

Isolated Juvenile Xanthogranuloma in Thoracic Spine: Intraoperative Cytological Diagnosis of a Rare Presentation

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Abstract

Juvenile Xanthogranulomas (JXG) are benign proliferative disorders of non-Langerhans histiocytes, which present in children as multiple, self-limited, cutaneous lesions. The extracutaneous manifestations of JXG are uncommon, and isolated JXG involving the spinal column is extremely rare.

We report here a case of isolated juvenile xanthogranuloma in thoracic spine correctly diagnosed intraoperatively on crush smear cytology and later confirmed by histopathological and immunohistochemical studies.

This case report draws attention to the fact that isolated xanthogranuloma should be considered among possible diagnoses of spinal tumor in children. Also, since the long term survival depends on complete surgical resection, a correct intraoperative diagnosis is extremely important for optimal management and prognosis of the patient.

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INTRODUCTION

Juvenile Xanthogranuloma (JXG) is a benign proliferative disorder of non-Langerhans histiocytes, occurring in vast majority of cases as multiple, self-limited, cutaneous lesions of children in the first two decades of life [1]. Extracutaneous manifestations of JXG are uncommon, and isolated JXG involving the spinal column is extremely rare [2]. We hereby report the clinicopathological features of a 9 month old boy and characterize a case of isolated JXG in the thoracic spine. This case report draws attention to the fact that isolated xanthogranuloma of the central nervous system should be considered among possible diagnosis of subdural extramedullary spinal masses in children and young adults.

CASE REPORT

A 9 month old boy presented in neurology outpatient department of our hospital with complaints of weakness in both lower limbs for past fifteen days.

Physical examination of the patient revealed that the child was unable to sit without support of his hands. There was no other positive finding on neurological examination. The CT scan of skull showed no significant abnormality and MRI demonstrated a 16x14 mm intradural extramedullary homogenous solid space occupying lesion measuring on left side of spinal cord at C4 and C5 levels causing displacement, compression and edema of the cord.

Intraoperative crush smears were prepared from the tumor by placing a tiny portion (1-2 mm³) of tissue between two glass slides and smearing it. These smears were fixed in 95% ethanol and stained with rapid Hematoxylin and Eosin (H&E). The smears revealed multinucleated Touton giant cells and foamy histiocytes containing abundant delicate cytoplasm, well defined round nuclei and hazy cell borders in a background of spindle cells (Figure 1a). No atypical histiocytes, necrosis or mitoses was observed. Since no epithelioid cells were identified and none of the histiocytes showed folded, lobulated or kidney shaped

nucleus, the differential diagnosis of Langerhans cell histiocytosis was ruled out and a diagnosis of Juvenile Xanthogranuloma was suggested. This guided the surgeons to perform a gross total excision of the tumor.

The resected tumor specimen was fixed in formalin and sent for histopathological examination. On microscopic examination of paraffin embedded sections, the tumor was composed of Touton giant cells and large number of foam cells (histiocytes), in a background of spindle cells, eosinophils, lymphocytes and macrophages (Figure 1b). These histological features were similar to

those seen on cytology. The tumor was labeled as Juvenile Xanthogranuloma after looking into all these features.

Immunohistochemically the histiocytes and giant cells were strongly positive for CD68 (Figure 1c), but negative for S100 and CD1a. These features further confirmed our intraoperative diagnosis of Juvenile Xanthogranuloma.

The postoperative course of the patient was uneventful and the patient remains asymptomatic one year after surgery.

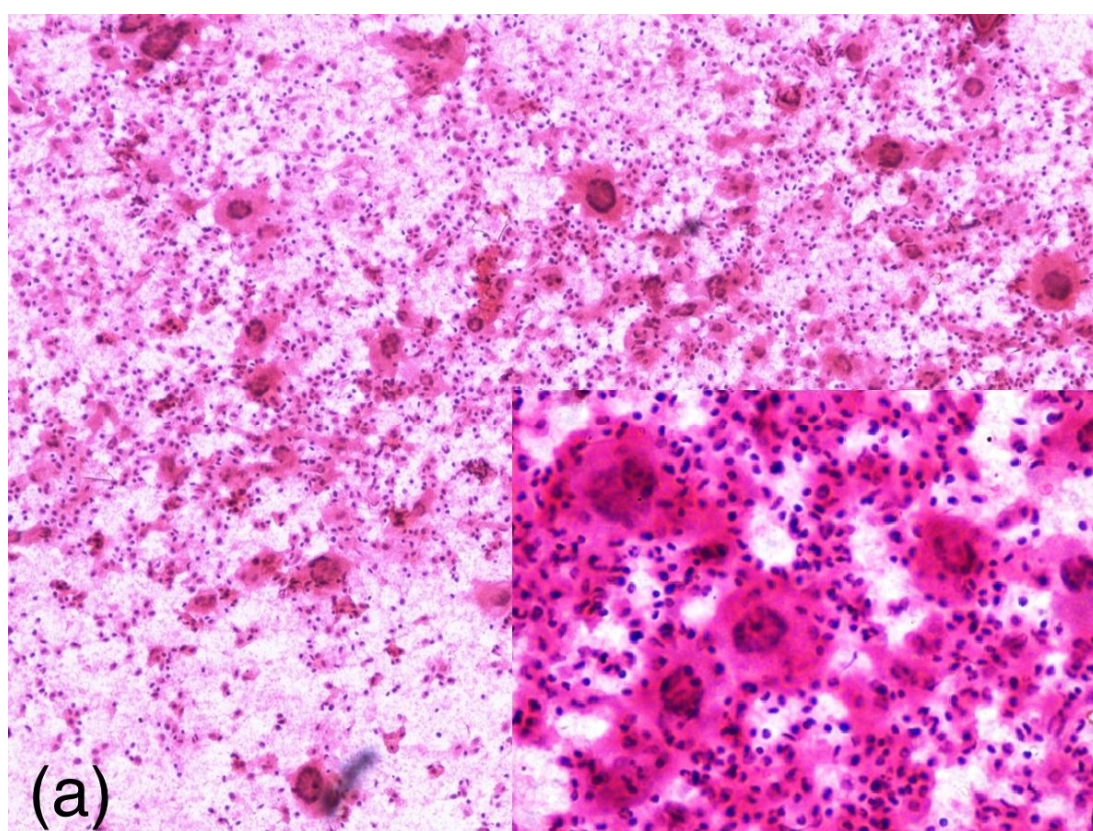


Figure 1a: Intraoperative crush smear from space occupying lesion in thoracic spine showing multinucleated Touton giant cells and clusters of foamy histiocytes in a background of inflammatory cells, suggestive of xanthogranuloma (H&E, x100); high power view in inset (H&E, x400).

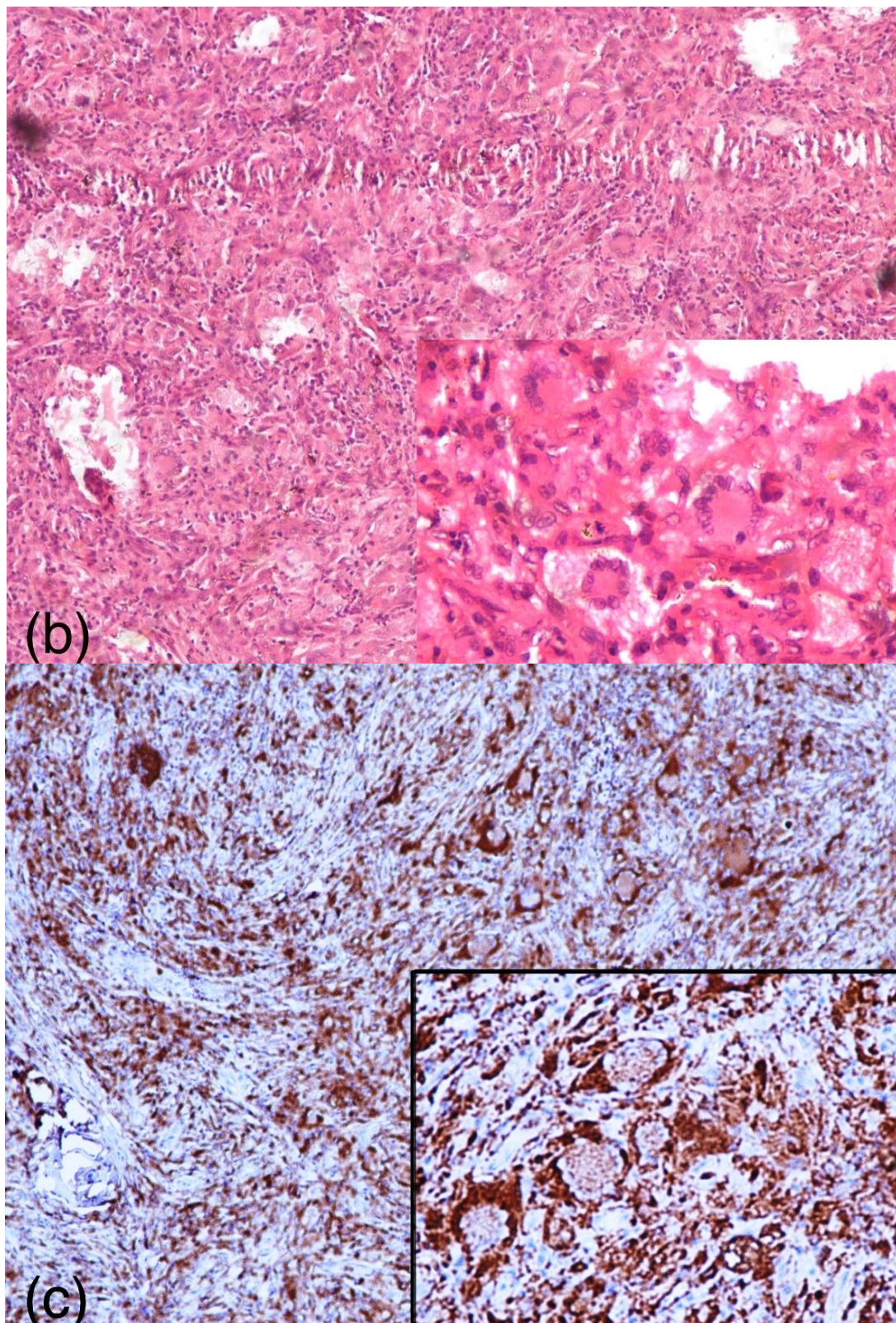


Figure 1 b-c: [b] Juvenile xanthogranuloma showing Touton giant cells and histiocytes with mononuclear cells in background (H&E, x100); high power view in inset (H&E, x400). [c] CD68 positive multinucleate giant cells and histiocytes (CD68, x100); high power view in inset (CD68, x400).

DISCUSSION

Juvenile Xanthogranuloma (JXG), the most common form of non-Langerhans cell histiocytosis, primarily is a self-limited dermatologic disorder that is associated rarely with systemic manifestations. Infants and small children are mainly affected [3]. The etiology is unknown and the tumors represent accumulations of differentiated histiocytes of non-Langerhans' cell type.

The most frequent occurrence of the JXG lesion is on the skin of head and neck. Extracutaneous involvement has been reported in only about 5-10% of all JXG cases. The eye, particularly the uveal tract, is the most frequent site of extracutaneous involvement, followed by the oropharynx, heart, lung, liver, spleen, adrenals, muscles, subcutaneous tissues, and the central nervous system, but the involvement of spine is extremely rare [4].

In all xanthogranuloma cases involving the spine, the clinical features are mostly related with the anatomic localization as slow-growing tumors in general [5]. On MRI, the lesion may appear hypointense, isointense, or slightly hyperintense in T1WI and T2WI [5].

Clinically JXG is difficult to distinguish intraoperatively from other tumors of neural origin (e.g., schwannoma, neurofibroma, nerve sheath myxoma, malignant nerve sheath tumor). Therefore, the pathological and immunohistochemical studies remain the gold standard for achieving a diagnosis of JXG [5].

Limited literature is available on the cytological features of JXG. Grenko et al first reported the cytologic features of cutaneous JXG in 1996 [6]. According to Li et al, cutaneous JXG can be diagnosed by its Fine Needle Aspiration Cytology (FNAC) and immunocytochemistry findings [7].

The characteristic cytological features of JXG include presence of numerous finely vacuolated histiocytes, eosinophils, multinucleated giant cells and scattered Touton giant cells [6]. There are two cytological findings that enable pathologists to discriminate between JXG and Langerhans cell histiocytosis (LCH) [8]. On meticulous examination, nuclear grooves in histiocytic cells [6] and the presence of epithelioid cells points to a diagnosis of LCH, whereas foamy cells and the absence of necrotic cellular debris or mitotic figures are indicative of JXG [8]. This was very well demonstrated in our case.

Grossly, the tumor is typically a round lesion, with a yellowish to grayish appearance on the cut surface. Microscopically, a diffuse and/or nodular pattern of growth is apparent at low magnification. As in our case, the typical cellular composition of the lesion consists one or more of the three basic cellular types:

mononuclear cells, multinucleated cells with or without Touton features, and spindle cells [9].

Immunohistochemistry has an important role in the diagnosis of JXG. Regardless of the cellular composition of a JXG, the mononuclear cells, giant cells, and spindle cells are consistently immunoreactive for CD68 [9]. And in most cases, S-100 protein is also non-reactive, but scattered cells may show weak cytoplasmic reactivity, unlike the more diffuse and intense reaction of Langerhans cells [5]. However, CD1a is consistently negative and an important diagnostic criterion for distinguishing JXG from LCH [10]. The pathological differentiation of a lesion as LCH or non-LCH is of considerable clinical importance, since unlike the majority of non-Langerhans histiocytoses, LCH is associated with considerable morbidity and mortality, often requiring aggressive therapy [5].

Currently, there is no standard treatment for JXG. The severity and location of the lesion dictates the course of treatment. Spontaneous regression of the skin lesions is the natural course, but in cases involving the spine, there is no spontaneous regression documented so far. Whenever total surgical resection alone is possible, it seems to be curative [4]. The recurrence of the tumor is unlikely after the total resection, and neurological functions can be well preserved if no damage to the neural structures had been done during tumor resection.

To conclude, JXG has characteristic diagnostic cytological features and this case has been presented to highlight the importance of cytology in intraoperative diagnosis of this very rare lesion. Since the long term survival depends on complete surgical resection, a rapid and correct intraoperative diagnosis is very important for optimal management and prognosis of the patient.

CONFLICTS OF INTEREST

Authors declare that there are no conflicts of interest.

REFERENCES

1. George DH, Scheithauer BW, Hilton DL, Fakhouri AJ, Kraus EW. Juvenile Xanthogranuloma of peripheral nerve: a report of two cases. *Am J Surg Pathol* 2001; 25:521-526.
2. Dehner LP. Juvenile Xanthogranuloma in the first two decades of life. *Am J Surg Pathol* 2003; 27(5):579-593.

3. Okubo T, Okabe H, Kato G. Juvenile Xanthogranuloma with cutaneous and cerebral manifestations in a young infant. *Acta Neuropathol* 1995; 90(1):87-92.
4. Rampini PM, Alimehmeti RH, Egidi MG, Zavanone ML, Bauer D, Fossali E, Villani RM. Isolated Cervical Juvenile Xanthogranuloma in Childhood. *Spine* 2001; 26(12):1392-1395
5. Dong C, Junming M, Xinghai Y, Jianru X. Solitary juvenile xanthogranuloma in the upper cervical spine: case report and review of the literature. *J Eur Spine* 2008;17 suppl 2: 318-323.
6. Bandyopadhyay A, Gangopadhyay M, Chakraborty S. Juvenile xanthogranuloma-diagnostic challenge on fine-needle aspiration cytology. *J Cytol* 2011;28:217-218.
7. Li HC, Roux JJ, Pranikoff T, Kilpatrick SE, Cappellari JO. Deep juvenile xanthogranuloma presenting as a chest wall mass: a case report. *Acta Cytol* 2008;52:607-611.
8. Fassina A, Olivotto A, Cappellesso R, Vendraminelli R, Fassan M. Fine-needle cytology of cutaneous juvenile xanthogranuloma and langerhans cell histiocytosis. *Cancer Cytopathol* 2011; 119(2): 134-140.
9. Lesniak MS, Viglione MP, Weingart J. Multicentric parenchymal xanthogranuloma in a child: case report and review of the literature. *Neurosurg* 2002; 51:1493-1498.
10. Jain A, Mathur K, Khatri S, Kasana S, Jain SK. Rare presentation of juvenile xanthogranulom in the thoracic spine of an adult patient: case report and literature review. *Acta Neurochir* 2011; 153:1813-1818.

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