Case Report
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MRI Findings of Juvenile Xanthogranuloma of the Spinal Cord: A Case Report[1]

척수에 발생한 연소성황색육아종의 자기공명영상 증례 보고[1]

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Juvenile xanthogranuloma (JXG) is a proliferative histiocytic disorder experienced during childhood and adolescents. JXG commonly presents as a solitary cutaneous lesion. Despite the term "juvenile", development of the disease during adulthood is possible, although spinal JXG is extremely rare in adults. We describe a 67-year-old female patient who presented with an intradural-extramedullary (IDEM) tumor of the spinal cord. Magnetic resonance imaging (MRI) findings indicative of JXG of the spinal cord were seen, which was then confirmed pathologically. A lumbar spinal MRI with contrast enhancement showed an oval-shaped, well-defined IDEM tumor at the L1 level. This tumor had mixed signal intensity on the T1-weighted image and high signal intensity on the T2-weighted image. Central homogenous enhancement was observed after contrast administration.

Index terms
Juvenile Xanthogranuloma
Spinal Cord Neoplasm
Magnetic Resonance Imaging

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INTRODUCTION

Juvenile xanthogranuloma (JXG) is a proliferative histiocytic disorder experienced during childhood and adolescents, and most commonly presents in the first two decades of life (1). JXG is not a true neoplasm, but rather a reactive proliferation of histiocytes, and belongs to the category of non-Langerhans dendritic cell disorders. However, the etiology and pathogenesis of JXG are still unknown. Most cases present as a solitary cutaneous lesion followed by a soft tissue mass in the head and neck region. Extracutaneous JXG, especially when involving only the spinal column, is extremely rare in adults. Through a review of the literature, we identified only three previous cases, which were in a 38-year-old male, 29-year-old male, and 41-year-old female (2-4). We describe a 67-year-old female who presented with an intradural-extramedullary (IDEM) tumor of the spinal cord. To our knowledge, this case report represents the oldest patient with a new presentation of spinal JXG. We report magnetic resonance imaging (MRI) findings of JXG of the spinal cord, which were then pathologically confirmed.

CASE REPORT

A 67-year-old female presented with a complaint of lower back pain with a tingling sensation in the left leg running from her left thigh to left calf of two months in duration. She had no other complaints. Her only significant past medical history was controlled hypertension. General physical and neurologic examination did not reveal any abnormalities apart from slight hyperactivity of the deep tendon reflex in both knee jerks. Muscle strength in all limbs, gait, and co-ordination were normal, and there was no bowel or bladder dysfunction. Cranial nerve ex-

amination and higher executive functions were normal. Lumbar spinal MRI with contrast enhancement showed an approximately 1.7 × 0.9 cm oval-shaped well-defined IDEM lesion at the L1 level. This lesion exhibited mixed signal intensity on the T1-weighted image (WI) and high signal intensity on T2-WI. Central homogenous enhancement was observed after contrast administration (Fig. 1).

The patient underwent a left hemilaminectomy at L1 and a partial hemilaminectomy at T12 for removal of the tumor. A yellowish soft mass was located in the intradural and extramedullary areas, and a total tumor resection was performed.

Histologic review demonstrated a collection of mononuclear cells with dilated blood vessels and areas of focal hemorrhage. The mononuclear cells were moderate to large round cells with small and uniform nuclei and lipid-laden cytoplasm (Fig. 2A). The cells stained positively for CD68, a histiocyte marker (Fig. 2B). The pathologic diagnosis was JXG.

Postoperatively the patient's pain and tingling sensation were relieved. A postoperative MRI obtained at 1 and 3 years after surgery did not reveal any residual or recurrent tumor.

DISCUSSION

JXG is the most common form of non-Langerhans cell histiocytosis. The exact etiology or pathogenesis of JXG has not been determined, although it is regarded as a reactive process rather

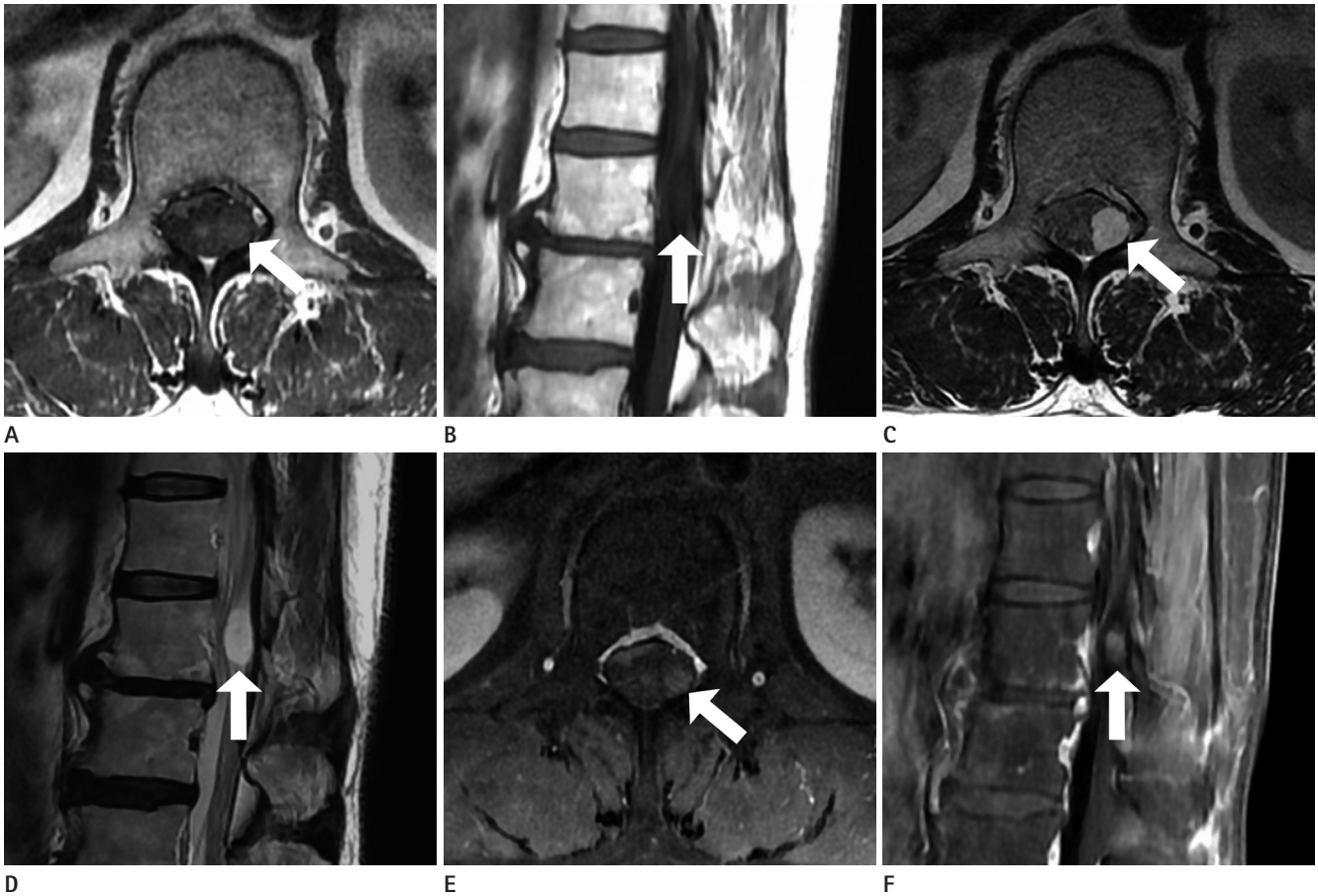
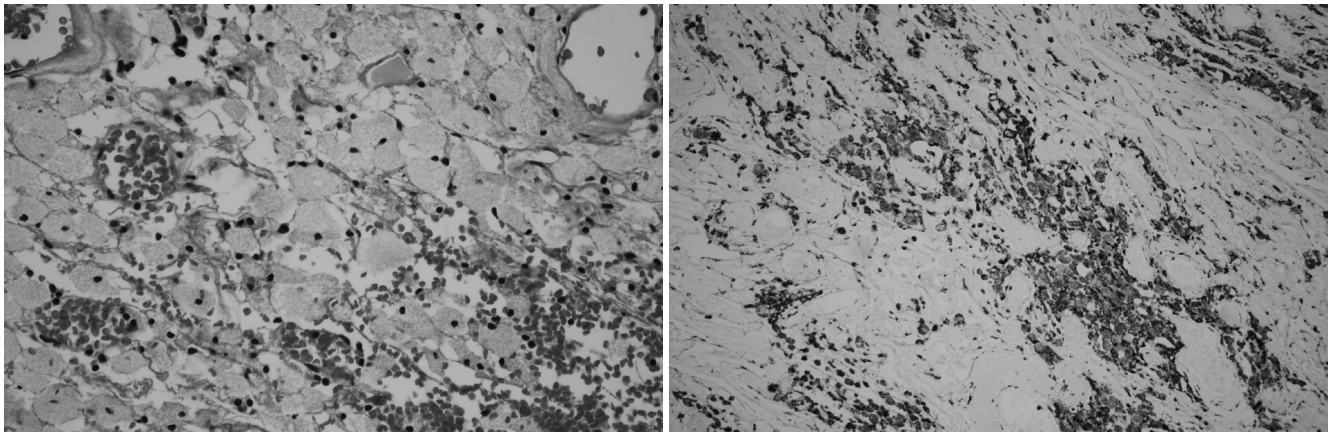


Fig. 1. Lumbar spinal magnetic resonance imaging with contrast enhancement shows a 1.7 × 0.9 cm oval-shaped, well-defined intradural-extramedullary tumor at the L1 level.
A, B. Fast spin echo axial T1-weighted image (TR 758, TE 22) and sagittal T1-weighted image (TR 786, TE 12) shows an oval-shaped lesion with mixed signal intensity (arrow).
C, D. Fast spin echo axial T2-weighted image (TR 2000, TE 100) and fast spin echo sagittal T2-weighted image (TR 2765, TE 100) shows a well-defined oval-shaped lesion with high signal intensity (arrow).
E, F. Contrast-enhanced fat-suppressed fast spin echo axial T1-weighted image (TR 842, TE 18) and contrast enhanced fat-suppressed fluid attenuated inversion recovery sagittal T1-weighted image (TR 668, TE 13) shows central homogenous enhancement (arrow).
Note.—TE = the value of the echo time, TR = the value of the repetition time



A B

Fig. 2. Histologic review of the juvenile xanthogranuloma.
A. Microphotograph of the lesion. The lesion is composed of foamy macrophages with a hemorrhagic background. The macrophages contain cytoplasmic lipids (H&E stain, × 400).
B. Cells stained positive with CD68 stain, which is a histiocyte marker (CD68, immunohistochemistry, × 400).

than as a neoplasm. The incidence of JXG has not been estimated, but may be higher than is generally recognized as JXGs occur early in life and usually resolve spontaneously. Based on an autopsy series, Ayres and Haymaker (5) suggested that the incidence varies from 1.6% to 7% of the general population. Despite the use of the term "juvenile", development of this disease during adulthood is possible, and generally occurs in the late 20s to early 30s (6). Predominance in males has been reported in childhood cases, but there is no difference by sex in adults. JXG typically presents as cutaneous lesions. Extracutaneous involvement is not common, with an incidence of only 5%. The eye is the most common site of extracutaneous involvement, and other involved organs include the oropharynx, heart, lung, liver, spleen, adrenal glands, muscles, subcutaneous tissues, and the central nervous system (7, 8). Solitary JXG involving the spinal column is extremely rare in adult patients. Thus far, only three cases have been reported in the English-language literature. Two of the reported cases involved the cervical spine and one was a lumbosacral lesion. The symptoms in all cases were a result of cord compression by the lesion (2-4).

MRI has been demonstrated to be the best imaging modality for the localization of spinal tumors and evaluation of their correlation with adjacent structures. The differential diagnosis contains some IDEM tumors, including spinal ependymoma, astrocytoma, and schwannoma. Spinal ependymoma and astrocytoma can be considered in the location of the lesion. Ependymoma is a tumor that arises from the ependyma, a tissue of the central nervous system. Generally, in pediatric cases the location is intracranial, while in adults it is spinal. Their occurrence seems to peak at age 5 years and then again at age 35. Ependymomas are generally hyper-intense on T2-WI and hypo-intense on T1-WI, which is often heterogeneous. Ependymomas nearly always show contrast enhancement, though are not always homogenous (9). Astrocytomas are the most common glioma and can occur in most parts of the brain and occasionally in the spinal cord. People can develop astrocytomas at any age. Astrocytomas are iso- to slightly hypo-intense on T1-WI, hyper-intense on T2-WI, and commonly have associated cysts. They enhance less intensely and are more eccentric than ependymomas (10). Schwannomas can be considered in the shape of the lesion. Schwannomas frequently affect the eighth cranial nerve most commonly. Less commonly, schwannomas occur in other nerves that contain Schwann cells. These tumors are most common among people who are 50 to 60 years of age. Schwannomas are generally hypo or iso-intense on T1-WI and hyper-intense on T2-WI which is often heterogeneous, and are frequently associated with hemorrhage, intrinsic vascular changes, cyst formation, and fatty degeneration. The location of the spinal JXG is variable from the cervical spine to the sacrum. The age spectrum of the adult spinal JXG is also wide from twenty nine to sixty seven. Spinal JXG may appear with variable signal intensity, that is, a mixture of hypo-, iso-, and hyper-intense in T1-WI and T2-WI. Furthermore, the lesion may exhibit homogenous enhancement after contrast media administration (Table 1).

Table 1. Four Cases of Xanthogranuloma of the Spinal Canal Reported on World Literature Previously

Location	Author	Year	Age/Sex	Size (cm)	Shape	MRI			Ref.
						T1	T2	Cont.	
L1	Present	2012	67 yr/F	1.7 × 0.9	Oval-shaped	Mixed	Hyper	Homo	
C1-2	Lee et al.	2012	29 yr/M	-	Dumbbell-shaped	Iso	Hypo	Hyper	4
C8 root	Inoue et al.	2011	38 yr/M	2.5 × 2.4	Round-shaped	Iso	Iso	Homo	2
Cauda	Iwasaki et al.	2001	41 yr/F	-	Diffuse enlarged mass filling the spinal column	-	-	Hyper	3

Note.—Cauda = cauda equina, Cont. = contrast enhancement, Ref. = reference

Upon gross anatomic examination, the JXG is a well-encapsulated and round lesion with a yellowish surface. JXG is confirmed by the microscopic pathologic identification of foamy histiocytic cells and an immunohistochemical finding of mononuclear cells, giant cells, and spindle cells positive for lysozyme stain and CD68 but negative for CD1a and S-100 proteins, which are all reactive markers of Langerhans cells (Fig. 2B).

These tumors may grow slowly without regression, and thus the symptoms worsen gradually. This is the distinguishing and important characteristic of JXG in adult patients compared with that found during childhood and adolescent JXG, which regresses spontaneous. Despite being a pathologically benign tumor, it is conventional to remove as much of the tumor as possible. There have been no reports of recurrence after total mass excision (4).

We have described the case of a 67-year-old female with a solitary tumor identified as a JXG involving the spinal column. Although JXG is very rare, JXG should be considered in the differential diagnosis of IDEM tumors of the spinal cord in adult patients.

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척수에 발생한 연소성황색육아종의 자기공명영상 증례 보고[1]

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연소성황색육아종(Juvenile xanthogranuloma; 이하 JXG)은 유소년기에 발생하는 조직구 증식성 질환이다. JXG는 주로 단독 피부병변으로 나타난다. 그러나 "연소성"이라는 명칭에도 불구하고, 성인에게서 발생할 수 있다. JXG가 피부 이외의 병변으로 나타나는 경우는 드물며, 특히 성인에게서 척수에 발생하는 경우는 극히 드물다. 저자들은 67세 여성에서 척수에 발생한 경수막내-수외의 종양을 발견하였다. 이 병변은 조직학적으로 확진되었으며, 이 환자에 대한 자기공명영상의 증례를 보고한다. 조영증강 자기공명영상에서 1번 요추 높이에서 경계가 분명한 타원형의 경수막내-수외의 종양을 볼 수 있었으며, 이 종양은 T1 강조영상에서 혼합된 신호강도를 보였고, T2 강조영상에서 고신호강도를 보였다. 조영증강시에는 종양 중앙부분에서 균질한 조영증강을 보였다.

성균관대학교 의과대학 강북삼성병원 [1]영상의학과, [2]병리과