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Atypical Case of Spinal Cord Ependymoma Presenting as Recurrent Pain Abdomen in a Child with Post-Operative Follow UP

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Abstract

Background: Spinal cord ependymomas are the second most common tumor of spinal cord in pediatric age group. These are intramedullary tumors and required to be diagnosed at the earliest for the management to avoid neurological complications. The important issue is of epidemiology and overall survival factors. **Case presentation:** We present a 14-years old boy who was evaluated for recurrent pain abdomen for eight months of histpry.He was diagnosed to have celiac disease and was put on treatment for the same. The complaint of pain abdomen did not subside .He underwent ultrasonography(USG),barium studies, upper gastro-intestinal tract (GIT) endoscopy, contrast enhanced computerized tomography(CECT) abdomen and magnetic resonance imaging (MRI).He was provisionally diagnosed as having nerve sheath tumor of the distal spinal cord. He underwent surgical excision of the tumor and that turned out be ependymoma. **Conclusion:**Child presented with recurrent pain abdomen and was treated for different ailments, but the investigations brought out the underlying pathology as spinal cord tumor. These type of tumors can only be diagnosed by cross sectional imaging like MRI and the surgical excision can be done at the earliest before it causes neurological deficit.

Keywords: Spinal Cord Ependymomas, Neurological Complications, Overall Survival Factors, USG, CECT, MRI, Case Report.

INTRODUCTION

These are common spinal cord tumor in adults as well as children. Intramedullary ependymomas are the second commonest spinal tumor in pediatric age group and constitutes 30% of these tumors. There may be in association with syringohydromyelia. The recurrence rate is very low if complete resection is done.[1,2]The incidence of spinal cord tumors of collective primary central nervous system tumors is 5% to 10% in adults. The commonest among intramedullary tumors are eprndymomas.The surgical delay can

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cause many neurological deficits and myxopapillary variety has worst prognosis even after surgery.[3]

CASE REPORT

14-years old boy reported with the complaints of pain abdomen in the right lumbar region of week duration. The pain was dull,off and on and without nausea and vomiting. There was no history of any previous illness or trauma. On examination the boy was appropriate for his age and constitution [Figure1].

Systemic examination was unremarkable. His weight was 32kg.His Hb was 11g/dL but rest of the blood investigations were normal. He was advised plain X-ray abdomen and ultrasound of whole abdomen.Plain X-ray abdomen was unremarkable [Figure 2].

USG whole abdomen revealed mesenteric lymphadenopathy.There was no free fluid seen in the peritoneal cavity. There was mild concretion seen in right kidney [Figure 3].

He was further advised barium meal follow through study [Figure 4] which was normal.

He underwent CECT abdomen to rule out any other pathology on the grounds of mesenteric lymphadenopathy.This also revealed the same findings without any other pathology [Figure 5].

He was advised upper GI endoscopy on the basis of positive TTG report.The endoscopy had shown scalloping of second part of duodenum with mosaic pattern which confirmed of having celiac disease [Figure 6].
He was started treatment for celiac disease. There was no relief of pain after treating for the celiac disease. He was further advised MRI to rule out any abnormality in spine or the spinal cord.MRI revealed a long mixed solid cystic component lesion extending from D8 to D12 vertebrae. This was iso- to hypointense on T1WI,hyperintenseinhomogenous on T2WI.There was intense patchy enhancement in post contrast TIW1 with fat suppression. [Figure 7 and Figure 8].

The working diagnosis of nerve sheath tumor was made as per the location and findings. He underwent gross total resection (GTR) in asuperspeciality hospital. The histo-pathological examination of the specimen had showed cellular type of ependymoma.The pain subsided and he is coming for regular follow-up. Post-op MRI was done and that revealed the removal of tumor to the major extent [Figure 9].

The patient had shown much improvement but he is still under the treatment for celiac disease in the form of gluten free diet and supplements of vitamins and iron. He had much relief of the unexplained pain abdomen.

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Table 1: McCormick grading had been done (Grade I –V)

Grade	Reason
I	Ambulatory patients without any deficit
II	Mild motor or sensory deficit
III	Moderate deficit
IV	Severe motor or sensory deficit
V	Paraplegic or quadriplegic

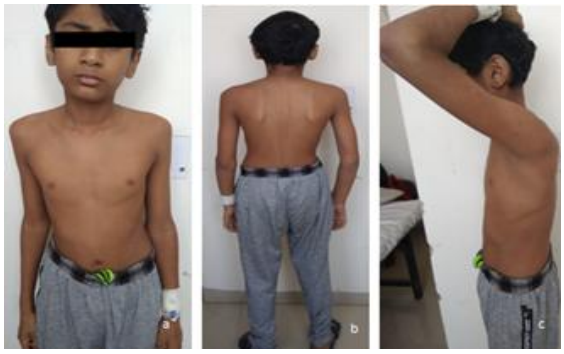

Figure1: Photo of 14-yrs old boy. a) front) back and c) profile views. There is no apparent abnormality in the body frame

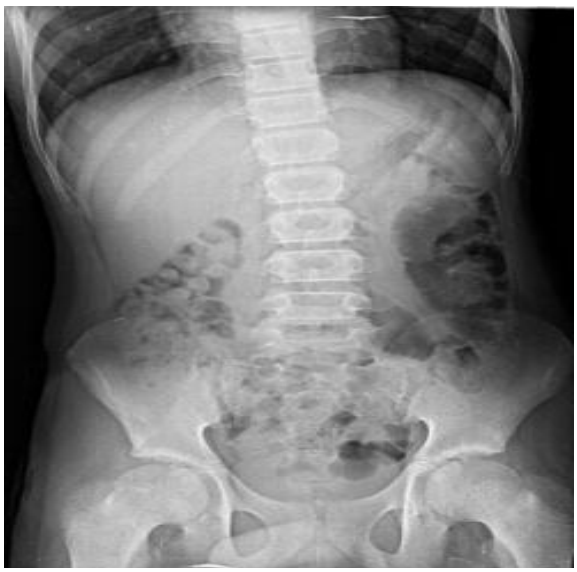

Figure 2.Plain X-ray abdomen. All the soft tissue, gas and bony landmarks are normal.

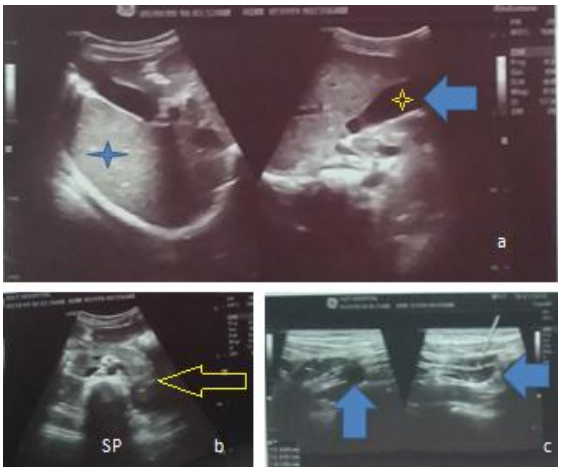

Figure 3: Ultrasound whole abdomen. a) right upper quadrant oblique section shows normal size and echotexture of the liver (blue star).b) axial section shows normal outline of pancreas (yellow arrow).c) High frequency abdominal scan shows mesenteric lymphadenopathy (blue arrows).

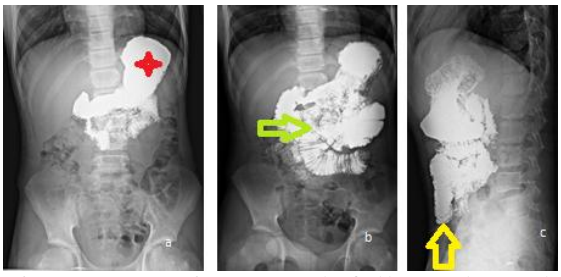

Figure 4: Barium meal follow through (BaMFT) study. a) initial film shows normal outline of stomach (red star).b) jejunal and ileal loops are normal (green arrow). c) distal gut loops are seen normal in outline and position in lateral film.

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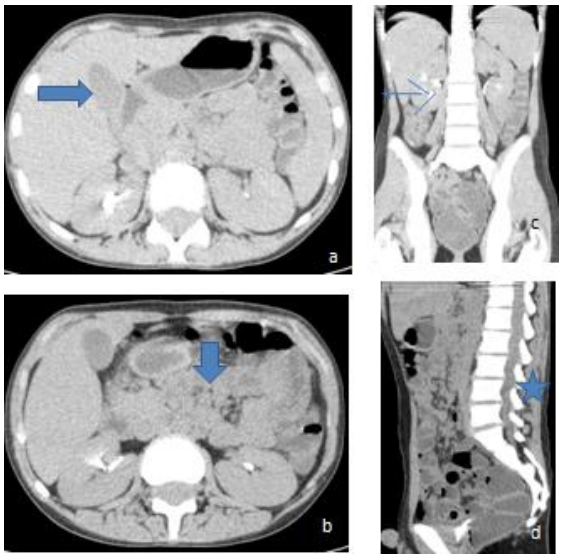

Figure 5: CECT abdomen. a) axial section at gall bladder level which is normal (blue arrow).b) axial section slightly lower to the previous section shows mesenteric lymphadenopathy (blue arrow).c) coronal reformatted section shows normal excretion in kidneys (blue thin arrow).d) sagittal reformatted section shows no abnormality of the vertebral column (blue star).

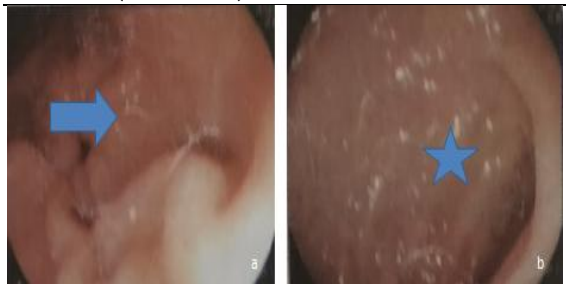

Figure 6: Upper GIT endoscopy. a) scalloping seen in second part of duodenum (blue arrow).b) mosaic pattern is highlighted (blue star).

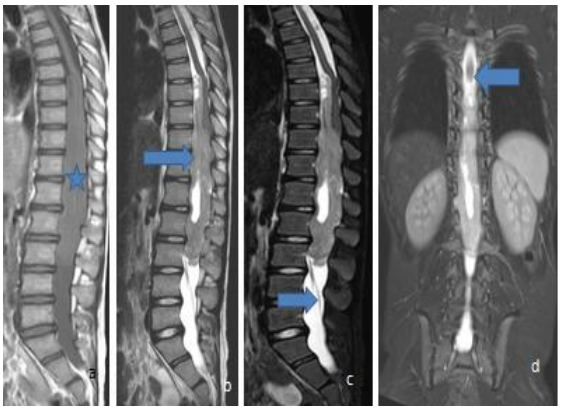

Figure 7: MRI of dorso-lumbar-spine.a) T1WI sagittal section shows expansion of the spinal canal with isointense mass in the lower dorsal vertebrae (blue star).b) T2WI sagittal section shows the mass lesion with cystic components extending from D8 to L1 level (blue arrow).c) STIR sequence shows the prominent cystic component and filumterminale (blue arrow).d) Coronal STIR section shows the extension of mass with the normal cord at superior region (blue arrow).

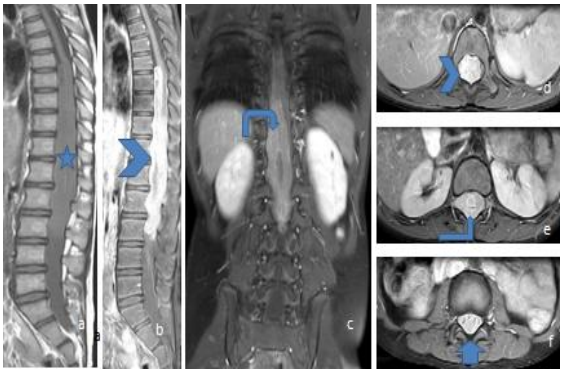

Figure 8: MRI spine contd…… a) T1WI sagittal section shows the expansion of the spinal canal (blue star).b) Post contrast T1WI fat suppressed images show patchy enhancement of the tumor with few cystic components without showing any enhancement (arrowhead).c) coronal post contrast section shows the same lesion (curved arrow).d) post contrast section axial T1WI at the

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proximal part of the tumor (arrowhead).e) same sequence at the mid portion of the lesion with patchy enhancement (blue arrow).f) same sequence at caudaequina (blue arrow).

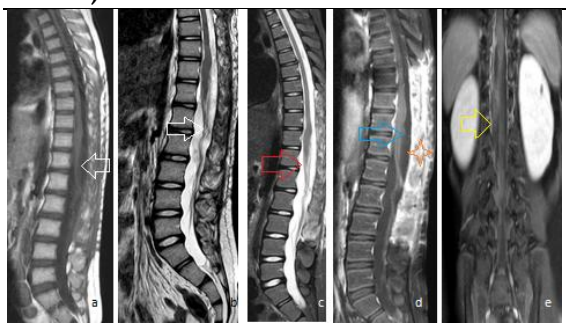

Figure 9: Post-op MRI scan. a) T1WI sagittal section shows no abnormal intensity in the tumoral region(white arrow).b) T2WI sagittal section shows spinal cord without any compression (white arrow).c) FFE sagittal section shows normal spinal cord (red arrow).d) post-gadolinium T1WI sagittal section with fat suppression shows with almost removal of the bulk of tumor except some soft tissue superficial fibrotic enhancement (red star).e) Same sequence in coronal section do not show any abnormal residual tumor (yellow arrow).

DISCUSSION

In the list of intramedullary spinal cord tumors (IMSCT) ependymomas comes next to pilocyticastrocytomas. These arise from ependymal cells of central canal or the cell rests in the filumterminale.Histologically there are six types of ependymomas.Cellular type is the most common type.[4]Although these can occur anywhere but the cervical cord is the most common site. The incidence increases with the advancement of age and found more frequently in adults and elderly. There are more occurrences in the patients of neurofibromatosis type 2.The most frequent clinical symptomatology is in the form of pain with gait deterioration, weakness and the compression symptoms as per the location of these tumors. The intramedullary tumors are very slow growing and may take on an average three years before presentation like in our present case.McGirt in 2008 confirmed that subtotal resection is sufficient for long term survival. The adjuvant therapy may or may not be the rule for this long term survival.[5]The clinical diagnosis is made as per McCormick grading. The patients used to report in higher grading before MRI was used as diagnosis. Now the patients are being diagnosed in grade I or II.The details of the grading had been given below in [Table 1].

Plain radiography can give the indirect clue for the tumors like spinal canal widening, scoliosis, vertebral body scalloping, laminar thinning and pedicle erosions.CT shows the bony changes in the form of canal widening, scalloping and erosions. The tumor can show intense enhancement in post contrast sequences. The diagnosis of intramedullary lesions was unveiled in details after MRI has come in diagnostic field. These are usually midline lesions, expanding the cord but features are quite variable. Some of the tumors are uncapsulated, eccentric and present as extramedullary component. The contrast enhancement pattern may

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be avid homogenous or patchy. The cystic component is well demarcated in contrast sequences with peripheral enhancement. 22% tumors show cyst formation but 62% may also show tumoral cysts.Non tumoral cysts lie in the Polar Regions. These tumors require to be differentiated from haemangioblastoma and astrocytomas on the basis of enhancement pattern. These tumors extend up to four vertebral segments.[6]T1WI are either isointense or hypointense when hemorrhagic component is not there.T2WI show hyperintense signal with peritumoraledema in 62%.20-30% cases show "cap sign" indicative of hemosidrin component of the hemorrhage. There is avid inhomogenous contrast enhancement. Invasive spinal ependymomas do not have good prognosis as compared to non-invasive higher grading. The key point lies in the early diagnosis and the corresponding surgical management with or without radiation therapy and chemotherapy.Myxopapillaryependymomas (MPE) are slightly uncommon and often found in the caudaacquina region. Those patients who underwent gross total resection (GTR) had better survival rate than those who had subtotal resection (STR).GTR is always better and this also requires long term follow- up to see any recurrence.[7]STR requires radiotherapy in addition to the surgery. Female children show less mortality.[8]

CONCLUSION

Spinal cord ependymomas are rare in pediatric age group. These are very slow growing tumors and are diagnosed quite late as the patients do not have much alarming symptomatology. These are diagnosed when patient present with higher McCormick grading stage .MRI is the most valuable tool for the diagnosis. Now the diagnosis is being made in early grading because of the cross sectional imaging tools especially MRI.GTR is more successful as compared to STR.The follow up of these case is quite long.

REFERENCES

1. Smith AB,SoderfundKA,Rushing EJ et-al.Radiologic-pathologic correlation of pediatric and adolescent spinal neoplasms:Part 1,Intramedullary spinal neoplasms.AJR Am J Roentgenol.2012;198(1): 34-43.doi:10.2214/AJR.10.7311
2. KuciaEJ,BambakidisNC,ChangSW,SpetzlerRF:Surgical technique and outcomes in the treatment of spinal cord epedymomas,part 1:intramedullaryependydomas.Neurosurgery 2011;68(1 Suppl Operative):57-63.doi:10.1227/NEU.0bo13e318208f181
3. BostromA,von LEHE M,HartmannW,PietschT,FeussM,BostromJP,etal:Surgery for spinal cord ependymomas:outcome and prognostic factors.Neurosurgery 2011; 68:302-309.doi:10.1277/NEU.0b013e3182004c1e
4. KoellerKK,RosenblumRS,MorrisomnAL.Neoplasms of the spinal cord and filumterminale:radiologic-pathologic correlation.Radiographics.2000;20(6):1721-49. Doi:10.1148/radiographics.20.6.g00nv151721.
5. McGritMJ,ChaichanaKL,AtibaA,AttenelloF,Yao KC,JalloGI:Resection of intramedullary spinal cord tumors in children:assessment of long-term motor and sensory deficits.JNeurosurgPediatr 2008;1:65-67.doi:10.3171/PED-08/01/063
6. Miyazawa N,HidaK,IwasakiY,KoyanagiI,Abe H:MRI at 1.5 T of intramedullary ependymoma and classification of pattern of contrast

enhancement.Neuroradiology 2000;42:828-832.https://doi.org/10.1007/s002340000386

7. Lucchesi KM,GHrantR,KahleKT,MarksAM,DiLunaML.Primary spinal myxopapillaryEpendymoma in the pediatric population:a study from the Srveillance,Epidemiology,and End Results(SEER) database.J Neurooncol.2016;130(1):133-140. doi:10.1007/s11060-016-2218-6.
8. Lin Y,LeaA,MelkonianSC,LamS.Treatment of pediatric Grade II spinal ependymomas:a population based study.JNeurosurgPediatr.2015;15(3):243-9.doi: 10.3171/2014.PEDS1473.

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