Spontaneous regression of infantile dural-based non-Langerhans cell histiocytosis after surgery: case report

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The authors report the case of a large left occipital mass lesion in an 8-month-old boy who presented with seizure. Neuroimaging demonstrated an approximately 5-cm extraaxial tumor, and the patient underwent partial resection. The tumor was strongly attached to the tentorium and falx. In the postoperative course the residual lesion regressed spontaneously, and after 5 years only a slight residual tumor remained along the tentorium. Histopathological examination of the tumor revealed non-Langerhans cell histiocytosis (non-LCH). However, the tumor was not diagnosed as juvenile xanthogranuloma (JXG) because it lacked Touton giant cells. Hence, the authors described this lesion as a fibroxanthogranuloma.

Most intracraniospinal non-LCHs have been reported as JXG; however, several cases of xanthomatous tumors with histopathological features resembling those of JXG have been described as fibrous xanthoma, xanthoma, fibroxanthoma, and xanthogranuloma. Among JXG and the xanthomatous tumors, a review of the literature revealed several cases of dural-based tumors; these dural-based tumors have had favorable courses, including the case described in this report. In addition, the patient in the present case experienced spontaneous regression of the residual tumor. The authors report this unique case and review the literature on isolated intracraniospinal non-LCHs, especially in cases of dural-based lesion.

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KEY WORDS non-Langerhans cell histiocytosis; dural-based lesion; spontaneous regression; infant; oncology

ISOLATED intracraniospinal non-Langerhans cell histiocytoses (non-LCHs) without cutaneous lesion are rare.^{5,10,11,14,19} The most frequently reported lesion in these cases is juvenile xanthogranuloma (JXG), with several cases reported as fibrous xanthoma, xanthoma, fibroxanthoma, and xanthogranuloma (XG).^{1–3,5–9,11–18,21–39,41–44} Most reported intracraniospinal non-LCHs have been pediatric cases, and histopathological investigations show that they are composed mainly of xanthomatous histiocytes. In addition, several cases of intracraniospinal non-LCHs presented with dural-based lesions. We present an 8-month-old boy with isolated non-LCH attached to the dura mater in the left occipital region. Interestingly, the residual tumor regressed without treatment after surgery. We report this unique case and review the literature on isolated intracraniospinal non-LCHs, especially in cases of dural-based lesion.

Case Report

History and Examination

An 8-month-old boy who had experienced seizures with loss of consciousness and cyanosis at the age of 5 and 7 months was admitted to our hospital due to convulsive status. His growth and development had been normal, including head circumference. He had no cutaneous lesions. Physical examination and chest radiographs showed no definite abnormality. Head CT scans demonstrated a mass lesion approximately 5 cm in diameter in the left occipital region. Portions of the lesion showed high radiographic attenuation, and there was strong medial and rim enhancement (Fig. 1). The cerebral parenchyma was compressed but surrounding edema was mild. Magnetic resonance imaging showed hyperintense areas in the medial portion and rim of the tumor on T1-weighted studies (Fig. 2A).

ABBREVIATIONS α -SMA = α -smooth muscle actin; EMA = epithelial membrane antigen; GFAP = glial fibrillary acidic protein; JXG = juvenile xanthogranuloma; LCA = leukocyte common antigen; LCH = Langerhans cell histiocytosis; NFP = neurofilament protein; XG = xanthogranuloma.

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FIG. 1. Preoperative CT scans. Plain CT scan (**left**) demonstrating a mass lesion with an approximately 5-cm diameter in the left occipital region. Enhanced head CT scan (**right**) showing a well-enhanced area in the medial portion and rim of the mass.

The mass demonstrated heterogeneous hypointensity on T2-weighted imaging (Fig. 2B). Diffusion-weighted imaging (Fig. 2C) showed hypointense areas, suggesting low cell density. The mass showed homogeneous enhancement contiguous with the dura mater, resembling meningioma (Fig. 2D and E).

Operation

The patient underwent tumor resection by left occipital craniotomy. The tumor was yellow colored, and strongly attached to the tentorium and falx; its consistency was very hard, and it was difficult to debulk even using an ultrasonic

aspirator. Considering the surgical stress on the infant, we performed a partial (approximately 60%) resection, and a staged operation was planned at that time.

Histological Examination

Macroscopically, the tumor was colored white-gray with areas of yellow, and appeared to arise from the cranial dura mater of the left occipital region.

On microscopic examination (Fig. 3), the tumor revealed abundant xanthomatous cells that had clear or slightly eosinophilic granular cytoplasm with thin fibrous stroma, aggregates of fairly uniform foamy histiocytes with scattered thin-walled blood vessels and occasional multinucleated giant cells (but not Touton giant cells), and dense fibrocollagenous tissue in which were embedded spindle-shaped cells occasionally arranged in a storiform pattern. No overt meningeal whorls were seen. No significant mitotic activity or necrosis was found in the tumor. Scattered inflammatory cells composed predominantly of eosinophils were noted in the tumor, and some plasma cells and lymphocytes were also observed. Although an accumulation of foamy histiocytes was observed in the granulomatous lesion of the tumor, no cholesterol cleft, hemosiderin deposits, or multinucleated foreign body giant cells were noted.

On immunohistochemical examination (Fig. 4), CD68, HAM56, and CD163 were diffusely positive for xanthomatous cells and foamy histiocytes. These lipid-laden cells were focally positive for S100 protein. Vimentin was strongly and diffusely positive for spindle-shaped cells, but negative for CD1a, S100 protein, CD207 (Langerin),

FIG. 2. Preoperative MRI studies. Axial T1-weighted MR image (**A**) demonstrating hyperintense areas in the medial portion and rim of the mass. The mass shows heterogeneous hypointensity on T2-weighted MR (**B**) and diffusion-weighted (**C**) images. Axial (**D**) and coronal (**E**) T1-weighted MR images obtained after contrast enhancement showing homogeneous enhancement contiguous with the dura mater.

FIG. 3. Microscopic findings. The tumor reveals abundant xanthomatous cells with clear or slightly eosinophilic granular cytoplasm (**left**) and dense fibrocollagenous tissue in which are embedded spindle-shaped cells (**right**). H & E, original magnification $\times 400$. Figure is available in color online only.

glial fibrillary acidic protein (GFAP), cytokeratin, epithelial membrane antigen (EMA), neurofilament protein (NFP), α -smooth muscle actin (α -SMA), CD34, and leukocyte common antigen (LCA). The MIB-1 labeling index was approximately 1%–2%.

Ultrastructurally, the tumor cells were relatively rich in cytoplasmic organelles, but lacked cell-to-cell attachments and intercellular junctions. No overt Birbeck granules were detected.

Based on these histological findings, the lesion was diagnosed as non-LCH, because it was positive for CD68, HAM56, and CD163, but negative for CD1a and CD207. The tumor did not present Touton giant cells, which are typical of JXG; hence, we described the lesion as a fibro-xanthogranuloma. Based on the results of the immunohistological examination, the lesion is unlikely to be xantho-

matous meningioma, solitary fibrous tumor, or pleomorphic xanthoastrocytoma.

Postoperative Course

Compared with a postoperative CT scan obtained at 1 week (Fig. 5A), the CT scan performed 3 months after surgery demonstrated spontaneous regression of the residual lesion (Fig. 5B). A follow-up CT scan performed approximately 1 year after surgery showed further spontaneous regression (Fig. 5C). The residual lesion continued to regress gradually, and as of approximately 5 years after surgery, only a slight residual lesion along the tentorium remained (Fig. 5D and E). The patient is presently achieving normal developmental milestones, is not receiving medications, and has experienced no seizures.

FIG. 4. Immunohistochemical findings. Staining for CD68 (**A**), HAM56 (**B**), and CD163 (**C**) demonstrates diffuse positivity for xanthomatous cells. Vimentin staining (**D**) shows strongly and diffusely positive results for the spindle-shaped cells, but negative results for CD1a (**E**) and CD207 (Langerin) (**F**). Original magnification $\times 400$. Figure is available in color online only.

FIG. 5. Postoperative neuroimaging. Axial CT scans performed 1 week (**A**), 3 months (**B**), and 1 year (**C**) after surgery. The residual tumor shows spontaneous regression. Gadolinium-enhanced MRI studies performed 5 years after surgery (**D** and **E**) demonstrate only a slight residual lesion remaining along the tentorium.

Discussion

Cases of intracraniospinal non-LCH without cutaneous lesions are rare.^{5,10,11,14,19} The most common form of isolated non-LCH is JXG.^{3,5} To our knowledge, 29 cases of isolated intracraniospinal JXG have been reported.^{2,3,5–8,11–15,17,18,24,25,29,31–36,38,39,41–43} Histopathologically, JXG exhibits abundant lipid-laden histiocytes associated with mononuclear inflammatory infiltrates and a variable quantity of Touton giant cells.^{10,19} The immunohistochemical examination is typically positive for CD68, but negative for CD1a and S100 protein.^{8,10,11,19} Of the 29 cases, 17 were intracranial,^{2,3,5,11,13–15,18,29,31,33,34,36,38,39,42} 9 were intraspinal,^{6,7,11,17,24,25,32,35,43} and 3 showed both intracranial and intraspinal lesions.^{8,12,41} Only one case was in an adult.¹⁷ Among the 29 cases, 22 followed a benign course, whereas 7 showed progression of the disease.^{8,12,29,31,33,39,41} These 7 cases occurred in Meckel's cave or presented with multiple lesions. Orsey et al.³¹ reported a JXG diffusely involving the CNS that subsequently underwent malignant transformation and dissemination to the peritoneum and bone marrow.

In addition to the 29 cases of JXG, 8 cases of isolated intracraniospinal xanthomatous tumors have been reported as fibrous xanthoma, xanthoma, and fibroxanthoma.^{1,9,21–23,27,28,44} Histopathological examination in these cases revealed a collection of foamy histiocytes with variable amounts of fibrous tissue. Four of these cases were immunohistochemically considered to be non-LCH, based on negative staining for CD1a or S100 protein.^{9,22,23,27} The pathological findings in the other cases also resembled those of JXG.^{1,21,28,44} Most cases occurred in children and essentially followed favorable courses.

Another related lesion is XG; we found 4 cases of XG that were histopathologically similar to JXG.^{16,26,30,37} These cases were positive for CD68 and negative for CD1a, or showed Touton giant cells. The location was intracranial in 2 cases,^{16,30} and intraspinal in the other 2 cases.^{26,37} One case was in an infant,³⁷ and the other 3 cases were in adults.^{16,26,37}

Among the 42 cases of isolated intracraniospinal non-LCHs (reported as JXG, fibrous xanthoma, xanthoma, fibroxanthoma, and XG) including our case (Table 1), 18 cases (43%) were dural-based lesions.^{1,2,5,9,11,16,21,22,25–28,30,32,35,37,38} Eleven of these 18 cases were intracranial^{1,2,5,9,16,21,27,28,30,38} and the other 7 were spinal.^{11,22,25,26,32,35,37} No cases showed recurrence during the follow-up period and essentially followed benign courses.

Interestingly, in our case, the residual tumor underwent spontaneous regression after surgery. Cutaneous JXGs are known to involute spontaneously.^{5,10} In addition, spontaneous resolution of extracutaneous JXG has been reported: these lesions occurred in the larynx,^{4,40} orbit,²⁰ temporal-petrous bone,¹⁰ and intraabdomen.¹⁰ Dehner suggested that JXG may have the capacity for self-healing or spontaneous regression. Of intracraniospinal cases, 1 case of regressive fibrous xanthoma has been reported.²¹ Non-LCHs may have the potential to regress spontaneously. Because all reported dural-based cases, including ours, have followed benign courses, they could be followed up without additional treatment after resection. However, close follow-up is mandatory in patients with lesions.

Among the non-LCHs that we reviewed, clinically similar tumors have been diagnosed as different pathological

TABLE 1. Previous reports of isolated intracraniospinal non-LCHs (JXG, fibrous xanthoma, xanthoma, fibroxanthoma, and XG)

Authors & Year	Reported Pathology	Age, Sex	Site	Dural- Based		Immunohistochemical Examination	Treatment	Outcome
				Touton	GC			
Abiko & Orita, 1981	Fibrous xanthoma	6 yrs, M	Bilat temporal	Yes	No	Not described	PR	Uneventful, 4 mos
Wertz et al., 1982	JXG	20 mos, F	ON	No	Yes	Not described	Resection	NR, 15 mos
Morimura et al., 1986	Fibrous xanthoma	46 yrs, M	Gasserian ganglion	Yes	No	(-) NSE	TR, RT	NR
Kamiryo et al., 1988	Fibrous xanthoma	6 yrs, M	Bilat temporal	Yes	No	(-) GFAP	PR	NR, 6 yrs; spontaneous regression
Yamataki et al., 1990	Fibrous xanthoma	77 yrs, F	Frontal, clival region	No	No	(-) GFAP	Biopsy	Died of cerebral infarction
Kimura et al., 1991	Xanthoma	9 yrs, M	Meckel's cave	No	No	(-) S100	PR	NR, 6 mos
Paulus et al., 1992	JXG	7 yrs, M	Meckel's cave	No	Yes	(+) CD68, HAM56; (-) S100, GFAP, EMA	PR	PD, 6 mos; STR, RT; NR, 6 mos
Shimosawa et al., 1993	XG	13 mos, F	T6-9	Yes	Yes	(+) lysozyme, vimentin; (-) S100	TR	NR, 9 mos
Kitchen et al., 1995	JXG	15 yrs, F	S-1 nerve root	No	Yes	(+) HAM56; (-) S100	TR	Not described
Kim et al., 1996	Xanthoma	16 mos, M	C7-T3	Yes	No	(+) CD68, lysozyme; (-) S100, GFAP	TR	NR, 3 mos
Oyama et al., 1997	JXG	18 yrs, F	C-1	Yes	Yes	(+) CD68, lysozyme; (-) S100, NSE	TR	NR, 6 mos
Schultz et al., 1997	JXG	13 yrs, M	Temporal	No	Yes	(+) HAM56, LCA; (-) S100	TR	NR, 12 mos
Miyazono et al., 1999	Fibroxanthoma	19 mos, M	Occipital	Yes	No	(-) S100	STR	NR, 6 mos
Bostrom et al., 2000	JXG	4 yrs, M	MF, lat vents, tentorium, orbita (xanthofibroma)	Yes	Yes	(+) CD68, vimentin, LCA; (-) CD1a, S100, EMA, GFAP	PR	NR, 7 yrs
Rampini et al., 2001	JXG	34 mos, F	C5-7	Yes	Yes	(+) CD68; (-) CD1a, S100, GFAP, CD34, vimentin, CK	TR	NR, 4 mos
Gutnik et al., 2001	JXG	15 yrs, F	Parietal	No	Yes	(+) CD68, S100; (-) CD1a	Resection	Not described
Ernemmann et al., 2002	JXG	18 yrs, F	Lat vent, spinal lesions	No	No	(+) CD68; (-) CD1a, S100	Biopsy, chemo, RT	PD, 1 yr
Ashley et al., 2005	JXG	7 yrs, M	Frontal	No	No	(+) CD68, factor XIIIa; (-) CD1a, S100	TR	NR, 1 yr
Nakasu et al., 2007	JXG	2 yrs, M	Meckel's cave	No	Yes	(+) CD68; (-) S100	PR	PD, 2 mos; RT; NR, 8 mos
Orsey et al., 2008	JXG	11 yrs, M	Brainstem, cerebral peduncles	No	Yes	(+) CD68, factor XIIIa; (-) CD1a, S100	Biopsy, chemo, RT	PD, dead at 29 mos
Cao et al., 2008	JXG	18 yrs, F	C-2 nerve root	No	Yes	(+) CD68, vimentin; (-) NSE	TR	NR, 12 mos
Fulkerson et al., 2008	JXG	8 yrs, M	Frontal	No	No	(+) CD68, factor XIIIa; (-) CD1a, S100	TR	NR, 3 yrs
Chung et al., 2009	Fibroxanthoma	6 mos, F	Occipital	Yes	No	(+) CD68; (-) CD1a, EMA, vimentin	Resection	Died just after surgery
Castro-Gago et al., 2009	JXG	14 yrs, M	Multi in spinal canal	No	Yes	(+) CD68; (-) CD1a, S100	PR, chemo	AWD, 12 mos
Sun et al., 2009	JXG	5 mos, M	Parietal	Yes	Yes	(+) CD68; (-) CD1a, S100	TR	NR, 6 mos
Rajaram et al., 2010	JXG	3 yrs, M	Frontal	No	Yes	(+) CD68, S100; (-) CD1a	Resection	NR, 6 mos
Naren et al., 2011	XG	40 yrs, M	Cerebellar	Yes	Yes	(+) CD68, vimentin; (-) S100, EMA	STR	Not described
Inoue et al., 2011	JXG	38 yrs, M	C-8 nerve root	No	Yes	(+) CD68; (-) S100, GFAP, EMA, CD34	TR	NR, 2 yrs

(continued)

TABLE 1. Previous reports of isolated intracraniospinal non-LCHs (JXG, fibrous xanthoma, xanthoma, fibroxanthoma, and XG) (continued)

Authors & Year	Reported Pathology	Age, Sex	Site	Dural-Based	Touton GC	Immunohistochemical Examination	Treatment	Outcome
Husain et al., 2012	XG	38 yrs, F	Frontoparietal	Yes	Yes	(+) CD68, vimentin, S100; (-) CD1a, GFAP, EMA, α -SMA	TR	Not described
Lee et al., 2012	XG	29 yrs, M	C1–2	Yes	No	(+) CD68, lysozyme; (-) CD1a, S100, GFAP	TR	NR, 2 yrs
Vijapura & Fulbright, 2012	JXG	13 yrs, M	Posterior fossa, spinal cord	No	Yes	(+) CD68, S100, factor XIIIa; (-) CD1a	PR, chemo	PD; chemo, RT; NR, 11 mos
Abla et al., 2013	JXG	9 yrs, F	Occipital	Yes	Yes	(+) CD68, factor XIIIa; (-) CD1a, S100	Biopsy, chemo	NR, 22 mos
Deisch et al., 2013	JXG	14 yrs, F	Gasserian ganglion	No	Yes	(+) CD68, LCA; (-) CD1a, S100	PR, chemo	NR, 9 mos
	JXG	15 yrs, F	T-9	Yes	Yes	(+) CD68, factor XIIIa, S100; (-) CD1a	TR	NR, 8 mos
Tamir et al., 2013	JXG	3.5 yrs, M	Occipital	No	No	(+) CD68, factor XIIIa; (-) CD1a, S100	TR	NR, 28 mos
	JXG	3.5 yrs, F	Meckel's cave	No	Yes	(+) CD68, vimentin, factor XIIIa; (-) CD1a, S100, EMA, GFAP, α -SMA	PR	PD, 1 yr; chemo; NR, 2.9 yrs
Chiba et al., 2013	JXG	4 yrs, M	Multi intracranial, spine	No	No	(+) CD68, vimentin, S100; (-) CD1a, GFAP	Biopsy, chemo	PD
Gressot et al., 2013	JXG	6 wks, M	Brainstem, thalamus, BG, ONS, lat vents	No	No	(+) CD68, S100; (-) CD1a	Biopsy, chemo	NR, 7 mos
Wille et al., 2013	JXG	6 mos, ?	C3–7	No	No	(+) CD68; (-) CD1a, S100	TR	NR, 2 yrs
Jain & Mehta, 2013	JXG	8 yrs, M	Lat vents	No	Yes	(+) CD68, S100; (-) CD1a, GFAP	PR	Not described
Konar et al., 2014	JXG	18 yrs, M	C3–5	Yes	Yes	Not described	TR	NR, 3 mos
Present study	Fibroxanthogranuloma	8 mos, M	Occipital	Yes	No	(+) CD68, HAM56, CD163, vimentin, S100; (-) CD1a, CD34, LCA, CK, GFAP, NFP, α -SMA, CD207, EMA	PR	NR, 4 yrs; spontaneous regression

AWD = alive with disease; BG = basal ganglia; chemo = chemotherapy; CK = cytokeratin; GC = giant cell; MF = middle fossa; multi = multiple; NR = no recurrence; NSE = neuron-specific enolase; ON = optic nerve; PD = progressive disease; PR = partial resection; RT = radiation therapy; STR = subtotal resection; TR = total resection; vent = ventricle; ? = unknown.

entities. Moreover, one pathological diagnosis, especially in JXG, has included both benign and malignant cases. Further investigations and the accumulation of more cases are needed to elucidate the pathogenesis of the non-LCHs.

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