

Primary pleomorphic xanthoastrocytoma of the spinal cord: A case report and review of literature

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ABSTRACT

Primary pleomorphic xanthoastrocytoma (PXA) of the spinal cord is a rare slow growing tumor. To our knowledge, only five such cases have been reported in the literature till date. We report the clinical, radiological, and histopathological features of a spinal PXA in a 23-year-old female previously operated 5 years back for a spinal tumor, presented with weakness in lower limbs, sphincter incontinence and low back pain. Magnetic resonance imaging scan with contrast reveals an intramedullary lesion in the spinal cord from D8-D10 level. The patient was operated with reexploration of the previous incision, and gross total excision was achieved. Histopathology confirmed the diagnosis of PXA. Clinical and radiological follow-up is required to detect early recurrence. Adjunct radiotherapy or and chemotherapy should be considered only when there is postoperative residual or recurrence, however there are no definite guidelines in view of the rarity of this condition.

Key words: Pleomorphic, primary, spinal, xanthoastrocytoma

Introduction

Primary spinal pleomorphic xanthoastrocytoma (PXA) is a rare entity. PXA was first described by Kepes *et.al.*^[1] in 1979, in which he described 12 cases with distinct histopathological features which were termed as PXAs. This entity was added to the WHO classification of central nervous system tumors in 1993.^[2] PXAs are usually located in the temporal lobe followed by the cerebellum, Spinal location is very rare.^[1-5] They are slow growing lesions which usually affect young patients between the first and third decades of life.^[6]

Only five such cases of spinal PXAs have been reported in the literature, this is probably the sixth case in the literature [Table 1].

We present a very rare case of primary PXA of the thoracic spinal cord in a 23-year-old female presenting with weakness, sphincter incontinence and back pain.

Case Report

A 26-year-old housewife presented in June 2010 with gradual progressive weakness in both lower limbs and back pain (at the operative site) since last 3 months. The patient was able to perform her routine activities without support prior to the onset of these symptoms. Weakness has gradually progressed to the extent that at the time of presentation patient was not able to move her limbs and was bedridden. Back pain was not associated with fever, and there were no aggravating and relieving factors. Sphincter incontinence was present.

In the past surgical history, the patient was admitted in June 2005 with a history of gradual progressive weakness in both the lower limbs of 1-month duration. There was no sensory and sphincter dysfunction. The onset of weakness was not associated with back pain, fever or trauma. According to the medical charts, bulk and nutrition of the muscles were normal and tone was reduced in the lower limbs. However, the power was reduced to 2/5 (Medical Research Council Scale) at the hip and to 3/5 at the knee and ankle on the right side. The power in the left lower limb was 2/5 at the hip, knee and the ankle. The power of the extensor hallucis longus and the flexor hallucis longus was 3/5 on both the sides. Rest of the neurological examination was normal at that time. Magnetic resonance imaging (MRI) dorsolumbar spine demonstrated an intramedullary mass measuring 3.3 cm × 7 cm × 2.1 cm in the spinal canal at D9-D10 causing severe compression and anterior displacement of the spinal cord [Figure 1]. The lesion was isointense on T1-weighted, intermediate to hyperintense on T2-weighted and showed diffuse enhancement on gadolinium contrast, and the provisional preoperative

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diagnosis was intramedullary astrocytoma or ependymoma. The patient was operated by D9-10 laminectomy and subtotal total excision of the tumor was achieved. The spinal cord was expanded. A midline durotomy and myelotomy was done. Intra-operatively, the lesion was intramedullary, grayish white in color, soft in consistency, suckable, and vascular. Histopathological examination was consistent with a diagnosis of astrocytoma (WHO grade 2). In the postoperative period, there was improvement in power to 4/5 at all the joints in both the lower limbs. The patient was gradually mobilized with the Taylor's brace, and physiotherapy was continued. The patient was planned for the MRI thoracic spine at 6 weeks. However the patient lost to follow-up.

During the second admission in June 2010, neurological examination revealed that the tone was increased, and power was 0/5 in both the lower limbs. Deep tendon reflexes at all the joints in both the lower limbs were 3+ and planters were up going on both the sides. Sensory examination was normal. On local examination, operative scar was healthy, and there were no signs of inflammation.

Magnetic resonance imaging dorsolumbar scan revealed recurrent intramedullary tumor involving the entire spinal cord extending from D8-D10 levels, measuring 1.46 cm × 5.92 cm × 1.57 cm with hydromyelia superior to the lesion at D6 and D7 levels. The lesion was isointense on T1-weighted, mildly hyperintense on T2-weighted and heterogeneously hyperintense on postcontrast study [Figures 2 and 3].

The patient was operated upon with an exploration of the previous incision and D8 laminectomy. Intra-operatively, thick calcified postlaminectomy membrane adherent to the dura over the tumor was present which was excised. The tumor was intramedullary in location, firm, grayish white, vascular and not easily suckable. There were dense adhesions between arachnoid membrane and pia mater of the spinal cord at the level of tumor due to previous surgery. The tumor was firm in consistency, moderately vascular and diffusely infiltrated the spinal cord with ill-defined margins. In view of above findings, gross total excision of the tumor was achieved using ultrasonic aspirator.

Histopathology (hematoxylin and eosin stain) showed highly cellular and fibrillary background. Cellular areas consist of small cells with highly pleomorphic and hyperchromatic nuclei. Numerous multinucleated giant cells and few xanthomatous cells with foamy cytoplasm were present with no evidence of mitosis or necrosis. These features were suggestive of PXA (WHO grade 2) [Figures 4 and 5].

The patient's low back pain had improved immediately following the surgery. However, the power in the lower limbs and sphincter incontinence did not improve at the time of discharge.

At the follow-up of 1-year, the power was 1/5 with flickering movement of toes in both lower limbs and no improvement in sphincter functions.

Discussion

Pleomorphic xanthoastrocytoma of the spinal cord is a rare entity at very uncommon location.^[1-5] These tumors are thought to arise from the subpial astrocytes due to their superficial and subpial location.^[1,4-6] These tumors usually occur between 7 and 25 years of age, however older patients were also reported.^[6] Temporal lobe is the most common site followed by cerebellum. Only five cases of spinal cord origin have been reported in the literature. The demographic of the patients, location, clinical presentation, extent of resection, adjunct therapy, and follow-up are shown in Table 1. The most common clinical presentation of supratentorial PXAs are headaches and seizures.^[4,7] Retina being the other uncommon location reported.

Hematoxylin and Eosin stain of these tumors reveal pleomorphic and multinucleate giant cells. There are vacuolization and xanthomatous changes within these cells. Some cells may be fusiform with hyperchromatic features. Features of malignancy such as necrosis, mitotic figures and vascular proliferation are typically absent. Reticulin stain shows granular deposits between tumor cells and fibers around blood vessels. The tumor shows strong positivity for glial fibrillary acidic protein (GFAP).^[4,6,9] In our case the histopathological findings were similar to this classical description of PXA. Despite these relatively benign histopathological features, cases of anaplastic and aggressive variants with widespread cranio-spinal dissemination have been reported.^[10] Most of these tumors show reactivity with GFAP and S-100, on immunohistochemical analysis. Moreover, immunoreactivity of these tumors to synaptophysin and neurofilament protein ranges from 38-100% and 8-71% respectively.^[3,11]

On computed tomography scan, these lesions are hypo to isodense on plain scans and show enhancement with contrast. MRI typically reveals hypo to isointense on T1-weighted, hyperintense on T2-weighted and enhancement on gadolinium contrast.^[12] Our case showed similar features on MRI spine.

There are no definite management protocols for spinal PXAs due to the paucity of cases. According to Herpers *et al.*^[6] spinal PXAs are more aggressive as compared to PXAs at other locations. These tumors are astrocytic in origin with a favorable prognosis.^[3] They have a tendency to recur and 15-20% of the recurrent tumors undergo anaplastic transformation with less pleomorphism and more invasive in nature making complete resection difficult.^[2,5] Therefore complete resection of the primary tumor provides the best chances of cure with favorable prognosis. The extent of tumor resection and the mitotic index are the two most important predictors of tumor recurrence and prognosis.^[3]



Figure 1: Preoperative magnetic resonance imaging T2-weighted (sagittal view) images before the first surgery showing hyperintense lesion at D9-D10 level showing expansion of the spinal cord

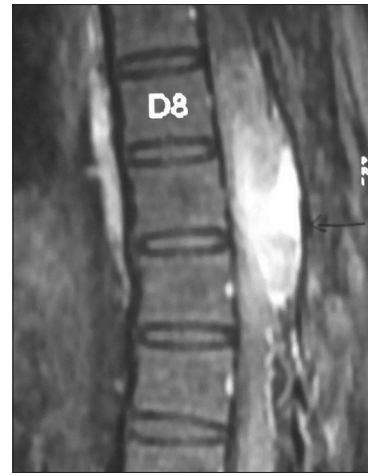


Figure 2: Preoperative magnetic resonance imaging scan before the second surgery, postgadolinium images (sagittal view) - showing heterogeneously enhancing lesion at D8-D10 level

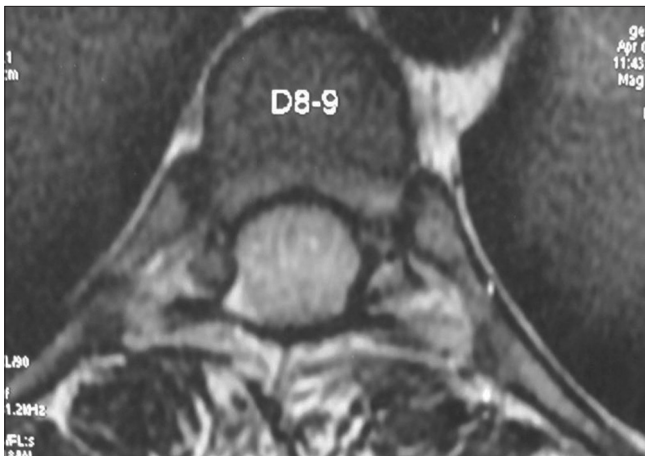


Figure 3: Preoperative magnetic resonance imaging scans before the second surgery T2-weighted (axial view) showing hyperintense lesion at D8-D10 level

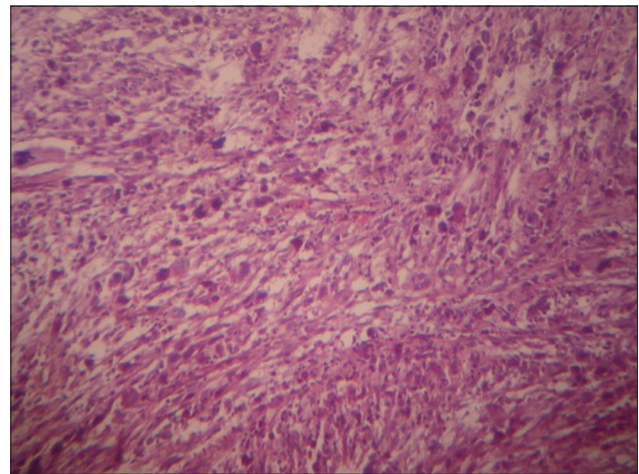


Figure 4: Low power view (x10): Histopathology (H and E) showing highly cellular and fibrillary background. Cells are highly pleomorphic with hyper chromatic nuclei. Multinucleate giant cells and few xanthomatous cells with foamy cytoplasm can also be seen. There is no evidence of mitosis or necrosis suggestive of pleomorphic xanthoastrocytoma

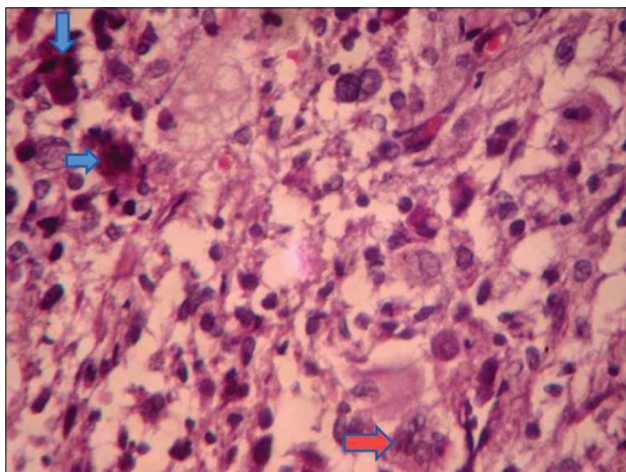


Figure 5: High-power view (x40) showing histological features mentioned in Figure 4 using H and E, Red arrow showing a multinucleate giant cell and blue arrows showing cells with hyperchromatic nuclei

Recommendations for adjunct therapy in cases of spinal PXAs are not available in the literature. Fouladi *et.al.*^[7] reported good outcome with prolonged disease control in 85% of the patients with gross total resection without radiotherapy. Some authors have recommended chemotherapy alone or chemotherapy with radiotherapy especially for malignant tumors with widespread leptomeningeal metastasis.^[4,9,12] However the results of these therapies for cranial PXAs cannot be extrapolated to spinal PXAs and further studies will be required to guide the management.

Follow-up of all the reported cases is shown in Table 1. Our patient had no recurrence till last follow-up of 12 months. The prognosis of spinal PXAs cannot be determined due to the paucity of cases.

Table 1: Demographics, presentation and management of spinal PXAs reported in the literature

Authors	Gender/age (years)	Location	Presentation	Management	Recurrence
Herperts <i>et al.</i> 1994 ^[6]	Male/66	Dorsal spine (D2-4)	Hypoesthesia (T1-12)	Complete excision with radiotherapy after 2 nd surgery	After 8 months
Fouladi <i>et al.</i> 2001 ^[7]	Male/12.9	Dorsal spine	Headache	Subtotal excision with radiotherapy	Died of disease at 22.7 months follow-up
Nakamura <i>et al.</i> 2006 ^[5]	Female/33	Cervical spine (C5-T1)	Hypoesthesia	Complete excision with no radiotherapy	No recurrence 3 years after surgery
Gill <i>et al.</i> 2010 ^[4]	Female/23	Dorsolumbar spine (D12-L2)	Weakness	Complete excision with no radiotherapy	No recurrence at 6 months follow-up
Simal-Julian <i>et al.</i> 2010 ^[8]	Female/60	Cervical spine (C4-5)	Weakness	Complete excision with no radiotherapy	No recurrence at 36 months follow-up
Present report	Female/26	Dorsal spine (D8-10)	Weakness	Subtotal excision with no radiotherapy	No recurrence at 12 months follow-up

PXAs – Pleomorphic xanthoastrocytomas

Therefore, spinal PXAs are rare tumors and complete excision is required for better survival and to prevent recurrence. Clinical and radiological follow-up is required to detect early recurrence. Adjunct radiotherapy or and chemotherapy should be considered only when there is postoperative residual or recurrence, however there are no definite guidelines in view of the rarity of this condition.

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