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LEPTOMENINGEAL DISSEMINATION OF PILOCYTIC ASTROCYTOMA OF CONUS MEDULLARIS IN A 16-YEAR-OLD MALE
Ibtihal Ahalli, Department of Medical Oncology, Hassan II University Hospital, Fez, Morocco, Lamiae Amaadour, Department of Medical Oncology, Hassan II University Hospital, Fez, Morocco, Younes Dkhissi, Department of Radiology, Hassan II University Hospital, Fez, Morocco, Kaouthar Messoudi, Department of Medical Oncology, Hassan II University Hospital, Fez, Morocco, Raihana Boujarnija, Department of Medical Oncology, Hassan II University Hospital, Fez, Morocco, Anas Lahlou Mimi, Department of Radiology, Hassan II University Hospital, Fez, Morocco, Karima Oualla, Department of Medical Oncology, Hassan II University Hospital, Fez, Morocco, Zineb Benbrahim, Department of Medical Oncology, Hassan II University Hospital, Fez, Morocco, Samia Arifi, Department of Medical Oncology, Hassan II University Hospital, Fez, Morocco, Meryem Boubbou, Department of Radiology, Hassan II University Hospital, Fez, Morocco, Nawfel Mellas, Department of Medical Oncology, Hassan II University Hospital, Fez, Morocco.

Corresponding author: Ibtihal Ahalli, Medical Oncology Department, Hassan II University Hospital, Faculty of Medicine and Pharmacy of Fez, Morocco, Postal Code: 30070 *Fès.* Email: ahalli.ibtihal@gmail.com

LEPTOMENINGEAL DISSEMINATION OF PILOCYTIC ASTROCYTOMA OF CONUS MEDULLARIS IN A 16-YEAR-OLD MALE

I.Ahalli, L. Amaadour, Y. Dkhissi, K. Messoudi, R. Boujarnija, A. L. Mimi, K. Oualla, Z. Benbrahim, S. Arifi, M. Boubbou and N. Mellas

ABSTRACT

Pilocytic astrocytoma (PA) is a slow growing brain tumor that tend to occur in young patients. Although PA is often a benign tumor, dissemination to leptomeninges has been described in the literature. We describe a sixteen-year-old boy who initially presented with paraplegia and progressed to severe neurological features. An intradural intramedullary spinal cord tumor was identified in medullar MRI, and diagnosis of PCA by a surgical biopsy of the lesion was performed. Subsequently, patient developed an intracranial hypertension syndrome and Craniospinal MRI revealed an intracranial and spinal leptomeningeal dissemination. The patient was treated with courses of cytotoxic consisting of carboplatin – vincristine-based chemotherapy. He died of disease two months later. Our case emphasizes the lack of benefit of chemotherapeutic agents in PA with diffuse leptomeningeal dissemination and review existing literature.

INTRODUCTION

Pilocytic Astrocytomas (PAs) is a low-grade tumor of the Central Nervous System (CNS), occurring predominantly in children and young adults. The tumor may arise in various parts of the CNS but has a predilection for the cerebellum, thalamus, third ventricle, optic tracts, brain stem, and medical temporal lobe (1,2). The PAs have usually a benign behavior however several cases of PA dissemination throughout central nervous system have been described. Primary spinal PA with leptomeningeal dissemination is even more uncommon, and to date, only four cases have been described in the medical literature (3,4). Since leptomeningeal dissemination of PA is a very rare entity, the most ideal treatment of the disease is still debated.

We report the case of a 16-year-old boy with a pilocytic astrocytoma of conus medullaris with leptomeningeal dissemination. A review of the literature on low grade gliomas of the spinal cord revealed few cases of primary spinal PCA with intracranial dissemination.

Case presentation:

A 16-year-old boy presented to our department with severe neurological impairment. Three months before his admission to hospital, he presented unsteadiness and loss of balance as well as an inability to stand or walk. Physical examination revealed Grade 3/5 bilateral strength in the muscles of lower extremities with concomitant hypoesthesia in the territory of the internal popliteal sciatic nerve and vibratory hypoesthesia in the lower left limb with abolition of patellar and Achilles reflexes.

Laboratory tests including Alpha Foeto - Protein (AFP) and B-human Chorionic Gonadotropins (HCG) were within the normal limit. The Medullar magnetic resonance imaging (MRI) revealed an intramedullary conus medullaris tumor extended for a length of 70 mm from T-11 to L-1. Surgical biopsy of the lesion was performed. Pathologic examination showed spindle shaped of tumor cells and the presence of brightly eosinophilic and corkscrew-shaped fibers (Rosenthal fibers) consistent with and confirmed the diagnosis of PA (Figures 1,2,3). Ten days later, functional neurological symptoms worsened, and the patient presented with intracranial hypertension syndrome.

Craniospinal MRI revealed multiple leptomeningeal enhancing nodules in the brain and total spinal cord suggestive of intracranial and spinal leptomeningeal dissemination (Figures 4,5).

Subsequently the patient received chemotherapy consisting of ten courses of; carboplatin (175mg/m2) and vincristine (1.55 mg/m2), schedule 4/2.

In the first assessment after completion the induction regimen, there was no change in the overall clinical status of the patient. In addition, brain and spinal cord MRI revealed progression of the intramedullary and brain lesions (Figures 6,7). One month later the patient died of disease progression.

DISCUSSION

This case manifested characteristics of Pilocytic astrocytoma in terms of clinical presentation, histology result and progression. Like most brain tumors these are low-grade gliomas corresponding to WHO grade I with an excellent prognosis, particularly if complete resection can be achieved. The PAs predominantly affect children and young adults in their early decades of life, with a predilection for males [5].

Despite the fact that PAs are grade 1 tumors, occasionally show an aggressive behavior such as malignant transformation, recurrence or even more rarely: Leptomeningeal dissemination [3,11].

However, spread mechanisms of low-grade gliomas to leptomeninges is rarely encountered. It was initially speculated that tumor manipulation during resection contributed to dissemination through the release of tumor cells to the cerebrospinal fluid [7,8]. This mechanism may play a role in some cases, it is most likely not the entire explanation primary origin, given that there are numerous reports of dissemination at initial presentation [9,10].

Since leptomeningeal dissemination of PA is considered uncommon, the best therapeutic approach is still complete surgical resection when feasible. Radiotherapy alone or in combination with Chemotherapy have shown beneficial effects in some studies [12].

Since in our case all the spinal and brain meninges were involved, complete surgical resection was not possible. Therefore, a decision was made by the pediatric oncology team to opt for chemotherapy. However, just like in most published cases, chemotherapy alone did not seem to provide significant benefit to this patient [6].

Conclusion:

In this report we described a child with a Leptomeningeal Dissemination of pilocytic astrocytoma of conus medullaris not amenable to surgical resection and no significant response to chemotherapy. This highlights the poor outcome when leptomeningeal dissemination of PA is diffuse.

APPENDIX

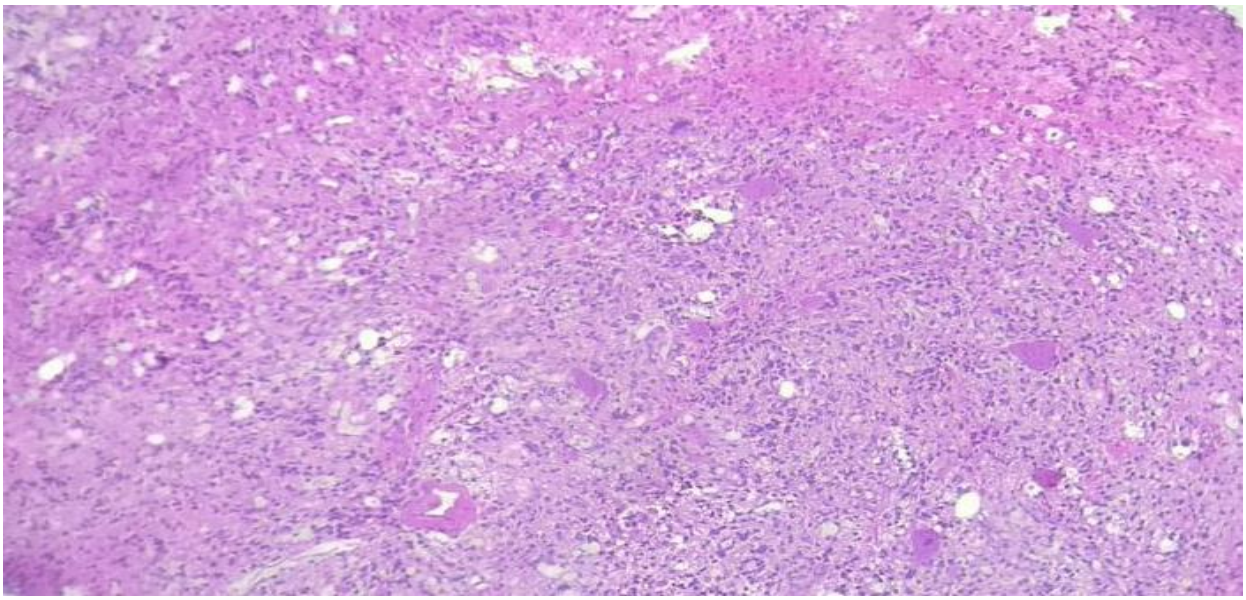

Fig 1: Image that shows a biphasic appearance with cellular areas alternating with hypocellular, microcystic areas (H&Ex10).

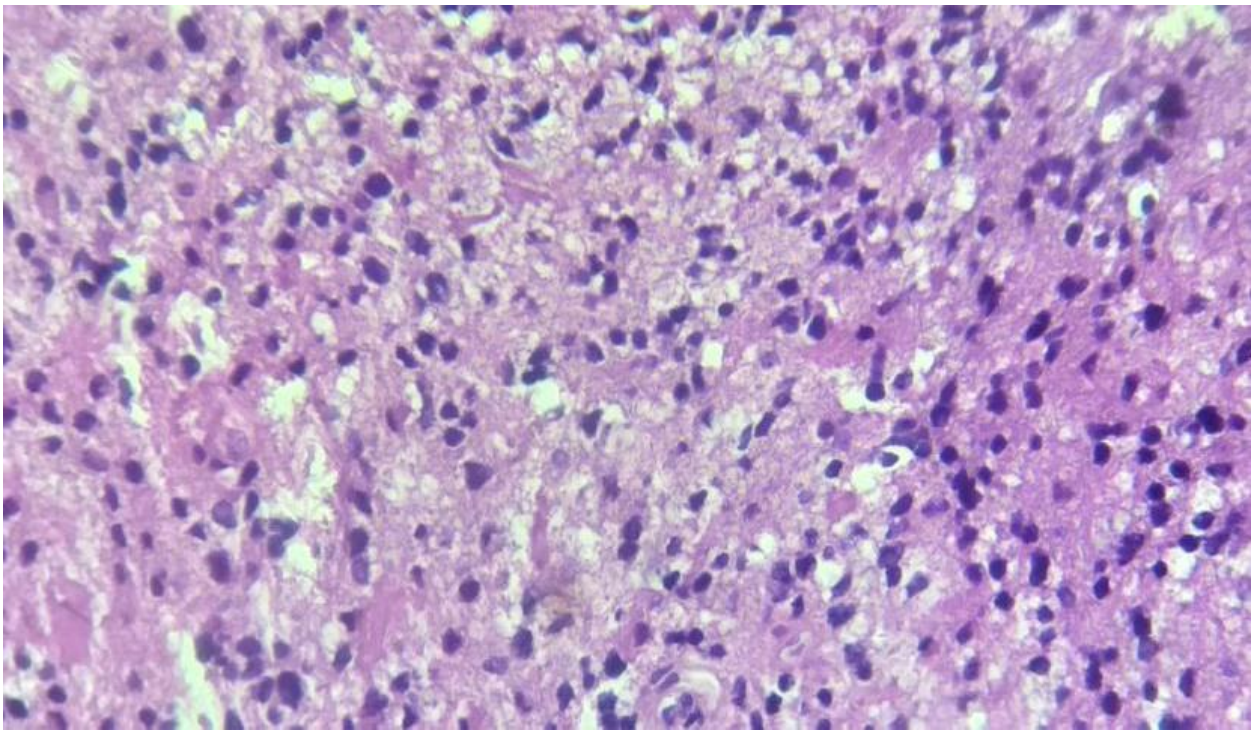

Fig 2: The tumor cells are spindle shaped. The nuclei are oval with bland chromatin and a low mitotic activity (H&Ex40)

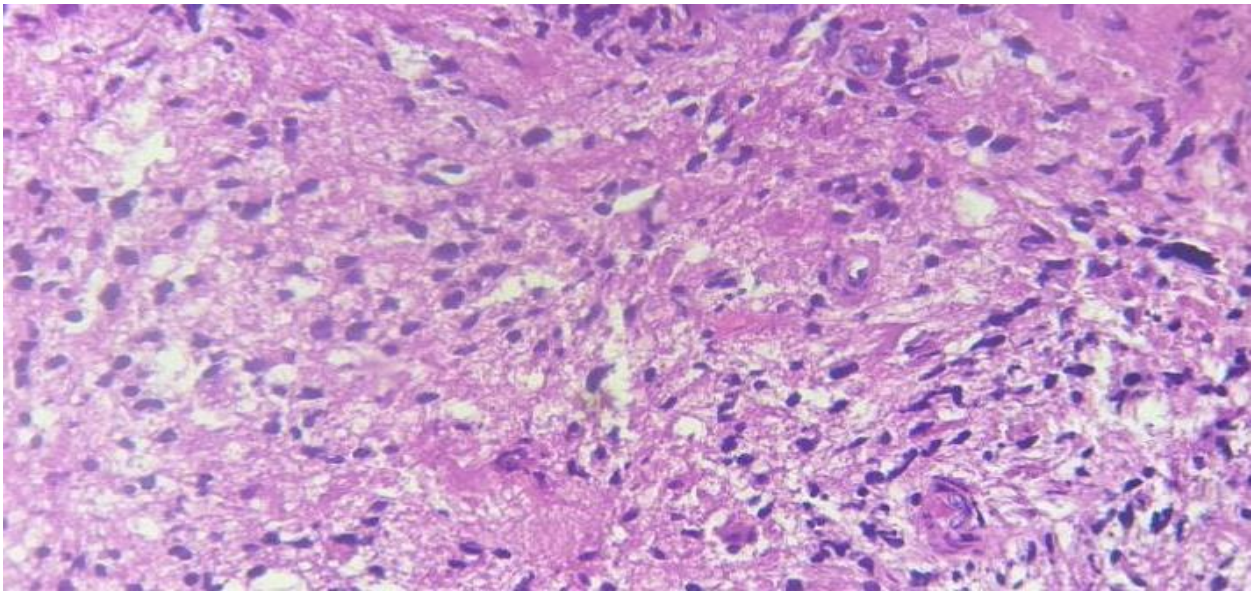

Fig 3: this image shows the presence of brightly eosinophilic, corkscrew-shaped fibers (Rosenthal fibers) (H&SX40).

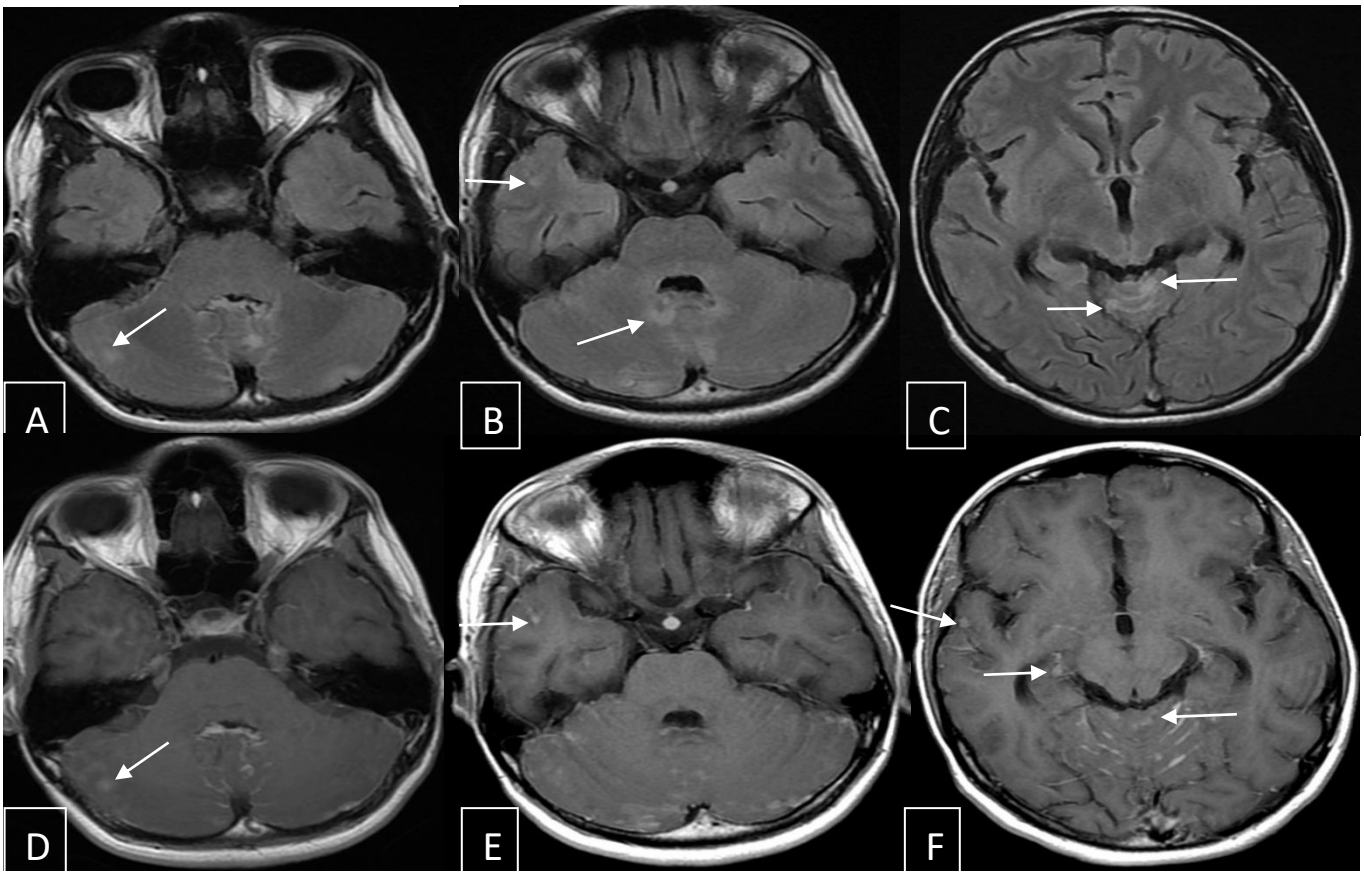

Fig 4: *Brain MRI in FLAIR weighted axial views (A,B,C) and gadolinium-enhanced T1-weighted axial views (D,E,F) : Intracerebral hyperintense flair weighted nodular lesions, enhanced after gadolinium injection.*

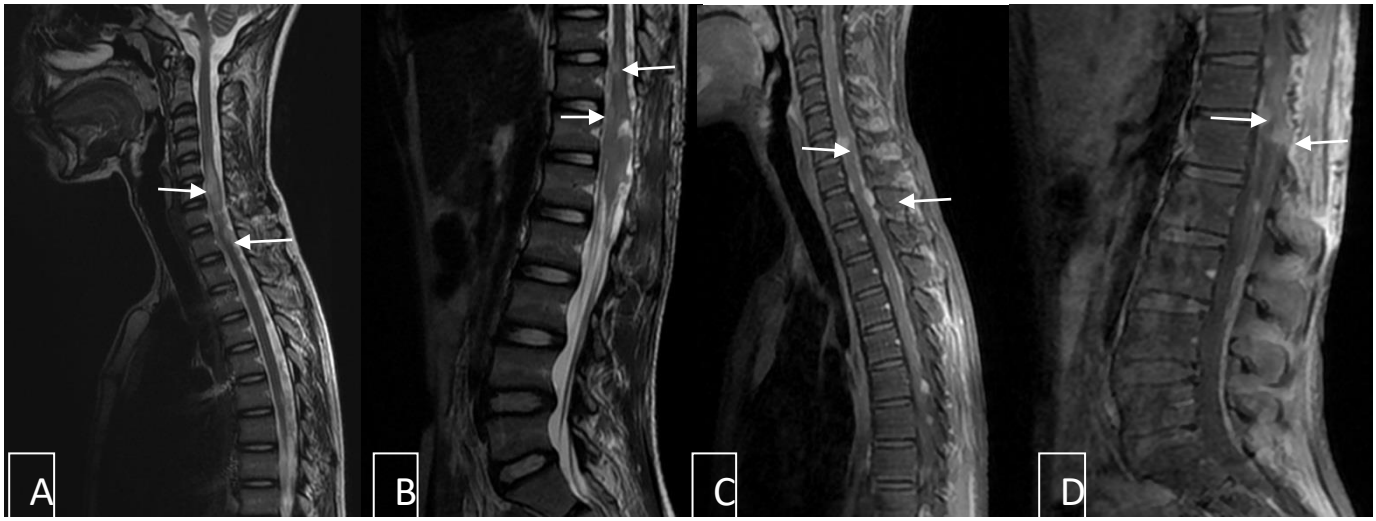

Fig 5: Sagittal sections MRI of the whole spine in T2-weightred views (A-B) and Gadolinium-enhanced T1-weighted views (C-D) : Hyperintense T2 intramedullary and dura nodular lesions, enhanced after gadolinium injection.

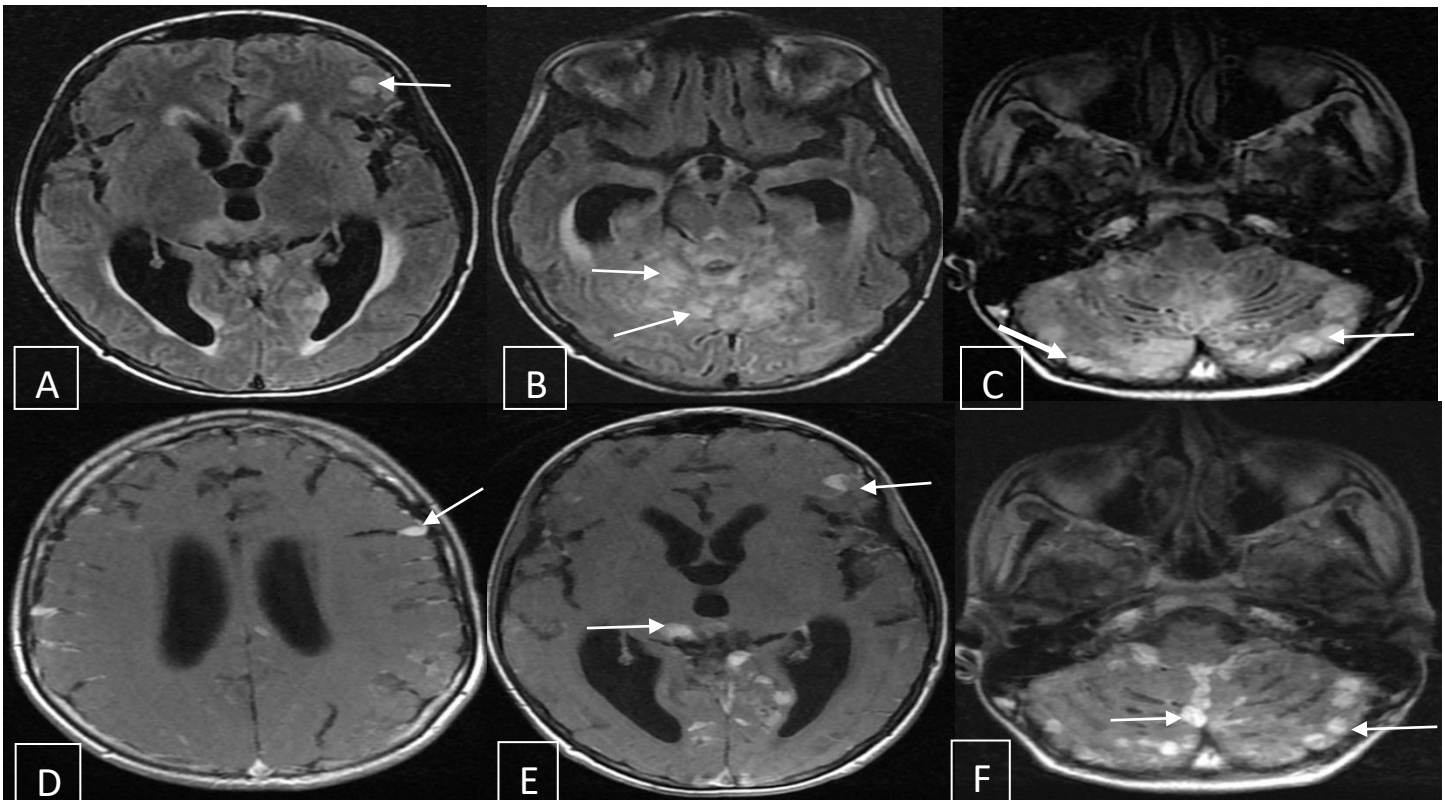

Fig 6: Brain MRI in FLAIR weighted axial views (A,B,C) and gadolinium-enhanced T1-weighted axial views (D,E,F) : Increase in size and number of intracerebral hyperintense flair weighted nodular lesions, enhanced after gadolinium injection.

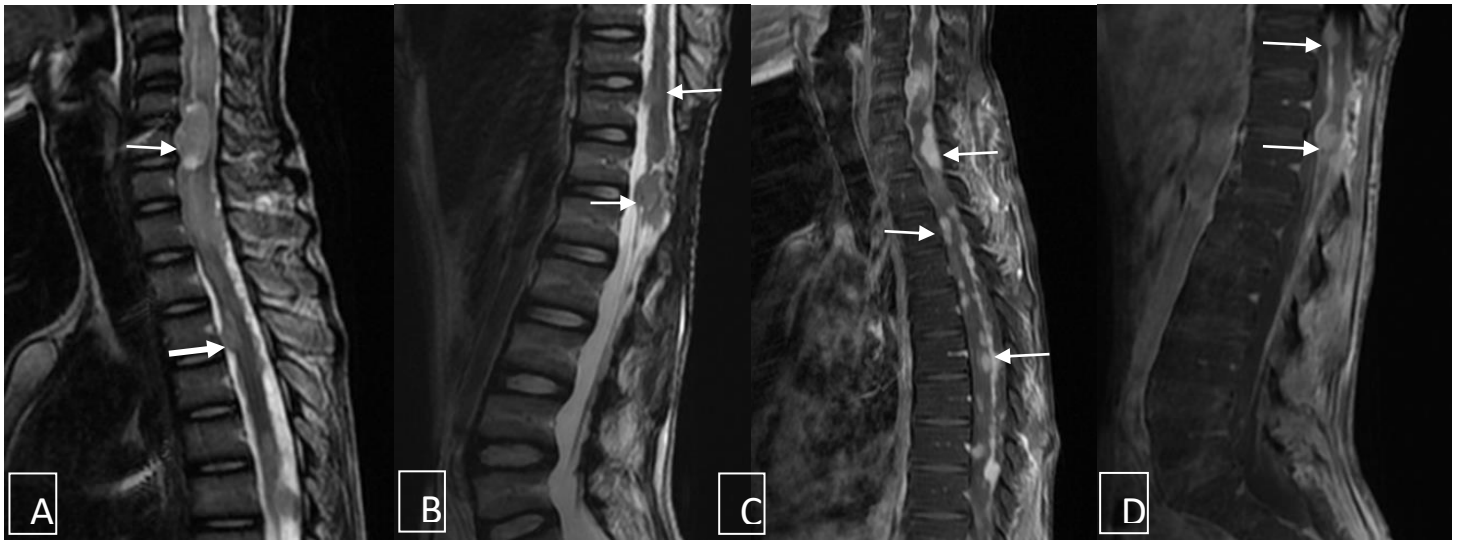

Fig 7: Sagittal sections MRI of the whole spine in T2-weightred views (A-B) and Gadolinium-enhanced T1-weighted views (C-D) : Increase in size and number of the intramedullar and dural nodular lesions.

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