis, opsomyoclonus with or without cerebellar ataxia, intractable secretory diarrhea and raccoon eyes.\(^4\)

Since the child did not have any fever or meningeal signs lumbar puncture was not done. We feel that this papilloedema as mentioned in the literature is a paraneoplastic papilloedema which is associated with neuroblastoma.

The exact mechanism of papilloedema due to neuroblastoma is uncertain. A defect in axoplasmic transport \(^5\) due to the effect of a toxic metabolite secreted by the tumor has been postulated.

We report this case to create awareness among pediatricians and oncologists about this rare complication of paraneoplastic papilloedema in neuroblastoma.

**References**