

## Case Report

# Spinal pleomorphic xanthoastrocytoma accompanied with periventricular tumor

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**Abstract:** Pleomorphic xanthoastrocytoma (PXA) is a low grade tumor that occurs in supratentorial area of children and young adult. In the previous reports, PXA of spinal cord or multicentre was extremely rare. A 60-year-old patient of spinal PXA and periventricular tumor presented with waist pain and weakness of double legs for one month. Neuroimaging showed that a lesion at the level of L2-L3 and periventricular tumor. Postoperative microscopy indicated that WHO grade II PXA. Photomicrograph of the lesion showed spindle cells, marked nuclear and cytoplasmic pleomorphism, with foamy cytoplasm. Immunohistochemical staining showed that GFAP and S-100 were positive. This is a rare case of synchronous multicentric PXA. Physicians should be realized multicentric dissemination by meninges or cerebrospinal fluid in PXA patients. It is important to describe the particular case in order to better understanding of clinical features.

**Keywords:** Pleomorphic xanthoastrocytoma, spinal tumor, multicentric tumor

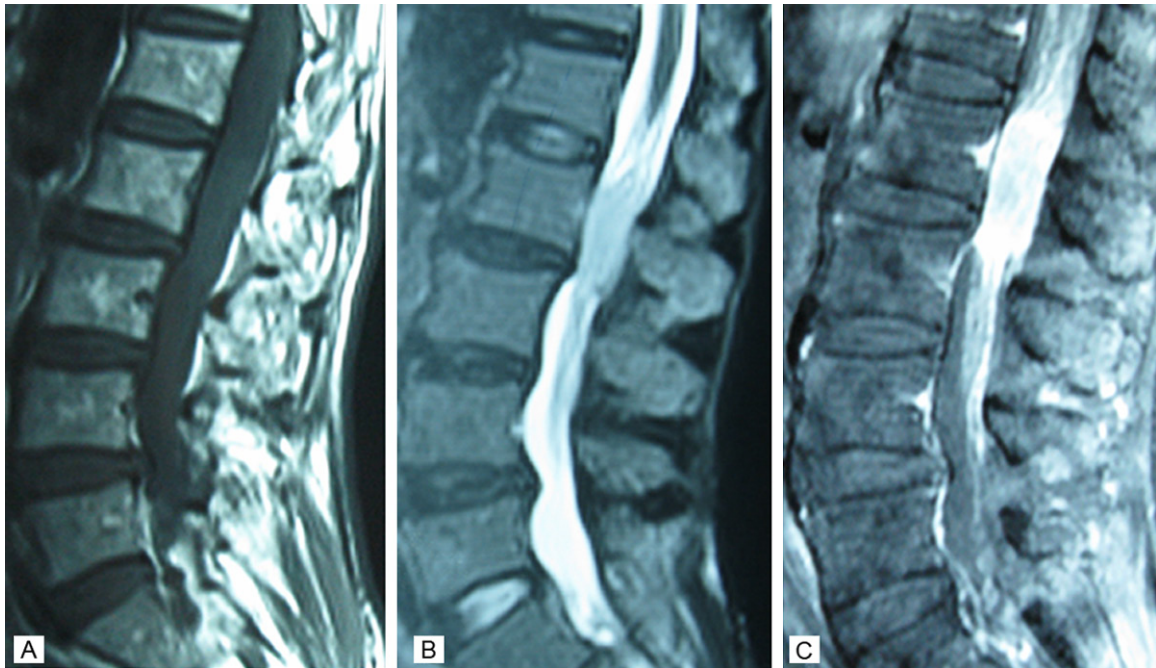
## Introduction

Pleomorphic xanthoastrocytoma (PXA) is a rare tumor of central nervous system, which accounts for less than 1% of brain tumors. It was firstly reported and named by Kepes in 1979 [1]. Later, literatures of PXAs have been reported. PXAs are mostly located in supratentorial area. PXAs of cerebellum, peneal body and spinal cord have been reported sporadically [2-4]. However, spinal or multicentric PXAs are extremely rare. In view of the rarity of the entity, a case is reported and literatures are reviewed.

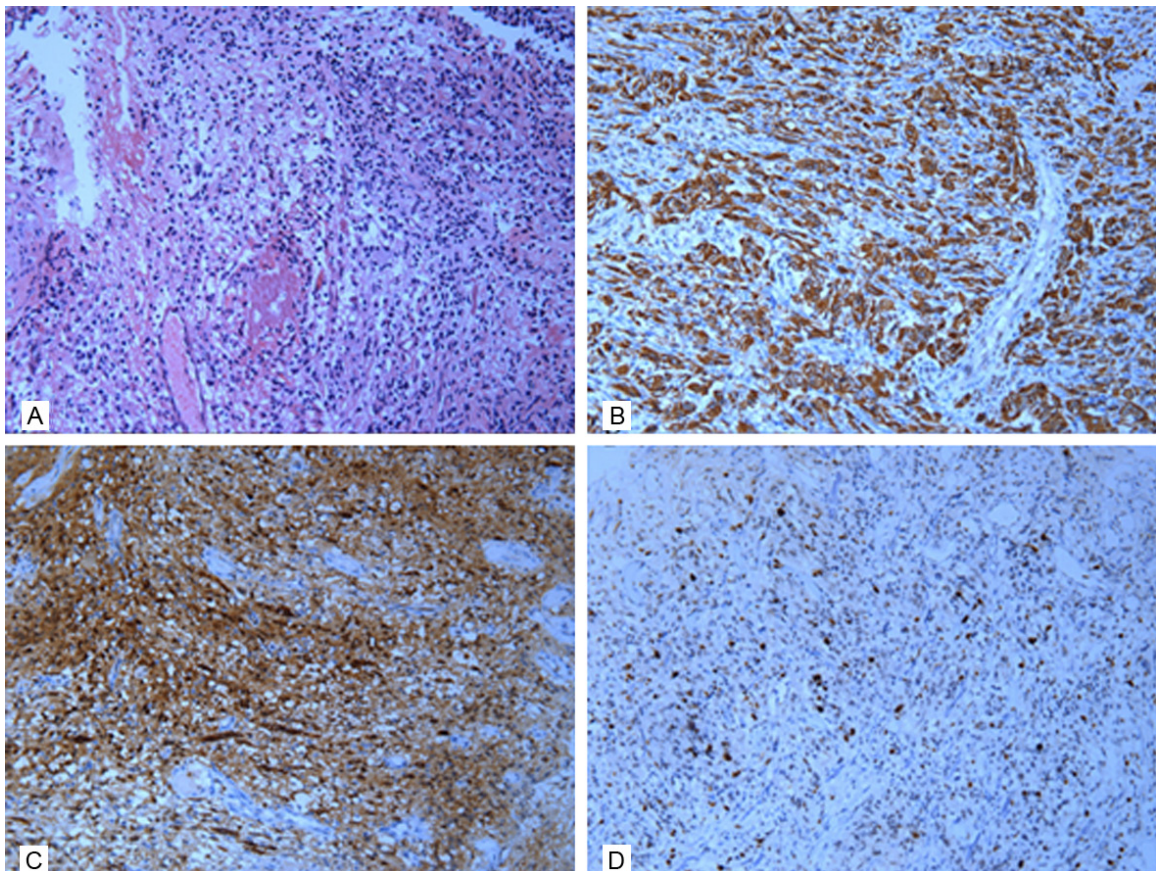
## Case report

A 60-year-old male patient suffered from waist pain and weakness of double lower limbs for one month. The patient had no other serious disease. Body examination showed that muscle strength of both legs was achieved grade 3. Lumbar magnetic resonance imaging showed isointense signal on T1 weighted imaging and slightly hyperintense signal on T2 weighted imaging at the level of L2-L3. The lesion showed significantly homogeneous enhancement (Figure 1). During operation, we found

that most of the cauda equina were wrapped by lesion and appeared swollen. The tumor appeared as a soft grayish-red mass. Partial resection was performed in this patient, and the dural matter was not sutured in order to relieve the intraspinal pressure. Postoperatively, muscle strength was the same as preoperative. But the patient still had severe waist pain. Therefore, at the nine days after operation complete resection was performed. Cauda equina invaded by lesion was resected totally. Muscle strength of both legs declined to grade 0. And sensation disappeared. He also suffered from defecation difficulty. Postoperative microscopy indicated that WHO Grade II PXA. There is no necrosis and nuclear fission. There are spindle cells, marked nuclear and cytoplasmic pleomorphism, foamy cytoplasm. Immunohistochemical staining showed that GFAP and S-100 were positive. Approximately 2% tumor cells were positive for Ki-67 (Figure 2). Sometimes the patient complained of dizziness postoperatively. On CT scanning, periventricular low-density tumor was found. Therefore MRI scanning was performed. The periventricular lesion showed hypointense on T1 weighted imaging, and hyperintense on T2 weighted imaging and



**Figure 1.** Lumbar MRI features. A: Isointense signal on T1 weighted imaging at the level of L2–L3; B: Slightly hyperintense signal on T2 weighted imaging; C: Significantly homogeneous enhancement on MRI enhancer.

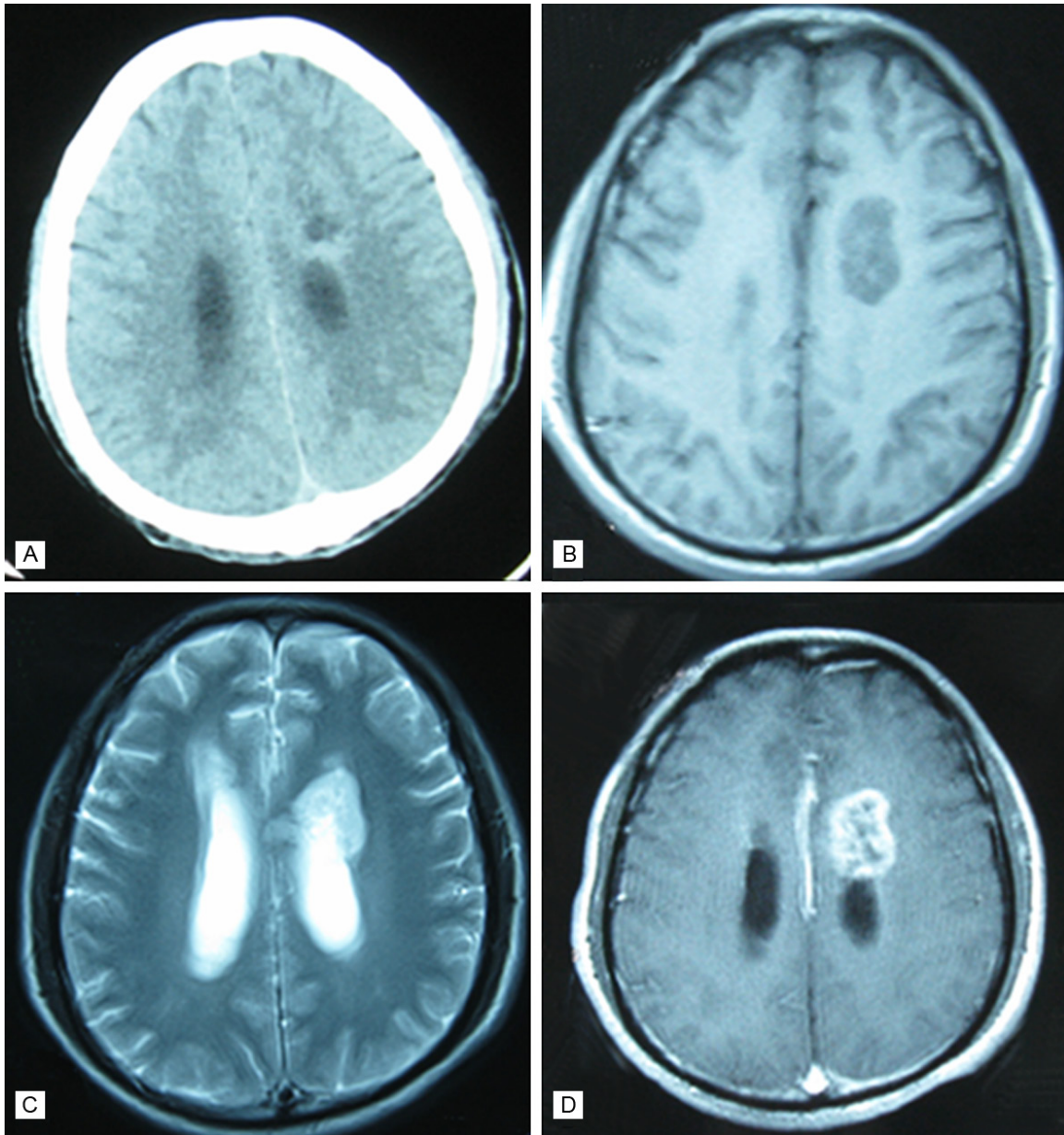


**Figure 2.** Histopathological examination of lesion obtained from the first resection of intraspinal tumor. A: Photomicrograph of the lesion showing spindle cells, marked nuclear and cytoplasmic pleomorphism, foamy cytoplasm



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(hematoxylin and eosin; original magnification,  $\times 200$ ); B: Immunohistochemical examination shows that GFAP was positive (original magnification,  $\times 200$ ); C: Immunohistochemical examination shows that S-100 was positive (original magnification,  $\times 200$ ); D: Approximately 2% of tumor cells are positive for Ki-67 (original magnification,  $\times 200$ ).



**Figure 3.** Cerebral radiology features. A: Periventricular hypodense lesion of on CT scanning; B: Hypointense signal on T1 weighted imaging; C: Hyperintense signal on T2 weighted imaging; D: Enhanced MRI scanning showed a heterogeneous and annular enhancement.

T2 fluid-attenuated inversion recovery. Enhanced MRI scanning showed a heterogeneous and annular enhancement (**Figure 3**). The lesion was not treated. He also didn't receive any adjuvant therapy. At 3 months of follow-up, the muscle strength and sensation of both legs did not rehabilitate.

### Discussion

PXA is a rare glial neoplasm, originating from subpial astrocytes, which has relative favorable outcomes due to WHO grade II designation. The overall survival rate at 5 years was 74% [5]. PXA often occurs in the first three decades of life. There is no sex predilection. PXA commonly

occurs in supratentorial location, especially the temporal lobe [5]. Seizure is the most common symptom, and headache may also present.

Histologically, PXA is characterized by spindle cells, multinucleated giant cells and foamy lipid-laden xanthomatous cells. PXA is positive for GFAP by immunohistochemistry. Mitosis and necrosis is commonly absent. Typical radiologic findings of supratentorial lesions are cystic neoplasm accompanied by mural nodule. On CT scanning, PXA shows iso-dense to hypodense. Calcification is uncommon. The cystic mass shows hypointense on T1 weighted imaging, and hyperintense on T2 weighted imaging. Mural nodule is strongly enhanced after the administration of gadolinium. Peritumor edema was minimal.

Supratentorial PXAs are the most common. Lesions of other locations were also reported, such as cerebellum, spine and pineal body. Spinal PXAs are extremely rare. Herpers firstly reported a 66-year-old patient with spinal PXA in 1994 [6]. Only six patients were presented in the previous literatures until now [2, 5-9]. The age distribution was 12.9-66 years old with an average age of 42.0 years old. Most of the lesions were located in cervical or thoracic cord.

Single tumor is the most common. Most of the reports thought that surgery should be the first choice. The patients could have a favorable outcome after gross total resection [6]. However, some patients suffered recurrence and anaplastic transformation. And some patients suffered dissemination by meninges or cerebrospinal fluid.

Multicentric PXAs were reported rarely at diagnosis. In 2005, Saikali firstly reported multicentric PXA in a 30-year-old patient with NF1 [10]. At the same year, McNatt reported a 13-year-old synchronous multicentric PXA [11]. In 2012 Gardiman firstly reported a multicentric PXA with cerebrospinal dissemination to cauda equine [3]. It is impossible to confirm that the two lesions represent the same histopathological entity in our patient. But the intracranial lesion is similar to PXA by radiology, and the intracranial lesion was periventricular. We conclude that this case can be considered as multicentric tumor. To our knowledge, this is the first case with dissemination to cauda equine at first diagnosis in an aged patient.

### Conclusion

Though poor growth in biological behavior and relative favorable outcome, physicians should be realized multicentric dissemination by meninges or cerebrospinal fluid in PXA patients. It is important to describe the particular case in order to better understanding of clinical and radiological features.

### Disclosure of conflict of interest

None.

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