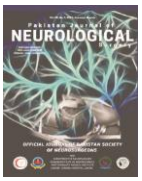

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Case Report

11 Years Old Female with Synergistic Presentation of Cranial and Spinal Pathologies: Myxopapillary Ependymoma with Hydrocephalus - A Rare Entity

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ABSTRACT

The correlation between hydrocephalus and spinal cord tumor was first described by Kyrieleis et. al. 1% concomitant presence was documented by Mirroni et.al. Moreover, the understanding of the pathophysiological association between these two conditions is still lacking. 11 years old girl presented in the outpatient department of Jinnah hospital Lahore with complaints of difficulty in walking for the last 2 years, numbness of right leg for the last one year, and severe bilateral sciatica for 2 months. On further inquiry, she told that she is also suffering from headaches and blurred vision for the last 6 months. Her neuroimaging revealed hydrocephalus and mixed density heterogeneously enhancing intradural lesion at the conus. She underwent ventriculoperitoneal shunting for hydrocephalus and laminoplasty followed by excision of spinal tumor reported being myxopapillary ependymoma grade II. The presence of headache along with signs & symptoms of raised intracranial pressure in these patients warrant cranial imaging. CSF diversion for hydrocephalus and gross total excision of the spinal lesion is the standard acceptable treatment.

Keywords: Cerebrospinal fluid (CSF), Ventriculoperitoneal Shunt, Myxopapillary Ependymoma, Primary Intra Dural Spinal Cord Tumor.

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INTRODUCTION

The correlation between hydrocephalus and spinal cord tumor was first described by Kyrieleis et al.[1] The prevalence of Malignant spinal tumors in the pediatric population is higher.[2] Hydrocephalus in children along with spinal tumors is a rare entity. 1% concomitant presence was documented by Mirroni et al.[3] Moreover, the understanding of the pathophysiological association between these two conditions is still lacking. It is observed that there is no obvious

obstructive pathology on neuroimaging in such cases of hydrocephalus and spinal tumor.[4,5,6] CSF examination of these patients showed a high concentration of proteins which can be the cause of hydrocephalus.[7-8] The presentation of hydrocephalus in these patients is usually covert. It is essential to keep in mind the association between hydrocephalus and spinal tumor in adolescent patients, who usually present with spinal symptoms. The presence of headache along with signs & symptoms of raised intracranial pressure in these patients warrant cranial neuroimaging. The management plan for patients with concomitant hydrocephalus and spinal tumor needs specialized care and treatment planning. Hydrocephalus is usually managed on priority but the decision varies from case to case.

Case Report

11 years old girl presented in the outpatient department of Jinnah hospital Lahore with complaints of difficulty in walking for the last 2 years, numbness in her right leg for the last one year, and severe bilateral sciatica for 2 months. On further inquiry, she said that she is also suffering from headaches and blurred vision for the last 6 months. On examination, she had right L2 hypoesthesia and weakness of right extensor hallucis longus along with absent right ankle jerk. Surprisingly she also had grade II Papilledema on Fundoscopy. She underwent neuroimaging for the brain and spine. Her CT brain revealed non-obstructive hydrocephalus whereas MRI Dorsolumbarspine showed T1W Hypo intense and T2W Hyper intense mass extending from D11 to S2 with solid and cystic components and intense enhancing solid mass from D11 to L3 with a non-enhancing cystic portion at the distal end. She was admitted to Neurosurgery Department and after a pre-operative workup, underwent a cerebrospinal fluid diversion procedure with a medium pressure ventriculoperitoneal shunt. Her CSF examination was revealed as normal (sample taken at the time of VP shunt). She planned for microsurgical excision of the spinal tumor during the same admission. She was operated on for spinal tumor 10 days after the VP shunt. She was positioned prone on the Wilson frame and after fluoroscopic guidance, an incision was made from D9 to L4 followed by Laminoplasty of D11 to L3. The gross total excision of the tumor arising from Conus was achieved under a microscope. The postoperative course was uneventful and she was discharged with the same neurological deficit as pre-op. Immunostaining of biopsy tissue revealed positive for GFAP & S-100, consistent with CNS WHO Grade 2 Myxopapillary Ependymoma. She was planned for multidisciplinary management and she is on regular follow-up at the Department of Oncology Jinnah Hospital Lahore along with the Neurosurgery department.

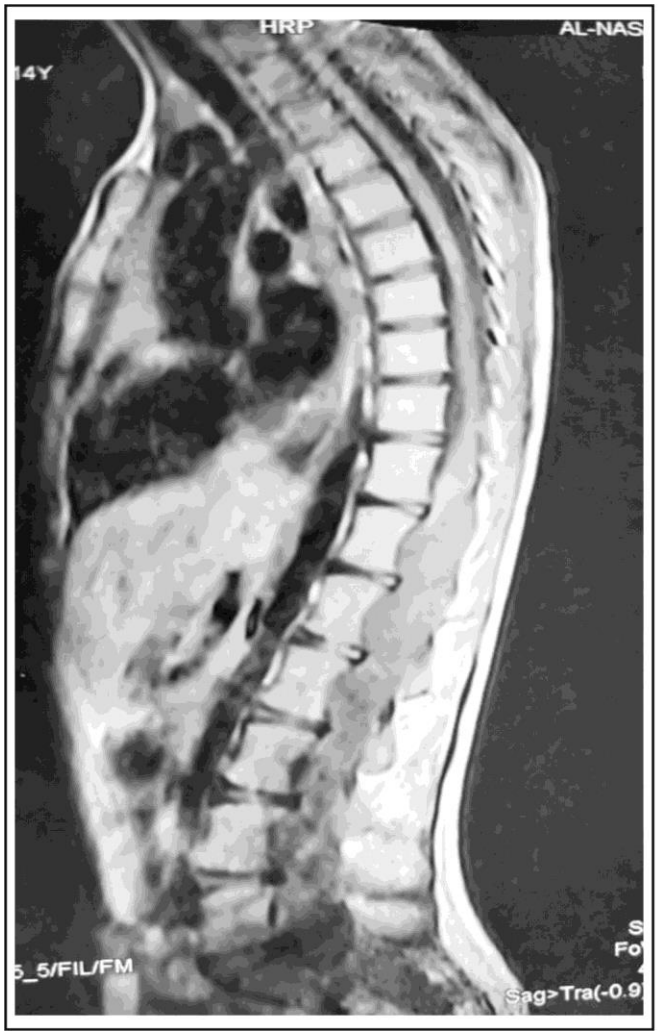

Figure A: Sagittal view T1.

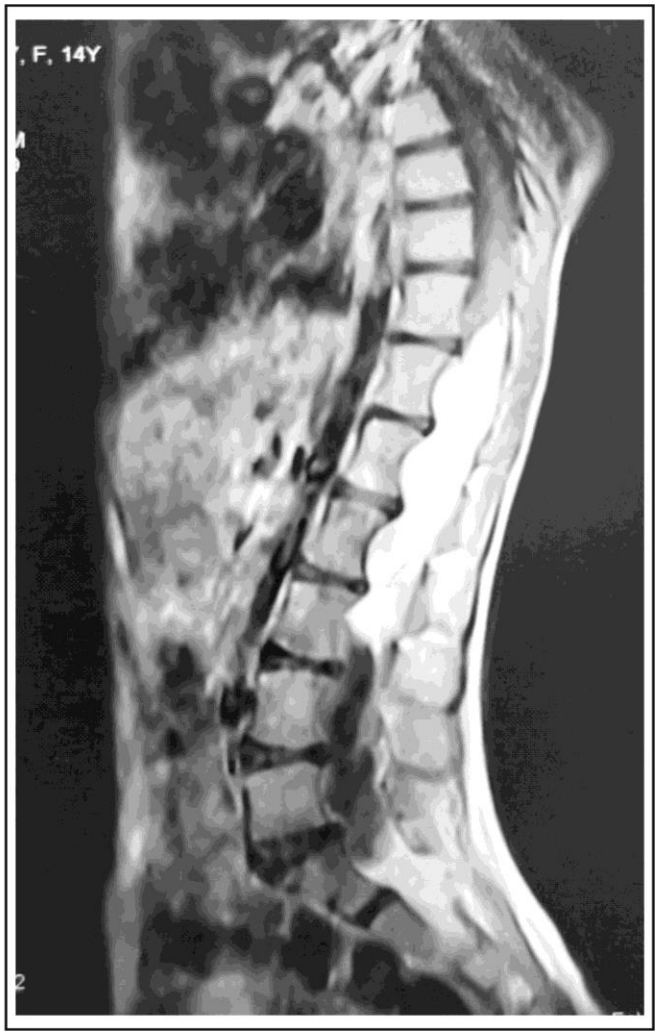

Figure B: Sagittal view T1 with contrast.

(Images used with patient's permission)

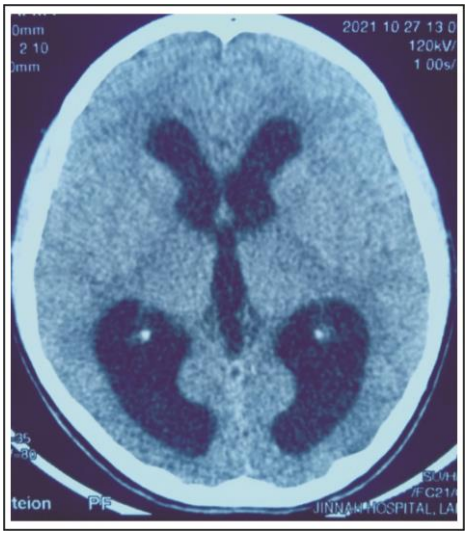

Figure C: CT Brain plain (Images used with patient's permission).

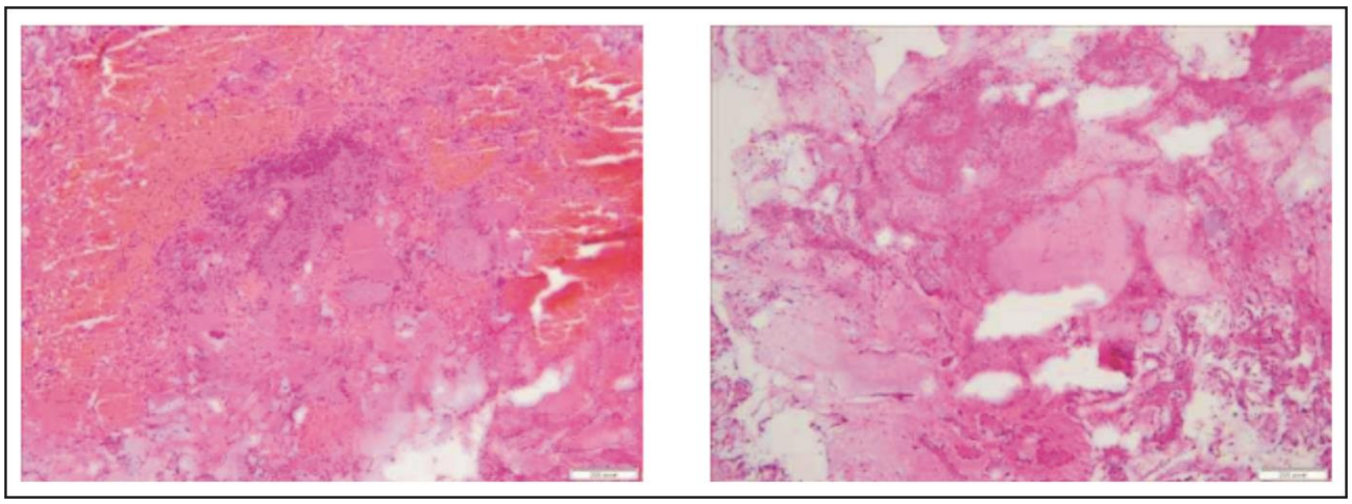

Figure D: Histological examination of the sections revealed a malignant neoplasm composed of well-differentiated cuboidal to elongated tumor cells radially oriented around blood vessels having hyalinized, myxoid walls. The tumor cells show no atypia, mitosis, or necrosis. Excision is complete. (Images used with patient's permission).

DISCUSSION

The presence of signs and symptoms of raised intracranial pressure along with spinal complaints secondary to Primary intradural spinal cord tumor in adolescent age must undergo detailed cranial neuro examination and neuroimaging to rule out the presence of hydrocephalus. The pathophysiology of hydrocephalus in such patients is still under debate. The spinal tumor at the cranio-cervical junction results in direct mechanical obstruction of CSF flow resulting in hydrocephalus, but the conus tumors are not usually responsible for such hydrocephalus. The rise in CSF proteins results in increased CSF viscosity that can lead to hydrocephalus in conus and intramedullary tumors.[9] The concept of conversion of fibrinogen to fibrin is considered to be responsible for increased CSF outflow resistance which results in hydrocephalus.[10,11,12] Rifkinson et.al. documented that a rostral tumor cyst at the cervicomedullary junction is responsible for obstructive hydrocephalus in 12 cases.[13] Neoplastic arachnoiditis as a result of intracranial seeding due to the spinal tumor is described by Lucey et al.[13-21] Rubinstein et. al. proposed the subarachnoid adhesions leading to hydrocephalus in patients with a leptomeningeal spread from glial intramedullary tumors.[22] The presence of hydrocephalus in conus ependymoma in our case resulted from fibrinogen pathogenesis, CSF proteins or neoplastic arachnoiditis is not clear. The presence of headache and blurring of vision in a patient with severe sciatica and numbness should guide us to do a workup for the cranial association.

Myxopapillary ependymoma represents 23% of the cauda equina region tumors[23-25] presenting with nonspecific symptoms and radiculopathy.[24-25] Although the presence of hydrocephalus in such patients is a known association it is still proved to be a rare entity.[23-29] The progression and synergy of symptoms are variable and it varies from case to case. In our case, headache preceded the spinal symptoms but they remain unnoticed and the patient was diagnosed with both pathologies at the same time. Buge et al, kalff et al, Roulades et al., and Riverez et al. reported the cases with concomitant hydrocephalus and conus myxopapillary ependymoma.[23-2]. The standard treatment for such patients is the concomitant management of both pathologies at the same time if diagnosed synergistically. Hydrocephalus is treated with CSF diversion procedures[29]

followed by standard microsurgical excision of spinal lesion.

CONCLUSION

The presence of signs and symptoms of raised intracranial pressure in a patient with spinal pathology must undergo cranial neuroimaging. The synergy of cranial and spinal symptoms is a rarity. The pathogenesis of the presence of hydrocephalus is still controversial. CSF diversion for hydrocephalus (VP shunt or endoscopic third ventriculostomy) and gross total excision of the spinal lesion is the standard acceptable treatment.

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Additional Information

Disclosures: Authors report no conflict of interest.
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Human Subjects: Consent was obtained by the patient in this case report.
Conflicts of Interest:
In compliance with the ICMJE uniform disclosure form, all authors declare the following:
Financial Relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work.
Other Relationships: All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

AUTHOR CONTRIBUTIONS

Sr. No.	Author's Full Name	Intellectual Contribution to Paper in Terms of
1..	Usman Ahmad Kamboh	Study design, and methodology.
2.	Muhammad Ashraf	Paper writing.
3.	Mehreen Mehboob	Data calculation and data analysis.
4.	Nabeel Chaudhary	Analysis of data and interpretations.
5.	Mehwish Manzoor	Literature review and referencing.
6.	Manzoor Ahmad	Editing and quality insurer.