CASE REPORT

Juvenile Xanthogranuloma of adult spine: A rare case and review of literature

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ABSTRACT

Juvenile Xanthogranuloma (JXG) is a rare disorder of central nervous system. It rarely produces compressive myelopathy. On reviewing world literature, we could find only nine cases of this disease involving spine and of which only four cases were in adults' i.e., 18 years and above. We are presenting a case of Spinal JXG in an18-year-old male with thoracic compressive myelopathy presenting as short duration progressive paraparesis. Magnetic Resonance Imaging of Spine showed mass lesion in epidural space compressing cord from behind without any bony involvement at D7 to D10 vertebral segment. It was isointense on T1 and hyperintense on T2 with no contrast enhancement. D7 to D10 Laminectomy with complete excision of firm epidural mass was carried out. The histopathology with tumor markers confirmed the diagnosis of JXG. Post-operative neurological recovery in this patient was good. His power improved to grade 5/5 with decreased spasticity. Follow-up MRI at 3 months showed no residual tumor. This case appears to be the first in the series with entirely extradural component in adult thoracic spine.

Key words: Adult, compressive myelopathy, epidural, juvenile xanthogranuloma, langerhanscell histiocytosis, thoracic spine

Introduction

"Xanthogranulomatous tumors of Central Nervous System are rare. Juvenile Xanthogranuloma (JXG) is a histiocytic disorder of unknown etiopathogenesis belonging to the category of non-langerhans dendritic cell disorders and probably originating from dermal dendrocytes. It usually affects children in the first two decades of life and most commonly present as solitary cutaneous lesion followed-up by subcutaneous and deep soft-tissue mass in head and neck region and can show spontaneous regression (Presence of solitary extracutaneous soft-tissue lesion is very uncommon, study of all JXG in a large series) and described in children in variety of sites such as Temporal bone, Base of skull, Tongue, Nasal cavity, orbit etc. Among the extracutaneous sites CNS involvement is rare. In CNS the most common location is Choroid plexus of lateral ventricle, others being Hypothalamus, Pituitary gland and Duramater. Involvement of spine alone is rare."

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Case Report

"An18-year-old male presented with a short history of 8 days of weakness of both lower limbs. There was no history of trauma or tuberculosis or skin lesions in form of papules or macules. No other complaint was associated and the past history was of no significance. The cranial neurological examination was normal. His Bulk, tone, and power in both upper limbs were normal. On examination of both lower limb, his bulk was normal, had bilateral spasticity with muscle power of grade 4 in all groups of muscles. His deep tendon reflexes were exaggerated with bilateral extensor plantar. No sphincter involvement and all routine investigations including lipid profile were normal. Magnetic Resonance Imaging of Spine showed mass lesion in epidural space compressing cord from behind without any bony involvement atD7 to D10 vertebral segment. Mass lesion was isointense on T1 and hyper-intense on T2 with no contrast enhancement [Figure 1a, b].

D7 to D10 Laminectomy with complete excision of firm epidural mass, which could be separated from dura with difficulty, was carried out. Histopathology revealed fibro-collagenous tissue with dense infiltration of lymphocytes, histiocytes, plasma cells, eosinophils, and multinucleated Touton giant cells. Histiocytic granulomas were seen. Presence of arteritis and panniculitis noted. Overall morphology was suggestive of JXG. Immune marker study was proposed and revealed Viamentin positive, Cluster Differentiation-68 positive in Touton giant cell and negative for S-100, CD1a, and Actin smooth muscle [Figure 2a-e].

Post-operative, patient improved neurologically. His power improved to grade 5/5 with decreased spasticity. At 9 month

follow-up patient had mild spasticity with extensor plantar. His MRI carried out at 3 months shows post-Laminectomy status with no residual tumor [Figure 1c]."

Discussion

"JXG as initially described by Adamson,[1] is an uncommon histiocytic disorder in which the principal pathological cells are macrophages and dendritic cells.[2] JXG is one of the dendritic cell-related disorders, and has been classified as a non-Langerhans cell histiocytosis(LCH) disease together with several other histiocytic entities including papularxanthoma, benign cephalic histiocytosis, sinus histiocytosis with massive lymphadenopathy (Rosai–Dorfman disease), and hemophagocytichistiocytosis.[3] Exact etiology being unknown. Still, it is believed to result from a disordered macrophage response to a non-specific tissue injury, resulting in a granulomatous reaction.[4]

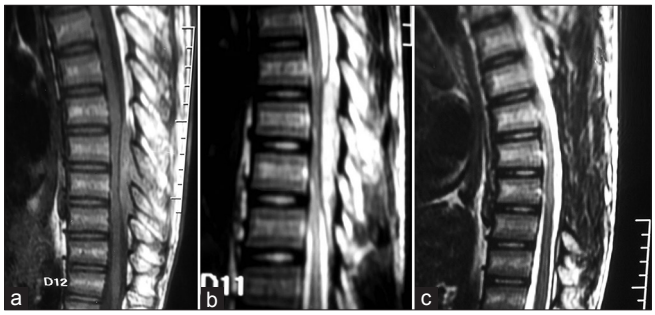

Figure 1: (a) Sagittal T1 weighted MRI scan, Isointense mass located epidurally posterior to cord from D7-10 with spinal cord compression. (b) Sagittal T2 weighted MRI scan, hyperintense mass located epidurally posterior to cord (pre-operative scan). (c) Sagittal T2 weighted MRI scan, post-operative scan with no features of residual tumor

JXG is essentially a cutaneous disorder, although, rarely, it has been described arising elsewhere. Extracutaneous involvement occurs in about 5-10% of all JXG cases. The most frequent extracutaneous site is the eye (eyelid, corneosclerallimbus, iris, optic nerve and disc), however, other rare sites include the heart, the lung, the testis, the oral mucosa, and CNS involvement is uncommon. Isolated cases of Xanthogranulomatous degeneration of choroid plexus have been reported, whereas involvement of spine is still rarer.

Classic JXGs are benign, usually asymptomatic, and solitary or multiple red-to-yellowish papules and nodules composed of histiocytic cells that predominantly occur in infancy and childhood, with self-limited course.[5] No spontaneous resolution ever reported in spinal involvement. In cases involving the spine, the clinical features were mostly related with the anatomical localization as slow-growing tumors in general hence, the symptomatology is gradual in onset. Search for spinal involvement yielded only nine cases of spinal JXG [Table 1].[2,4,6-12] Here, we report the tenth in the series and fifth case to be reported in adult spine. As reported, spine can be involved at any location but an overall predilection for thoracic spine can easily be noticed. Including our case 4/10 cases had thoracic spine involvement with two cases of lumbar and three of cervical spineand one case of sacral nerve root involvement reported. Spine involvement can be in any form ranging from nerve root involvement to bony involvement or intraduralextramedullary mass lesion. In our case, it was exclusively extradural without bony involvement, which was never reported previously. Neither MRI nor intraoperative finding of any hemorrhage in the tumor was found. So the exact cause for sudden deterioration could not be explained.

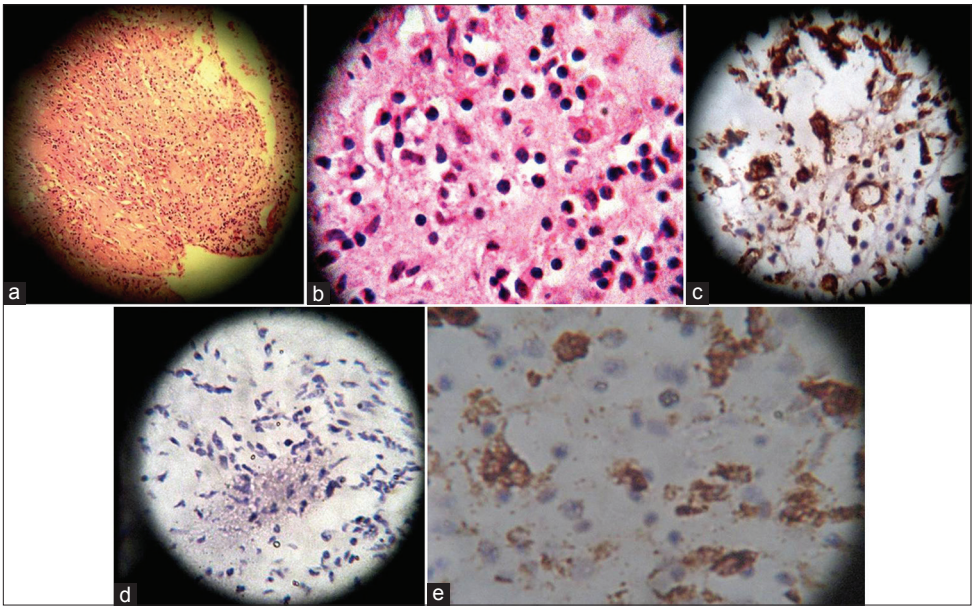

Figure 2: (a) H and E, in low power (×10). (b) H and E, in high power (×40) showing histiocytes, multinucleated giant cells, lymphocytes. (c) Immunohistochemical stain showing positivity for Viamentin. (d) Immunohistochemical stain showing negativity for S-100. (e) Immunohistochemical stain showing positivity for CD68

Table 1: Review of all cases of spinal Juvenile xanthogranuloma reported in literature

Case reported	Shimosawa *et al.* (1993)	Kitchen *et al.* (1995)	Kim *et al.* (1996)	Rampini *et al.* (2001)	Dehner (2006)	Cao *et al.* (2008)	Agabegi *et al.*. (2011)	Ayusijain *et al.* (2011)	Hirokazu *et al.* (2011)	Present case
Age/sex	13 m/F	15 year/F	16 m/M	34 m/F	14 year/F	18 year/F	47 year/F	22 year/F	38 year/M	18 year/M
Location	T6-T9 IDEM	S1 nerve root sheath	T1-T2 IDEM	C5-C7 IDEM	L3 Vertebral body	C2 nerve root sheath	L2 vertebral body with intradural extension	T7 vertebral body	C7 IDEM and extradural with paravertebral extension	T7-T10 Extradural dorsally located
Clinical features	Mild spastic paraparesis	Lower back and leg pain	Spastic paraparesis	Pain shoulder with spastic quadriparesis	Back pain	Intermittent back pain	Sphincter dysfunction, back pain, difficulty walking with diminished sensation	Progressive back pain	C8 radiculopathy with compressive myelopathy	Mild spastic paraparesis
Duration	NM	14 months	1 month	NM	NM	1 week	2-3 months	6-7 months	3 months	8 days
MRI	Hypointense on T1 and T2	Isoentense on T1 and hyperintense on T2	Isointense on T1 and hyperintense on T2	Isointense on T1andT2 with contrast enhancement	Collapse vertebral body with vertebrae plana	Iso to hypointense on T1andT2 with contrast enhancement	NM	Hypointense on T1 and T2	Isointense on T1 and T2 with contrast enhancement	Hypointense on T1 Hyperintense on T2 with no contrast enhancement
Histology	FC+TGC+IHC: Vim+S100-Lysozyme+ ACT+EM: no birbeck granules	FC+TGC+IHC: HAM56+S100-	FC+TGC–IHC: CD68+S100-Lysozyme+ EM: no birbeckgranules	FC+TGC+IHC: Vim-S100-CD1a-, CD68+	FC-TGC+OCGC+ IHC: Vim+S100-CD68+CD1a-factorXIIIa+	FC+TGC+IHC: Vim+CD68+ CD163+	FC+TGC+OCGC+ IHC: Vim+CD68 focal S100	FC+TGC+ OCGC+IHC: Vim+S100-CD68+CD1a-	FC+TGC+IHC: KL-1-, desmin-, ALK-1-, EMA-, CD34-, GFAP-, S-100-, CD68+	FC+TGC+IHC: Vim+S100-CD68+SMA-CD1a-
Intervention	Laminectomy with TR	L5 with TR	Laminectomy with TR	Laminectomy with TR	NM	Laminectomy with TR	Laminectomy with PR	Laminectomy with TR	Laminectomy with TR	Laminectomy with TR
Follow-up	Improved at 6 months	Complete resolution	Recovered in 3 months	Resolution by 4 months	NM	No recurrenceat 2 year	Post-op RT, improved clinically at 8 months	Improved post operatively with no recurrence at 5 month	Improved post operatively with no recurrence at 2 years	Clinical and radiological improvement at 3 month follow-up

ACT1 – Antichymotrypsin; EM – Electron microscopy; F – Female; FC– Foam cells; IHC – Immunohistochemistry; M – Male; NM – Not mentioned; OCGC – Osteoclastic giant cells; PR – Partial resection; RT – Radiotherapy; SMA – Actin smooth muscle; TGC – Touton giant cell; TR – Total resection; Vim – Vimentin; IDEM – Intradural extramedullary

MRI is the best method for obtaining details of the localization of the tumors and their relation to adjacent structures. The lesion may appear hypo-intense, iso-intense, or slightly hyper-intense in T1WI and T2WI. Homogeneous enhancement with gadolinium was observed in most cases. However, such contrast uptake was absent in our case. Because of this array of presentation, radiology apart from localization offers a little help in achieving at a diagnosis of these lesions. JXG is also difficult to distinguish intra-operatively with other tumors of neural origin (e.g. schwannoma, neurofibroma, nerve sheath myxoma, and malignant nerve sheath tumor), epidural lesions like lymphoma, plasmacytoma, metastasis, and other granulomatous conditions like tubercular, fungal. Therefore, the pathological and immunohistochemical studies remain the golden standard for achieving a diagnosis of JXG.

Tumor is typically grayish to yellowish in appearance. On microscopy, it consists of cellular component ranging from mononuclear cells, multinucleated cells with or without Touton features, and spindle cells. With bony involvement, osteoclast like giant cells can also be seen.[13]

Immunohistochemistry plays a key role in diagnosis. Cellular components of JXG shows consistent imunoreactivity to Viamentin, CD68, CD163, fascin, CD14 and factor XIIIa and non-reactive for CD1a and S-100 protein.[4,13-15] LCH need to be considered in differential diagnosis and to be excluded at earliest. As disease course is more aggressive and prognosis guarded in case of LCH. Immunohistochemically Langerhans cells are S-100 and CD1a positive and electronmicroscopically defined by presence of Birbeck granules (pentilaminar cytoplasmic Inclusions).[8,16]

No treatment protocol still available for the management of JXG. Spontaneous regression of the skin lesions is the natural course, however, in cases involving the spine, there is no regression documented so far. Total excision is the recommended method forfavorable outcome. This was also documented in follow-up of this case till date. The recurrence of the tumor is unlikely after total resection. In few reported cases partially excised cases were post-operatively subjected to adjuvant radiotherapy with favorable outcome. As recommended by Cao *et al*., follow-up and close medical observation can also be opted after a subtotal resection.[2] And if the symptom recurs, a second operation may be appropriate."

Conclusion

JXG of spine is a rare condition and can mimic in presentation similar to any Extradural or Intradural extramedullary mass lesion causing Compressive Myelopathy, making it an important consideration in differential diagnosis. Though MRI is very helpful in tumor localization, exact pathology can be defined only by histopathology and immunohistochemistry. Surgical excision provides a good neurological outcome. Complete excision of the tumor is advisable to prevent recurrence.

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